North American Neuro-Ophthalmology Society

42nd Annual Meeting

February 27 - March 3, 2016

JW Starr Pass Marriott • Tucson, Arizona

Poster Session I: Clinical Highlights in Neuro-Ophthalmology

Sunday, February 28, 2016 • 12:30 pm – 2:00 pm

Authors will be standing by their posters during the following hours:

Odd-Numbered Posters: 12:30 - 1:15 pm

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**Please note that all abstracts are published as submitted.*

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Poster 1
Compressive Optic Neuropathy from Salivary Gland Tumor of Sphenoid Sinus

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Introduction:
We present compressive optic neuropathy from salivary duct carcinoma of minor salivary gland origin, arising primarily from the sphenoid sinus.

Methods:
Case Report

Results:
A 59-year-old Hispanic man presented with an acute right, pupil-sparing CN III palsy. PMH included tubular adenoma of the colon, HTN, DM II, and NPDR. BCVA was 20/70 OD and 20/50 OS without RAPD. He had bilateral NPDR. Non-contrast CT head showed a non-expansile ground glass trabeculation of the right sphenoid sinus felt to represent interrupted pneumatization. Contrasted brain MRI/MRA redemonstrated this lesion with no other intracranial or vascular abnormality. CRP, TSH, ESR, ACE, ANA, Lyme Ab, SSA/B were normal. He was diagnosed with an ischemic CN III palsy and strict glycemic control was recommended. He presented to neuro-ophthalmology clinic 4 months later with one-month of blurry vision OD. His 3rd nerve palsy had resolved. He now had CF vision OD and stable acuity OS. A right RAPD was present. DFE showed right optic atrophy. Contrasted MRI of the brain and orbits showed a heterogeneous, enhancing, expansile mass emanating from the sphenoid sinus involving the right orbital apex. CT orbits was concerning for osteosarcaoma. Pathology showed a carcinoma that stained positively for pancytokeritin, CAM 5.2, androgen receptors, PSAP, PSA, and CK-7. CD31, CD34, PAX8, TTF1, chromogranin, P63, S-100, CK5-6, and synaptophysin were negative. PSA was 3.54. Clinical examination revealed a 35-gram prostate without nodularity. CT and PET/CT showed diffuse osteoblastic metastatic disease. There were no other lesions seen systemically. Positive staining for CK-7 and androgen receptors supported a salivary duct origin[1]. His tumor was determined to be an adenocarcinoma of minor salivary gland origin, arising from the sphenoid sinus, stage T4dN0M1.

Conclusions:
This is an uncommon case of prostate marker positive ductal adenocarcinoma, arising from minor salivary glands of the sphenoid sinus, resulting in compressive optic neuropathy.

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Keywords: Neuroimaging, Tumors, Skull Base, Optic Neuropathy

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Poster 2

Unexpected Pathologic Diagnosis of Primary Dural B Cell Marginal Zone Lymphoma

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Introduction:
Several infiltrative and neoplastic conditions may have similar features on MRI as meningioma. A biopsy should be obtained when clinically indicated to exclude diagnoses that may require a different treatment from meningioma.

Methods:
We present a single case report of severe vision loss of the left eye. Clinical, neuro-radiologic, and histopathologic findings will be presented.

Results:
A 54 year-old man presented with six weeks of progressive, painless decrease in vision of the left eye. The right eye lost vision to no light perception four years prior secondary to an ophthalmic artery occlusion. Brain MRI with contrast at that time was negative. Cataract extraction with intraocular lens implant of the left eye was performed locally, but post-operatively vision continued to decline to count fingers at one foot. Repeat MRI showed a suprasellar mass compressing the chiasm and intracranial optic nerves on the right more than the left and meningeal tail. A craniotomy for the presumed planum meningioma was performed. Histologic examination and immunohistochemical stains were consistent with a low-grade marginal zone B-cell lymphoma of mucosa-associated lymphoid tissue. No evidence of concomitant meningioma could be found. The patient received radiation therapy for the remainder of the unresectable tumor as well as intrathecal chemotherapy. Vision of the left eye improved to 20/250 post-operatively with a remaining paracentral inferotemporal visual field defect.

Conclusions:
This case highlights the difficulty in evaluation of a patient with a previous ophthalmic artery occlusion of the right eye with a previously negative MRI complicating clinical evaluation of new vision loss in the left eye. Repeat imaging and acquiring tissue for pathology should be obtained in complex cases of vision loss. In addition, extranodal marginal zone B-cell lymphoma should be considered in the differential diagnosis of a lesion radiographically appearing as a meningioma.

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Keywords: Neuroimaging, Tumors, Optic Neuropathy

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 3
Bilateral Epstein-Barr Virus Optic Neuritis in a Lung Transplant Patient

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Introduction:
In organ transplant patients, atypical optic neuritis necessitates a broad differential, including post-transplant lymphoproliferative, inflammatory, toxic, and infectious causes.

Methods:
Case report

Results:
A 57-year-old female presented with acute onset headache, malaise, and left eye vision loss. Medical history was significant for systemic scleroderma necessitating lung transplantation six years prior. Daily medications included prednisone and sirolimus, and prophylactic sulfamethoxazole/trimethoprim and valganciclovir. On examination, BCVA was 20/20 OD and 20/400 OS, with a 3+ left APD. Fundus examination revealed bilateral disc edema with nerve fiber layer hemorrhages, more severe on the left. Brain MRI showed enlargement of the retrobulbar left optic nerve and sheath along with subtle nerve enhancement. Whole body PET/CT was unremarkable. Serum testing, including for NMO-IgG, RPR, ACE, and ANCA, was unremarkable. CSF analysis showed mild lymphocytic pleocytosis, elevated protein, and negative cytology. With viral PCR results pending, vision in the right eye dropped steadily to counting fingers, and the APD was now detected on that side. Right disc edema worsened and lipoproteinaceous macular fluid appeared. Cytomegalovirus, herpes simplex virus, and varicella zoster virus DNA was absent from CSF and serum. PCR analysis showed 10,400 copies/mL of Epstein-Barr virus (EBV) DNA in the CSF, but none in the serum. The patient was treated with intravenous acyclovir for six weeks, followed by long-term oral valacylovir. After initiating targeted treatment, vision improved slightly as CSF EBV titer decreased steadily, most recently to 2,600 copies/mL at 6-month follow up.

Conclusions:
EBV optic neuritis is rare with few reported cases[1-3], mostly involving immunocompetent patients developing optic neuropathy after contracting infectious mononucleosis. In patients with atypical optic neuritis, it is important to include EBV in the differential, even when the patient is immunocompetent or has negative blood titers. This case is unique given the bilaterality with both neuroretinitis and retrobulbar optic neuritis.

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Keywords: Optic Neuritis, Epstein-Barr Virus, Neuroretinitis

Financial Disclosures: The authors had no disclosures.

Grant Support: None

Poster 4

Purtscher's Retinopathy as a Manifestation of Hemophagocytic Lymphohistiocytosis

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Introduction:

Hemophagocytic Lymphohistiocytosis (HLH) is an inflammatory disease whose diagnostic criteria include fever, cytopenia, and hemophagocytosis in the bone marrow. Often incited by viral infections, it leads to uncontrolled activation of macrophages and a subsequent cytokine storm. We describe the case of a 52-year-old previously healthy Hispanic woman who presented with bilateral vision loss and purtschers retinopathy secondary to HLH.

Methods:

Upon presentation, the patient underwent comprehensive ophthalmologic assessment, including color fundus photography and spectral-domain optical coherence tomography (SD-OCT).

Results:

The patient was admitted for worsening fever and found to have elevated transaminases, a pericardial effusion, and pulmonary edema. MRI of the brain revealed bilateral, near-symmetric subcortical white matter T2/FLAIR hyperintensities of the occipital lobes. She developed progressive pancytopenia, elevated serum ferritin, triglycerides and LDH as well as elevated soluble Interleukin 2 receptor alpha (sIL2Ra) levels. Epstein-Barr virus and Cytomegalovirus levels were elevated on quantitative PCR. Bone marrow biopsy showed hemophagocytosis, confirming the diagnosis of HLH.The fundus exam revealed patchy, polygonal areas of retinal whitening and edema with associated flame-shaped hemorrhages scattered throughout the posterior pole. The areas of retinal whitening corresponded to inner retinal edema on SD-OCT. These findings were consistent with purtscher's retinopathy. On subsequent exam when the patient was more alert, she was noted to have hand motion vision in both eyes.

Conclusions:

This report represents a rare case of purtscher's retinopathy as a manifestation of HLH. Since patients with HLH can develop DIC secondary to cytokine storm, it is reasonable to propose that they are also at risk of developing a purtscher's retinopathy from this consumptive coagulopathy. HLH has previously been shown to have various neurologic and neuro-ophthalmologic manifestations, including encephalopathy and nystagmus. This disorder, along with other cytokine storm diseases, should be included in the differential of purtscher's retinopathy as well.

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Keywords: Neuro-Ophth & Systemic Disease, Retina, Vascular Disorders

Financial Disclosures: The authors had no disclosures.

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Poster 5

Central Retinal Vein Occlusion, Paracentral Acute Middle Maculopathy, and Cilioretinal Vein Sparing with Acquired Shunt in a Patient with Antiphospholipid Syndrome and Cryoglobulinemia

Dov B. Sebrow[1], Jeffrey G. Odel[1], Jesse J. Jung[1,2], Jason D. Horowitz[1]

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Introduction:
Paracentral Acute Middle Maculopathy (PAMM) is a recently described entity resulting from ischemia in the intermediate and deep capillaries of the retina, and can be associated with central retinal vein occlusion (CRVO). We describe the first reported case of CRVO with PAMM and cilioretinal vein sparing in the setting of two pro-thrombotic diseases; antiphospholipid syndrome (APS) and type II cryoglobulinemia.

Methods:
A 50-year-old man presented with intermittent loss of vision in his right eye. Color photography, SD-OCT and fluorescein angiography were performed to confirm the diagnosis.

Results:
On presentation, visual acuity was 20/HM OD, 20/30 OS with an RAPD OD. Fundus examination OD revealed vascular tortuosity, few scattered flame-shaped hemorrhages, as well as retinal whitening around the macula. SD-OCT revealed thickening of the middle layers of the retina that correlated with these areas of whitening. Laboratory testing revealed that the patient had APS and type II cryoglobulinemia.A discrete area superotemporal to the disc OD was spared from the PAMM-induced macular whitening. This area corresponded to the distribution of a single vein on FA leading to the superior edge of the optic disc where it anastamosed to an optociliary shunt vessel that had developed since previous fundus photos in 2008. As the patient's CRVO progressed and subsequently stabilized after treatment in the following months, this area of venous sparing remained the only functional, non-ischemic retinal tissue in his macula.Presumably, this vein possessed privileged and uncompromised blood flow by circumventing the occluded venous circulation and emptying instead through the shunt vessel at the disc margin.

Conclusions:
PAMM should be considered in the differential diagnosis of retinal whitening and is thought to occur in CRVO secondary to altered capillary pressure. This is also the first description of venous sparing of the retina, which can occur in a CRVO if a cilioretinal vein is present.

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Keywords: Retina, OCT, Vascular Disorders

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 6
A Case of Wyburn-Mason Syndrome

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Introduction:
Wyburn-Mason syndrome is rare Phakomatosis which characterizes neuro-oculo-cutaneous involvement. The symptoms and signs are variable according to the location and size of arterio-venous malformation (AVM). We present 6 year-old boy with near blind vision in his right eye which reveals marked vascular malformation on the optic disc and extend to the orbit, nasal cavity and perichiasmal area.

Methods:
Case presentation
- Ophthalmologic exams: VA. Ref. fundus photo, FAG, OCT, VEP
- Neuro-radiologic evaluation: Transfemoral cerebral angiography

Results:
On fundus examination, right optic disc and posterior pole are blocked with severely tortuous and engorged retinal vessels which show early filling in FAG. OCT reveals vascular shadow underneath the ganglion cell layer and disrupted retinal layer contour. Trans-femoral cerebral arteriography finds 2.5x4.0cm AVM nidus which extends to the right orbit, perichiasm and hypothalamus.

Conclusions:
Multidisciplinary approach is needed to diagnosis Wyburn-Mason syndrome. In case of optic nerve and macular involvement, vision threatening should be considered. With full understanding of this entity, we can differentiate diagnosis from Von Hippel-Lindau disease, Stuger-Weber syndrome, Retinal cavernous hemangioma and vasoproliferative retinal tumor.

References: None.

Keywords: Phakomatosis, AVM, Optic Disc

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 7

Debulking Optic Nerve Gliomas for Disfiguring Proptosis: A Globe-Sparing Approach by Lateral Orbitotomy Alone

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Introduction:

Optic nerve gliomas (ONG) with disfiguring proptosis and severe vision loss usually require a combined intraorbital and intracranial tumor resection. Incomplete tumor removal (Spicer et al.) and associated morbidity using this surgical approach suggests that a less invasive approach may be preferred.

Methods:

A retrospective case review of 3 patients with sporadic (not secondary to NF1) ONG manifesting profound vision loss and disfiguring proptosis who were treated at our institution. All patients underwent a lateral orbitotomy alone with a globe sparing resection of just the orbital intraconal portion of the ONG

Results:

Three ONG patients, all previously treated with chemotherapy, underwent debulking surgery between 12 and 13 years of age. One patient's ONG extended from the orbit to the chiasm. The second patient had intracanalicular extension, and the third case was isolated to the orbit. All were treated via a lateral orbitotomy alone for removal of the orbital, intraconal portion of the optic nerve and tumor, leaving a stump of tumor 2-3 mm posterior to the globe and tumor in the prechiasmal optic nerve. All 3 tumors were pilocytic astrocytoma (WHO Grade I). The average follow-up was 24 months (range 8-40 months). All patients had a significant reduction in proptosis with outstanding cosmetic results, blindness in the operated side, but normal retinal perfusion. Temporary post-operative complications which improved included esotropia and decreased corneal sensation. Follow-up imaging thus far reveals no tumor progression.

Conclusions:

Disfiguring proptosis due to ONG can be improved via a lateral, orbitotomy-only, debulking surgical procedure utilizing a less invasive approach rather than attempting complete tumor removal. The implication of leaving tumor near the globe or at the proximal optic nerve or chiasm following orbital debulking warrants further study

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Keywords: Optic Nerve Glioma

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Grant Support: None.

Poster 8

Longitudinally Extensive Spinal Cord Lesion in Leber's Hereditary Optic Neuropathy Due to the M.3460A Mitochondrial DNA Mutation

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Introduction:
Leber's hereditary optic neuropathy (LHON) is an inherited mitochondrial disorder characterized by optic nerve degeneration leading to progressive visual loss. Prior reports have described multiple sclerosis (MS) associated with all three primary LHON mutations (m.11778A>G, m.3460A>G, and m.14484T>C. Furthermore, neuromyelitis optica (NMO) has been reported in a patient with nt11778 LHON mtDNA mutation.

Methods:
We report a patient with 3460- point mutation for LHON who developed a longitudinally extensive spinal cord lesion with negative work-up including anti-NMO antibodies.

Results:
A 43-year-old female presented with progressive bilateral visual loss over 8 weeks. She reported a family history of visual loss. Brain MRI showed a mild central pontine enhancement. Genetic testing confirmed the 3460-point mutation for LHON. She had progressive decline in vision over a year with visual acuities of 20/300 OU and bilateral optic nerve pallor. The patient received Idebenone on a compassionate basis and recovered some visual acuity, improving to 20/250 OD and 20/150 OS after one year of treatment.Two years after her initial vision loss, she complained of lower limbs spasms, and neurological examination revealed a decrease in position and vibration sense in the lower limbs. MRI of the spine showed a posterior predominant longitudinally extensive spinal cord lesion that extended throughout the cervical cord and most of the thoracic cord. CSF composition was within normal range with negative oligoclonal bands. An extensive work-up was done to rule out other possible causes of her spinal cord lesion. Anti-NMO antibodies, inflammatory, metabolic, infectious, and paraneoplastic work up was negative.

Conclusions:
We report a case of LHON positive mutation associated with a spinal cord lesion. Our case report raises the possibility that the spinal cord lesion and even the previously reported MS-like lesions could be related to mitochondrial dysfunction and secondary axonal transport failure rather than an acquired inflammatory etiology.

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Keywords: Neuroimmunology

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Poster 9
Growth of an Optic Disc Vascular Anomaly for Twelve Years

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Introduction:
Idiopathic retinal vasculitis, aneurysms, and neuroretinitis (IRVAN) is an uncommon idiopathic disorder in otherwise healthy young women in the third and fourth decades. It is characterized by aneurysmal dilations of the arteries of the disc and retina with peripapillary exudates, arterial sheathing and irregular venous dilation. The clinical course is unpredictable, ranging from benign self-limited form to severe visual loss due to an exudative or ischemia maculopathy. The long-term course of the disorder is not well documented

Methods:
Case report.

Results:
A 52 years-old female presented with normal acuity but an inferior field defect OS in April 2002. Funduscopic examination was normal OD but revealed a large vascular structure involving the optic disc with an overlying small hemorrhage and a large exudate adjacent to the disc OS. FA showed late staining of the disc. MRI/MRA and blood studies for systemic inflammatory disorders and infections were negative. The retinal abnormalities and visual field loss resolved leaving the vascular anomaly of the disc. The patient was seen in 2010 with normal visual testing but enlargement of the disc abnormality; OD was normal. In 2014 symptoms recurred OS. Exam showed Va 20/40, Ishihara 11/12 and nonspecific depression on perimetry. There was a dilated elongated segment of artery at the disc OD. OS showed vitreous hemorrhage obscuring the disc. FA revealed staining of the disc, the superior macroaneurysm and a segment of an inferior arteriole OD. There was staining of the optic disc with marked capillary nonperfusion peripherally OS.

Conclusions:
This is the longest followup of a patient with IRVAN. There was slow progression of the vascular abnormalities of the disc OS and development of similar abnormalities on the disc OD before a vitreous hemorrhage and marked capillary nonperfusion developed in the originally involved eye 12 years after initial presentation.

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Keywords: Retina, Vascular Disorders, Visual Fields, Optic Neuropathy, Tumors

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Poster 10
Retreatment with Ethambutol After Toxic Optic Neuropathy

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Introduction:
Ethambutol is an important component of the treatment for mycobacterial infections. Toxic optic neuropathy is a dose-dependent side effect of ethambutol treatment, occurring in approximately 5-6% of patients at 25mg/kg/day and in 1% at 15mg/kg/day. When optic neuropathy is suspected, ethambutol is typically withdrawn. Some patients require retreatment for mycobacterial infections, but the safety of ethambutol retreatment in a patient who previously developed and recovered from ethambutol-induced optic neuropathy or chiasmopathy has not been established.

Methods:
We reviewed the medical record of a 59-year-old woman who was diagnosed with pulmonary Mycobacterium avium complex (MAC) infection and developed ethambutol-induced optic neuropathy 7 months into treatment, recovered vision after ethambutol was discontinued, and required retreatment for pulmonary MAC infection 10 years later.

Results:
The patient's initial ethambutol toxicity was noted 7 months into therapy at a dose of 22mg/kg/day. She developed dyschromatopsia and decreased acuity to 20/80. She required retreatment for pulmonary MAC infection 10 years later. At time of neuro-ophthalmologic consultation before reinitiation of ethambutol, her vision was 20/20 OU with no residual dyschromatopsia. She was retreated with ethambutol at 25mg/kg 3 days per week for 14 months and copper supplementation without recurrence of optic neuropathy.

Conclusions:
Retreatment of mycobacterial infections with ethambutol after prior ethambutol-induced optic neuropathy is a controversial topic with very little data in the literature and most neuroophthalmologists recommend against retreatment. Though further research is required, our case shows that recurrent toxicity can be avoided when a lower dose is used.

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Sadun AA, Wang MY. Drug-related mitochondrial optic neuropathies. J Neuroophth. 2013;33:172-178.

Keywords: Optic Neuropathy, Neuro-Ophth & Infectious Diseases, Ethambutol

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Poster 11
Multifocal Electroretinogram Findings in Two Patients with Idiopathic Neuroretinitis

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Introduction:
Neuroretinitis is a rare pathology of the optic disc vasculature, resulting in edema of the disc and macula with intra- and sub-retinal effusions. To date, full field electroretinogram (ERG) analysis has been described in several patients with neuroretinitis. Our purpose is to describe multi-focal ERG findings in two patients with idiopathic neuroretinitis, which to our knowledge has not been previously described.

Methods:
A retrospective analysis of two cases.

Results:
Two patients, ages 59 (female) and 31 (male), presented with unilateral subacute vision loss and eye pain. Both were found to have a visual acuity of 20/200 OD and optic disc edema on initial examination and subsequently developed macular exudates in a stellate pattern one to two weeks after presentation. Laboratory testing was negative for Bartonella Henselae and Quintana antibodies, rapid plasma reagin (RPR), Tuberculosis (Quantiferon Gold), and Borrelia Burgdorferi. Optical coherence tomography (OCT) showed intraretinal exudates and sub-retinal fluid accumulation in both patients. Automated visual field analysis showed significant hemifield deficits in both patients. Patient 1 had a dense inferior altitudinal defect, while patient 2 had a dense superior altitudinal defect. Multifocal ERG in both patients showed significant waveform amplitude and latency abnormalities centrally and nasally in the distribution of edema and subretinal fluid found on fundus exam and OCT.

Conclusions:
While full-field ERG analysis in patients with neuroretinitis may be within normal limits, multifocal ERG analysis is likely to show decreased waveform amplitudes and prolonged latencies. Multifocal ERG may contribute to a fuller understanding of the pathogenesis of neuroretinitis, and could potentially provide an avenue to earlier diagnosis and treatment of this rare condition.

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Keywords: Neuroretinitis, Multifocal Electroretinogram, Stellate Maculopathy

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 12

Neuromyelitis Optica Spectrum Disorder Presenting as Bilateral Anterior Optic Neuritis with Venous Stasis

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Introduction:
We present a case of bilateral anterior optic neuritis complicated with bilateral retinal vein stasis in the setting of neuromyelitis optica spectrum disorder.

Methods:
This is a descriptive case report based on clinical records, patient observation and analysis of diagnostic studies.

Results:
A 31 year-old man with no significant medical history presented with new onset of bilateral decreased vision and pain on eye movements. Vision in the left eye dramatically decreased a week prior to presentation and he started noticing blurry vision in the right eye 5 days thereafter. On examination, visual acuity was 20/30 OD and 20/1000 OS. He had a dense left afferent pupillary defect. Fundus examination showed crowded and swollen optic discs, dilated veins and numerous macular hemorrhages with pale center. There were ceco-central scotomas on visual field examination. Fluorescein angiogram disclosed delayed filling of the central retinal veins. An MRI of the brain and orbits showed enlarged enhancing optic nerves and no other abnormalities. AQP4 antibodies were positive in the blood and cerebro-spinal fluid. Patient was treated with a 3-day course of intravenous corticosteroids and underwent 5 plasma exchanges. Vision improved to 20/20 OD and 20/40 OS a week later. Optic disc swellling, dilation of the retinal veins and macular hemorrhages resolved within a month.

Conclusions:
Central retinal vein occlusions (CRVO) have been reported in severe papilledema, and optic disc swelling is a common feature of CRVO. However, venous stasis can also occur in mild optic disc swelling, especially when there is prominent enhancement of the optic nerves on the MRI. Retinal vein stasis with mild optic disc swelling should raise suspicion about a severe retrobulbar process impeding the venous outflow, especially when visual impairment is incommensurate with the fundus findings.

References: None.

Keywords: Neuromyelitis Optica, Anterior Optic Neuritis, Optic Disc Swelling, Venous Stasis, Central Retinal Vein Occlusion

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Grant Support: None.

Poster 13
Orbital Apex Syndrome Secondary to Scedosporium Boydii

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Introduction:
Orbital Apex Syndrome (OAS) consists of any combination of 3rd, 4th or 6th nerve paresis, V1 distribution sensory loss, Horner's syndrome and visual loss due to optic nerve involvement. OAS secondary to mucormycosis in the immunocompromised patients is well described. Unlike *Aspergillus*, which is the most commonly reported, *Scedosporium boydii* has not been previously reported as a cause of OAS. *Scedosporium boydii* is now increasingly recognized as a cause of resistant life-threatening infections in immunocompromised patients.

Methods:
Case Report.

Results:
We report the case of an 89-year-old female with a remote history of lymphoma who presented with rapidly progressive infiltrative optic neuropathy. She also had a right OAS primarily manifested as a pupillary-involving right oculomotor palsy, right superior oblique palsy, and right abducens palsy. MRI of the brain showed a right intraconal orbital cellulitis with extension to the orbital apex. CT scan of the sinuses showed mucosal thickening or mass within the right sphenoid sinus extending into the right posterior ethmoid air cells. The patient underwent nasal endoscopy with a complete right ethmoidectomy and sphenoidectomy. She had a postoperative diagnosis of a right sphenoid mycetoma. H&E staining showed acute angled branching septate, compatible with *Aspergillosis*. However, fungal cultures were positive for *Scedosporium boydii*.

Conclusions:
We present the first reported case of OAS secondary to *Scedosporium boydii*. Neuroimaging demonstrated a soft tissue right intracranial mass with extension to the right orbital apex. The biopsy and initial staining were consistent with *Aspergillosis*, however additional fungal culture and additional staining were consistent with a final diagnosis of *Scedosporium boydii*. Diagnosis of *Scedosporium* infection is difficult, because clinical features and histopathology are similar to those of *Aspergillus*. The clinician should consider *Scedosporium boydii* in the differential diagnosis of an orbital apex syndrome.

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Keywords: Orbital Apex Syndrome, Scedosporium, Aspergillosis, Oculomotor Nerve Palsy

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Poster 14
Electrophysiologic Analysis of the Foster Kennedy Sign Due to a Pituitary Mass

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Introduction:
The Foster Kennedy sign is defined by unilateral optic nerve atrophy due to direct compression by an intracranial mass lesion with resultant contralateral papilledema. It varies in accompanied clinical presentation, and may include headaches or vision loss. Increased knowledge of disease progression and objective diagnostic options, including electrophysiologic tests, are needed to further address and monitor such pathologic conditions. This case is unique as it is due to a pituitary macro-adenoma, which is a rare precipitant of Foster Kennedy sign.

Methods:
A 44-year-old woman with a history of severe, post-partum pre-eclampsia experienced visual changes and was found on imaging to have a compressive pituitary macro-adenoma. She presented for neuro-ophthalmology evaluation where initial exam showed decreased visual acuity of the left eye, atrophy of the left optic nerve, and papilledema of the right optic disc without evidence of pupillary defects, nystagmus, or compromised gaze. Such presentation was consistent with the Foster Kennedy sign. Further diagnostic workup included visual field (VF) testing, ocular coherence tomography (OCT), visual evoked potential (VEP), full field electroretinogram (ERG).

Results:
30-2 VF indicated visually significant bilateral temporal hemianopsia. VEP and ERG resulted in subnormal amplitudes, which aided to objectify the pathogenesis behind the Foster Kennedy sign and its relationship to the patient's space-occupying pituitary mass.

Conclusions:
The Foster Kennedy sign is a rare finding resulting from different intracranial etiologies that compromise the integrity and function of the optic nerves, and thus, vision. In our case, the anterior visual pathway was directly affected, which supports the involvement of a direct pathogenic mechanism, specifically, pituitary macro-adenoma. Both clinical evaluation and quantifiable diagnostic testing proved valuable tools for the diagnosis and plan for future monitoring of this very rare case and hold promise for others with a similar mechanism.

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Lai, Chiu, Lin, Sanders, Foster Kennedy Syndrome: Now and Then, Journal of Neuro-Ophthalmology, Volume 34, Page 92-94, 2014.

Keywords: Tumors, Visual Fields, Optic Neuropathy, Orbit/Ocular Pathology

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Poster 15
Pembrolizumab-Related Panuveitis/Choroiditis

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Introduction:
There have been several documented cases of Vogt-Koyanagi-Harada (VKH) like symptoms associated with interferon-alpha treatment in chronic hepatitis C patient. There has also been at least two reports of similar presentations with the use of ipilimumab, another monoclonal antibody used in the treatment of melanoma. We present a case of bilateral optic disc swelling and panuveitis with choroiditis and multifocal serous retinal detachments in a patient being treated with Pembrolizumab and Pegylated interferon alfa-2b for metastatic melanoma.

Methods:
A single case report.

Results:
A 68 year old male being treated with pembrolizumab and pegylated interferon alfa-2b for metastatic melanoma presented with a two week history of worsening headaches and progressive blurry vision. He was found to have bilateral disk edema, relative hypotony, panuveitis, and multifocal serous retinal detachments. Fluorescein angiography revealed bilateral optic disc leakage, diffuse leakage and pooling in the posterior pole consistent with active choroiditis and serous retinal detachments but no definite vasculitis. Optical coherence tomography imaging demonstrated significant optic disc swelling, diffuse retinal edema with multifocal pockets of sub-retinal fluid and choroidal thickening in both eyes. The decision was made in consultation with the patient's oncology team to stop his anti-cancer medications. The patient was then treated aggressively with posterior sub-Tenons triamcinolone injections in both eyes and oral prednisone on a slow taper. The patient made a rapid recovery with resolution of his iritis and vitritis within one week and almost complete resolution of his subretinal fluid and disc swelling within one month. His vision also improved significantly over this period of time.

Conclusions:
Our patient likely represents the first reported case of a VKH-like presentation in a patient being treated with pembrolizumab. His course was notable for rapid improvement of symptoms after the initiation of steroids and cessation of the concerning medications.

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Keywords: Pembrolizumab, Vogt-Koyanagi-Harada (VKH), Disc Swelling

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Poster 16
"Here Kitty, Kitty, Kitty": Optic Neuropathy in the Setting of Exposure to Bartonella Henselea

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Introduction:
Cat scratch disease caused by Bartonella henselea has traditionally been associated with unilateral neuroretinitis consisting of optic disc swelling and a macular star. Larger published case series show features such as bilaterality, retinal and choroid lesions, and enhancement of the optic nerve head on MRI are now being recognized as more common. We present a case with these features accompanied by high resolution OCT images and fundus photos documenting progression and resolution.

Methods:
Case report and review of literature.

Results:
A 16 year old female presented with complaint of 2 day history of fever and blurry vision in her right eye. Her past medical history was significant for recent exposure to stray kittens with fleas, barn yard animals, a close contact who recently returned from Moldova, and unprotected sexual activity. Her corrected visual acuity at near was 20/70 OD 20/20 OS with no afferent pupillary defect and full colors. Humphrey visual field 30-2 demonstrated enlarged blind spot in the right eye. Fundus exam showed bilateral optic disc swelling although the right disc was more prominent and atypical in appearance with an adjacent white retinal lesion. OCT confirmed disc swelling OU with circumpapillary NFL thickening and subretinal and intraretinal fluid tracking from the disc OD. The patient's vision rapidly declined to 20/200 OD and was therefore started on high-dose IV steroids and doxycycline which resulted in rapid improvement of retinal fluid and disc swelling. Extensive workup was conducted for both bilateral optic nerve swelling and neuroretinitis which eventually revealed infection with Bartonella henselea.

Conclusions:
Our case provides evidence to support a much less expected, but well documented ocular manifestation of cat-scratch disease, and anecdotally shows positive results for use of steroids and antibiotics in cases of worsening clinical presentation. It additionally provides detailed imaging of the progression and resolution of clinical findings.

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Keywords: Cat Scratch Disease, Bartonella Henselea, Neuroretinitis, Macular Star

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Poster 17

Hemodialysis as a Possible Risk Factor for Ischemic Optic Neuropathy: Case Report

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Introduction:
Nonarteritic Anterior Ischemic Optic Neuropathy (NAION) is presumed to result from circulatory insufficiency within the optic nerve head. NAION has been reported in association with many conditions that may predispose to decrease its perfusion. The hemodynamic compromise such as the one generated in systemic hypotension, blood loss and anemia may produce optic nerve ischemia. Hypotensive episodes in the setting of chronic anemia without blood loss, as in chronic hemodialysis, may result in Ischemic Optic Neuropathy (ION).

Methods:
Single Case Report

Results:
A 38 year-old caucasian male presented with painless visual loss OS. The patient reported chronic hemodialysis due to a past medical history of glomerulonephritis 18 years prior and hypertension. Laboratory evaluation revealed a low red blood cell count. In the neuro-opthalmologic examination the best corrected visual acuity was OD 20/20 and OS 20/200 with a relative afferent papillary defect. Color vision was OD 5/8 and OS 0/8. The fundus examination revealed a pale optic disc edema with peripapillary retinal hemorrhage in OS. The OD disc was normal. Visual fied evidenced a nasal depression with a centrocecal defect and the Optical Coherence Tomography showed a decrease average retinal nerve fiber layer thickness OS.

Conclusions:
Chronic hemodialysis can be considere as a risk factor for ION. Related to hypotension most often presents in patients with chronic renal failure and dialysis. The accelerated diffuse vasculopathy that occurs in this disease may predispose to the development of ION, as may previus chronic hypertension, with possible arteriosclerosis and impaired autoregulation oft the optic disc vascular supply.

References: None.

Keywords: Ischemic Optic Neuropathy, Hemodialysis

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Grant Support: None.

Poster 18
Neuroretinitis After a Bee Sting

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Introduction:
To report and review a case of toxoplasma neuroretinitis after a bee sting

Methods:
This is a descriptive case report based on clinical records, patient observation and analysis of diagnostic studies.

Results:
A 17-year-old woman presented with 3-day history of decreased vision and pain on eye movement in the left eye. The patient had history of a bee sting about two weeks ago and had an epi pen injection for allergic skin reaction, otherwise she was healthy and had no recent vaccination or no contact with animals. On evaluation, 5 days after the development of symptom, the patient complained of eye pain and pain on eye movement in the left eye with seeing black spot. The examination showed clinical signs of optic neuropathy with decreased visual acuity (20/400), dyschromatopsia, visual field defect and relative afferent pupillary defect in the left eye and also showed swelling of inferior optic disc and papillomacular bundle in the left eye. MRI orbit with and without contrast with fat suppression showed slightly increased signal around the left optic disc with no enhancement of optic nerve or sheath. Further investigation with serologic test was considered. CBC, ESR, CRP, ANA, ANCA, ACE, Lyme, quantiferon gold, FTA-ABS, Bartonella were performed and they were unremarkable. Given the poor visual sensory function in the left, systemic corticosteroid was administered and showed minimal improvement of eye pain. Additional serologic test was considered for toxoplasmosis, HSV, histoplasmosis and toxoplasma IgG was positive and otherwise negative. Under the suspicion for activated toxoplasmosis neuroretinitis in the left eye, Bactrim with prednisone was started and in the four weeks with the treatment, her visual acuity has improved to 20/70 in the left eye.

Conclusions:
This is a rare case of toxoplasma neuroretinitis after a bee sting. The acquired ocular toxoplasmosis may be considered in clinical setting of optic neuritis or neuroretinitis after a bee sting.

References: None.

Keywords: Optic Neuropathy, Retina, Neuro-Ophth & Infectious Disease

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 19

Analysis and Outcome of Ocular Syphillis Manifesting as Optic Neuritis – A Report of Five Cases From a Tertiary Referral Eye Care Centre

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Introduction:
Syphilis is a sexually transmitted disease caused by the spirochete *treponema pallidum* with protean manifestations. Ocular syphilis can masquerade as anterior uveitis, panuveitis, retinitis, retinal vasculitis, vitritis or papillitis. We aim to describe clinical features and outcome of patients with ocular syphilis who presented with disc swelling at a tertiary referral eye care centre.

Methods:
Retrospective cohort study of 8 eyes of 5 patients from 2004-2012.

Results:
All 5 patients were male, with mean age of 42 years (range 35-50). 3 patients had bilateral disc swelling. There were no other retinal or choroidal lesions. Neuroimaging was performed in all patients to rule out space occupying lesions. Laboratory diagnosis was supported with two serological tests – a positive venereal disease laboratory test (VDRL) or rapid plasma reagin (RPR) as well as either treponema pallidum particle agglutination (TP-PA) or syphilis immunoglobulin G (IgG). Three of them had positive retrovirus. Lumbar puncture was performed in all patients, with cerebrospinal fluid (CSF) VDRL positive in 3 (60%) patients. All patients received 14 days of intravenous benzylpenicillin with 4 (80%) patients receiving concurrent systemic steroids Average time from presentation till initiation of penicillin treatment was 5.2±3.7 days. Mean logmar visual acuity at presentation was 0.68 with improvement to mean logmar VA of 0.04 at 1 year post presentation.

Conclusions:
Ocular syphilis can present with papillitis that may be unilateral or bilateral. Although life threatening diseases should be ruled out first, one must not forget infection as an important differential – particularly in patients with an atypical presentation. In this series of patients with syphilitic optic neuritis, appropriate prompt treatment together with judicious use of systemic steroids tends to produce good visual outcomes.

References: None.

Keywords: Optic Neuropathy

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Poster 20

Retinal Hyper-excitability and Ischemic Optic Neuropathy Following Oxaliplatin and Capecitabine Chemotherapy for Carcinoma of the Appendix

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Introduction:
Oxaliplatin and capecitabine together are a primary therapy for advanced colorectal cancer. The combination is relatively new but increasingly common without fully delineated side effect profiles, though there have been case reports of ocular toxicities including optic disc edema, transient vision blurring and blepharoptosis.

Methods:
Case report and literature review of a 64 year old man undergoing adjuvant oxaliplatin and capecitabine therapy for locally advanced appendiceal carcinoma who experienced episodic peripheral whitening of his vision ("white tunnel vision") lasting 5-30 seconds triggered by light following an oxaliplatin infusion.

Results:
The white tunnel vision episodes decreased in frequency over time following his oxaliplatin infusion. The patient demonstrated anatomically crowded optic discs with disc edema bilaterally, but no other observable fundus pathology. Fluorescein angiography and temporal artery biopsies were not consistent with temporal arteritis, and MRI of the brain and orbits was unremarkable. His symptoms and optic disc edema entirely dissipated after eight weeks without further chemotherapy. Given the timing and negative work-up his optic disc edema and his white tunnel vision are thought to be chemotherapy toxicities.

Conclusions:
Oxaliplatin's dose-limiting neurotoxicity is characterized by hyper-excitability of peripheral pain receptors. This patient's episodic white tunnel vision may reflect a similar retinal hyper-excitability toxicity preferentially involving rods versus cones or magnocellular versus parvocellular ganglia. The optic disc edema could be from axonal edema in an already small anatomic disc secondary to oxaliplatin (axonal swelling seen in lab studies), or due to short posterior ciliary artery vasospasm secondary to capecitabine which exhibits vasospastic side effects like 5-fluorouracil. The optic disc pathology could conceivably have caused breakdown in the retinal-blood barrier allowing oxaliplatin to induce the retinal hyper-excitability toxicity. This is the first report of this phenomenon, and it is important to alert oncologists to this possible side effect given the regimen's expanding usage.

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Keywords: Chemotherapy and Radiation Injury, Optic Neuropathy, Retina

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Poster 21
Phenotypic Spectrum of Optic Neuropathy Associated with Primary Sjögren's Syndrome

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Introduction:
Optic neuropathy associated with primary Sjögren's syndrome (SS) is seldom reported and little is known of its variable spectrum of clinical presentations.

Methods:
We described 3 different cases of optic neuropathy associated with primary SS.

Results:
Case 1: A 50-year-old woman was referred for incidentally found bilateral optic atrophy. She had dry eye symptoms for a month without subjective visual decline. Visual acuities (V/A) were 20/30 in both eyes. Color vision and visual field defects were found. Brain MRI was normal. Schirmer's test was below 5 mm, anti-Ro/SSA and anti-La/SSB antibodies were positive, whereas anti-aquaporin-4 (AQP4) antibody was negative. Salivary gland biopsy showed lymphoplasmacytic infiltration. Chronic optic neuropathy associated with primary SS was diagnosed. Plasmapheresis with immunosuppressants improved V/A up to 20/20, as well as color vision and visual fields. Case 2: A 67-year-old woman showed acute monocular V/A decrease with optic disc edema. MRI showed optic nerve enhancement. Schirmer's test was 5-10 mm bilaterally without sicca symptoms. Anti-Ro/SSA was positive and salivary gland biopsy revealed primary SS. Due to steroid dependence after high dose treatment, immunosuppressants were added. Case 3: A 43-year-old woman was newly diagnosed with SS- associated encephalitis. One month later, acute visual loss to counting fingers occurred in the right eye with pain. Based on positive serum anti-Ro/SSA, antinuclear and AQP4 antibodies and enhancement of the optic nerve on MRI, neuromyelitis optica spectrum disorders (NMOSD) associated with SS was diagnosed. Plasmapheresis fully recovered visual functions, which was maintained with immunosuppressants.

Conclusions:
Optic neuropathy associated with primary SS can present as acute optic neuritis, insidious progression of chronic optic atrophy, or in the context of NMOSD. Optic neuropathy may present initially without sicca symptoms, which makes the diagnosis difficult. Specific antibodies are supportive for the diagnosis and treatment in atypical cases of optic neuropathy.

References: None.

Keywords: Optic Neuropathy, Sjögren's Syndrome

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Poster 22
Syphilitic Perineuritis with Preserved Visual Function

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Introduction:
Syphilis has reemerged since the past 20 years. Designated as great masquerader, it can have multiple clinical manifestations in the eye: anterior uveitis, chorioretinitis, exudative retinal detachments and papillitis, to name a few. We report a case of syphilitic optic perineuritis, rare presentation of the disease.

Methods:
Retrospective case report.

Results:
58 year-old man presented with four weeks history of intermittent blurry vision in the right eye. He also reported having a recent history of skin rash on the upper arms and trunk as well as recurrent tinnitus. His past medical history was significant for well-controlled type 2 diabetes and hyperthyroidism. Best corrected visual acuity was 20/20 in each eye. There was very subtle right relative afferent pupillary defect. Biomicroscopic exam was normal in each eye. Posterior segment exam revealed few vitreous cells and a florid optic nerve edema with multiple peripapillary flame hemorrhages in the right eye. Fundoscopy was normal on the left. Formal visual field testing (Humphrey 24-2 algorithm) was normal in each eye. Optical coherence tomography (OCT) of the peripapillary retinal nerve fiber layer (RNFL) demonstrated marked elevation of the peripapillary RNFL on the right. MRI of the brain and orbits was normal. CBC, ESR, CRP, ANA, ACE were normal, HIV testing was negative. The rapid plasma reagin (RPR) test though was strongly positive (titer 1:512) and Treponema pallidum particle agglutination (TP-PA) testing was positive as well.

Conclusions:
Optic perineuritis, an unusual presentation of syphilis, is a form of inflammation of optic nerve sheath with sparing of axons comprising the optic nerve itself and thus, preserved visual function. This case is unique in that optic nerve head edema was quite pronounced with multiple peripapillary hemorrhages yet the patient was completely asymptomatic with normal visual function. It underscores the importance of including syphilis serologies in the work up of patients with optic nerve head edema.

References: None.

Keywords: Syphilis, Perineuritis

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Poster 23
Diagnosis of Optic Nerve Sheath Meningioma by Optic Nerve Sheath Decompression

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Introduction:
Optic nerve sheath meningioma (ONSM) is typically diagnosed based on clinical suspicion and imaging characteristics, and is most often treated with radiation. Historically, biopsy, optic nerve sheath decompression, and debulking surgeries have been avoided for fear of optic nerve vascular disruption and tumor spread into the orbit. Recently, however, a limited number of case reports have reexamined the utility of optic nerve sheath decompression (ONSD) with biopsy for suspected ONSM.

Methods:
We present a patient with unilateral optic disc edema and initial questionable findings of enhancement on MRI orbit. Clinical, neuroradiologic, and histopathologic findings will be presented confirming the diagnosis of ONSM.

Results:
A 48-year-old man presented with unilateral optic disc edema (ODE) and an enlarged blind spot. Initial imaging demonstrated questionable right optic nerve sheath enhancement vs. motion artifact. Subsequently, a lumbar puncture demonstrated an elevated opening pressure of 25 cm H_2O and he was diagnosed with idiopathic intracranial hypertension (IIH). The unilateral ODE initially resolved with acetazolamide. Following weight reduction and acetazolamide discontinuation, the ODE recurred and his vision subsequently worsened to 20/30. Repeat imaging 11 months after initial presentation demonstrated clear right optic nerve sheath enhancement. Optic nerve sheath decompression (ONSD) with biopsy was performed, simultaneously to decompress the nerve and secure a pathologic diagnosis, which confirmed ONSM. He was treated with radiation, but the visual acuity and fields improved after ONSD alone.

Conclusions:
The utility of optic nerve sheath decompression with biopsy should be reconsidered in cases of suspected ONSM with a complicated differential diagnosis, both for restoration of vision through relief of axoplasmic stasis and for definitive pathologic diagnosis.

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Keywords: Optic Nerve Sheath Meningioma, Optic Nerve Sheath Decompression, Optic Disc Edema

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Poster 24
Isolated Optic Disc Metastasis as the Presnting Sign of Adenocarcinoma of the Lung

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Introduction:
The optic disc is an uncommon location for metastasis and rarely the presenting sign of an underlying malignancy. There were 13 previous cases reported in the literature of lung carcinoma metastatic to the optic disc, however no cases of documented adenocarcinioma of the lung were found with an extensive literature review.[1,2] Most cases were a result of small cell lung cancer and others lacked pathological details on the type of lung cancer.[1]

Methods:
Clinical examination and biopsy of lung lesion.

Results:
A 67-year old woman noticed decreased vision in the right eye (OD) for 3 weeks. She was treated for presumed giant cell arteritis after an optometrist noted disc edema and an elevated ESR of 80mm/h; however, subsequent temporal artery biopsy was negative and she was referred to neuro-ophthalmology. On exam, her visual acuity was 20/40 in each eye with a right relative afferent pupillary defect. Visual field testing revealed an enlarged blind spot and infero-nasal defect OD (MD -7.22dB). Fundoscopy revealed a well-circumscribed white mass with fine blood vessels on its surface originating from the optic nerve head. MRI of the brain demonstrated 4 small enhancing lesions in the parietal lobes and cerebellum. A quest for underlying primary neoplasm was initiated and a CT of the chest revealed a left hilar mass measuring 6cm. A subsequent biopsy of the lesion demonstrated adenocarcinoma of the lung. The patient underwent stereotactic radiotherapy for the brain mestastases and systemic therapy with premetrexed-cisplatin. Her vision was closely monitored and at her 6 month follow up she had a decline in her vision to 20/60 with a worsening of her visual field defect (MD -9.18dB). She underwent low-dose stereotactic radiotherapy to the right optic nerve.

Conclusions:
Familiarly with the clinical features of optic disc metastasis can lead to the search for an underlying malignancy and allow for the rapid initiation of treatment at an earlier stage.

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Keywords: Metastasis, Optic Disc, Lung Cancer, Neoplasm, Radiotherapy

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Grant Support: None.

Poster 25

Purkinje Cell Antibody (PCA)-2 Positive, Collapsin Response-Mediating Protein-Associated (CRMP)-5 Negative Paraneoplastic Optic Neuropathy

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Introduction:
Purkinje cell antibody (PCA)-2 is a marker of neurological autoimmunity related to cancer that has not been previously associated with visual pathway dysfunction.[1]

Methods:
Clinical examination and biopsy of lung lesion.

Results:
A 57-year old female presented to the emergency room with bilateral sequential vision loss. Examination by an ophthalmologist demonstrated bilateral optic disc edema and panuveitis. MRI of the brain revealed several small FLAIR and T2-hyperintensities within the subcortical, deep and periventricular white matter, and corpus callosum. MRI of the spine performed because of suspicion of demyelinating disease was normal but revealed a lung lesion that a subsequent biopsy confirmed as small cell lung cancer. Further workup did not demonstrate any evidence of metastatic disease. The patient underwent 4 rounds of chemotherapy with cisplatin and etoposide, 15 sessions of radiotherapy to the lung area, and 10 sessions of radiotherapy to the brain. She was then referred to neuro-ophthalmology where her examination revealed a visual acuity of 20/25 with superior optic nerve pallor and corresponding inferior altitudinal visual field defects in both eyes. A paraneoplastic panel revealed positive PCA-2 antibody titers (1:64,550) and negative CRMP-5 titers confirmed with western blotting. Extensive workup for other autoimmune, inflammatory and infectious causes of optic neuropathy were negative. The patient continued to do well and was stable 8 months after diagnosis.

Conclusions:
Previous patients positive for PCA-2 presented with limbic encephalitis, cerebellar ataxia, Lambert-Eaton myasthenic syndrome, motor or autonomic neuropathy associated with small cell lung cancer.[1] Although PCA-2 often coexists with CRMP-5, the latter antibody was negative in our patient after she underwent treatment.[2] Discovery of the PCA-2 antibody in patients with optic disc edema and vitritis may suggest an underlying malignancy, which would allow for early detection and initiation of treatment.

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Keywords: Paraneoplastic, Optic Neuropathy, Uveitis, Lung Cancer, Autoantibodies

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Poster 26

Features Neuroprotective Support for the Compressive Optic Neuropathy Traumatic Conditionality Bone Fractures Orbit

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Introduction:
When injury to the optic nerve canal according Rajinikanth MG, 2003, causing compression of blood vessels, which leads to the cessation of nerve impulses, causing blindness. This explains the usefulness of decompression surgery with a combination of conservative and surgical methods. It is believed that decompressive surgery is most advisable to use up to 7 days from date of injury and methylprednisolone injection should be started as soon as was first diagnosed tone. Also showing dekompreyvnyh to endoscopic operations is the lack of a positive effect from the use of methylprednisolone over 72 hours, during the progressive reduction of corticosteroids or confirmed by CT or MRI signs of compression of the optic nerve blindness

Methods:
We examined 6 patients with compressive lesions of the optic nerve. The most effective was the use of neuroprotective support before, during surgery and in the early postoperative period.

Results:
So the patient, despite the heavy multiple fracture of the lower and outer walls of the orbit with displacement, the crack in the apex of the orbit, low preoperative visual acuity (within 0.04 on the affected side and 0.6 on the opposite side), 3 months after injury visual acuity in both eyes is 1.0.

Conclusions:
Consequently, the use of neuroprotective support for compression of traumatic optic neuropathy caused by fractures of the orbit will help protect the optic nerve from the possibility of re-injury, and improving ways to use it will improve the effectiveness of the treatment as a whole.

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Keywords: Traumatic Impairment, Optic Nerve, Neuroprotective Therapy, Methylprednisolone, Compresion Optic Neuropathy

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Grant Support: None.

Poster 27
A Case of Bilateral Optic Neuritis Due to NMO in the Setting of HIV

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Introduction:
NMO is an autoimmune disorder that causes ON and LETM. HIV is one of the most common causes of acquired immunodeficiency but is not commonly associated with ON. This case highlights a rare HIV patient with acute bilateral optic neuritis due to NMO IgG antibodies.

Methods:
A case study report.

Results:
A 52 year old AA female with HIV presented with sudden acute bilateral vision loss. She first noted sudden vision loss in her left eye and two days later it involved the right eye. The vision loss was associated with left eye pain that was worse with movement. She was hospitalized after she was seen by an ophthalmologist and found to have swollen optic nerves OU. She was treated with three days of IV solumedrol 1000mg with no improvement. One week later, visual acuities were CF OD and HM OS. Pupils were 5mm with no APD. Dilated funduscopic exam still showed mildly swollen ON OU. The patient was readmitted and treated with plasmapharesis. An MRI brain showed diffuse T2 signal abnormality involving the bilateral optic nerves throughout their extension as well as the optic chiasm associated with abnormal enhancement. A spinal tap showed an opening pressure of 18 cm of water. The CSF cultures were negative. PCR for CMV, HSV and VZC were negative. Cryptococcal antigen was negative. Toxoplasmosis IgG antibody was negative. The CSF had 14 white cells (94% lymphocytes), 0 red cells, glucose 48 and protein 79. The IgG synthesis rate was elevated and there were oligoclonal bands present in the CSF that were absent in the serum. HIV RNA PCR was 124 and CD4 count was 736. Serum NMO IgG was 59.4 (normal <1.6). After treatment with plasma exchange and rituximab her vision improved to CF OD and OS.

Conclusions:
This is the fifth reported case of NMO occurring in the setting of HIV. Treatment in these cases is challenging due to the immunosuppressed status of the patients.

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Keywords: Neuromyelitis Optica, Optic Neuritis, HIV, Demyelinating Disease, Infectious Disease

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Poster 28
Acute Optic Neuritis Associated with Subdural Hematoma

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Introduction:
A 70-year old man was referred to neuro-ophthalmology for subacute right monocular vision loss. One week prior to evaluation, he developed a dull right peri-orbital headache, which was followed 3-days later by progressive monocular vision loss. A few days before the onset of his headache, a fifth wheel trailer hitch had struck the top of his head without loss of consciousness. He had also traveled the Western United States in the months before presentation, however, he denied any location-specific infectious symptoms. He denied any history of demyelinating disease. Review of systems was positive for stable idiopathic left hearing loss of several years duration.

Methods:
Case report.

Results:
Visual acuity was counts fingers right eye (OD) and 20/25 left eye (OS). Color vision was 0/10 OD and 9/10 OS with a right relative afferent pupillary defect. Extra-ocular movements were full without evidence of nystagmus. Fundus examination of both eyes revealed flat, orange discs with sharp margins and C/D ratio of 0.3. The right eye demonstrated arteriole attenuation with superior arcade venous sheathing without leakage on fluorescein angiogram. Neurologic exam confirmed left hearing loss and wide based gait without cerebellar dysmetria. Contrasted MRI of the brain and orbits was significant for right optic nerve enhancement and subacute bilateral subdural hematomas up to 1.1-centimeters in thickness on the right. Cerebrospinal fluid studies showed a leukocytosis of 5/mm^3 (53% lymphocytes) and elevated protein at 92 mg/dL. Serum evaluation for infectious and inflammatory markers was negative. Three weeks after receiving intravenous steroids, visual acuity OD improved to 20/30.

Conclusions:
Subdural hematoma associated optic *neuropathy* has been previously described, but to the authors' knowledge, this is the first reported case of a subdural hematoma associated optic *neuritis*. The exact pathophysiologic mechanism remains elusive, but we postulate that subdural hematomas may trigger a reactive, immune-mediated T-cell response, which may rarely result in optic neuritis.

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Tao Z, Ding S, Hu M, Li J. "Delayed Hypersensitive Process in Subacute Subdural Hematoma." J Craniofac Surg. 26(5):e443-5. 2015.

Keywords: Optic Neuritis, Subdural Hematoma

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Poster 29
NAION, BRAO, Tumor or All!

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Introduction:
Capillary hemangiomas of the optic disc are vascular hamartomas that may rarely arise from the optic nerve.

Methods:
Case report and review of literature.

Results:
A 61-year-old male presented in August of 2014 for evaluation of persistent right optic nerve swelling. He reported noticing distortion in the superior field of his right eye in October of 2012. He was diagnosed locally with an epiretinal membrane (ERM), and subsequently underwent vitrectomy with ERM membrane peel in May 2013. Post-operative vision did not improve, and exam revealed optic nerve head edema. Humphrey visual field testing revealed a superior altitudinal defect in the right eye, and a normal left field. His local ophthalmologist administered multiple courses of oral prednisone over the course of a year, without improvement in his optic nerve edema and unchanged visual field defect. He underwent multiple neuroimaging studies that were normal. He eventually referred to Neuro-Ophthalmology consultation. Examination revealed 20/25 acuity in the right eye compared to 20/20 in the left. Color perception was normal in each eye, with subtle early release of the right pupil. Dilated ophthalmoscopy revealed a vascular growth of the right optic nerve nasally, with associated inferior pallor and gliosis. Optical Coherence Tomography (OCT) of the right eye exhibited relative inferior altitudinal macular thinning and mild inferior temporal RNFL loss. Fundus fluorescein angiography revealed a vascular optic nerve mass consistent with a capillary hemangioma, with delayed filling of the inferior temporal arterial and venous vasculature.

Conclusions:
Capillary hemangioma of the optic nerve is a rare enitity. The hemangioma may cause vision loss by axonal compression, or compromise of the optic nerve and/or retinal vasculature. This case illustrates visual field loss secondary to a combination of these 3 mechanisms. Optic nerve hemangioma should not be confused with other optic nerve pathology such as NAION to avoid unnecessary intervention.

References: None.

Keywords: Hemangioma, NAION, BRAO

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Grant Support: None.

Poster 30
Documented Progression of "Impending NAION" to Permanent Visual Loss

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Introduction:
The primary risk factor for NAION appears to be a small, crowded optic disc. Asymptomatic disc swelling in patients with this disc appearance has been observed and called “impending NAION” but progression to full-blown disease has not been described. We present 3 cases of documented asymptomatic disc swelling with progression to NAION and permanent vision loss.

Methods:
Retrospective clinical case series

Results:
Case 1: A 69 year-old man with presented with sudden OS vision loss. Visual acuity was 20/20 OD and 20/40 OS with an inferior altitudinal scotoma OS, and normal perimetry OD. Bilateral optic disc swelling with peripapillary hemorrhage was noted. Right optic disc swelling improved slightly 5 weeks later with normal right eye vision, but 6 days later new visual field loss OD and recurrent optic disc swelling OD occurred. Case 2: 7 years after NAION OD, a 52 year-old man presented with transient visual obscuration OS. Visual acuity and perimetry were normal OS with left optic disc swelling and peripapillary hemorrhage. Several days later, he presented with new visual field loss and reduced visual acuity OS and worse optic disc swelling. Within 8 weeks he had permanent visual field defects and optic disc pallor OU. Case 3: A 69 year-old woman had temporal right optic disc swelling with normal visual acuity and visual fields. Fellow eye had “disc at risk.” She had sudden onset of vision loss 1 month later, and subsequent exam showed pallor at the site of prior swelling. The left eye now has similar swelling with normal visual function.

Conclusions:
The pathogenesis of non-arteritic anterior ischemic optic neuropathy (NAION) remains uncertain; our case series demonstrates how optic disc swelling without visual acuity or field loss may precede NAION onset by days or weeks. Patients with this presentation may benefit from early intervention once therapeutic agents are available.

References: None.

Keywords: Optic Neuropathy, Visual Fields, Vascular Disorders

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 31
Leber's Hereditary Optic Neuropathy?

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Introduction:
Leber's hereditary optic neuropathy (LHON) is a mitochondrial disease primarily affecting the optic nerve. Pattern electroretinogram characteristics of LHON include prominent reduction of N95 with a normal P50 component (1). Optical coherence tomography (OCT) typically shows thickening of the retinal nerve fiber layer (RNFL) (2). Here we present a case of LHON with unusual OCT and ERG findings.

Methods:
Review of medical chart.

Results:
An 11 year old girl with normal developmental history presented with decreased vision in the left eye (OS) for 3 months that began after a fainting episode. She denied head trauma, pain, or progression of visual loss. Visual acuity was 20/20 in the right eye (OD) and 4/200 OS. A 3+ afferent pupillary defect and loss of gross colors to 2/4 were seen OS. Intraocular pressures, extraocular motility, and slit lamp exam were normal. Fundus exam showed a 0.6 cup-to-disc ratio with 1+ optic atrophy OS, and a 0.4 cup-to-disc ratio with a pink appearing rim OD. MRI of the orbits, ACE, ANA, NMO-IgG, Lyme, and syphilis testing were negative. LHON teseting revealed a 14484 mutation. She was started on Idebenone 900 mg daily. Follow-up exam 3 years later showed progression of visual loss to counting fingers and temporal pallor of the optic disc OS. OCT showed decreased macular volume wihtout optic atrophy. Inner RNFL thinning was seen and the outer RNFL was intact. Electroretinography showed normal rod function bilaterally but did reveal photoreceptor loss and inner retinal dysfunction, indicative of cone system dysfunction. Pattern electroretinography showed severe reduction of P50 and N95.

Conclusions:
The mixed genetic, OCT, and ERG findings make it unclear whether a retinal or optic nerve pathology is responsible for vision loss in this patient. Among patients with LHON who do not develop RNFL thinning, one should consider macular OCT and ERG testing to evaluate alternative diagnoses.

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Keywords: Leber's Hereditary Optic Neuropathy, LHON, Retinal Disease

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Grant Support: None.

Poster 33
Chiasmal Neuritis in Seropositive Neuromyelitis Optica Spectrum Disease

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Introduction:
Inflammation of the visual pathways affects most commonly the optic nerves, in association with multiple sclerosis, infections and other autoimmune disorders. The aim of this study was to determine the frequency of chiasmal involvement in patients with neuromyelitis optica spectrum disorder (NMOSD).

Methods:
Retrospective study, including patients with chiasmal involvement on clinical and neuroimaging criteria, in a seropositive NMOSD population, seen at our institution between January 2012 and April 2015.

Results:
Among 83 patients with optic neuritis who underwent aquaporin 4 (AQP4) antibody testing, 18 patients were seropositive. Of these, 3 patients (16.7%) showed radiologic and clinical signs of optic chiasm involvement. All 3 patients presented with severe bilateral visual loss of counting fingers or worse visual acuity. Contrast-enhanced magnetic resonance imaging showed enhancement of the optic chiasm in all three patients and contiguous involvement of both optic nerves in two of the patients. In none of the remaining 65 patients who tested negative for AQP4 antibody was optic chiasmal involvement noted, either clinically or on neuroimaging.

Conclusions:
Chiasmal involvement is not uncommon in patients with afferent pathway inflammation associated with seropositive NMOSD. An acute inflammatory chiasmal syndrome may be the presenting sign of NMOSD, prompting specific management.

References: None.

Keywords: Neuro-Ophth and Systemic Disease, Optic Chiasmitis, Neuro-Imaging, Visual Fields, Demyelinating Disease

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Poster 34

CSF-JC Virus PCR-Negative Progressive Multifocal Leukoencephalopathy presenting with Visual Loss

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Introduction:

Progressive Multifocal Leukoencephalopathy (PML) is an infectious demyelinating disease of the brain caused by JC virus.[1] Typically, the diagnosis of PML is established when JC virus DNA is detected in the cerebrospinal fluid (CSF) of a susceptible individual with characteristic clinical and radiological findings. However, CSF-JC virus PCR testing[1,2] can result in false negatives.

Methods:

A 63-year-old man presented with two months of painless visual loss in both eyes. Just over one year prior to presentation, he was diagnosed with Chronic Lymphocytic Leukemia/Small Lymphocytic Lymphoma (CLL/SLL), and underwent six cycles of chemotherapy with bendamustine and rituximab, ending seven months prior to presentation. His visual acuity was finger counting at 2 feet in the right eye and hand motion in the left eye. His pupils were briskly reactive with no relative afferent pupillary defect. There was very mild optic disc pallor bilaterally. The remainder of his exam was normal.

Results:

A contrasted brain MRI revealed nearly symmetric, bilateral parieto-occipital white matter T2 hyperintensities without mass effect and with mild, mostly peripheral enhancement. There were several small scattered areas of T2 hyperintensity and enhancement in the frontal white matter. CSF analysis showed 0 white blood cells, 69 mg/dL total protein, and 49 mg/dL glucose. By PCR, no DNA was detected in the spinal fluid for JC virus or other viruses. A brain biopsy revealed gliotic areas of brain parenchyma with scattered lymphocytes. SV-40 staining demonstrated nuclear positivity in scattered cells, confirming the presence of JC virus and the diagnosis of PML.

Conclusions:

CSF PCR for JC virus is often considered to be the principal diagnostic test for PML, however it can be negative in the disease[1,2]. Caution should be taken when excluding the diagnosis on the basis of this test.

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Keywords: Neuro-Ophth and Infectious Disease, Demyelinating Disease

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Poster 35
Left Homonymous Quadrantanopia Due to an Aggressive Culprit

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Introduction:
To report the case of an otherwise healthy 38 year-old male presenting with left homonymous quadrantanopia due to an unusual culprit.

Methods:
This is a case report of a patient presenting to the Ophthalmology emergency room with one month of headaches and subjective blurry vision. The patient denied nausea, vomiting, seizures, or gait imbalances. Vision was 20/25 OU without APD or gaze limitations. The patient was found to have a left homonymous quadrantanopia. Anterior segment examination was unremarkable while the posterior segment was notable for Grade 2 papilledema.

Results:
Imaging demonstrated a large, avidly enhancing, dural based, 5.3 cm x 7.1 cm x 7.0 cm right temporo-parieto-occipital lobe mass, with 1 cm of right-to-left midline shift and transtentorial herniation with mass effect on the brainstem. The patient underwent preoperative embolization of the lesion. He then underwent a craniotomy for presumed meningioma. Pathology, however, showed a WHO grade II hemangiopericytoma. Metastatic workup was significant for a heterogeneously enhancing 1.4 cm liver mass and a 0.9 cm aortocaval lymph node. There were no postoperative complications.

Conclusions:
We report a 38 year old man who presented with left quadrantanopia due to hemangiopericytoma, a relatively rare intracranial soft-tissue sarcoma clinically and radiographically indistinguishable from meningioma. This diagnostic determination can only be made through immunohistochemical study. It is important to distinguish these two entities as hemangiopericytomas tend to recur aggressively and have a high metastatic potential. Hemangioperictomas are best treated with total resection and adjuvant radiotherapy. These tumors require diligent follow-up by neurosurgery and ophthalmology because of their recurrence potential.

References: None.

Keywords: Tumors, Visual Fields, Neuroimaging, Neuropathology

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Poster 36
Pourfour du Petit Precipitated by Neosynephrine Drops

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Introduction:
Reverse Horners (RHs) or Pourfour du Petit syndrome (PdPs) is a rare.The most common clinical presentations include mydriasis, eyelid retraction, exophthalmos and hyperhidrosis. The pathophysiology of this disorder is not well explained;it is suggested to be secondary to cervical sympathetic chain hyperexcitability, or oculosympathetic spasm.

Methods:
This is a case report and review articles

Results:
A 38 year-old female with no PMH was presented with a 10-year history of intermittent right lid retraction. These events occur once a month with a recent increase in frequency. During this event she gets a tightening sensation of periorbital muscles and significant lid retraction. She is not aware of any pupil enlargement, excessive sweating. Neuro-Ophthalmologic examination was positive for RUL crease was higher OD 11 mm and OS 8 mm. MRD 1 OD 2.0, OS 1.75 mm, MRD 2 OD 8 OS 7. 10 minutes after 2.5 % neosynephrine MRD 1 OD 7 OS 2.5. Pupils were in light OD 2.5mm OS 3.0mm and in dark OD 4.5 mm OS 4.75- 05.0mm. There was no dilation lag and no APD. 30 minutes after 2.5 % neosynephrine pupils OD 5.0 mm and OS 4.0 mm. CTA of neck and brain and MRI of brain and cervical spine to T4 were normal.We reviewed 18 cases with RHs. RHs have been reported following penetrating and non- penetration trauma and in conjunction with mass lesions such as tumors or vascular abnormalities.

Conclusions:
RHs can be precipitated by neosynephrine which might be due to longstanding preganglionic 2nd order neuron dysfunction with transynaptic degeneration of the 3rd order neuron, leading to denervation supersensitivity or due to a 3rd neuron dysfunction resulting in denervation supersensitivity resulting in a severe lid retraction on the affected side. However there is dysfunction of the sympathetic on the right side the baseline function of the sympathetic nervous system remain intact.

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Keywords: Eyelid & Adnexal Disease, Pupils, Neuroimaging, Orbit/Ocular Pathology, Vascular Disorders

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 37
Seizing to See

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Introduction:
Epilepsy is a spectrum disease with wide variety of neurologic manifestations. We report a case of multifocal epilepsy whose initial manifestation was homonymous hemianopia.

Methods:
Case Report

Results:
We present a 12-year-old boy who initially presented with transient left homonymous hemianopia. His symptoms progressed to complex partial seizures. His MRI brain was normal. After failing numerous anti-epileptic drugs and VNS placement, he had a pre surgical evaluation which showed left temporal lobe seizures. He eventually underwent inferior temporal corticectomy. Complex partial seizures continued even after corticetomy. At 26, he had an evaluation for revision surgery, and was found to have a persistent homonymous visual field defect with seizures. At the epilepsy monitoring unit, he had a focal seizure with bilateral arm posturing and postictal left homonymous defect that lasted 20 minutes. This was confirmed by confrontational testing but not by Humphrey visual field (HVF). Unfortunately, the transient nature of his visual field defect made HVF testing difficult. On EEG, seizures were seen in the right frontal hemisphere, which spread to the right posterior quadrant specifically the occipital lobe followed by attenuation. This precluded him from surgery due to the multifocal sources of his seizures.

Conclusions:
This is a unique case of epilepsy where the seizures are characterized by homonymous hemianopia in addition to complex partial seizures. The semiologic and electrographic features localized the seizures spread to the right posterior quadrant. This case supports the localization value of visual changes in correlation with ictal patterns.

References: None.

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 38

Homonymous Ganglion Cell Layer Thinning following Temporal Lobectomy Demonstrating Retrograde Transsynaptic Retinal Degeneration in an Adult Patient with Refractory Seizures

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Introduction:
Optical coherence tomography (OCT) is a non-invasive imaging technique which provides transpupillarily cross-sectional scans of the retina and the optic disc. With the improvement of the resolution of optical coherence tomography, the ganglion cell complex (GCC) has also become routinely accessible in the clinic. Retrograde trans-synaptic degeneration of retinal GCC may result from post-synaptic lesions of the visual pathway.

Methods:
Case report, and review of literature.

Results:
A 53-year-old female reports right superior homonomous field defect sustained during traumatic brain injury with resultant epidural hematoma and left temporal lobectomy 32 years prior to presentation. Her examination revealed visual acuity of 20/20 in each eye with normal color vision and pupillary reactions. Dilated fundoscopy revealed healthy appearing optic disks, without frank pallor. Humphrey visual field testing revealed right superior homonomous visual field loss.

Conclusions:
In our case, the damage to the post-synaptic temporal lobe visual axons resulted in a left inferior homonomous GCC thinning noticed on OCT through trans-synaptic retrograde retinal degeneration. The pattern of GCC loss matches with the right superior homonomous visual field loss. This finding supports the use of GCC thickness as an imaging marker of trans-synaptic degeneration in the visual pathway secondary to brain lesions that can help localize the lesion.

References: None.

Keywords: Ganglion, Retina, Seizures

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Grant Support: None.

Poster 39

Bilateral Vision Loss and Optic Tract Edema on MRI after Bilateral Internal Carotid Artery Coil Embolization

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Introduction:
Delayed visual loss after paraophthalmic internal carotid artery (ICA) aneurysm coil embolization has been documented. The putative cause is coil-induced perianeurysmal inflammation affecting the visual pathway[1,2,3,4]. We present the first case of subacute binocular visual loss associated with striking bilateral optic tract T2/FLAIR hyperintensity.

Methods:
Case Report

Results:
A 41 year-old woman presented with progressive bilateral vision loss starting 2 weeks after right ICA aneurysm coil embolization. Fourteen months earlier, she had undergone left ICA aneurysm coil embolization and placement of a flow-diverting stent without any damage to vision. Previous brain MRI had disclosed no abnormality of the visual pathway. But after right aneurysm coiling, visual acuity dropped to 20/100 OD and 20/80 OS with Humphrey perimetry suggesting bilateral optic tract damage. Brain MRI disclosed coiled suprasellar aneurysms displacing the distal optic chiasm and proximal optic tracts and marked T2/FLAIR hyperintensity of both optic tracts. After high-dose corticosteroid treatment, visual acuity improved to 20/30 OD and 20/25 OS.

Conclusions:
This case adds to the small reported series of visual pathway damage after coil embolization. Our case is unusual in showing T2 signal hyperintensity of both optic tracts. Other reports have described high T2 signal in the optic tract[1], optic nerves[2], and chiasm[2]. Perianeurysmal inflammation as the cause is suggested by visual improvement after corticosteroid treatment[1,2,3]. Coated bioactive coils have been implicated, but a similar phenomenon occurs after coiling with bare platinum coils. Vision loss may also be caused by mass effect from enlargement of the aneurysm or coil compaction causing a water hammer effect. Recognition that vision loss is a potential risk of coil embolization is important. Prompt corticosteroid treatment may be critical in reversing it. Alternative approaches include repeat coiling, surgical coil mass extraction, and carotid artery occlusion.

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Keywords: Interventional Neuroradiology, Neuroimaging

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 40
A Rare Case of Contrast-Related Transient Cortical Blindness

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Introduction:
Neurotoxicity from contrast agents is a rare complication of angiographic procedures. Reported clinical effects include transient cortical blindness, encephalopathy, seizures, and focal neurologic deficits. Imaging may show enhancement in the occipital lobes of patients with cortical blindness due to contrast deposition in the tissue. Most cases return to baseline visual function within a few days.

Methods:
Case report and review of the literature.

Results:
A 50 year old woman presented to the emergency department with worsening headaches, where imaging revealed a large basilar artery tip aneurysm. She underwent dual crossing-Y stent construct and embolic coiling with neurosurgery. The morning after the procedure she reported bilateral vision loss. Initial ophthalmologic exam found generally depressed visual fields and light perception vision. CT brain, MRI brain, and repeat angiogram showed scattered embolic changes throughout both cerebral hemispheres, but did not identify a lesion that explained the severe visual deficits. Vision rapidly improved over the next four days with intravenous fluids and dexamethasone, but visual fields continued to show checkerboard defect. At four-week follow-up, visual acuity was 20/25 OU and visual fields were full.

Conclusions:
We present a case of transient cortical blindness following endovascular basilar aneurysm repair. Posterior circulation intervention has the highest risk of this rare complication. This case was unique due to a lack of imaging changes that correlated with the vision loss, either due to ischemia or the reversible encephalopathy syndrome related to contrast deposition. Hyperosmotic contrast agents may cause fluid shifts that disrupt the blood-brain barrier and allow contrast deposition in the brain tissue, leading to cerebral edema. Our patient's procedure was performed with an isosmolar agent (iodixanol) diluted with saline. Other etiologies leading to a disrupted blood-brain barrier must be considered, such as a reperfusion syndrome due to redirected blood flow from the large aneurysm.

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Keywords: Interventional Neuroradiology, Higher Visual Cortical Functions, Vascular Disorders, Neuroimaging

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Poster 41
Cilioretinal Artery Occlusion: A Rare Complication of Severe Papilledema

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Introduction:
Central vision loss in the setting of papilledema can result from optic neuropathy, macular pathology (e.g., subretinal fluid, choroidal folds), or a combination of optic neuropathy and macular pathology.[1] The central vision loss is largely reversible when due to macular pathology, whereas it is not reversible when due to optic neuropathy.[1] We report 3 patients who developed irreversible central vision loss due to cilioretinal artery occlusion associated with severe papilledema in the setting of idiopathic intracranial hypertension (IIH).

Methods:
A retrospective review of our database of patients with IIH identified 3 patients with central vision loss in one eye due to cilioretinal artery occlusion. All patients were assessed with kinetic or static perimetry, fundus photography, optical coherence tomography (OCT) of the optic disc and macula, and fluorescein angiography. One patient was also studied with laser speckle blood flowgraphy imaging.

Results:
All three patients had an arcuate visual field defect corresponding to the distribution of the cilioretinal artery in the affected eye. In all cases, the cilioretinal artery occlusion occurred in the setting of severe papilledema (modified Frisen grade III or higher). In all cases, the visual field defect was permanent.

Conclusions:
Cilioretinal artery occlusion is rarely reported as a complication of papilledema. [2,3] Approximately 20% of the population has a cilioretinal artery, which arises from the short posterior ciliary arteries rather than the central retinal artery.[3] We propose that cilioretinal artery occlusion might result from compression of the cilioretinal artery by the edematous retinal nerve fiber layer or decreased perfusion of blood from the cilioretinal artery due to increased venous pressure[2,3,] similar to the mechanism in venous stasis retinopathy[5].

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Keywords: High Intracranial Pressure, Pseudotumor Cerebri, Vascular Disorders

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Poster 42
To Be or Not To Be, IIH is the Question

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Introduction:
Idiopathic intracranial hypertension (IIH) is mainly characterized by headaches and elevated intracranial pressure with normal cerebrospinal fluid (CSF) content. Moreover, brain and orbit magnetic resonance imaging studies image are invariably unremarkable. Typically, patients with IIH present clinical signs and symptoms that differ from conditions that mimic disc edema or other secondary cause of nerve swelling. However, in patients lacking definite clinical criteria this may be a difficult task; particularly cases were absence of true optic nerve edema is a differential diagnosis. Optic nerve head drusen (ONHD) can pose a diagnostic challenge in cases of suspected disc edema without symptoms. Nonetheless, despite similarities in optic nerve head appearance, etiology and visual outcome of these two conditions is quite different.

Methods:
A 37 year old obese female with hypertension and hypothyroidism presents for a routine eye examination. Although mainly asymptomatic, she does report occasional frontal headaches. She has a pre-existing diagnosis of pseudopapllidema secondary to ONHD. Examination is unremarkable except for tilted discs along with superior and inferior blur disc margins, but without evidence of ONHD. Bilateral optic disc elevation is observed with optical coherence tomography (OCT), but presence of ONHD is not clearly delineated. Given patient's profile and clinical findings the diagnosis of IIH cannot be disregarded and further diagnostic work up is warranted.

Results:
No abnormalities on brain MRI, with and without contrast. Bilateral scleral flattening is noted in axial T2 weighted images (WI) of orbits. MRV is normal. Opening pressure of 35 cm H2O with normal CSF composition is obtained. Diagnosis of IIH is recognized and oral acetazolamide treatment is started.

Conclusions:
The case accentuates the importance of a detailed history and thorough examination before consenting to the diagnosis of pseudopapilledema, which led to the suspicion of IIH.

References: None.

Keywords: Neuroimaging, Pseudotumor Cerebri, OCT, High Intracranial Pressure, Headache

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Poster 43
Pseudo-Susac Syndrome Due to Elevated Intracranial Pressure

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Introduction:
We present a complicated case of autonomic instability, quadriparesis and encephalopathy followed by vision loss and hearing loss to illustrate the potential morbidity of secondary elevations of intracranial pressure (ICP).

Methods:
Case report and review of the literature

Results:
A previously healthy 44-year-old male acutely developed tachycardia, hypertension, headaches, confusion, hallucinations, flaccid quadriplegia, bowel and bladder incontinence. Within one month he had acute bilateral visual loss, followed by acute bilateral hearing loss. Outside work-up included unrevealing brain MRI, C-spine MRI, bland CSF and extensive normal blood work. EMG showed axonal degeneration. He was treated with plasma exchange for presumed auto-immune etiology with moderate improvement of both mental status and quadriplegia, but neither vision nor hearing. Upon presentation to our institution four months later visual acuity was NLP in both eyes with exam findings of bilateral central retinal vein occlusions(CRVO) and chronic atrophic papilledema. MRI showed non-enhancing T2 signal in both optic nerves, increased optic nerve sheath width, and bilateral meningooceles in the tegmen tympani extending into the petrous apices and inner ear cavities. MR venography showed narrowing of multiple dural venous sinuses. Lumbar puncture opening pressure was 42 cm H2O and CSF was bland. He was treated with intravenous immunoglobulin and methylprednisolone for presumed ongoing autoimmune contribution to his illness. He underwent ventriculoperitoneal shunt (VPS) placement with rapid mild improvement of hearing, but not vision. Digital subtraction angiogram showed complete reconstitution of flow in cerebral veins and normalization of their diameters. One week after VPS placement optic disc swelling had resolved and audiometry testing had improved.

Conclusions:
Elevated intracranial pressure presumed secondary to neuro-inflammatory disease accounts for sensorineural hearing loss, papilledema, CRVO and dural venous sinus narrowing in our patient. It is important for physicians to be aware that elevated ICP can cause both hearing and vision loss.

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Keywords: Idiopathic Intracranial Hypertension, Central Retinal Vein Occlusion, Hearing Loss, Papilledema, Meningocele

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Grant Support: None.

Poster 44
Acquired Tonsillar Ectopia in a Case of Pseudotumor Cerebri

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Introduction:
The development of tonsillar descent is a rare but reported complication after lumboperitoneal shunting in pseudotumor cerebri. Fatal tonsillar herniation following lumbar puncture(LP) has also been rarely reported in pseudotumor cerebri . We report a case of pseudotumor cerebri with a minor tonsillar descent who developed worsening of the descent after repeated lumbar punctures.

Methods:
A single case report and review of literature.

Results:
41 year old Caucasian woman with headache and papilledema was diagnosed with pseudotumor cerebri when she had unremarkable brain MRI (except minor tonsillar ectopia <4 mm), unremarkable MR venogram and an opening pressure of 29 cm H2O on LP with normal CSF contents. She had stable visual functions following medical management with acetazolamide- visual acuity 20/20 OU, grade 1 papilledema OU and scattered peripheral defects on perimetry with MD <3 db. However she had mild metabolic acidosis. Attempts to wean acetazolamide led to return of headaches. Repeat LPs performed as temporizing measures showed opening pressures above 30 cm of H2O but with normal CSF contents.
4 years later when she reported occipital headache worsening with cough and an unsteady gait repeat brain MRI showed worsened tonsillar descent 8 mm below the foramen magnum. Repeat MR venogram showed severe right mid transverse sinus focal stenosis and long segment of pronounced stenosis in the left transverse sinus. She underwent suboccipital craniectomy and C1 laminectomy which stabilized her symptoms.

Conclusions:
This report highlights an unusual and rare presentation of acquired tonsillar descent in pseudotumor cerebri. Metabolic acidosis has led to fatal tonsillar herniation following LP in pseudotumor cerebri. We believe that repeated LPs and metabolic acidosis with a contribution from intracranial hypertension might have led to the tonsillar descent. Fortunately this was recognized before it became fatal

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Keywords: Tonsillar Ectopia, Pseudotumor Cerebri, Lumbar Puncture, Papilledema

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Poster 45
A Case of Perineural Invasion from Squamous Cell Carcinoma

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Introduction:
Squamous cell carcinoma (SCC) is the second most common cutaneous malignancy, accounting for 5% to 10% of periocular cutaneous tumors (1).Literature on SCC with perineural invasion (PNI) is scarce (2). Incidence ranges from 2.6 to 43.4% (3,4).Typical features for SCC with PNI are older patients, with recurrent and/or multiple incompletely excised SCC.Two groups of patients with PNI have been described: one without neurological symptoms (incidental finding on histological examination) and a second group with clinical symptoms of damaged nerves, which has an increased risk of local recurrence and regional and distant metastases (2,5).MRI provides the most sensitive radiographic methodology to detect PNI (6).Management guidelines for PNI are evolving. Radiotherapy for the potential proximal and distal extent of involvement is the mainstay of treatment. Combined chemo-radiotherapy has shown improved outcome in bulky disease. Role of surgery is mostly diagnostic (biopsy) and palliative (7).

Methods:
Single Case Report

Results:
A 65 year-old man presented with 45 day long permanent diplopia. He brought a previous orbital MRI which showed a supraorbital tumor that involved orbital soft tissues.Examination revealed left hemicranial anesthesia, a supraciliar tumor, and an abduction restriction in the left eye. A new MRI was ordered. It showed a left supratroclear tumor, with perineural dissemination through the supraorbital nerve that reached Meckel´s cavum, and involvement of the cavernous sinus lateral wall. Left lateral muscle was atrophic. Biopsy revealed SCC infiltration. Chemotherapy and radiotherapy were provided.

Conclusions:
PNI from SCC should be considered in the presence of an orbital tumor in association with sensory deficit and diplopia. Even though MRI provides the most sensitive radiographic methodology to detect it, PNI may still be present without neuroimaging evidence. The importance of such diagnosis relies on poorer prognosis PNI implies.

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Keywords: Perineural, Invasion, Squamous, Cell, Carcinoma

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Poster 46
Juvenile Nasopharyngeal Angiofibroma with Intracranial Extension

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Introduction:
Juvenile nasopharyngeal angiofibroma (JNA), the most common benign tumor of the nasopharynx, accounts for 0.5% of all head and neck neoplasms.This vascular tumor is found in adolescent males. Its aggressive local growth may lead to paranasal sinus, orbital and intracranial extension, even without detectable clinical signs and symptoms(1). Ocular findings are uncommon (2). Exophthalmos (14%), decreased vision (5%) partial ophthalmoplegia (2%), and facial deformities were reported (1) .Diagnosis is based on clinical findings and neuroimaging (3). Pre-operative angiography should be routinely performed, and tumor feeder vessels embolized before tumor resection (4).Treatment of choice is surgical excision, combined with radiotherapy in large tumors. Cure ranges between 70 and 100%. (2,4). Follow-up is mandatory due to high relapse rate (3, 4).

Methods:
Single Case Report

Results:
A 20 year old man presented with a 6 year vision loss of the left eye after the second embolization prior to resection of JNA. Ocular evaluation revealed a normal right eye, and light perception with an afferent pupillary defect on left eye. On external examination prominence of the nose and left cheek, and monocular exophthalmos were observed. Fundus of left eye showed a pale disc. OCT of left eye showed a peripapillary retinal nerve fiber layer thickness significantly decreased. MRI showed injury centered in the left pterygopalatine fossa, and extended infiltrating the extraconal orbital space (proptosis of the eyeball), and ipsilateral masticatory parapharyngeal space, cavum, both nostrils and skull base. It also extended to the endocranium in the suprasellar cistern, with compression of the optic chiasm and affection of both carotid cavernous sinuses.

Conclusions:
JNA may produce significant ocular complications due to orbital or intracranial extension, including exophthalmos, optic atrophy and ocular motor palsy. Early diagnosis, interdisciplinary evaluation and treatment are needed in order to improve prognosis.

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Keywords: Juvenile, Nasopharyngeal, Angiofibroma, Intracranial, Extension

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Poster 47
Choroidal Folds Secondary to Cavernous Hemangioma

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Introduction:
Choroidal folds are associated with papilledema, hypotony, thyroid disease, and inflammation of the orbit. However, little is known about its association with orbital tumors and its prognosis after tumor resection. Our purpose is to describe a case of choroidal folds and its associated findings caused by an orbital cavernous hemangioma.

Methods:
We followed a patient diagnosed with an intraconal cavernous hemangioma of the left orbit and conducted diagnostic studies including multifocal ERG, OCT, visual fields, and fundus photography. The patient underwent surgical resection of the hemangioma, and the same studies were conducted post-operatively to compare the results.

Results:
Pre-operative:
Visual acuity was 20/25 OD, 20/250 and 20/80 with pinhole OS. OCT of the left eye showed thickening of the macula and choroidal folds, which appeared as light and dark parallel lines in the temporal retina and macula. Multifocal ERG showed decreased amplitudes in the left eye. Post-operative:
Visual acuity was 20/40 and 20/20 with pinhole OD, and 20/200 and 20/25 with pinhole OS. OCT of the left eye showed choroidal folds and improvement of macular thickening. Multifocal ERG showed a 46% improvement in the average amplitudes.

Conclusions:
In this patient, an orbital cavernous hemangioma led to the development of choroidal folds, which resulted in vision loss likely due to the effect of choroidal folds on the neurosensory retina. Additionally, the vision was almost certainly affected by hyperopic shift as a result of the intraconal location of the tumor. However, the difference in pre- and post-operative pinhole acuity demonstrates a difference in potential acuity which is not fully explained by resolution of a hyperopic shift. Resection of the tumor via anterior orbitotomy showed improvement in vision, but the choroidal folds remained and the vision was not restored completely at one month follow-up.

References: None.

Keywords: Diagnostic Tests, Mferg, Retina, Tumors, Orbit, Orbit/Ocular Pathology

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 48
High Definition Fiber Tractography of a Giant Macroadenoma

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Introduction:
High-definition fiber tracking (HDFT) is a novel method of neuroimaging post-processing that allows precise delineation of white matter fibers in the anterior and posterior visual pathway. This technique contributes to pre-operative planning for resection of large sellar lesions.

Methods:
Case report and review of literature.

Results:
A previously healthy 37 year old man presented with three months of progressive loss of peripheral vision and headaches which persisted despite wisdom teeth extraction. He presented for urgent Ophthalmology examination complaining of two days of horizontal binocular diplopia. Examination revealed acuity of 20/30- in each eye, with a prominent right afferent pupillary defect (APD). Motility exam revealed decreased abduction bilaterally. Dilated ophthalmoscopy revealed bilateral optic disk pallor. Formal visual fields revealed a complete bitemporal hemianopsia. MRI of the skull base identified a large, 6.4 cm sellar and suprasellar mass with enlarged sella turcica, invasion of the sphenoid sinus and complete encasement of both parasellar internal carotid arteries. HDFT was performed and imaging reviewed prior to endoscopic endonasal approach to anterior, posterior, and bilateral middle skull base and bilateral cavernous sinus tumor removal with reconstruction and vascularized nasal flap, using stereotactic image guidance. Biopsy revealed a sparsely granulated growth hormone producing pituitary macroadenoma, with characteristic keratin staining pattern. Visual fields 12 days post op revealed significant improvement in bitemporal field loss.

Conclusions:
High-definition fiber tracking (HDFT) delineates white matter fibers in the anterior and posterior visual pathway. The identification of disruption of optic nerve, tract, and chiasmal fibers by sellar tumors contributes to significant post-operative improvement in visual fields and visual function.

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Keywords: NeuroImaging, Skull Base, Tumors, Optic Neuropathy, Visual Fields

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 49
Double Vision Followed By Oscillopsia

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Introduction:
To report and review MRI and a case presented with double vision followed by oscillopsia

Methods:
This is a descriptive case report based on clinical records, patient observation and analysis of diagnostic studies.

Results:
A 35-year-old right handed man presented to Neuro-Ophthalmology clinic with double vision after the recent brain surgery. The patient had medical history of von Hippel-Lindau disease, cerebellar hemangioblastoma and surgical history of two resections of cerebellar hemangioblastoma and ventriculoperitoneal shunt placement. On examination at 3 months after the last surgery, the patient had good visual sensory function in both eyes and the finding showed limitation of infraduction (about 30% of normal) on adduction in the right eye and 4 prism diopter hypertropia which followed three steps test with excyclotorsion on Double Maddox Rod test, which were suggestive of fourth cranial nerve palsy in the right eye. On seven months after the surgery, MRI T2/FLAIR showed increased signal in the left pons which was suggestive of hypertrophic olivary nucleus in the left. On twelve months after the surgery, the patient complained of oscillopsia and examination showed vertical pendular nystagmus with palatal myoclonus.

Conclusions:
This is a case of fourth cranial nerve palsy followed by oculopalatal myoclonus. The patient developed oscillopsia and palatal myoclonus after the latent period of about 12 months. The signal change in MRI suggesting hypertrophic olivary nucleus was observed prior the clinical symptoms and signs. The development of oculopalatal myoclonus should not be mistaken for recurrence of tumor or stroke.

References: None.

Keywords: Neuroimaging, Nystagmus, Ocular Motility

Financial Disclosures: The author had no disclosures.

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Poster 50

Transcranial Orbitotomy After Superselective Coil Embolization Sacrifice of Intracranial Ophthalmic Artery Preserves Vision in Resection of Recurrent Large Orbital Apex Solitary Fibrous Tumor, Via Collateral Retrograde Orbital Circulation

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Introduction:
We report the case details of a 51-year-old male with a recurrent, progressive solitary fibrous tumor (SFT), engulfing proximal ophthalmic artery, and the pre-operative endovascular and surgical methods that promoted preservation of vision despite sacrifice of the ophthalmic artery.

Methods:
A retrospective and radiographic chart review

Results:
The SFT (1.7cm X 1.2 cm X 1.1 cm), originally resected via Kronlein orbitotomy, recurred 39 months after resection (9.9 mm X 6.8 mm X 7.9mm) growing over 26 months (2.9cm X 2.0cm X1.65 cm). Super-selective endovascular coil embolization occluded the intracranial ophthalmic artery immediately distal to its C6 intracranial origin, and resulted in retrograde ophthalmic artery filling through ECA and ICA collateral vessels, maintaining CRA filling. Post-angiographic neuro-ophthalmic examination revealed no change in ocular motility, vision and visual field. One week later, the tumor was excised via right frontal pterional craniotomy with lateral orbitotomy, extended transfacial muscle (temporalis) dissection, and transcranial orbital dissection. The approach provided excellent direct visualization of the tumor from orbital apex to globe, minimized the difficulty of operating in the previously scarred lateral operative bed, and allowed gross total resection. Despite intraoperative sacrifice of the tumor-encased proximal orbital segment of ophthalmic artery and meningo-lacrimal branch, CRA filling was maintained via other ECA and ICA collaterals despite sacrifice of the major lateral collaterals and the patient retained normal vision and visual field. Transient abduction deficit was readily explained by post-operative muscle edema, and a partial frontal branch facial nerve palsy was transient and continues to improve.

Conclusions:
Extended transcranial orbitotomy and super-selective angio-embolization are valuable techniques that may promote favorable outcomes in deep orbital surgery of lesions affecting the orbital apex, by relying on collateral circulation than may maintain retrograde ophthalmic artery and thereby CRA flow.

References: None.

Keywords: Solitary Fibrous Tumor, Transcranial Orbitotomy, Ophthalmic Artery, Retrograde Filling, Collateral Circulation

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Poster 51
An Unusual Orbital Foreign Body

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Introduction:
Needlefish are slender fish with pointy beaklike jaws. In this report we describe a patient in whom a needle fish lodged the tip of its jaw in his optic canal.

Methods:
Case report.

Results:
A 53 year-old male, scuba diving off the coast of the United Arab Emerits (UAE), surfaced and removed his mask. A needlefish jumped out of the water, piercing his left medial canthus and upper eyelid. He immediately lost his vision. Computed tomography obtained at a local emergency room demonstrated a bony foreign body within the orbit apex. He underwent emergent "cleaning out". No foreign material was noted to have been found. He was given a three week course of prophylactic antibiotics and presented to our institute one month following injury. At that time he reported no light perception OS. There was a scar extending from the medial canthus to the upper eyelid margin. Ocular motility was abnormal with reduction in supraduction (50%) and adduction (25%). Exophthalmometry measurements were symmetric at 21 OU. IOP was normal and symmetric. Slit lamp evaluation was unremarkable. Funduscopic evaluation was notable for optic atrophy OS, and otherwise normal (Figure 1). Magnetic resonance imaging (MRI) indistinctly demonstrated the fish jaw extending through the left optic canal into the prechiasmatic suprasellar cistern with the left optic nerve displaced inferomedially. No other abnormality was identified. Due to the location of the jaw with intracranial extension, and improbability of visual recovery no further intervention was recommended. The patient remained in stable condition when last contacted three months following injury.

Conclusions:
This case demonstrates the radiographic appearance (MRI and CT) of an unusual injury: a needle fish jaw lodged in the optic canal. This case is also important in that it demonstrates that such cases can be managed without removal of the foreign body.

References: None.

Keywords: Orbital Foreigh Body, Truama, Fish, Optic Nerve, Imaging

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Grant Support: None.

Poster 52
Multifocal ERG in MEWDS and BRAO

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Introduction:
Spectral domain optical coherence tomography (SD-OCT) has become the most widely available and easily accessible retinal imaging technique in ophthalmology. In the era of SD-OCT, multifocal electroretinogram (mERG) still has value especially in cases where the retina appears normal or cannot be well visualized.

Methods:
Case series.

Results:
Case 1. 31-year-old woman with acute, painless, visual loss in the right eye (OD) associated with photopsias and nyctalopia. She has no medical history and takes no medications. Her examination shows an acuity of 20/40 OD and 20/20 in the left eye (OS), normal color vision in both eyes (OU). The visual field shows a temporal superior scotoma OS. Pupils are normally reactive without an RAPD. Ocular motility is normal. The fundus appears normal, and a mERG shows a defect which corresponds to the visual field abnormality OS in keeping with multiple evanescent white dot syndrome (MEWDS). Case 2. 69-year-old woman experiences positive visual phenomena OS for 2 days with a resultant scotoma OS. Her medical history includes a uveal melanoma OD with an ocular prosthesis in that eye, AMD, renal failure and dyslipidemia. Her examination shows normal acuity and color vision OS. The visual field exhibits a nasal superior scotoma OS. The left pupil is normally reactive. Ocular motility is normal OS. The fundus shows dry AMD changes with numerous small and intermediate sized drusen intermixed with an area of retinal whitening inferiorly. The mERG shows a defect which corresponds to the visual field and the area of retinal whitening, in keeping with a branch retinal artery occlusion (BRAO).

Conclusions:
In addition to SD-OCT, mERG remains an important technique in detecting retinal pathology particularly in its ability to specifically localize and correlate the retinal defect with the visual field.

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Oh KT, Folk JC, Maturi RK, Moore P, Kardon RH. Multifocal electroretinography in multifocal choroiditis and the multiple evanescent white dot syndrome. Retina. 2001;21:581-9.

Keywords: mERG, SD-OCT, BRAO, MEWDS, Scotoma

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Poster 53

Myocarditis and Arteritic Anterior Ischemic Optic Neuropathy Secondary to Giant Cell Arteritis

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Introduction:
Giant cell arteritis (GCA) is a primary vasculitis of medium and large vessels with a broad range of clinical presentations. Myocarditis is a rare known manifestation of GCA. We present a case of a 67 year old man who presented with acute systolic heart failure of unknown etiology. Development of arteritic anterior ischemic optic neuropathy lead to the diagnosis of myocarditis secondary to GCA.

Methods:
Case Report

Results:
A 67 year old man with a history of asthma, migraine, post-polio syndrome, and hepatitis B complained of new dizziness and fatigue after sustaining a tick bite. After empiric treatment with doxycyline for tick borne illness he developed an intractable headache with intermittent vertical binocular diplopia. Outside CSF studies, brain MRI, tick borne illness laboratory investigation, and ophthalmological examination were reportedly normal. One month later he developed acute chest pain and was found to be in atrial fibrillation with rapid ventricular response with elevated troponins. His initial transthoracic echocardiogram was normal, but two days later repeat exam revealed marked systolic heart failure. CT chest revealed mediastinal and hilar adenopathy. He was transferred to our institution for evaluation for presumed myocarditis. A cardiac MRI did not show signs of myocarditis so alternative diagnoses were pursued. Bronchoscopy was non-diagnostic. He developed sudden painless loss of vision in his left eye with elevated platelets, ESR, and CRP and was started on systemic steroids with improvement of his vision. Temporal artery biopsy revealed findings consistent with GCA. Cardiac PET revealed diffuse FDG uptake consistent with myocarditis and a diagnosis of myocarditis secondary to GCA was made.

Conclusions:
Myocarditis is a rare manifestation of giant cell arteritis. In this case, the cause of acute systolic heart failure was determined based on neuro-ophthalmologic findings.

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Keywords: Giant Cell Arteritis, Arteritis Ischemic Optic Neuropathy, Myocarditis, Headache

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Poster 54
Creudzfeldt-Jakob Disease an Important Differential Diagnosis of Progressive Supranuclear Palsy

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Introduction:
Creutzfeldt–Jakob disease (CJD) is a prion disorder characterized by rapidly progressive dementia, myoclonus, ataxia, visual disorientation, and motor dysfunction. Criteria for diagnosis of CJD include CSF markers, 14-3-3 and tau, EEG and MRI.

Methods:
Single case report

Results:
A 71 year-old man presented with left veering ambulation and mild left leg weakness. Lumbar spine MRI and EMG were unremarkable. One month later he noticed intermittent horizontal diplopia, a 10-pound weight loss, worsening handwriting and worsening left sided weakness. There were no behavioral changes. Visual acuity was 20/25 OU, HVF 24-2 revealed superior arcuate defects consistent with symmetric thinning of the optic disc OU. He had normal anterior segments, pupillary reactions, IOP and fundi OU. He had convergence insufficiency, reduced vertical more than horizontal saccades. Strength was 5/5 throughout with 4/5 quadriceps on the left. Reflexes were decreased throughout. There was dysmetria. His gait was shuffling. He had mild memory impairment and no disinhibition. Myoclonic jerks were noted when he was asleep. Antibodies to gm1, gd1a and gd1b in the CSF were positive and paraneoplastic panel was negative. EEG showed bitemporal slowing. He was treated with 5 days of IV methylprednisolone followed by 60mg prednisone for concern of encephalitis. He developed worsening memory and was treated with IVIG for 5 days. The patient continued to decline and developed visual hallucinations. MRI revealed DWI positivity in the bilateral basal ganglia, thalamus and anterior gray matter. CSF 14-3-3 and tau were highly elevated. He was given a likely diagnosis of CJD. The family declined brain biopsy. He passed away three weeks.

Conclusions:
CJD has been misdiagnosed as Progressive Supranuclear Palsy. Diplopia is a common complaint in early CJD, most commonly due to abnormalities of vertical gaze. Given the rapid decline of these patients it is essential to diagnosis CJD as early as possible to provide the family and patient with the time required to absorb this diagnosis.

References: None.

Keywords: Creuzfeldt-Jakob, Progressive Supranuclear Palsy, Diplopia

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Poster 55

A Case of HSV-2 Presenting with Encephalomyelitis and Diffuse White Matter Lesions

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Introduction:
Disseminated HSV-2 presenting with a relapsing, protracted, encephalomyelitis has not been well described.

Methods:
Case report from a tertiary medical center.

Results:
A 39 year-old woman, 8 months post-partum, presented with 3 weeks of progressive leg numbness. Exam showed bilateral mild iliopsoas weakness and patchy leg numbness. She had also noted mild hearing loss. MRI revealed scattered periventricular white matter lesions and enhancing spinal cord lesions. She was treated with IV steroids. A week later she developed right-sided hearing loss and vertigo, a positive head impulse test, and right cochlear enhancement on repeat MRI. Days later, she developed a diffuse morbiliform and vesicular rash, and was treated with one week of IV acyclovir. CSF revealed a lymphocytic pleocytosis with elevated protein, PCR was negative for HSV-1 and VZV and positive for HSV-2 with elevated IgM (1.15 IV) and IGG titers (27.91 IV, reference range <0.8 IV). She then developed leg numbness, worsening vertigo and gait imbalance. Repeat MRI showed new atypical appearing subcortical, pontine and cerebellar T2 lesions. She was treated with 6 weeks of IV acyclovir, plasmapheresis and IVIG. She was found to have low natural killer (NK) cells (47 cells/uL, reference range 59-401) that later returned to normal. Clinically, she has become less ataxic, and CSF studies have demonstrated declining HSV-2 antibody titers.

Conclusions:
An unexplained relapsing course of atypical white matter brain and spinal cord lesions may rarely complicate HSV-2 encephalomyelitis, and is exceedingly rare in immunocompetent adults.[1] Steroids may suppress NK cell levels,[2] which are known for their innate defense against herpesvirus infections.[3] It is likely that the combination of transient steroid-induced immunosuppression as well as the post-partum state allowed the virus to flourish and widely disseminate.[4,5]

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Keywords: HSV-2, Encephalomyelitis, Head-Impulse Test, Immunodeficiency, Natural Killer

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Poster 56
Relapsing Polychondritis Causing Inflammatory Optic Neuropathy, a Case Report

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Introduction:
Relapsing Polychondritis (RP) is a severe, episodic, and progressive systemic inflammatory condition involving cartilaginous structures of the eyes, ears, nose, and laryngotracheobronchial tree.[1,2] It is potentially fatal disease, that often goes misdiagnosed and mismanaged, that can cause wide-spread systemic manifestations including irreversible vision loss.[1,2] Our goal is to elucidate this rare condition as it relates to the ophthalmic care of patients and provide a clinic vignette illustrating the clinical presentation, diagnosis and management of a patient with RP.

Methods:
A 62-year-old man with no significant ocular history presented with painful and itchy eyes as well as flashes and floaters in both eyes for 2 days. He had also noted red, swollen ears over the same time frame. He had recently suffered from a bout of pneumonia, which was treated with oral antibiotics and steroids. His visual acuity and color vision were normal, but automated perimetry revealed bilateral visual field constriction. He had trace conjunctival injection, 1+ anterior chamber inflammation, and bilateral optic disc edema.

Results:
Cranial MRI and MRV were unremarkable, including no stigmata of increased intracranial pressure. His serologic workup was unremarkable except for an elevated sedimentation rate, C-reactive protein, and positive rheumatoid factor. He was started on high dose oral steroids, which resulted in significantly improved vision, discomfort, and redness of the eyes and ears. He was then started on methotrexate and the prednisone was tapered accordingly.

Conclusions:
In review articles of patients with RP, the average elapsed time from patient presentation to correct diagnosis was 2.9 years, with one third of patients being evaluated by 5 or more physicians before receiving appropriate treatment.[1,3,4] Despite these challenges, prompt diagnosis is of the utmost importance as treatment with systemic steroids and immnomodulators can preserve vision and prevent potentially fatal damage to major organ systems.

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Keywords: Relapsing Polychondritis, Optic Disc Edema, Uveitis, Red Ears, Episcleritis/Scleritis

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Poster 57
Double Take

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Introduction:
A 13 year old Afghani boy was referred with an apparent isolated optic nerve sheath meningioma. The outside imaging report stated that "intracranial contents appear normal". A careful dilated fundus exam, however, uncovered an epiretinal membrane in the left eye and lesions resembling retinal astrocytic hamartomas in the right eye. These associated findings alerted us to a possible underlying systemic diagnosis of neurofibromatosis type 2 (NF2) and prompted a review of the original MRI images. Upon re-reading by a neuro-radiologist, bilateral trigeminal and vestibular schwannomas were identified, establishing a provisional diagnosis of NF2 by the Manchester Criteria (1).

Methods:
Subsequent investigations included an audiogram, imaging of the entire CNS, and genetic testing. The audiogram was normal. The MRI spine revealed a very large paraspinal mass (70 mm x 65 mm x 87 mm) with intradural extension into the spinal canal. An biopsy was done, and the paraspinal mass was pathologically confirmed as a WHO grade I schwannoma. Genetic testing identified a truncating mutation in exon 1 of the NF2 gene. The patient has since been enrolled in a multi-disciplinary surveillance program headed by neuro-oncology.

Results:
This case represents a 'near miss' for delayed diagnosis of NF2. The original MRI report was falsely reassuring, identifying only the orbital tumour, and missing multiple intracranial tumours pathognomonic for NF2. Furthermore, our patient lacked the most common ophthalmic association of NF2 (cataract), and exhibited findings that were either non-specific (epiretinal membrane), or very subtle (retinal astrocytic hamartomas). Finally, despite harbouring multiple intracranial tumours and a large paraspinal tumour, he was neurologically intact.

Conclusions:
Young patients diagnosed with primary optic nerve sheath meningioma have a high likelihood of comorbid NF2; knowledge of this association may minimize time to diagnosis.

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Keywords: Pediatric Neuro-Ophthalmology, Genetic Disease, Neuro-Ophth & Systemic Disease, Orbit

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Poster 58

A Brain on Fire! A Case of Neuromyelitis Optica Mimicking Progressive Multifocal Leukoencephalopathy

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Introduction:
Neuromyelitis optica is an aggressive form of demyelination that typically causes recurrent optic neuritis and transverse myelitis. Atypical presentations can be difficult to diagnose.

Methods:
Case report and review of literature.

Results:
A 66-year-old woman with history of longstanding rheumatoid arthritis (currently on methotrexate and infliximab) presented with acute imbalance, fever and left-sided weakness. She underwent head CT that showed a hyperdense lesion within right corona radiate (CR) and basal ganglia (BG) with possible involvement of R. thalamus. It was suspected that she had an acute stroke. Few hours later she developed an episode of involuntary, non rhythmic jerking of left upper and lower limbs that lasted a few minutes. Her mental status started to wax and wane the next morning. Serological testing including Lyme, SPEP, ANCA, RPR, and FTA ABS were negative. IGG level in CSF was remarkably elevated at 13.20 mg/dL with an IGG index of 0.70. Her paraneoplastic panel was positive for AChR Ganglionic Neuronal Abs. MRI revealed T2 signal abnormalities with mild mass effect at right temporal lobe, white matter changes of right frontal parietal lobe, with involvement of R. thalalmus and posterior limb of Internal capsule(IC), also signal abnormalities of right pons and midbrain. There was no diffusion restriction and minimal patchy enhancement was noticed. MRI-guided brain biopsy showed parenchymal inflammation without signs of infection. A large, T2 hyperintense, non-enhancing mass with some mild associated internal reduced diffusion and associated expansion of the right temporal stem was again noted. A repeat LP was still suggestive of an inflammatory process, however, CSF HSV-2 IgM positive from encephalitis panel. Infectious disease medicine recommended restarting acyclovir. PCR was negative for HSV. Aquaporin-4 antibodies were positive in serum and CSF. Patient was put on 5 day course of IV solumedrol with improvement in her mental status and muscle strength. Patient was started on Rituximab.

Conclusions:
NMO can mimick progressive multifocal leukoencephalopathy (PML) and acute disseminated encephalomyelitis (ADEM).

References: None.

Keywords: Neuromyelitis Optica, Aquaporin-4, Demyelination

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Poster 59
Langerhan's Cell Histiocytosis Masquerading as Optic Glioma

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Introduction:
Langerhans cell histiocytosis (LCH) is a rare disease of uncertain etiology. LCH possibly related to clonal proliferation of CD1a-positive langerhans cells forming a pseudotumoral growth. LCH may present as unifocal, multifocal with a benign or fatal course. LCH disorders are classified into 3 groups: eosinophilic granuloma, Hand-Schüller-Christian disease and Letterer-Siwe disease. The incidence of LCH ranges from 1 to 9 cases per million/ year. Of all cases of LCH, temporal bone involvement has been reported in 20% to 30%. Herein we present an unusual case of LCH masquerading as optic glioma. The clinical, radiological and histopathological results are discussed.

Methods:
This is a case report with retrospective chart review including radiographic and histopathological studies. The literature search was performed using the following search terms: langerhans' cell histiocytosis of optic chiasm, central nervous system and langerhans' cell histiocytosis.

Results:
A 21-year-old male with no significant past medical history presented with progressive hypersomnolence, headache, blurred vision, and 20 pounds weight gain. Neuro-ophthalmological examination revealed a VA of 20/50 on the right and 20/25 on the left. Pupils were equal, and there was no RAPD. EOM was full. Humphrey visual field showed bitemporal hemianopsia. MRI of the brain revealed hypothalamic- suprasellar mass with impingement on both optic nerves and the pituitary stalk consistent with an optic glioma. Due to the associated risks biopsy was deferred. Three weeks later he presented with left otorrhea and CT scan of the temporal bone showed destruction of the mastoid process. Surgical exploration revealed fragile, fleshy grape-like cluster lesions. The diagnosis of LCH was confirmed by *histopathological* and immunohistochemical *staining*. Chemotherapy was subsequently initiated.

Conclusions:
This report describes an unusual mode of presentation masking the presence of LCH thereby delaying diagnosis and management. Although LCH is uncommon in this location it should be considered in the differential diagnosis of a suprasellar mass.

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Keywords: Optic Neuropathy, Neuroimaging, Tumor, Neuro-Ophth & Systyemic Disease, Visual Field

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Poster 60
Profound Bilateral Optic Disc Edema One Month Post-Partum: A Rare Etiology

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Introduction:
Intracranial subdural hematoma formation is a rare, yet potentially fatal complication of spinal anesthesia, as well as inadvertent dural puncture of various iatrogenic etiologies.[1-16] The proposed pathophysiologic mechanism involves significant cerebrospinal fluid loss with resultant caudal displacement of the brain and rupture of bridging cerebral vessels. We present a case of profound bilateral optic disc edema secondary to intracranial subdural hematomas, presumably due to unintentional dural penetration during epidural anesthesia for labor.

Methods:
CASE REPORT: A 27-year-old Ethiopian female, with no significant past medical history, was referred for neuro-ophthalmic evaluation of bilateral disc edema. Patient reported intractable periorbital and frontal headaches, with accompanying photophobia, since vaginal delivery four weeks prior. Childbirth was uncomplicated, though epidural anesthesia required two attempts for correct catheter placement. Upon initial examination, visual acuity was 20/20 OU, with no rAPD. Dilated fundoscopic examination revealed Grade IV disc edema with numerous peripapillary hemorrhages bilaterally. Humphrey visual field testing indicated an enlarged blind spot OU, and RNFL OCT substantiated significant disc edema, with average thicknesses of 381 µm OD and 452 µm OS. Emergent MRI imaging revealed bilateral complex intracranial subdural hematomas. Consequently, the patient underwent successful burr-hole evacuation of the largest right frontoparietal component, which measured 7.7 cm in AP diameter and 2 cm in transverse thickness.Though headaches temporarily recurred 10 days post-operatively, repeat MRI at that time indicated a considerable decrease in the size of the subdural hematomas and no significant new hemorrhaging. The patient began treatment with oral acetazolamide for headache control. Vision remains at 20/20 OU, now with marked reduction in the degree of papilledema (Grade 1 OU).

Conclusions:
Intracranial subdural hemorrhaging status post inadvertent dural puncture may occur within the obstetric population. This clinical entity has not been sufficiently reported in neuro-ophthalmic literature to date, thus improved awareness of this potentially life-threatening complication is critical.

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Keywords: High Intracranial Pressure, Headache, Neuroimaging

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Poster 61

Supranuclear Palsy and Ataxia Related to Anti-Ma2 Paraneoplastic Syndrome: a Diagnostic and Therapeutic Dilemma

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Introduction:
Anti-Ma2 paraneoplastic syndrome is characterized by limbic, diencephalic or brainstem encephalitis, alone or in combination, with oculomotor deficits in 92% of patients with brainstem dysfunction. In young males, this syndrome is highly associated with germ cell tumors, and commonly presents with MRI abnormalities as well as abnormal CSF and EEG atudies.[2] The recommended management approach includes treatment of the underlying malignancy as well as immunotherapy,[3] with neurologic stabilization or improvement in approximately 54% of patients.[1]

Methods:
Case report and literature review.

Results:
A 29 year old male presented with several weeks of difficulty looking down, head tremor and gait instability. Neuro-ophthalmic examination was significant for absent vertical saccades with preserved smooth pursuit on oculocephalic and optokinetic maneuvers. MRI brain with and without contrast revealed no intracranial abnormalities. Routine EEG was also normal. CSF examination was normal except for the presence of oligoclonal bands. Testicular exam revealed a mass later identified as a mature teratoma with metastasis to one paraaortic lymph node. The patient was treated with 5 cycles of plasma exchange as well as high-dose corticosteroids and orchiectomy, followed by two cycles of bleomycin/etoposide/cisplatin. Initial serum test for Anti-Ma1/Ma2 was indeterminate, but was felt to represent a positive result in the setting of recent plasma exchange. However, repeat testing at 3 and 9 months was negative. Despite treatment of the underlying malignancy and negative whole-body PET-CT scan, the patient's symptoms continued to worsen, and repeat brain MRI 9 months following symptom onset showed new midbrain atrophy and enlargement of the 3rd ventricle.

Conclusions:
This is an unusual presentation of Anti-Ma2 paraneoplastic syndrome, with normal initial brain MRI and negative repeat serologies for Anti-Ma2 antibodies, as well as worsening symptoms despite treatment of the underlying malignancy. These characteristics make definitive diagnosis and successful treatment particularly challenging, and highlight a need for further investigation into therapeutic interventions earlier in the disease course.

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Keywords: Paraneoplastic, Supranuclear, Brainstem, Autoimmune, Malignancy

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Poster 62
Painful Opthalmoplegia Due to Burkitt´s Lymphoma (BL) in an HIV-Patient

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Introduction:
To raise awareness on an uncommon etiology of cavernous sinus syndrome, although more frequent in an HIV population, but poorly taken under consideration. The diagnosis was stablished with certainty by means of surgical biopsy.

Methods:
Clinical report of an HIV-patient who consulted with complete and sudden painful opthalmoplegia in his left eye that developed in 10 days, associated with lymphadenopathy and increased liver and spleen´s size. He has been diagnosed of HIV in 2012, and was undergoing retroviral treatment. 2 months prior his symptoms: CD 4+173 cells/mm^3. Normal VA 20/20 OU and normal fundus oculi. His etiological investigation arrouse BL after biopsy of the cavernous sinus and anatomopathological analysis. A brief summery of the patient will be presented, and also the studies carried out to diagnose this rare etiology.

Results:
Our patient underwent the basic neuroopthalmologic examination, brain MRI that revealed an occupying mass in left cavernous sinus, interconsultations with Neurology and Neurosurgery department. The decision of surgical biopsy was conclusive of BL, and intratecal chemotherapy was performed. After treatment, the patient refered diplopia only in extreme abduction of his left eye, and a persistent mydriasis could be noticed.

Conclusions:
Although BL affects the central nervous system very rarely, BL should be considered in any immunosuppressed patient presenting with diplopia or ophthalmoparesis.

References: None.

Keywords: Tumors, Ocular Motility, Neuroimaging, Neuro-Ophth & Infectious Disease, Neuro-Ophth & Systyemic Disease

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Poster 63
Cut From the Same Cloth

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Introduction:
We describe a patient with both HLA-B27 positive anterior uveitis and idiopathic CNS inflammatory syndrome responsive to steroids.

Methods:
Case report.

Results:
A previously healthy 15 year-old African-American boy was diagnosed with bilateral HLA-B27 positive anterior uveitis, successfully controlled with topical difluprednate and oral prednisone. Eight months later, he experienced the acute onset of headaches, left eyelid ptosis, and binocular diplopia. Visual acuity was 20/20 bilaterally. Pupils were reactive without relative afferent pupillary defect. There was trace cell in both anterior chambers and intraocular pressure was normal. Extraocular motility was full with small esodeviation on balance testing. He had mild left upper eyelid ptosis.Brain MRI showed enhancing and T2 hyperintense lesions in the right midbrain, superior colliculus, cerebellar peduncles, cerebellar vermis, and leptomeningeal enhancement along the vermian folia. CSF showed a mild lymphohistiocytic pleocytosis with negative cytology and ACE. Serum ACE, ferritin, TSH, FT4, TPO, TSI, Lysozyme, ESR, CRP, complement factors, aquaporin-4, PPD, ANA, anti-ds DNA, anti-Sm, anti-RNP, anti-SSA, anti-SSB, cystercercus, and HIV labs were within normal limits or negative. Chest X-ray was negative. He received 5 days of 1g IV methylprednisolone without symptomatic improvement; however, repeat MRI showed reduced enhancement of his CNS lesions. He was discharged on 20mg of oral prednisone daily. His symptoms resolved in one month. Repeat MRI two months after presentation showed almost complete resolution of his CNS lesions.

Conclusions:
This is a novel case of idiopathic CNS inflammatory disease in the rare setting of bilateral HLA-B27 positive anterior uveitis.[1,2,3] Given his lesion distribution and response to steroids, a presumed diagnosis of chronic lymphocytic inflammation with pontine perivascular enhancement responsive to steroids (CLIPPERS) was made.[4,5] While not previously reported with CLIPPERS, HLA-B27 positivity may represent a novel association with this new and still incompletely defined CNS disease.

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5. Dudesek A, Rimmele F, Tesar S, Kolbaske S, Rommer PS. CLIPPERS: chronic lymphocytic inflammation with pontine perivascular enhancement responsive to steroids. Review of an increasingly recognized entity within the spectrum of inflammatory central nervous system disorders, Journal of Clinical and Experimental Immunology, 175, 385-396, 2013.

Keywords: CLIPPERS, Uveitis, Neuroinflammation, HLA-B27, Brainstem

Financial Disclosures: The authors had no disclosures.

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Poster 64
Diffuse Large B-Cell Lymphoma Mimicking the Presentation of a Carotid-Cavernous Fistula

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Introduction:
To report an alternative cause of subacute progressive unilateral proptosis with dilated conjunctival vessels suggestive of a carotid-cavernous fistula.

Methods:
Background:Carotid-cavernous fistula (CCF) symptoms typically include unilateral proptosis and dilated conjunctival vessels. While these symptoms may also portend thyroid eye disease, orbital neoplasm or inflammation, dilated vessels point toward a vascular origin. We report a case of an orbital lymphoma initially presenting with signs suggestive of a CCF.

Results:
A 70-year-old male presented with subacute progressive dilated episcleral vessels with subsequent periorbital edema OD, followed by binocular vertical diplopia. Neuro-ophthalmologic examination revealed visual acuity of 20/30 OD and 20/20 OS and normal color vision with 6/6 HRR plates OU. Pupils were 5mm with normal reaction without RAPD OU. Conjunctival injection with corkscrew episcleral blood vessels were noted at the limbus OD. Intraocular pressure was 16 mmHg OD and 19 mmHg OS, and proptosis of 3mm OD was observed. Visual field testing was unreliable with non-specific changes OU. The motility exam revealed decreased supraduction OU, along with a 3-4 diopter left hypertropia with upgaze that converted to a 10-12 diopter right hypertropia with downgaze, suggestive of a partial oculomotor nerve palsy OD. Ophthalmoscopy showed venous distention OD.Cerebral angiography showed no carotid-cavernous fistula. MRI revealed several enhancing cerebral lesions in left cingulate gyrus, right parietooccipital lobes, and superior right orbit with superior rectus involvement. Inferior displacement of the globe was noted, likely impinging venous outflow from right orbit. An orbital biopsy revealed an aggressive diffuse large B-cell lymphoma.

Conclusions:
This case illustrates an alternative diagnosis to explain limited orbital venous outflow in the absence of a CCF. Increased venous pressure occurred secondary to mass effect from the neoplasm on the orbital venous channels. Motility defects may be neuropathic in etiology, with mechanical components possibly related to the superior orbital mass.

References: None.

Keywords: Orbit/Ocular Pathology, Tumors, Vascular Disorders

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Poster 65

Optic Nerve Head Features of a Multiple Sclerosis Patient Masquerading as Glaucomatous Cupping

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Introduction:
The purpose of this abstract is to illustrate a case of Multiple Sclerosis with bilateral temporal pallor of optic nerve heads and Cup/Disc ratio of 0.6 that was initially diagnosed and managed as glaucomatous cupping

Methods:
A case report of a 16 year old female patient who presented with a one month history of ocular pain on eye movement associated with photopsias. Ophthalmic and Neurological evaluation, Automated Perimetry, OCT (ONH and RNFL), MRI (Brain and Cervical spine), VEP, Colored Fundus Photographs and Laboratory Investigations (CBC, CRP, ESR, ALT,AST, Creatinine, Potassium, Sodium,Urea, Anticardiolipin, Lupus anticoagulant, Rheumatoid Factor, TORCH, ACE, CSF examination, TB-DNA, NMO (Aqua Purin - 4Abs)Oligoclonal bands in Serum and CSF) were performed for the patient

Results:
Fundus examination revealed bilateral temporal pallor of optic nerve heads and enlarged cup/disc ratio of 0.6. Average Intraocular pressure was 20mmHg in both eyes on repeated measurements. Automated Perimetry revealed bilateral paracentral defects. OCT revealed bilateral enlarged cup/disc ratio of 0.6 and reduced thickness of RNFL and Ganglion Cell complex
MRI (Brain and Cervical Spine) reported bilateral cerebral, callosal and cerebellar as well as pontine foci and patches of altered signal intensity, likely Multiple Sclerosis with one active lesion at the left parietal region .VEP reported bilateral optic nerve dysfunction and impaired retinocortical transmission mostly attributed to Multiple Sclerosis . Laboratory investigations confirmed the clinical and radiological diagnosis of Multiple Sclerosis

Conclusions:
The presence of optic nerve head pallor, particulary temporal pallor, in the presence of suspicious glaucomatous cupping should be investigated for the possibility of Multiple Sclerosis

References: None.

Keywords: Temporal Pallor of Optic Nerve Head, Glaucomatous Cupping, Multiple Sclerosis

Financial Disclosures: The author had no disclosures.

Grant Support: None.

Poster 66

Transcalvarial Spread of Lymphoma Causing Venous Sinus Congestion, Papilledema, and a Scalp Lump

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Introduction:
The bony cranial vault usually prevents intracranial tumors from spreading extracranially. Transcalvarial extension of a tumor without significant destruction of skull bone is rare.

Methods:
A 46 year old man had five months of worsening blurry vision, transient visual obscurations, and pulsatile tinnitus. A CT head was unremarkable. Subsequent neuro-ophthalmic examination was notable only for decreased visual acuity in the left eye (20/40 left eye versus 20/20 right eye), and moderately severe bilateral papilledema. Visual fields showed blind spot enlargement, left eye more than right. Opening pressure on lumbar puncture was elevated at 38 cm of water. Routine CSF studies (not including cytology) were unremarkable. He was started on acetazolamide. An MRI/MRV head showed an unusual intracranial enhancing mass overlying the right parietal convexity and invading the superior sagittal sinus and incompletely occluding it. This mass extended through the skull into the extracranial scalp, where it formed a right parietal soft tissue scalp mass. Abnormal marrow signal was focally evident between the extracranial and intracranial components of the tumor, but no definite bony destruction of the calvarium was seen. Radiological differential diagnosis included meningioma and lymphoma. The patient carefully palpated his scalp and, indeed, discovered a new right scalp lump (photograph).

Results:
Two consecutive ultrasound-guided needle biopsies of the scalp mass were nondiagnostic; therefore, a right parietal craniotomy was undertaken, and lesional tissue was sent to pathology. Histopathologic examination established a diagnosis of diffuse large B cell lymphoma (80%) arising from a follicular lymphoma (20%). Our patient had a complete response to treatment with five cycles of CHOP-R chemotherapy and declined radiation treatment. His symptoms resolved, and follow-up neuro-ophthalmic examinations and visual fields were normal.

Conclusions:
Intracranial lymphoma may sometimes breach the bony confines of the calvarium and have extracranial extension without significant bony erosion of intervening skull.

References: None.

Keywords: High Intracranial Pressure/Headache, Neuro-Ophth & Systemic Disease, Neuroimaging, Pseudotumor Cerebri, Tumors

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Grant Support: None.

Poster 67
Clinical Variability in Wolfram Syndrome: Report of a Sample of 16 Patients

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Introduction:
Wolfram syndrome is an autosomal recessive disease characterized by the presence of diabetes mellitus (DM), optic atrophy (AO), central diabetes insipidus, hearing loss and neurological disorders such as cerebellar ataxia and psychiatric manifestations. It is caused by mutations in the WFS1 gene. The aim of this study is to present a sample of 16 patients with Wolfram syndrome.

Methods:
We present a sample of 16 patients with Wolfram syndrome, analyzing mutations in the WFS1 gene and describing the clinical manifestations. We performed a complete neurological and ophthalmological examination of these patients, including visual perimetry, fundus and OCT (optical coherence tomography) of optic nerve, when the age of the patients allowed a proper cooperation.

Results:
All individuals in the sample have mutations in the WFS1 gene, communicating in this work at least 5 new mutations not described in the scientific literature. All patients have optic atrophy and ceco-central visual defects in perimetry. However, other typical clinical manifestations present in Wolfram syndrome varies greatly in its clinical expression. In this manner in this sample are presented in one hand patients with mild visual disturbances secondary to optic atrophy, without any other clinical alteration; and patients with severe optic atrophy, diabetes mellitus, diabetes insipidus, deafness and severe ataxia.

Conclusions:
The sample presented here shows that the spectrum of clinical expression of this syndrome is very variable, without an adequate phenotype-genotype correlation.

References: None.

Keywords: Wolfram Syndrome, Congenital Optic Neuropathy, Diabetes Mellitus, Diabetes Insipidus, Hearing Loss

Financial Disclosures: The author had no disclosures.

Grant Support: None.

Poster 68
The Elusive Embolus: Hiding in Plain Sight

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Introduction:
Branch retinal artery occlusion, BRAO is a vision threatening condition caused by occlusion of one or more of the major arteries in the retina, frequently caused by an embolus. Identification of an embolus has important implications to identifying a cause, determining a treatment, and preventing other end-organ damage. Proximal emboli on the optic nerve head are more difficult to visualize compared to distal emboli, which are more easily seen against the contrast of the pigmentary retina.

Methods:
We present 4 cases of patients referred to our clinic with BRAO who had proximal emboli on the optic nerve head. Emboli were invisible or overlooked by biomicroscopy, but easily visualized by red-free and autofluorescence imaging.

Results:
Provided fundus images show emboli that are imperceptible or nearly so on optic nerve heads, but easily visualized with red-free and/or autofluorescence imaging.

Conclusions:
We encourage clinicians to use red-free and autofluorescence imaging when there is suspicion of BRAO but no embolus visualized upon biomicroscopy. Further investigation is warranted and necessary to determine the sensitivity of this relatively inexpensive, noninvasive, and often readily available tool to diagnose proximal embolic BRAO on the optic nerve head.

References: None.

Keywords: Branch Retinal Artery Occlusion, Elusive Embolus, Cilioretinal Artery Occlusion

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Poster 69
Aortic Aneurysm is Common in Treated Giant Cell Arteritis

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Introduction:
Giant Cell Arteritis (GCA) typically affects the vasa vasorum supplying the cranial-cervical arteries. However, any medium to large vessel can be involved. Aortic aneurysm secondary to aortitis may be an unexpected complication of GCA. Corticosteroid related weakening of an already widened aortic wall is also a possibility. We review underlying mechanisms and screening guidelines for aneurysms in GCA.

Methods:
Single case report and literature review

Results:
A 72 year old female complained of a sharp unilateral headache, intermittent fever, and scalp tenderness. Physical Examination revealed visual acuities of 20/20 OU, visual fields full to confrontation. Funduscopic exam was unremarkable. Serologic panel revealed elevated CRP and SED rates. The patient began high dose steroid therapy and temporal artery biopsy confirmed the diagnosis of GCA. CRP and platelets normalized and prednisone was tapered over the next year. After one year, thoracoabdominal contrast enhanced MRA revealed an aneurysm of the ascending aorta 3.6 cm in diameter.

Conclusions:
Investigators report patients with GCA are up to 17.3 times more likely to develop thoracic aortic aneurysms and up to 2.4 times more likely to develop an isolated abdominal aneurysm. Pathologic examination of ruptured anuerysm generally reveals active arteritis. Long term treatment with glucocorticoids may potentially add to the risk of aneurysm development. The present case emphasizes even patients with well controlled GCA may have silent aortic involvement. All patients need to be screened for the presence of aortic pathology.

References: None.

Keywords: Vascular Disorders, Neuro-Ophth & Systyemic Disease

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 70
Chiasmal Tract Neuritis Secondary to Neuromyelitis Optica

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Introduction:
Both neuromyelitis optica (NMO) and multiple sclerosis (MS) can present as acute optic neuritis. The involvement of more posterior structures of the optic nerve, including the chiasm tends to be more indicative of NMO. Distinguishing between NMO and MS is of great importance in guiding treatment.

Methods:
Case Report.

Results:
A 37-year-old female presented with the sudden onset of bilateral visiual loss, headache, and pain with extraocular movement. Initial examination on 9/15/15 showed visual acuity of 20/30 OD and 20/20 OS, color vision was markedly diminished, 0 of 6 HRR plates, flicker-fusion was decreased bilaterally, measuring 17 hz OD and 21 hz OS. Humphrey visual field OD showed a dense central scotoma. HVF OS showed a preponderant temporal hemifield defect. Initially, the fundoscopic examination was normal. Neuroimaging studies demonstrated enhancement of the right optic chiasm and also edema involving the right optic tract. Laboratory studies showed a markedly elevated NMO- IgG antibody at 27.9, (positive range > 5.1). The patient was aggressively treated with high dose intravenous Solu-Medrol followed by an oral steroid taper, six treatments of plasmapheresis and four doses of Rituxan. The patient was reevaluated two weeks later. VA improved to 20/20 OU, HRR plates 0.5/6 OD and 1/6 OS. Repeat fundoscopic examination showed bilateral optic nerve pallor. Flicker-fusion improved to 21 hz OD and 24 hz OS. Visual field dramatically improved with resolution of the central scotoma OD and clinically improved left temporal hemifield defect OS.

Conclusions:
Consideration of NMO was given at the initial evaluation, which was confirmed with the serology. It is noteworthy that in this patient the diagnosis of NMO was established within 48 hours after presentation. We instituted aggressive management with steroids, plasmapheresis and Rutixan, which greatly improved the outcome. Delay in the diagnosis of NMO would have protended a poorer prognosis for full recovery.

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Keywords: Neuromyelitis Optica, Chiasm, Chiasmatic Neuritis

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 71

A Report and Electrophysiologic Analysis of an Isolated Case of Bilateral Optic Atrophy Due to Ocular Graft-Versus-Host Disease

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Introduction:
Hematopoietic stem cell transplantation (HSCT) is a successful treatment for many hematologic malignancies. Despite successes, its known risks include graft-versus-host disease (GVHD). Ocular manifestations occur in up to 90% of patients with chronic GVHD, most commonly affecting the ocular surface, though some cases involve the posterior segment. This unique case reveals a sole clinical exam finding of bilateral optic nerve atrophy in a patient not treated with cyclosporine A. This presentation is completely unique. Furthermore, this study provides objective data from electrophysiologic studies.

Methods:
A 41 year old female with acute myeloid leukemia (AML) in full remission status-post HSCT and chronic GVHD presented with 5 months of deteriorating, blurry vision in both eyes, 1 year after HSCT. She did not receive cyclosporine A for immunosuppresion. Visual acuities fluctuated throughout evaluations and posterior exams showed only bilateral optic nerve pallor.

Results:
Visual field testing fluctuated, ultimately resulting in bilateral central deficits. She had poor color vision with color plate testing. Ocular coherence tomography (OCT) of both maculas showed no edema. OCT of the optic nerves showed diffuse nerve fiber layer thinning bilaterally. OCT of the retina showed generalized retinal thinning bilaterally with nasal sparing of the right eye. Conventional VEP showed decreased amplitudes with normal latencies bilaterally.

Conclusions:
Due to the patient's onset of symptoms following HSCT, the GVHD chronicity and repeatedly negative infectious and malignancy workups, optic atrophy secondary to GVHD was ultimately favored over other proposed etiologies. This unique case highlights the clinical course of a patient who presented with bilateral optic atrophy alone in an atypical diagnosis of surface-sparing ocular GVHD. Clinical evaluation and quantifiable diagnostic testing proved valuable to diagnose and plan for future monitoring of this very rare case and hold promise for others with a similar mechanism.

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Walter, Bertz, Gerling, Bilateral optic neuropathy after bone marrow transplantation and cyclosporin A therapy, Graefe's Archives of Clinical and Experimental, 238, 472-476, 2000.

Keywords: Graft-Versus-Host-Disease, Acute Myeloid Leukemia, Hematopoietic Stem Cell Therapy, Optic Atrophy, Visual Evoked Potential

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 72
IV Bevacizumab: Friend or Foe of the Optic Nerve?

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Introduction:
I present a woman who suffered unilateral blindness from optic neuropathy, after radiation and IV bevacizumab for glioblastoma of the temporal lobe

Methods:
observational/case report

Results:
A 57 y.o. female received a diagnosis of grade 3 astrocytoma of the the right temporal lobe.She completed radiation to the tumor bed after gross total tumor resection two and a half months after the surgery for a total of 59.4 Gy.Temodar was given once but tumor recurrence led to further surgery 5 months after the initial surgery and now pathology showed grade 4 malignant glioblastoma.She underwent q 2 weeks IV bevacizumab, supplemented by NOVO-TFF therapy for a total of 6 bevacizumab treatments. Eight months after completing radiation therapy and after the six bevacizumab treatments she noted blindness in her right eye.She was NLP right eye and 20/20 left eye with a left superior quadrant defect compatible with the priot tumor location and surgeries. The right optic nerve was pale and tthe right pupil was amaurotic. MRI showed enlargement of the right pre-chiasmal optic nerve with T2 hyperintensity and trace enhancement. Bevacizumab related optic neuropathy was diagnosed and treatment was stopped. Bevacizumab is used to treat choroidal neovascularization, diabetic macular edema and even radiation related retinopathy.Could this adverse event occur because of radiation sensitizing the optic nerve to vaso-occlusive events.?Is it purely an anti-VEGF complication we all need to be aware of? How should we treat this?

Conclusions:
Rarely bevicizumab has been reported to cause optic neuropathy after radiation for CNS tumors.I will discuss the literature and suggest possible etiologies and possible treatment options.

References: None.

Keywords: Optic Neuropathy, Chemotherapy and Radiation Injury

Financial Disclosures: The author had no disclosures.

Grant Support: None.

Poster 73
Autoimmune Optic Neuropathy, Case Series

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Introduction:
Auto-immune optic neuropathy represents a heterogeneous group of conditions, some with recognized systemic autoimmune disease and various autoimmune markers in serologic testing including NMO Abs. Others may present with recurrent visual loss and steroids dependency, but lack any evidence of other systemic inflammatory disease. The aim of this study is to characterize the clinical course of patients with recurrent and/or progressive autoimmune optic neuropathy.

Methods:
Retrospective chart review. Patients who were subsequently defined as MS or NMO positive were excluded.

Results:
There were 4 males and 2 females. The mean age of presentation was 47 (range 35-58y). Mean follow up was 4.9 years (range 1-12y). Two patients had simultaneous bilateral onset, 3 had sequential involvement of the second eye, and 1 had pure unilateral disease. All patients had normal brain and cervical imaging (except isolated optic nerve enhancement), and normal lumbar puncture content. Comprehensive inflammatory work up was negative in all of them. Two patients had few episodes of recurrent optic neuritis and could be weaned of steroid after several months. Both had good final visual acuity. 4 patients had chronic relapsing course (fulfilling CRION criteria) and required prolonged treatment with steroid sparing agent.

Conclusions:
Autoimmune optic neuropathy might be a distinct variety of optic nerve inflammation which is not secondary to one of the described demyelinating syndromes. A thorough investigation is necessary to exclude other systemic autoimmune disease and comorbidities. Early recognition of patients is important because they require aggressive and long-term immunosuppressive therapy in order to prevent substantial visual impairment.

References: None.

Keywords: Optic Neuropathy, Inflammtory, NMO, Steroid, CRION

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 74
Sneeze-Induced Amaurosis

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Introduction:
We present the first case of a patient with recurrent amaurosis fugax (AF) induced by sneezing. He was found to have a flow limiting stenosis in the ipsilateral ophthalmic artery.

Methods:
A 74 year old man with a history of hypertension and hyperlipidemia presented to the ER with a temporary loss of vision in his left eye. His CT head had no abnormalities and he was discharged home on Aspirin. He had another spell of transient vision loss in his left eye a few days later and Clopidogrel was added. After this second ER visit he continued to have four to five additional spells. All but one spell were preceded by a sneeze. The first spell was described as a central darkness but for all others he noted a diffuse darkness and blurred vision. None had any altitudinal quality. Fundoscopic examination was unremarkable with no vascular changes suggestive of embolic disease vascular insufficiency.

Results:
Carotid ultrasound, MRI and MRA brain were normal. After another episode he was switched to Aspirin/Extended-Release Dypiridamole and since then has not noted any further events. A diagnostic cerebral angiogram revealed a short segment flow-limiting stenosis in the distal left ophthalmic artery, just proximal to the origin of the central retinal artery. He continues on the medication with no further spells.

Conclusions:
We suspect that the abrupt changes in blood pressure and distal blood flow due to the Valsalva effects of a sneeze resulted in symptomatic retinal ischemia. A transient increase in intraocular pressure with a secondary decrease in retinal perfusion pressure may be another contributing factor in this situation. It is likely that these improved due to mainly the vasodilatory effect of Dipyridamole.

References: None.

Keywords: Amaurosis Fugax, Interventional Neuroradiology, Intracranial atherosclerosis, Retina

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Grant Support: None.

Poster 75

Morbidity and Mortality in Patients After Skull Base Osteomyelitis. A Report of 3 Cases and Review of the Literature.

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Introduction:
Skull base osteomyelitis (SBO) is an uncommon but life threatening condition, most commonly secondary to otitis externa (1). We report 3 cases of SBO which presented with neuro-ophthalmic manifestations without aural pathology. Knowledge of SBO by the neuro-ophthalmologist is essential.

Methods:
Retrospective case series and review of literature.

Results:
Three cases of non-otogenic SBO are presented. All presented with neuro-ophthalmic features. Detailed history revealed an insidious onset over several months. Initial radiological abnormalities were subtle and easily overlooked. The neuro-ophthalmologist is essential in monitoring the efficacy of treatment. One patient had a fatal outcome whereas the other 2 recovered after an extended cause of antibiotics.

Conclusions:
It is important that neuro-ophthalmologists are familiar with the presentation as well as the subtle radiological features in central or non-otogenic SBO. These patients are usually elderly and immuno-compromised (2), present with optic neuropathy and/or cranial nerve palsies and commonly have a prior history of headache(2) or neck pain. The onset is insidious and a high index of suspicion is required.

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2) Pang A, Chong K. Skull base osteomyelitis originating in the middle ear or mastoid: A review of 3 cases. Ear, Nose and Throat Journal;90(3):121-4, March 2011.

Keywords: Skull Base Osteomyelitis, Optic Neuropathy, Sinusitis, Cavernous Sinus Syndrome, Orbital Apex Syndrome

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Poster 76

Anti-Phospholipid Syndrome and Neurofibromatosis 1- New Association or Coincidence?

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Introduction:
Structural vascular anomalies and ischemia associated with neurofibromatosis 1 (NF1) are well reported and are thought to result from neurofibromin dysfunction. Documented cases of associated anti-phospholipid syndrome (APS) that fulfill the accepted diagnositic criteria for APS are exceptionally rare.

Methods:
We present a patient with NF1 and asymptomatic cavernous meningioma and contralateral optic atrophy who also had clinical and laboratory evidence of APS.

Results:
A 40 year old woman with known NF1 presented with sudden painless right vision loss and was diagnosed with central retinal vein occlusion. MRI of the brain revealed a left cavernous sinus meningioma. Past history was significant for three spontaneous abortions. Two of her three children had dermatologic stigmata of NF1. Examination revealed multiple café au lait spots and visual acuities of 20/300 OD and 20/25 OS. Color vision was 1/15 OD and 15/15 OS. The pupils were reactive with right relative afferent papillary defect. The anterior segments were normal with one Lisch nodule OS. Ophthalmoscopy revealed right optic atrophy and a normal left nerve. Octopus VF was not possible OD but showed mild constriction OS. OCT measured average retinal nerve fiber layer thickness of 52µm (right) and 97µm (left). Positive lab results include increased anti-β_2 glycoprotein 1 IgA and anti-β_2 glycoprotein 1 IgM antibodies, C4 and ESR (51mm). Repeat tests at 3 months revealed persistently elevated ESR and anti-β_2 glycoprotein 1 IgA. She fulfulled Sydney Criteria for diagnosis of APS with a least one clinical and laboratory evidence of APS: frequent abortions, a vascular occlusive episode and persistently elevated anti-β_2 glycoprotein1 IgA antibodies.

Conclusions:
The association of NF1 and APS has been reported previously and this case, to our knowledge, documents the first ophthalmic manifestation. Although the concurrence of these disorders, both with wide phenotypic spectra, might be coincidental, it might represent a new unrecognized association.

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Carr ME Jr, Howe CW. Lupus anticoagulant and cerebrovascular accident in a patient with neurofibromatosis. South Med J 1989; 82: 921-3.

Keywords: Neurofibromatosis 1, Anti-Phospholipid Syndrome, Anti-Glycoprotein 1, Optic Atrophy, Meningioma

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Poster 77

Headache in Multiple Sclerosis May Be Due to Idiopathic Intracranial Hypertension

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Introduction:

Headache is common in individuals with proven MS and generally considered either migraine or muscle tension type. Idiopathic Intracranial Hypertension (IIH) with Multiple Sclerosis (MS) is rarely reported. We examined three patients with IIH. In two cases papilledema was absent, confounding the correct diagnosis.

Methods:

Case series of women with IIH and MS and literature review.

Results:

Three women with definite relapsing remitting MS complained of generalized headaches. None experienced visual obscurations, tinnitus, nor diplopia. Visual acuity was preserved, and visual fields showed only big blind spots. Papilledema was absent in two out of three cases. Spinal fluid pressures were significantly elevated in all cases. In one individual, headache and associated positive MRI scans preceded by one year the development of the first clinical sign of MS, a left hemi-paresis. Two patients responded to conventional treatment; one required ventriculoperitoneal shunt for headache control.

Conclusions:

More than 50% of patients with MS complain of frequent headaches, typically classified as migraine or tension type. 3 patients previously reported present with unremitting headaches. Each case demonstrated disc head swelling. In two of our three patients, fundus findings were consistently normal, confirmed by optical coherence tomography and fundus photos. The cause of IIH in our patients in uncertain. IIH reportedly occurs with other autoimmune conditions, including Systemic Lupus Erythematous(SLE), Sjogren's syndrome, Crohn's disease, Guillain-Barre syndrome, and Ankylosing Spondylitis. Similar to our patients, several SLE patients with IIH reportedly had no papilledema. Also, most reported cases had no additional predisposing risk factors. Although, most reports were not case controlled for co-morbidities. Recently investigators demonstrated elevated inflammatory markers and cytokines in patients with IIH. It seems plausible inflammatory states caused by autoimmune conditions like MS and SLE cause dysfunction of the arachnoid villi. IIH is a consideration in patients with MS and headache.

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Keywords: High Intracranial Pressure/Headache, Neuro-Ophth & Systyemic Disease (Eg. MS, MG, Thyroid), Demeylinating Disease

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Poster 78

Multifocal Electroretinography and Visual Evoked Potential Findings in a Case of Optic Disc Pit

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Introduction:
To present multifocal electroretinography (mfERG) and visual evoked potential (VEP) findings in a case of a unilateral optic disc pit.

Methods:
ISCEV guidelines were followed while performing mfERG and VEP. Color fundus photography, SITA-standard Humphrey Visual Field (HVF), and spectral-domain Optical Coherence Tomography (OCT) were also obtained.

Results:
A 34 year-old woman complaining of decreased vision was found to have a left optic disc pit. Visual acuity was 20/80 in the right eye and 20/100 in the left eye. Vision improved with pinhole to 20/25 in the right eye and 20/30 in left eye. Intraocular pressures and anterior segment examination were unexceptional. Dilated fundus examination revealed mild temporal optic disc pallor, and a localized oval depression at the temporal border of optic disc in the left eye. No macular abnormalities were observed. OCT supported the clinical findings with mild temporal thinning of the nerve fiber layer in the region of the optic disc pit. HVF 30-2 findings showed a superior arcuate defect. HVF 10-2 revealed a trace subtle defect. MfERG findings revealed differences in voltage amplitude and waveform latencies in the specific region of the optic disc pit when compared with corresponding regions in the right eye. VEP findings were abnormal in both eyes, with poorly defined n135 waveforms at 15 minutes check size of stimulation. Latencies and amplitudes of n75 and p100 waveforms in the left eye were slightly prolonged and reduced, respectively.

Conclusions:
Optic disc pit is a rare congenital condition, first reported by Weithe in 1882.[1] Although OCT is commonly used to describe the morphology of optic disc pits,[2] few studies have reported mfERG and VEP findings. Both mfERG and VEP appear to have a role in defining the electrophysiology characteristics of optic disc pits.

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Keywords: Optic Disc Pit, Electroretinography, Visual Evoked Potentials

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Poster 79
Right in Front of Your Face

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Introduction:
Neurosarcoidosis is a rare but well documented cause of vision loss and should be considered when neuroimaging shows multiple enhancing intracranial masses.

Methods:
Case report

Results:
A 54-year-old African American woman with diabetes and hypertension presented with a 2-week history of painless left-sided vision loss and a 6-month history of a red-brown cutaneous lesion at the right nasal bridge. Best-corrected visual acuity was 20/25 on the right and counting fingers on the left with a left relative afferent pupillary defect. Anterior segment and dilated fundus examinations were unremarkable. MRI with contrast showed irregular, nodular enhancement of both optic nerve sheaths with a 4x4x2 mm-nodular enhancing mass abutting the medial aspect of the left optic nerve just posterior to the orbital apex. There were an additional 3 extra-axial enhancing lesions along the superior margin of the left tentorium, the medial aspect of the right middle cranial fossa, and the vertex. Workup revealed normal CBC, ANA, ACE level, and lysozyme. CSF analysis showed a mild lymphocytic pleocytosis with normal glucose and negative cytology. Punch biopsy of the cutaneous lesion revealed non-caseating granulomas consistent with sarcoidosis. CT of the chest showed hilar adenopathy. A diagnosis of neurosarcoidosis was made. Treatment with 60 mg of oral prednisone led to rapid improvement of visual acuity to 20/30 in the left eye, reduction in the size of all of the intracranial masses, and complete resolution of the orbital apex mass compressing the left optic nerve. She is now stable on azathioprine and a slow steroid taper.

Conclusions:
Neurosarcoidosis can cause vision loss via direct optic nerve sheath involvement, and is important to consider in the differential diagnosis of enhancing CNS lesions. Prompt identification of systemic manifestations and biopsy of a skin nodule characteristic of cutaneous sarcoidosis aided in this patient's prompt diagnosis and successful treatment.

References: None.

Keywords: Neuro-Ophth & Systemic Disease, Neuroimaging

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Poster 80

Visual Evoked Potential Findings in a Case of Posterior Ischemic Optic Neuropathy Following NSAID-Induced Gastrointestinal Hemorrhage

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Introduction:
This case report aims to add to the paucity of clinical data on posterior ischemic optic neuropathy (PION) by presenting the electrophysiological findings in an unusual case of PION following NSAID-induced gastrointestinal (GI) hemorrhage.

Methods:
ISCEV guidelines were followed to record single channel VEP (visual evoked potential) findings. Results of SITA-standard Humphrey visual field (HVF) and spectral-domain OCT were also obtained.

Results:
A 56-year-old male presented with history of decreased vision in left eye for 3 years following an acute episode of GI hemorrhage secondary to NSAID use for chronic shoulder pain. Upon initial examination, visual acuity was 20/20 in the right eye (OD) and hand motion in left (OS). Intraocular pressures and anterior segment exam were unexceptional. A dilated fundus exam revealed bilateral disc pallor, 1+ in OD and 2+ in OS. HVF results revealed an inferior arcuate defect in OD, and non-specific peripheral visual field loss, with infrotemporal preservation in OS. OCT of optic nerves showed bilateral temporal thinning of the nerve fiber layer and other regions consistent with areas of visual field loss. VEP findings were abnormal, with prolonged latencies and decreased amplitudes observed bilaterally.

Conclusions:
With anterior etiologies accounting for 90% of cases of ischemic optic neuropathy, little has been reported about PION[1]. Unlike anterior ischemic optic neuropathy, PION is difficult to recognize due to its rarity and relatively subtle clinical exam findings. Additionally, the vast majority of PION cases occur perioperatively [1] rather than in association with medication use as in our patient. To the best of our knowledge, this is the first reported use of VEP findings to assess PION in NSAID induced GI hemorrhage. [1] (Biousse & Newman, NEJM, June 2015)

References:
Biousse, Newman, Ischemic Optic Neuropathies, New England Journal of Medicine, Vol. 372, pages 2428-2436, June 2015

Keywords: Posterior Ischemic Optic Neuropathy, Visual Evoked Potentials, Ischemic Optic Neuropathy, Optic Neuropathy, Electrophysiology

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Poster 81
Neuro-Ophthalmologic Manifestations of Cholangiocarcinoma: A Case Series

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Introduction:
Cholangiocarcinoma is a rare liver malignancy originating in the biliary epithelium. Patients may present with signs and symptoms of biliary obstruction, or nonspecific complaints such as malaise, weight loss, and night sweats. It is uncommon for this malignancy to spread distally and our review of the literature identified only 16 cases of brain metastasis from cholangiocarcinoma none of which presented neuro-ophthalmologic manifestations. Metastasis to the brain and orbit can result in neuro-ophthalmologic deficits such as diplopia, proptosis, and decreased vision. We present the first and largest series of the neuro-ophthalmologic manifestations of cholangiocarcinoma.

Methods:
We completed a retrospective chart review of recent cholangiocarcinoma patients presenting to ophthalmology clinics within two tertiary care centers in the Texas Medical Center.

Results:
Chart review identified four patients with neuro-ophthalmologic symptoms related to cholangiocarcinoma. One patient presented with diplopia due to metastasis to the left medial rectus, two had involvement of the brain resulting in sixth nerve palsy and hemianopsia, and one presented a hypercoagulable state that predisposed the patient to a stroke causing homonymous hemianopsia and visual hallucinations. The clinical presentation of these patients included symptoms of partial to complete vision loss, diplopia, and visual hallucinations.

Conclusions:
Neuro-ophthalmic manifestations of cholangiocarcinoma depend upon both mechanism and localization. We report 4 cases of cholangiocarcinoma with neuro-ophthalmologic findings. To our knowledge, this is the largest such series reported in the English-language ophthalmic literature.

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Fujimoto, Kenji, Jun-Ichiro Kuroda, Keishi Makino, Yu Hasegawa et al. Skull Metastasis From Intrahepatic Cholangiocarcinoma: Report of 3 Cases and Review of the Literature. Neurol Med chir. p. 717-21. 2013.

Keywords: Tumors, Optic Neuropathy, Visual Fields, Neuro-Ophth & Systemic Diseases

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Poster 82

Tacrolimus Optic Neuropathy: A Case Series and Review of the Literature

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Introduction:
Tacrolimus is a potent immunosuppressant that inhibits cytokine synthesis and blocks T-cell development. Optic neuropathy from tacrolimus toxicity is very uncommon with only a few case reports in the literature. We report three cases of tacrolimus optic neuropathy following bone marrow transplant complicated by graft versus host disease. These three cases demonstrate differing clinical and radiologic presentations of tacrolimus optic neuropathy.

Methods:
Case series and review of the literature

Results:
The first case presented with unilateral optic nerve head swelling and peripapillary hemorrhages similar in appearance to NAION with a normal appearing MRI. The second case demonstrated bilateral optic nerve head edema with flame shaped hemorrhages and the MRI demonstrated subtle enhancement of both optic nerves. The third case had normal appearing optic nerves with a clinical presentation of bilateral optic neuropathy, however the MRI demonstrated profuse T2 hyper intensity throughout the anterior medulla, pons, midbrain and internal capsule in addition to T2 hyper intensity of the optic nerves. This patient's marked clinical and radiographic findings were noted to improve following cessation of the medication

Conclusions:
Tacrolimus optic neuropathy can present as a unilateral, bilateral, anterior or posterior optic neuropathy. Enhancement of the optic nerve may or may not be present and additional radiologic findings can include diffuse intracerebral lesions. Lastly, all of our patients had graft versus host disease which raises the suspicion of whether patients with GVHD have a higher propensity to develop optic neuropathy from tacrolimus. Being aware of the spectrum of clinical presentation will help the neuro-ophthalmologist to detect this vision threatening toxicity.

References: None.

Keywords: Tacrolimus Toxicity, Optic Neuropathy

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Poster 83
Intraocular-Orbital-Cerebral Toxoplasmosis as an Initial Presentation of AIDS

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Introduction:
To Demonstrate Clinical Manifestations and Treatment Outcome of Intraocular-Orbital-Cerebral Toxoplasmosis in Newly Diagnosed HIV-Infected Patient

Methods:
Case report

Results:
A 67-year-old Thai man presented with acute painful visual loss of his right eye for 5 days. External examination of right eye showed visual acuity of no light perception (NLP), complete ptosis, generalized chemosis, axial proptosis and severe ocular motility restriction. External examination of left eye was unremarkable. Slit-lamp bio-microscopy of right eye revealed active panuveitis with large area of infiltration involving entire posterior segment, particularly in the posterior pole. Slit-lamp bio-microscopy of left eye was significant only for senile cataract. MRI of the brain and orbit showed profound right optic nerve sheath enhancement, right orbital inflammation and multiple targetoid lesions in the brain. He was subsequently diagnosed with HIV infection according to the presence of anti-HIV antibody in his serum. Lumbar puncture was not performed regarding high risk of brain herniation. PCR for *toxoplasma sp.* was positive from aqueous humor of right eye. After treatment with oral Trimethoprim/Sulfamethoxazole for 6 days, the patient demonstrated clinical improvement except for NLP of his right eye. This was confirmed by MRI which showed marked decrease enhancement of right optic nerve sheath and numbers of targetoid lesions.

Conclusions:
Concurrent intraocular-orbital-cerebral involvement is a rare clinical presentation of toxoplasmosis. Patient with new-diagnosed HIV infection can present with this uncommon manifestation. In our case, the diagnosis was confirmed by positive PCR for *toxoplasma sp.* from aqueous humor and significant improvement after treatment.

References: None.

Keywords: Neuro-Ophth & Infectious Disease, Neuroimaging, Optic Neuropathy, Orbit/Ocular Pathology

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Poster 84

Total Absence of Conjugate Horizontal Eye Movements and Facial Paresis as the First Presentation of Multiple Sclerosis

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Introduction:
Eye movement abnormalities are a common manifestation of Multiple sclerosis (MS). Here we report a patient with total absence of conjugate horizontal eye movements, and facial diparesis, as an initial presentation of MS.

Methods:
Case report and review of literature.

Results:
An 18 year old girl presented with a two-month history of blurry vision, diplopia and new left facial droop involving the upper and lower face. She had right facial droop one month before this admission that improved after receiving steroid treatment for presumed Bell's palsy. On exam, she had absent horizontal saccades, pursuit, VOR, and OKN, but spared convergence. Upper eyelids, pupils and vertical eye movements were intact. The initial differential diagnosis included a vascular or inflammatory etiology and Miller-Fisher syndrome. MRI showed a pontine, paramedian T2 hyper-intense lesion, with subtle post-contrast enhancement, involving bilateral sixth nerve nuclei and facial colliculi. In addition, multiple other T2 hyper-intense lesions were also observed, some enhancing, indicating dissemination in time and space satisfying MS diagnostic criteria. CSF studies revealed multiple oligoclonal bands. She was treated with five days of steroids, and Interferon Beta-1a. On follow up three weeks later, she regained horizontal ductions of the right eye. The abduction of the left eye was still impaired. Pursuit and VOR failed to improve left abduction.

Conclusions:
The abducens nucleus is the final common pathway for all horizontal conjugate eye movements. A complete bilateral conjugate gaze paralysis associated with preceding peripheral facial diparesis suggests a demyelinating lesion affecting the facial colliculi with secondary inflammation of the immediately adjacent bilateral abducens nuclei. To our knowledge, absent conjugate horizontal eye movements complicating acute bilateral facial colliculi demyelination have not been previously reported in MS.

References: None.

Keywords: Demeylinating Disease, Ocular Motility

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Poster 85
Bilateral Vision Loss Due to Chiasmal Neuropathy Secondary to Muslinoma

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Introduction:
Intracranial aneurysms are surgically managed by microsurgical clipping of the aneurysm neck, endovascular coiling of the aneurysm sac or balloon occlusion of the parent vessel. Cases with broad-based aneurysms may not be amenable to these techniques. Microsurgical wrapping of the aneurysm wall with muslin may be applied.

Methods:
Case report, and review of literature.

Results:
We report a case of 59-year-old female who initially presented with the incidental finding of multiple intracranial aneurysms on MRI. She underwent elective clipping of the right MCA bifurcation aneurysm and wrapping of a small anterior communicating artery aneurysm with muslin. After 40 months, she experienced progressive, bilateral visual loss, facial pain and retrobulbar headache. On examination, best corrected visual acuity was 20/25 in the right eye and counting fingers in the left eye, with color vision of 10/13 in the right eye and 0.5/13 in the left and a 0.6 log APD in the left eye. Visual field testing revealed a junctional scotoma, with dense central vision loss in the left eye. Cranial magnetic resonance imaging revealed multiple enhancing lesions surrounding both aneurysms and involving the optic chiasm. Intravenous methylprednisolone (1g/day) was administered over for 5 days, with significant improvement in acuity and fields within 36 hours. After 1 month, visual acuity improved to 20/20 in the right eye and 20/30 in the left with near-complete resolution of the visual field defect.

Conclusions:
A muslin-induced optic neuropathy is a rare but serious complication of a chronic inflammatory reaction in response to muslin wrapping. Treatment options include surgery, steroids and cyclophosphamide, but recovery of the vision is unpredictable. Muslin should only be utlilized with caution in neurovascular repairs in close proximity to the anterior visual pathyway. Patients with muslin in close proximity to the visual pathway need to be warned about and screened for vision loss.

References: None.

Keywords: Muslinoma, Aneurysm, Chiasmal, Neuritis

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Poster 86
Horner Syndrome in a Patient with Marfan Syndrome and Carotid Artery Tortiuousity

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Introduction:
Marfan syndrome (MFS) is an autosomal dominant disorder of the connective tissue characterized by early development of thoracic aortic aneurysms and/or dissections. The presence of MYH-11 mutation has been associated with Familial thoracic aortic aneurysm and dissection (TAAD) .Horner syndrome in a patient with MFS indicates carotid dissection until proven otherwise.

Methods:
Case report, and review of literature.

Results:
A 15-year-old male with a Marfanoid habitus presented for outpatient ophthalmic evaluation. His ocular examination revealed distance uncorrected visual acuity of 20/20 in each eye. He had no classical ocular signs of MFS with normal lenses and reasonable keratometry. However, he did have some retinal arterial tortuously more obvious in right eye than the left. He was found to have a variance of both the Fibrillin 1(FBN1) and the Myosin-11(MYH11) genes. Echocardiography revealed aortic root dilatation (38mm) but no other abnormalities. Four months later, he presented for an urgent evaluation of a droopy right eyelid and asymmetric pupils noticed by the parents. He was found to have 2mm ptosis and anisocoria in the pattern of right Horner syndrome. The diagnosis was confirmed pharmacologically using apraclonidine with reversal of anisocoria. He was referred for an urgent neurological evaluation and neuro-vascular imaging that revealed bilateral carotid and vertebral artery tortuously with no signs of dissection.

Conclusions:
Horner syndrome is one of the presenting neurologic findings in patients with carotid dissection (present in about 50% of patients). Early recognition of Horner syndrome, allows early diagnosis and prompt management of carotid artery dissection to prevent its neurovascular complications. Our case did not have any radiological findings of dissection. However, we theorize that oculosympathetic neurotemesis may have resulted from the repeated trauma of the carotid pulsations and daily head movements given the weak connective tissue support of the sympathetic chain.

References: None.

Keywords: Horner, Marfan, Taad, Aneurysm

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 87
Leukemic Infiltration of the Optic Nerve

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Introduction:
Leukemic infiltration of the optic nerve is rare and optic neuropathy as presenting symptom of relapse in patients with leukemia is rarer still. We report a patient with acute myelogenous leukemia (AML) who was in remission yet relapsed with an acute optic neuropathy in the left eye.

Methods:
A 30-year-old woman presented with a 1 month history of painless progressive visual loss in her left eye (OS). She denied any previous ocular history. She had a medical history of acute myelogenous leukemia (AML) for which she received induction chemotherapy and 4 cycles of consolidation chemotherapy resulting in complete remission 9 months ago. Examination showed a visual acuity of 20/25 in the right eye (OD) and count fingers OS. She had a relative afferent pupillary defect OS. Confrontation visual field was normal OD and severely constricted OS. Ocular motility was normal and there was no ptosis or proptosis. Anterior segment and posterior segment examinations were unremarkable including a sharp, pink, and healthy optic nerves.

Results:
Pattern VER was normal OD and non-recordable OS. With the flash VER, the P-100 was delayed at 120 ms OS. A gadolinium enhanced, fat-suppressed cranial and orbital MRI showed focal enlargement and enhancement of the intracranial segment of left optic nerve consistent with leukemic infiltration. The patient was admitted, prescribed high dose IV methylprednisolone and received systemic and intrathecal chemotherapy. Allogeneic transplantation and radiotherapy to the left optic nerve are being discussed at the time of submission of this case.

Conclusions:
In a patient with a history of leukemia and acute optic neuropathy, leukemic infiltration of the optic nerve is the main differential diagnosis. Contrasted fat-suppressed cranial and orbital MRI imaging is very helpful in the detection of the leukemic optic nerve infiltrate with the infiltrated optic nerve characteristically enlarged and enhancing. A variety of therapies have been proposed for leukemic infiltration of the optic nerve including intrathecal chemotherapy and radiotherapy. The optimal therapy for our patient remains to be seen.

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Keywords: Optic Neuropathy, Chemotherapy and Radiation, Neuroimaging, Leukemia

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Poster 88
Malingering and Secondary Gain in the Afghanistan and Iraq Conflicts

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Introduction:
Functional or non-organic visual loss (NOVL) is defined as a loss or decrease in visual acuity or visual field range with no identifiable organic cause. NOVL can be a difficult diagnosis to make and requires a high index of suspicion. This can be especially difficult in the setting of true organic pathology, as was the experience with several patients presenting to an Ophthalmology clinic after sustaining either ocular or non-ocular injuries. This case series examines several such patients with NOVL that were either injured or developed an ocular condition while serving in Iraq or Afghanistan. The aim of this series is to provide a review of several possible presentations of NOVL and the various modalities that the ophthalmologist can use to arrive at a diagnosis of NOVL.

Methods:
A retrospective chart review was conducted of patients with NOVL presenting to an Ophthalmology department after sustaining either ocular or non-ocular injuries during operations in Iraq or Afghanistan.

Results:
Seven patients were diagnosed with NOVL after sustaining either ocular or non-ocular injuries during operations in Iraq or Afghanistan. A variety of useful techniques are available, including binocular visual fields, 4 PD prism test, and refractive fogging, to better assess for a possible functional overlay.

Conclusions:
The process of diagnosing NOVL begins with a detailed history and a high index of suspicion. The exam and work-up must be thorough to exclude any possible organic pathology that could be contributing to the patient's symptoms. Once NOVL is suspected, the ophthalmologist has several simple examination techniques at their disposal to properly identify these patients in a timely fashion that may eliminate or reduce the need for additional and more costly testing. The early recognition and diagnosis NOVL, though often a tedious task for the clinician, is essential for the appropriate care and proper disposition of these patients.

References: None.

Keywords: Non-organic visual loss, Functional Visual loss, Malingering

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Poster 89
Congenital Hereditary Idiopathic Horner Syndrome in an Upstate New York Family

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Introduction:
Congenital hereditary idiopathic Horner syndrome has been rarely described in the literature, herein we present three affected first-degree relatives.

Methods:
Case series and literature review.

Results:
A 44-year-old woman was referred for possible pseudotumor cerebri. She incidentally reported a left “eye droop” present from birth. Pupils measured 4 mm OD, 3.5 mm OS in light, and 5 mm OD, 4.5 mm OS in dark. Marginal reflex distance (MRD) was 6 mm OD, 3 mm OS. Her palpebral fissure height was 8 mm OD, 6 mm OS. After the instillation of 0.5% apraclonidine, pupils measured 3.5 mm OD, 3.5 mm OS in light, and 4 mm OD, 4.5 mm OS in dark. MRD was 7 mm, 5 mm, respectively. Palpebral fissure height measured 10 mm, 11 mm, respectively. A CTA head and neck were normal with no carotid dissection. At a subsequent visit, her 10-year-old son and 76-year-old mother were examined and found to have ptosis and miosis, without heterochromia iridum or anhidrosis. The son exhibited reversal of ptosis and the mother exhibited reversal of ptosis and anisocoria to apraclonidine. In all cases, ptosis was noted since birth, and there was no history of birth trauma. A post-ganglionic lesion is surmised in all three cases. The patient reports that her sister, maternal grandmother, and a maternal first-cousin also exhibit ptosis. A review of literature identified five case reports of hereditary congenital Horner syndrome (1918 - 1992). They described an autosomal dominant inheritance with variable penetrance, consistent with the family we studied.

Conclusions:
Congenital hereditary idiopathic Horner syndrome is an autosomal dominant condition with incomplete penetrance. We report three first-degree relatives with ptosis and miosis. Further exploration including a genome wide association study of this family could improve our understanding of the developing sympathetic nervous system.

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Miller N R, Walsh F B, Hoyt W F, eds., Walsh and Hoyt's Clinical Neuro-Ophthalmolog, Disorders of Pupillary Function, Accommodation, and Lacrimation: Horner Syndrome, 2, 749-758, 2005.

Keywords: Congenital, Hereditary, Idiopathic, Horner Syndrome

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Poster 90

Rhinocerebral Mucormycosis in an HIV/AIDS Patient Presenting as Isolated Unilateral Sixth Nerve Palsy: Case Report and Literature Review

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Introduction:
Mucorales causes a rare but serious and potentially life-threatening fungal infection. Mucorales is invasive and angiotrophic. It causes significant vascular changes including thrombus, arteritis and aneurysm.[1] It occurs primarily in immunocompromised individuals with compromised neutrophil function. The occurrence in HIV/AIDS patients is rare since T cell is not the main defensive mechanism and neutrophil function is relatively preserved with those patients.[2] [3] [4]In this report, we have a case of rhinocerebral mucormycosis that presented as isolated sixth nerve palsy without Horner's syndrome due to intracavernous internal carotid artery (ICA) mycotic aneurysm in an HIV/AIDS patient. The case is unique due to the clinical presentation, location of the pathology and the uncommon risk factor.

Methods:
This is a case report

Results:
Rhinocerebral mucormycosis can be one of the causes of the isolated sixth nerve palsy without Horner's syndrome.

Conclusions:
As practitioners, we need to be vigilant for the presentation of mucormycosis in HIV/AIDS patient since prompt diagnosis and treatment can potentially significantly impact the outcome of mucormycosis.

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Keywords: Abducen, Mucor, HIV

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Grant Support: None.

Poster 91
A Jaw Dropping Case

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Introduction:
A 78-year-old woman presented with two months of intermittent diplopia associated with headaches and jaw pain that worsened with chewing.

Methods:
Case Report.

Results:
Her medical history was significant for hypertension, hyperlipidemia and breast cancer (s/p lumpectomy and chemotherapy 24 years prior). On initial presentation, she had normal visual acuity and full ductions. She had a small right hypertropia that worsened in left gaze and right head tilt. ESR and CRP were normal. A head CT and brain MRI were unremarkable. A left temporal artery biopsy revealed no vasculitis. EMG with repetitive stimulation was normal. Serum acetylcholine receptor, MuSK, and voltage-gated calcium channel antibodies were normal. One month later she presented with worsening headaches and diplopia. She also experienced jaw weakness so severe she had to use her hand to close her jaw in order to chew solid foods. On examination she had ptosis, exotropia, left adduction deficit, left hypertropia, inability to close her jaw, and weak eyelid closure. She had lower extremity ataxia and reduced deep tendon reflexes (absent in the lower extremities). Anti-GQ1B antibodies were not present, and a repeat brain MRI with contrast was unchanged. A lumbar puncture was performed. CSF analysis revealed WBC 91 (15% segs), RBC 3000, protein 160, glucose 13. Cytology was positive for metastatic carcinoma. Chest CT revealed a 0.5 cm pulmonary nodule and a right breast soft tissue mass.

Conclusions:
This elderly patient presented with fluctuating diplopia, headache and jaw pain. When GCA and a myasthenic syndrome had been ruled out, it was assumed she had a microvascular cranial nerve palsy. Instead of improving, however, she developed multiple cranial nerve palsies, ataxia and areflexia. Two MRI brain studies were non-diagnostic. Although the Miller Fisher variant of GBS was considered, lumbar puncture revealed a pleocytosis and hypoglycorrhachia, and CSF cytology confirmed the correct diagnosis. Final diagnosis: Meningeal carcinomatosis (primary breast cancer with metastatic disease).

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Keywords: Diplopia, Neuro-Ophth & Systemic Disease, Multiple Cranial Neuropathies, Meningeal Carcinomatosis

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Poster 92
Slowly Eye Turned

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Introduction:
In the setting of an otherwise isolated sixth nerve palsy without vascular risk factors, it is important to evaluate reflex tearing with a Schirmer test as the presence of ipsilateral decreased tearing can localize the lesion to the petrous apex.

Methods:
Case report.

Results:
A 63-year-old healthy man presented with three months of painless horizontal diplopia. He denied weakness, ptosis, trouble swallowing or difficulty breathing. His previous MRI brain was reportedly normal. Edrophonium testing reportedly improved his right abduction deficit, although the patient denied improvement. A single fiber EMG was supportive of the diagnosis of myasthenia gravis. His diplopia improved but did not resolve with 100 mg prednisone every other day.On examination, his vision was 20/20 bilaterally. 24-2 Humphrey automated SITA-fast perimetry was normal. His pupils were 3mm OU briskly reactive without RAPD with palpebral fissures of 8mm OU. He had visibly slowed and limited abduction OD with 2.5mm abduction OD compared to 9mm abduction OS. There was no nystagmus, lightening saccades, lid twitch or fatigue and no evidence of proptosis or retraction. He had a large esotropia in primary gaze that increased in right gaze. Slit lamp examination and dilated fundus evaluation were unremarkable. V1-V3 sensation was intact. A Schirmer test with anesthetic and intranasal tickle was performed with 2mm OD and 19mm OS. Review of previous MRI revealed a destructive right petrous apex mass, which was resected and found to be a chondroma. On evaluation six months later, his saccadic velocity and abduction normalized, but his tearing deficit persisted.

Conclusions:
Identification of an ipsilateral reflex tearing deficit in the setting of an otherwise isolated sixth nerve palsy localizes the lesion to the petrous apex. Furthermore, it is important to differentiate the supranormal saccadic velocity in myasthenia gravis from the slowed saccadic velocity in sixth nerve palsies.

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Keywords: Ocular Motility, Neuroimaging, Skull Base, Tumors, Myasthenia

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Grant Support: None.

Poster 93

A difficult day with double trouble

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Introduction:

A 59 year old woman with a history of hypothyroidism, an inner ear implant and lupus (complicated by autoimmune hemolytic anemia and raynaud's disease) presented with 1 week of intermittent horizontal binocular diplopia. She denied headache and giant cell arteritis symptomatology. Examination demonstrated normal afferent function, full ductions and an incomitant esotropia worse in right gaze. She was diagnosed with a subtle microvascular right sixth nerve palsy. Two weeks later, she developed intractable bifrontal headaches and photosensitivity. On neuro-ophthalmic evaluation she had bilateral limitation of abduction, left greater than right, and slight limitation of elevation of both eyes. A head CT with enhancement demonstrated slightly enlarged extraocular muscles and crowding of the orbital apex bilaterally. Bloodwork was significant for a slightly elevated WBC count with a longstanding microcytic anemia (Hb 10.4) and thrombocytosis. Thyroid function studies and myasthenia gravis work-up was normal. However, antithyroglobulin antibody was elevated at 59 (N<2). Thyroid orbitopathy was considered a probable diagnosis at this time. However, the patient's complaint of relentless headaches prompted persuing an MRI brain and orbits with contrast that demonstrated diffuse pachymeningeal enhancement, abnormal enhancing soft tissues in the orbital apices bilaterally particularly surrounding the intercanalicular segments of the optic nerves and enlarged enhancing extraocular muscles. Lumbar puncture demonstrated an opening pressure of 24 cm H20 with normal constituents. Blood ACE, lysozyme, RPR, VDRL, ANCAs, complement level and inflammatory markers were unremarkable. The patient improved significantly upon solumedrol although diagnosis was unclear. An immunoglobulin and IgG4 level was ordered and the serum IgG4 level was significantly elevated. A pachymeningeal/leptomeningeal biopsy demonstrated multifocal lymphoplasmacytic infiltrate containing elevated numbers of IgG4 plasma cells in keeping with IgG4 related disease hypertrophic pachymeningitis. The patient responded significantly to solumedrol with resolution of her headache and diplopia and is currently on Rituximab therapy.

Methods:

Case report.

Results:

This patient suffered from IgG4 related hypertrophic pachymeningitis and orbital disease. Her restriction of ductions, extraocular muscle enlargement and elevated antithyroglobulin antibody were most in keeping with thyroid orbitopathy. However, the patient's major complaint of severe headaches and atypical disease course raised suspicion for a more central etiology. Hypertrophic pachymeningitis was first described by Charcot and Joffrey as a condition in which "the neighboring leptomeninges always suffer as well becoming opaque and thick."[i] The disorder can present in a myriad of ways including chronic headache, hypopituitarism, cranial nerve palsies, papilledema, cerebellar ataxia and motor or sensory compromise. It can result from infectious, inflammatory and infiltrative disorders and at times may be idiopathic.[ii][iii] Recognition of IgG4 related disease (IgG4RD) occurred in 2001 in sclerosing autoimmune pancreatitis.[iv] Since then, the spectrum of the disease has significantly expanded to include involvement of multiple organ systems. Effects on the central nervous system by IgG4 occur secondary to hypertrophic pachymeningitis, hypophysitis and infiltration of cranial nerves. The clinical presentation of IgG4RD hypertrophic pachymeningitis is similar to other etiologies pachymeningitis; however, involvement of other organs may provide clues to the diagnosis. In our case, the presence of diffuse pachymeningeal enhancement in addition to diffuse enlargement of the extraocular muscles and orbital soft tissue involvement led to the consideration of this disease in the differential which was later confirmed with pathologic diagnosis.

Conclusions:

The patient's inner ear implant limited initial neuro-imaging to a CT scan that demonstrated enlargement of the extraocular muscles with an elevated antithyroglobulin antibody, which many would consider compatible with thyroid orbitopathy. However the history of the patient's relentless headache did not fit the clinical picture and prompted ordering of an MRI demonstrating diffuse hypertrophic pachymeningitis. The orbital and pachymeningeal findings ultimately led to the consideration of a number of diagnoses, most importantly IgG4 disease. Final Diagnosis: IgG4 related disease presenting as hypertrophic pachymeningitis with orbital involvement
References: None.

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[iv] Hamano H, Kawa S, Horiuchi A, Unno H, Furuya N et al. High serum IgG4 concentrations in patients with sclerosing pancreatitis. *N Engl J Med*. 344(10):732-738. 2001

Keywords: None.

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Poster 94

Resolution of Sixth Nerve Palsies Following Treatment with Primary Fractionated Stereotactic Conformal Radiotherapy of Petroclival Meningiomas

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Introduction:
Petroclival meningiomas are a recognised cause of sixth nerve palsy. Despite their benign pathology, petroclival meingiomas remain a surgical challenge due to the complex anatomy, close association with critical neurovascular structures and difficult exposure of this region.Fractionated Stereotactic Conformal Radiotherapy (FSCRT) is a process whereby radiotherapy is delivered via a fractionated schedule utilizing nonrigid immobilization, thereby providing the precision of stereotaxy while allowing adjacent normal structures to repair sublethal damage.To our knowledge there are no studies on the role of FSCRT on petroclival meningiomas.

Methods:
A review of the case notes of three patients presenting with unilateral (n=2) and bilateral (n=1) sixth nerve palsies secondary to radiologically diagnosed petroclival meningiomas treated with primary FSCRT, was undertaken.

Results:
All three patients were treated with FSCRT and received 50Gy in 30 fractions with 6MV Photons over a period of six weeks. MRI scans were performed at diagnosis, 3 months post treatment and annually thereafter. All three patients achieved a marked symptomatic and clinical improvement following treatment and this persisted throughout the five year follow-up period, obviating the need for Fresnel prisms. The mean percentage reduction in the distance angle of the esotropia was 99% (range 97%-100%). None of the patients experienced side-effects related to the treatment.

Conclusions:
Stereotactic radiosurgery (SRS) is considered a good first-line treatment for small tumours. FSCRT, however, is helpful in cases in which SRS has limitations, such as with tumours arising near critical neurovascular structures. The results of FSCRT in skull base tumours, with respect to tumour control and improvement of neurological deficits have been comparable to SRS.FSCRT shows promising results for the management of symptomatic sixth nerve palsies secondary to petroclival meningiomas and should be offered as first line therapy.

References: None.

Keywords: Adult Strabismus with a Focus on Diplopia, Ocular Motility, Neuroimaging, Tumors

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Poster 95
One-and-a-Half Syndrome Evolving Into an Eight-and-a-Half Syndrome

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Introduction:
One-and-a-half syndrome was first coined by C. Miller Fisher in 1967 to describe a combination of horizontal gaze palsy and internuclear opthalmoplegia caused by a lesion in the median longitudinal fasciculus (MLF) or parapontine reticular formation (PPRF).[1] The constellation of one-and-a-half syndrome plus fascicular cranial nerve VII involvement was referred to as eight-and-a-half syndrome by Eggenberger in 1998.[2] In this case, we describe a case one-and-a-half syndrome with a delayed onset of cranial nerve VII involvement.

Methods:
62 year old gentleman with a history of hypertension, hyperlipidemia, COPD, aortic insufficiency, current smoker, and history of alcohol abuse presented with new onset binocular, horizontal diplopia. On presentation, he had a right sided horizontal gaze palsy and a right sided INO. His seventh nerve was fully intact bilaterally. Lumbar puncture, CT head, as well as MRI brain and brainstem were all unremarkable. On follow-up examination 48 hours later, he was found to have developed a right sided LMN seventh nerve palsy, as well as exposure keratopathy. He subsequently underwent a temporary tarsorrhaphy. The patient was seen at follow-up 7 weeks later with resolution of his presenting signs and symptoms.

Conclusions:
One-and-a-half syndrome and eight-and-a-half syndrome differ by whether cranial nerve 7 is involved. This differentiation is paramount for many reasons, including prevention of exposure keratopathy. Here we present a unique case of delayed onset seventh nerve involvement in a patient with one-and-a-half syndrome caused by a small brainstem infarct, highlighting the importance of close follow-up of one-and-a-half syndrome.

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2. Eggenberger E,” Eight-and-a-half syndrome: one-and-a-half syndrome plus cranial nerve VII palsy.” J Neuroophthalmol. 1998;18(2):114-116

Keywords: One-And-A-Half Syndrome, Eight-And-A-Half Syndrome, Internuclear Ophthalmoplegia, Horizontal Gaze Palsy, Ocular Motility

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Poster 96
Brown Syndrome: An Uncommon Cause of Diplopia Following Levator Advancement

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Introduction:
Brown Syndrome, the inability to elevate an eye in abduction due to restriction of the superior oblique muscle tendon, can present as a congenital or acquired abnormality. Although uncommon, Brown Syndrome has been described following blepharoplasty.[1, 2] In this report, we describe a patient who developed Brown Syndrome following levator advancement surgery for blepharoptosis.

Methods:
Observational case report.

Results:
Case description: A 65 year old female presented in January of 2015 for assessment of diplopia. This was first detected in March 2014, two weeks following otherwise uncomplicated levator advancement surgery. The patient stated that the primary intent of the surgery was to elevate the eyelids and was unaware of fat being excised. The operative report was not available for review. Symptoms were described as vertical binocular diplopia when looking to the right. Her evaluation was normal with the exception of well-healed eyelid crease incisions and a 7 prism diopter right hypertropia in right gaze, which increased with supraduction, and decreased with infraduction. She was orthorphoric in primary and all other directions of gaze, consistent with Brown Syndrome. One and half years following surgery her evaluation remained unchanged. She was minimally symptomatic and no intervention was pursued.

Conclusions:
Brown Syndrome may result from injury to the superior oblique tendon-trochlea complex during levator advancement surgery.

References:
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2) Wesley, Pollard, McCord Jr. Superior oblique paresis after blepharoplasty, Plast Reconstr Surg, 66:283-6, 1980.

Keywords: Brown Syndrome, Blepharoptosis, Double Vision

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Poster 97
Long Term Follow Up of Children With Downgaze Nystagmus at Infancy

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Introduction:
Paroxysmal downgaze nystagmus of infant is an unsettling phenomenon that leads to extensive work-up, although benign outcomes have been reported in sporadic cases. However, long term outcome of these children was not published. Our aim is to report a case series of 7 healthy infants with long neurological and ophthalmological follow up. They all presented with acute onset of episodic paroxysmal tonic downgaze at the ages of 2-12 weeks of age. We describe their spontaneous resolution without neurologic or ophthalmological sequelae.

Methods:
A clinical retrospective case series from two tertiary pediatric ophthalmology centers.

Results:
Six of the seven infants included were born at term (39-41 weeks) with normal pregnancies and deliveries, and only one preterm birth (34th week).All children presented with tonic downgaze nystagmus, with increasing frequency per day, and subsequent gradual decrease until complete resolution within a maximum of 8 months. Each episode lasted a few seconds. In five children symptoms occurred while lying supine. Three infants had concurrent abnormal body movements. No other neurologic or ophthalmic pathologies were found.All infants underwent work-up to exclude metabolic, neurologic and anatomic etiologies. Diagnostic studies included EEG (5 cases), MRI (3 cases), brain US (3 cases), urinary VMA and HMA (5 cases). All tests were negative.The children were followed for up to 10 years (median 2.0 years). Six infants followed normal developmental steps whereas one infant had asymmetric development of motor skills which improved with time.

Conclusions:
Parents and clinicians are usually concerned when paroxysmal tonic downgaze occurs in normal infants born after uneventful delivery. In line with previous reports of sporadic cases, we found tonic downgaze nystagmus of infants to be a benign syndrome that occurs in healthy infants as part of the maturation of the visual system. We describe a good long term outcome. This phenomena may not require a comprehensive clinical investigation.

References: None.

Keywords: Nystagmus, Ocular Motility, Infants, Pediatric Neuro-Ophthalmology

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Poster 98

Complete Bilateral Ophthalmoplegia in the Setting of Elevated Intracranial Pressure that Improved with Transverse Venous Sinus Stenting

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Introduction:
Elevated ICP is known to cause vision loss, abducens nerve palsy, and rarely oculomotor and trochlear nerve palsies [1,2]. Severe dysmotility of both eyes in all gazes has not been reported. Venous sinus stenosis has been postulated as a possible pathophysiologic mechanism underlying elevated ICP in the setting of idiopathic intracranial hypertension, and stenting of the transverse sinus has been therapeutic [3,4]. The current report describes resolution of complete ophthalmoplegia in the setting of secondary elevated ICP treated with transverse venous sinus stenting.

Methods:
Case report and literature review.

Results:
A 32-year-old woman presented after sudden onset of profound vision loss and complete ophthalmoplegia in both eyes in all directions. Neurological exam was unremarkable, but visual acuity was light perception with pallid optic disc edema bilaterally. Brain MRI showed optic nerve edema, and MRV revealed severe stenosis of the right transverse sinus. CSF opening pressure was > 50 cm H2O. Extensive serum and CSF laboratory studies were all unremarkable. Conventional angiogram with venography and transverse sinus manometry showed venous pressure of 40 - 70 mmHg in the proximal transverse sinus and < 20 mmHg distal to the focal stenotic region. Pressures normalized to 21 - 26 mmHg following transverse sinus stenting, and repeat lumbar puncture showed an opening pressure of 14 cm H2O. The patient reported significant improvement in her headaches and also showed improvement in her ocular motility, although her vision remained poor at hand motion bilaterally.

Conclusions:
This is a unique case of EIP presenting with complete ophthalmoplegia in addition to vision loss. The patient's findings of transverse sinus stenosis and subsequent improvement after stenting are consistent with retrospective reports in the IIH literature, and suggest that venous sinus stenting may be a viable treatment approach in other forms of elevated ICP.

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Keywords: High Intracranial Pressure/Headache, Ocular Motility, Interventional Neuroradiology

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Grant Support: None.

Poster 99

Clinicopathologic Correlation in Lateral Medullary Stroke with Transient Ocular Motor Findings Mimicking Peripheral Vestibular Dysfunction

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Introduction:
Unilaterally decreased horizontal vestibulo-ocular reflex (h-VOR) gain occurs with peripheral vestibular lesions or lateral pontine strokes involving the fascicle or the medial vestibular nucleus (MVN). Vestibular reflexes are typically normal in (LMS). We describe clinicopathologic findings in an unusual LMS presenting transient unidirectional nystagmus and decreased h-VOR gain, mimicking peripheral vestibulopathy (PV). The MVN neurons, close to the stroke core (ischemic penumbra), appeared pathologically normal, suggesting that the initial vestibular signs related to transient ischemia.

Methods:
Clinicopathologic examination of a 61 year-old man admitted to ICU with an acute vestibular syndrome accompanied by aphonia and respiratory distress. He had right-beating nystagmus obeying Alexander's law, an abnormal leftward head impulse test (HIT) confirmed quantitatively by video-oculography, and no skew deviation. He had severe axial lateropulsion and Horner's syndrome. MRI showed a left LMS extending rostrally to the ponto-medullary junction and dorsally to near the MVN. The nystagmus subsided within two days and the horizontal HIT normalized clinically. Three weeks after discharge to rehabilitation, he died of sudden cardio-respiratory arrest. After autopsy, brainstem sections were stained with Luxol Fast Blue, H&E and neurofilament. To explain the ocular motor and vestibular findings, we hypothesized that the MVN was involved pathologically.

Results:
Neuropathological examination showed a left LMS whose extent matched that seen by imaging. Non-ocular motor signs correlated well with structures affected by the infarction. The nearby MVN, however, was spared. The left vertebral artery was occluded and the right was hypoplastic and 50% stenotic.

Conclusions:
We hypothesize that the ischemic penumbra of the LMS may have involved the MVN in the rostral medulla, leading to transiently abnormal h-VOR and unidirectional nystagmus. This is the first case of quantitatively proven abnormal horizontal-HIT during transient brainstem ischemia without pathologic infarction.

References: None.

Keywords: Nystagmus, Ocular Manifesttaions of Systemic Disorders, Stroke

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Poster 100
Chronic Progressive External Ophthalmoplegia in the Absence of Ptosis

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Introduction:
Chronic progressive external ophthalmoplegia (CPEO) is a manifestation of various disorders sharing a common pathophysiology of mitochondrial dysfunction leading to progressive extraocular myopathy. It may occur in isolation during adulthood or as part of a multi-organ mitochondrial cytopathy. Ptosis, presenting simultaneously with or preceding ophthalmoplegia, is usually the earliest clinical feature of CPEO.

Methods:
We present two cases of biopsy supported CPEO presenting with diffuse bilateral ophthalmoparesis in the absence of ptosis.

Results:
Case 1 was a 64-year-old Caucasian female who presented with intermittent binocular diplopia for five years. She had mild to moderate diffuse ophthalmoplegia bilaterally and a slightly incomitant 1-4 prism diopter intermittent esotropia. Superior eyelid margin-to-reflex distance (MRD1) measurements were 5 mm bilaterally. Quadriceps muscle biopsy showed scattered cytochrome oxidase (COX) negative fibers and moderate reductions in complex I and III enzymatic activities (25 and 27% of normal, respectively). Case 2 was a 53 –year-old Caucasian female with intermittent diplopia for eleven years. Her exam showed bilateral moderate diffuse ophthalmoplegia and a comitant 2-3 prism diopter intermittent esotropia. MRD1 measurements were 3.5 mm and 3 mm. Deltoid biopsy showed numerous fibers with increased succinate dehydrogenase and reduced COX staining. Alternative causes of ophthalmoplegia were excluded in both patients with lab testing, serial exams, and neuro-imaging.

Conclusions:
Clinical heterogeneity is a hallmark feature of mitochondrial diseases; CPEO without ptosis has been reported previously and likely represents one variant along the spectrum of atypical progressive external ophthalmoplegia phenotypes.[1,2] Timely diagnosis of CPEO with early detection of concurrent systemic disease is important, particularly given its association with cardiomyopathy and cardiac arrhythmias.[3] While CPEO almost always presents with myopathic ptosis at the time of diagnosis, clinicians should be cognizant of exceptions to the rule.

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Keywords: Chronic Progressive External Ophthalmoplegia, Mitochondrial Myopathy

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 101

Torsional Nystagmus Associated with Palatal Tremor in Vertebral Artery Dolichoectasia

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Introduction:

A 50 year-old woman presented with three-month history of oscillopsia. This accompanied with gait disturbance. Her previous medical history was significant for right-side hemiparesis for the last year, DM type2 and essential hypertension. She denied any history of drug abuse.On examination, BCVA was 20/20 in both eyes. Anterior segment and fundus exam were unremarkable. Binocular conjugated pure counter-clockwise torsional nystagmus synchronizing with palatal tremor was observed. The amplitude and frequency of nystagmus were similar in all gazes. Null point was absent. Ocular alignment was orthotropic. Extraocular muscles function, saccadic velocity and smooth pursuit eye movement were all within normal limit. Neurological examination showed right-sided hemiparesis and hyper-reflexia. Celebellar functions were impaired on the right-side including wide-base gaits, dysdiadokonesia and impaired Finger-to-Nose test. Thin slice axial T2 weighted MRI with fat suppression and Apparent Diffusion Coefficient(ADC) images show dolichoectatic left vertebral artery, exerting pressure effect to the left medulla. A hypersignal intensity T2 change with increased diffusion on the ADC image at the left medulla is also depicted. 3D Time of Flight (TOF) MRA of the posterior circulation reveals dolichoectatic left vertebral artery with redundancy to the right. Hypoplasia of the right vertebral artery is noted. To our knowledge, this is the first case of torsional nystagmus with palatal tremor in vertebral artery dolichoectasia.

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Keywords: Nystagmus, Ocular Manifestations of Vestibular Disorders, Ocular Motility, Vascular Disorders

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 102

Impaired Rightward Saccades with Preserved Rightward Pursuit Following Right Pontine Ischemic Stroke

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Introduction:
The pontine paramedian reticular formation (PPRF) is the brainstem saccadic generator. Pursuit axons pass through the PPRF but do not synapse there. In macaques, injection into the PPRF of neuronal toxins that spare axons selectively paralyzes saccades[1]. In humans, selective impairment of horizontal and vertical saccades with preserved horizontal pursuit and vestibulo-ocular reflex has been described following cardiac surgery with or without documented systemic hypotension[2, 3]. Imaging has been negative except for one case in which focal FLAIR hyperintensity appeared in the dorsomedial pons unilaterally 10 months after the event. In one autopsied case[2], there was neuronal necrosis, axonal loss, and astrocytosis in the pontine paramedian reticular formation bilaterally. Ocular motor nuclei and cerebral gaze-generating centers were histologically normal. In a second case[3], necropsy pathology was negative, suggesting submicroscopic damage to PPRF inputs as the explanation for saccadic paralysis. We present a patient who developed a selective right horizontal saccadic palsy in conjunction with ipsilateral dorsomedial pontine infarction in the region of the PPRF.

Methods:
Case Report

Results:
A 53 year-old woman had sudden imbalance and inability "to look to my right." Neurologic examination disclosed left upper extremity and gait ataxia. One day later, our first examination revealed slow and incomplete rightward saccades initiated by blinking. Pursuit was normal in all directions of gaze, as were leftward and vertical saccades. (VIDEO). Diffusion-weighted brain MRI showed restricted diffusion in the right dorsomedial pons and extensive cerebral white matter T2/FLAIR hyperintensity.

Conclusions:
This is the first case to show selective impairment of unilateral horizontal saccades with restricted diffusion in the region of the ipsilateral PPRF. It supports animal evidence that selective impairment of horizontal saccades may occur from a discrete pontine lesion. It is a reminder that complete clinical assessment of eye movements must extend beyond having the patient pursue your finger!

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Keywords: Ocular Motility, Neuroimaging

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Grant Support: None.

Poster 103

Complete Ophthalmoplegia and Extraocular Muscle Atrophy in a Patient with Craniofacial Scleroderma with CNS Involvement

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Introduction:

Craniofacial scleroderma includes a spectrum of disease from linear scleroderma ('*en coup de sabre*') to progressive hemifacial atrophy (Parry-Romberg syndrome). It is an uncommon cause of external ophthalmoplegia as a result of atrophic myopathy, the pathophysiology of which is incompletely understood. In addition, central nervous system changes are a not uncommon finding in this spectrum of disease.

Methods:

We report a case exhibiting extreme findings and a review of the literature.

Results:

We present a 61-year-old Caucasian woman with chronic external ophthalmoplegia of her right eye greater than her left, which was first noted at age 12, with the onset of diplopia. This gradually progressed, twice requiring strabismus surgery. Abnormalities on an unrelated brain MRI and subsequent LP revealing oligoclonal bands led to a diagnosis of multiple sclerosis. There is no history of focal neurologic deficits but a questionable seizure history. On examination her afferent visual function was normal except for a mild superior visual field defect OS on automated perimetry. Motility of the right eye was severely limited in all gazes. The left eye showed markedly limited supraduction and adduction but was otherwise relatively full. Dilated funduscopic examination revealed optic atrophy and enlarged cup-to-disc ratios. She had a linear indentation along her right fronto-parietal region including bone and overlying soft tissues without alopecia or significant fibrotic skin changes. MRI of brain and orbits showed multiple T2 and FLAIR white matter hyperintensities and severe atrophy of all right eye extraocular muscles and left eye medial rectus.

Conclusions:

This case highlights an unusual presentation of craniofacial scleroderma. It is atypical in both its severity of ophthalmoplegia and the contralateral involvement of both extraocular muscle atrophy and CNS changes. In addition, it underscores the need to consider craniofacial scleroderma syndromes in the differential of multiple sclerosis in atypical patients.

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Keywords: External Ophthalmoplegia, Craniofacial Scleroderma

Financial Disclosures: The authors had no disclosures.

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Poster 104
Preservation of Reading in a Patient with Acquired Impairment of Saccadic Function

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Introduction:
Saccadic disturbance after cardiac surgery is a well-recognized but a poorly understood phenomenon. Historical notions suggest that absence of saccadic function, at least in the horizontal plane, should significantly compromise the ability to read. We present a case of a 49-year-old man with clinically obvious loss of saccadic function after surgical repair of aortic dissection. Upon questioning, the patient reported no difficulty at work where he had to do considerable reading. This apparent discrepancy was analyzed with quantitative eye movement recordings.

Methods:
A case report with analysis of the clinical features and quantitative measures of eye movements that were obtained using Neuro Kinetics, Inc. I-Portal®-Neuro-Otologic Test Center. The test apparatus consisted of a rotatory chair, a visual stimulus, and a digital eye tracking system. We used a standard battery of tests and added one test to measure horizontal eye movements while reading text.

Results:
The clinical exam revealed essentially complete absence of volitional saccades in both the horizontal and vertical planes. Eye movement recordings showed marked slowing and hypometria of all voluntary saccades in both horizontal and vertical planes. His optokinetic quick phases of nystagmus were absent, while the optokinetic ocular pursuit reflex was normal. The horizontal and vertical sinusoidal pursuit was normal. When reading, the patient was able to generate a series of quite small, hypometric saccades without compensatory blinks and head trusts. These hypometric saccades also replaced the large-amplitude leftward return saccade that normal subjects make at the end of each line to re-orient the eyes to the beginning of the next line.

Conclusions:
The syndrome of saccadic palsy after cardiac surgery includes a spectrum of saccadic abnormalities. This case is unique in that the patient reported normal ability to read, and quantitative recordings documented small-amplitude saccades that generally sufficed for reading.

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Solomon D, Ramat S, Tomsak RL, Reich SG, Shin RK, et al. Saccadic palsy after cardiac surgery: characteristics and pathogenesis. Ann Neurol 63:355-65 (2008).

Keywords: Saccades, Reading, Cardiac Surgery

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Grant Support: None.

Poster 105

The Clinical Overlap of Long-Standing Optic Neuropathy and Retinal Arterial Occlusion – The Value of Optic Coherence Tomography to Establish the Anatomical Diagnosis and to Guide Management

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Introduction:
In the acute phase of blindness, the clinical distinction between an optic neuropathy and retinal arterial occlusion is straightforward. In the former, there is often edema of the optic nerve head, especially in non-arteritic anterior ischemic optic neuropathy (NAION). With occlusion of either the central or branch retinal arteries, there is usually edema of the respective area of the retina. After the acute phase, however, the clinical distinction between an old optic neuropathy and an old vascular retinopathy can be challenging given that optic nerve pallor can be present in either case. Establishing the correct anatomical diagnosis is critical because the management and systemic implications of these two conditions are quite disparate.

Methods:
We describe a case series of three patients whose initial clinical presentations were thought to be consistent with NAION. None of the patients had edema of the retina or optic nerve in the acute phase. Optic coherence tomography (OCT) was used to assess the thickness of the inner 2/3s of the retina to search for evidence of inner retinal ischemia.

Results:
All patients had reduced acuity, an afferent pupillary defect and optic nerve head pallor in the affected eye. In all three cases OCT of the retina demonstrated significant reduction of the thickness of the inner two thirds of the retina. This finding motivated a different approach to management.

Conclusions:
After the acute phase, it can be difficult to distinguish retinal from optic nerve ischemic, and one might be misled by the finding of optic nerve pallor. OCT of the retina can be used to identify evidence in support of a diagnosis of retinal ischemia. In such a case, a search for an embolic source should generally be performed. In the absence of such information, the diagnosis might be mistakenly assumed to be an optic neuropathy.

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Keywords: Retinal Artery Occlusion, Non-Arteritic Ischemic Optic Neuritis, OCT

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Grant Support: None.

Poster 106
MALT Lymphoma Masquerading as Thyroid Orbitopathy

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Introduction:
Clinical presentation appeared c/w compressive optic neuropathy secondary to thyroid eye disease. However imaging revealed a different story.

Methods:
Case Report

Results:
67 y/o WF referred for disc edema OS, complaining of distortion OS x 1 year. No headaches. PMH: hypertension and hypothyroidism. SH: 40 pack-years of smoking. Exam revealed 20/20 and 20/25. Visual fields were FTC, and color vision was normal. Pupils: 1+ reaction OU, and no RAPD. EOM revealed mild limitations. Exam demonstrated RLL and LLL edema, bilateral brow ptosis, bilateral Queen Anne sign, and lid retraction with an MRD of 6 mm OU. Hertel was 20 and 22 with a base of 97. DFE OS revealed 2+ disc edema. TED with compressive optic neuropathy OS was suspected. CT revealed bilateral apical orbital soft tissue masses with widening of the superior orbital fissures bilaterally and extension on the left side into the left pterygopalatine fossa and along branches of the left trigeminal nerve. MRI further characterized the masses limited to the posterior third of the orbits R>L, bilateral infiltration of the adjacent tendons of the EOMs with infiltration into the muscle bellies on the right, and extension along the left petrous ridge and left free edge of the tentorium. Left upper cervical adenopathy was identified by imaging and then biopsied by ENT. Results revealed a low-grade marginal zone lymphoma (MALToma) composed of CD20+ B-cells with abnormal co-expression of BCL2. Concurrent flow cytometry identified a population of CD5-/CD10- kappa light chain restricted B-cell population. CT chest, abdomen, and pelvis was unremarkable.

Conclusions:
Low-grade extranodal marginal zone lymphoma of mucosa-associated lymphoid tissue (MALT Lymphoma) was found occupying the posterior orbits bilaterally with widening of the superior orbital fissures, and expansion of cavernous sinus. Additionally, a cervical lymph node was luckily identified on CT. This finding obviated the need for a more difficult and risky posterior orbital biopsy or needle biopsy of the pterygopalatine fossa.

References: None.

Keywords: Lymphoma, MALT, Orbital Mass

Financial Disclosures: The authors had no disclosures.

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Poster 107

Back to Basics

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Introduction:

A 71 year-old female with an unremarkable medical history was referred for evaluation of progressive painless vision loss in her left eye. An outside corneal specialist had diagnosed Fuch's corneal dystrophy. She had undergone cataract extraction OS, DSEK OS, and then repeat DSEK OS after the initial graft failed. However, the patient's vision had not improved following the surgery. Rather, her vision had worsened. On examination, visual acuity was 20/30 OD and HM OS. External examination showed 4 mm proptosis OS and ptosis OS. Motility was full OU. There was a 2.8 log unit RAPD OS. Anterior segment examination showed changes consistent with Fuch's corneal dystrophy, an endothelial graft OS, and a partially dislocated posterior chamber intraocular lens OS. Dilated funduscopic examination showed mild temporal optic disc pallor OD and moderate optic disc pallor OS. Goldmann visual field testing showed an inferior arcuate defect with generalized depression OD and a small nasal island of vision OS. MRI orbits with contrast showed bilateral enhancing inferior orbital soft tissue masses involving both inferior recti and infraorbital nerves, extending into both cavernous sinuses. Although there was a broad differential diagnosis, the MRI findings were felt to be most consistent with orbital inflammatory disease, possibly IgG4-related disease. A laboratory work-up was obtained, including CBC, basic metabolic panel, ESR, CRP, ANCA, and syphilis serology. Most of the studies were unrevealing, but the ESR was elevated at 38 mm/hr. A diagnostic test was then performed.

Methods:

Case Report

Results:

Evaluation of serum IgG subclasses revealed an elevated serum IgG4 at 230 mg/dL (normal range: 7-89 mg/dL). Although the clinical picture was suggestive of IgG4-related disease, a left anterior orbitotomy was performed and the inferior orbital soft tissue mass was biopsied. On histopathology, the mass consisted of collagenous tissue containing infiltrative islands of atypical epithelial cells. There were focal areas of necrosis. In some areas, the tissue was noted to be forming tubules and ductules that were lined with goblet cells; these were highlighted on periodic acid-Schiff (PAS) and mucicarmine staining. Immunohistochemistry was positive for pancytokeratin, epithelial membrane antigen, and p63 in all tumor cells. Overall, these findings were consistent with adenosquamous carcinoma, likely metastatic with features to suggest an upper respiratory tract origin. CT chest/abdomen/pelvis with contrast and mammogram were unrevealing. A PET scan showed intense activity within the superior margin of the right nasopharynx, consistent with an upper respiratory tract origin. The final diagnosis was adenosquamous carcinoma arising from the upper respiratory tract with perineural spread into the orbits and cavernous sinuses. The patient went on to receive radiation therapy with adjuvant chemotherapy, since the extent of disease precluded primary surgical resection.

MRI orbits showed changes suggesting orbital inflammation extending into the cavernous sinuses, possibly due to IgG4-related disease. Furthermore, the serum IgG4 level was significantly elevated. However, tissue biopsy revealed adenosquamous carcinoma and further imaging with PET suggested an upper respiratory tract primary.

Conclusion: Primary adenosquamous carcinoma of upper respiratory tract with perineural spread into the orbits and cavernous sinuses.

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Keywords: None.

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Poster 108

The Stuck Eyelid Syndrome: A Benign Phenomenon of Brief Intermittent Unilateral Ptosis in Children

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Introduction:
Several children have been observed at our institution with a stereotypic intermittent unilateral ptosis, with one eyelid appearing stuck and fluttering in a ptotic position, for several seconds at a time, followed by complete resolution. No etiology was identified despite detailed evaluation. We present and identify the unifying clinical features of this phenomenon.

Methods:
This is a retrospective case series using records of children seen between 1994 and 2015. The cases were identified by a search for "stuck" and "eyelid." Patient charts were reviewed, then a phone follow–up in each case inquired about outcome.

Results:
Four (4) children, presenting between 18 months and 7 years of age, were identified with a very similar presentation of acquired, brief, intermittent episodes of unilateral ptosis during which the eyelid appeared to be stuck and faintly fluttering for several seconds. The reported ages of onset ranged from soon after birth to 7 years. The work-up for a cause of the ptosis, including acetylcholine receptor antibody testing, was negative for an underlying etiology in each case. Follow up ranged from 7 months to 5 years; one child had a complete resolution of symptoms at 12 months.

Conclusions:
The "stuck eyelid syndrome" appears to be a distinct, clinical entity of benign acquired unilateral intermittent ptosis in children. While the etiology is unclear, we speculate that it may be a type of tic involving the tarsal portion of the orbicularis oculi muscle.

References: None.

Keywords: Eyelid & Adnexal Disease, Pediatric Neuro-Ophthalmology

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 109
Anesthesia Dolorosa of Trigeminal Nerve as an Early Presentation of Orbital Tumor

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Introduction:
Anesthesia dolorosa is a syndrome of numbness and pain in the same sensory distribution. This combination of symptoms is thought to be caused by an injury to the first order neurons with spontaneous firing of the second order neurons. It has been described in association of injuries related to surgery or radiation, but may present as an early sign of cancer.

Methods:
A 90 year-old woman with history of squamous cell skin cancer presented with new face pain and scalp tenderness. The initial examination showed decreased sensation in the V1 distribution the left trigeminal nerve. Giant-cell arteritis was ruled out and an MRI/MR angiogram showed evidence of a vascular loop at the left trigemnial nerve. The patient did not tolerate medical therapy for pain, and attempts at nerve blockade didn't provide relief. Six months later she underwent an uncomplicated cataract surgery in the left eye with initial improvement in vision. One month later, at a follow-up visit she noted spread of her left face pain. There was a new esotro-deviation, with hand motion-only vision and a new APD in the left eye. At urgent neuro-ophthalmic consultation, there was immediate recognition of the ominous history, and follow-on imaging revealed the lesion.

Results:
This is a case of trigeminal neuralgia, with features of anesthesia dolorosa and an initially negative MRI/MRA. After progression of symptoms to include a larger sensory distribution, optic neuropathy and sixth nerve palsy repeat imaging showed a lesion. The tumor progressed from an undetectable lesion on initial MRI to a lesion involving the posterior orbit, cavernous sinus and Meckel's cave on the left. Given history of skin cancer concern was raised for neurocutaneous spread.

Conclusions:
Pain and numbness, "anesthesia dolorosa," is a rare but important sign suggestive of a potential infiltrative lesion, particularly in the absence of history suggesting an alternate mechanism.

Keywords: Anesthesia Dolorosa, Trigeminal Neuralgia, Orbital Tumors

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 110

Facial Venectasia 50 Years After Internal Carotid Artery Ligation for Traumatic Cavernous Sinus Fistula

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Introduction:
CCA (Common Carotid Artery) ligation was a commonplace procedure throughout the 19th century and was first used in CCSF (Carotid-Cavernous Sinus Fistula) in 1809 after it became evident that carotid artery compression reduced pulsating exophthalmos. The first description of ICA (Internal Carotid Artery) ligation for CCSF followed in 1904. Carotid artery ligation continued to be the mainstay of treatment until the introduction of balloon occlusion of the fistula in the early 1970s. While often effective in reducing symptoms and preventing the most severe complications, ICA ligation has usually not been successful in eliminating all shunt related clinical manifestations.

Methods:
Review of clinical history, neuro-imaging and relevant literature.

Results:
In 1953, a then 17 year old man had suffered cranial trauma complicated by a high flow CCSF. Surgical ligation of the ICA was performed but could not prevent the development of chronic exophthalmos and secondary glaucoma with progressive loss of vision and blindness. Over time the patient developed mild varicosities in the ipsilateral face illustrating the extent of facial venous drainage through the cavernous sinus. The patient's CT scan of the head obtained 50 years after the procedure demonstrates bony erosion in the sellar region apparently due to dilated vascular channels and shunting of blood from the posterior circulation into the cavernous sinus.

Conclusions:
This case demonstrates very long term extracranial and intracranial sequelae of a traumatic CCSF treated with ICA ligation. It also reveals the field of facial venous drainage via the cavernous sinus.

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Ringer AJ[1], Salud L, Tomsick TA, Carotid cavernous fistulas: anatomy, classification, and treatment, Neurosurg Clin N Am, 16 #, 279-95 #, 2005.

Keywords: Orbit/Ocular Pathology, Trauma, Vascular Disorders

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 111
Simbu Ptosis: Drooping Lids in the Highlands of Papua New Guinea

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Introduction:
This poster focuses on identification, description and treatment of a previously undescribed myogenic ptosis found in the Eastern Highlands of Papua New Guinea.

Methods:
During the course of a 10 day surgical camp, in collaboration with local center for inclusive education 97 patients were collected from the Simbu Province and brought to a rural hospital in the Eastern Highlands of Papua New Guinea. Three oculoplastic surgeons and one neuro-ophthalmologist were involved in careful identification, diagnosis and treatment of the patients' progressive bilateral ptosis. Affected patients were aged 9 to 75 and presented with varying degrees of ophthalmoplegia, dysphagia, dysphonia and orbicularis oris weakness. These patients were systematically examined and determined to be "mild", "moderate" and "severe" in the stage of ptosis and systemic findings. The moderate patients all underwent frontalis suspension (n=31), and high risk patients were provided ptosis crutches (n=13) to assist with clearing the visual angle for activities of daily living.

Results:
In total, 97 patients presented for evaluation. Thirtyone underwent bilateral frontalis suspension, 4 received unilateral frontalis slings. 13 patients were fitted with ptosis crutch glasses.

Conclusions:
High volume surgical camps are possible in remote areas. Work is ongoing in characterizing the genetic inheritance pattern of this ptosis.

References: None.

Keywords: Myogenic Ptosis, High Volume Intervention, Frontalis Suspension, Ptosis Crutch

Financial Disclosures: The authors had no disclosures.

Grant Support: Moran Eye Center and Himalayan Cataract Project gave grants for travel to remote area as well as provided instruments for the surgical camp.

Poster 112

Vision Loss Due to Gas Gangrene of Orbit Secondary to Clostridium bifermentans and Enterobacter Aerogenes

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Introduction:
We describe a case of orbital cellulitis in which the causative organisms (*Clostridium bifermentans* and *Enterobacter aerogenes*) are gas producing bacteria.

Methods:
Culture analysis along with CT imaging confirmed orbital cellulitis in a 25 year old otherwise healthy male presenting with hand motion acuity, intraocular pressure of 62 mmHg, and proptosis of the left eye. The patient was initially treated with broad-spectrum IV antibiotics. Intraocular pressure was controlled with timolol, dorzolamide, and latanoprost The patient then underwent endoscopic sinus tissue removal and drainage as well as direct drainage of the orbital abscess through blepharoplasty incision.

Results:
CT scan revealed gas within the left orbit, globe tenting, and extensive paranasal sinus disease. Orbital biopsy culture revealed *Clostridium bifermentans* and *Enterobacter aerogenes*. Early postoperative assessment revealed no light perception acuity, despite a successful incision and drainage procedure that resulted in decompression of the orbit.

Conclusions:
Most cases of orbital cellulitis are caused by streptococcus species, staphylococcus species, or *Haemophilus influenzae*, and result in relatively good patient outcomes when appropriate antibiotic therapy is initiated. However, it appears that gas-producing organisms such as *Clostridium bifermentans* and *Enterobacter aerogenes* may require more urgent surgical and antibiotic intervention to avoid severe loss of vision. Therefore, the presence of severe proptosis, gaseous accumulations within the orbit, decreased vision, and globe tenting should prompt more aggressive and timely surgical therapy.

References: None.

Keywords: Orbit/Ocular Pathology, Orbit, Neuroimaging

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 113
Historical treatment of Carotid Cavernous Fistula

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Introduction:
CCA (Common Carotid Artery) ligation was a commonplace procedure throughout the 19th century and was first used in CCSF (Carotid-Cavernous Sinus Fistula) after it became evident that carotid artery compression reduced pulsating exophthalmos. The first description of ICA (Internal Carotid Artery) ligation for CCSF followed in 1904. Carotid artery ligation continued to be the mainstay of treatment until the introduction of balloon occlusion of the fistula in the early 1970s. While often effective in reducing symptoms and preventing the most severe complications, ICA ligation has usually not been successful in eliminating all shunt related clinical manifestations.

Methods:
Review of clinical history, neuro-imaging and relevant literature.

Results:
In 1953, a then 17 year old man had suffered cranial trauma complicated by a high flow CCSF. Surgical ligation of the ICA was performed but could not prevent the development of chronic exophthalmos and secondary glaucoma with progressive loss of vision and blindness. Over time the patient developed mild varicosities in the ipsilateral face illustrating the extent of facial venous drainage via the cavernous sinus. The patient's CT scan of the head obtained 50 years after the procedure demonstrates bony erosion in the sellar region apparently due to dilated vascular channels and shunting of blood from the posterior circulation into the cavernous sinus.

Conclusions:
This case demonstrates the very long term sequelae of a traumatic CCSF treated with ICA ligation. It also illustrate the extensive facial venous drainage via the cavernous sinus.

References: None.

Keywords: Orbit/Ocular Pathology, Trauma, Vascular Disorders

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 114
Congenital Entropion and Hypotropia Secondary to Duplication of the Inferior Rectus Muscle!

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Introduction:
Congenital anomalies of the extraocular muscles, apart from congenital fibrosis, are rarely encountered. Duplication of extra-ocular muscles is a very rare congenital anomaly that has been seldomly reported in the literature as a cause of abnormal ocular motility. We present a case of unilateral congenital entropion and hypotropia resulting from duplication of the inferior rectus muscle.

Methods:
Case report and review of literature.

Results:
A 1-month-old caucasian male infant presented to our department for evaluation of right congenital entropion. On examination he was found to have 6 digits in each hand but otherwise normal. On ocular examination, he was found to have right hyotropia and limitation of supra-duction. On careful review of his orbital MRI, he was found to have a mass in the right inferio-temporal retrobulbar region with displacement of the optic nerve superiorly. Additionally, the inferior rectus muscle appeared to be bilobed and distorted. Heterogenous enhancement of both lobes was noted following gadolinium adminstration. On orbital exploration via an inferior transconjunctival approach, the mass was identified to be a skeletal muscle attached to the lateral margin of the inferior rectus and medial aspect of lateral rectus. The tissue was excised to restore normal extraocular muscle anatomy. The lower lid was allowed to heal without re-suspending it given the pre-operative lid tightness with satisfactory correction of the entropion and hypotropia. Histo-pathological examination of the excised tissues revealed skeletal muscle tissue with muscle fiber disarray, alternating hypotrophy and hypertrophy with fibrosis.

Conclusions:
We believe that the hyotropia and entropion are explainable by the attachment of the duplicate inferior rectus muscle to the globe and the lower lid retractors respectively. To the best of the authors' knowledge this is the first report of congenital entropion and hypotropia due to duplication of the inferior rectus muscle.

References: None.

Keywords: Extraocular Muscle, Entropion, Strabismus, Congenital, Anomaly

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 115
Orbital Apex Syndrome Caused By Rosai-Dorfman Disease

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Introduction:
Rosai-Dorfman Disease (RDD) is a rare idiopathic histioproliferative disease characterized by massive painless lymphadenopathy, fever and weight loss. Central nervous system (CNS) manifestations are extremely rare.

Methods:
Case report, and review of literature.

Results:
A 72 year old woman was evaluated in the emergency room for retroorbital pain and decreasing vision in the right eye to No Light Perception (NLP). She has been evaluated by her local ophthalmologis and neurologist for several months with negative workup. She reported right retroorbital pain for 3 months followed by binocular diplopia within a week. She later developed lid drooping with "resolution" of her diplopia. Her vision has been gradually worsening in the right eye until she lost it completely a month prior to presentation. She was sent to the ER to rule out giant cell arteritis (GCA).

She was found to have NLP vision in the right eye, 20/25 in the left and a 4+ right APD. The right optic nerve was diffusely pale with a pink, healthy looking left optic nerve. An MRI of the orbit was ordered that on preliminary read was normal. On careful review of the imaging, an enhancing lesion was noted along the right side of the skull base extending to the right orbital apex. An endoscopic endonasal transpterygoid biopsy and debulking of the mass was carried out using stereotactic image guidance. Biopsy revealed mixed inflammatory cell infiltrates suggestive of RDD. 3 Months later she underwent a skin biopsy for a lesion behind her left knee. The histopathology and and immunochemistry confirmed the diagnosis of RDD. Patient has been followed for 12 months with no progression of the mass.

Conclusions:
RDD should be included in the differential diagnoses for a dural or skull base mass. Although the pathology is benign elsewhere, as with other inflammatory lesions of the orbit and skull base, this case illustrates severe vision loss resulting from RDD,

References: None.

Keywords: Rosai Dorfman, Orbit, Histiocytosis

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 116
A Case of Adult T-Cell Leukemia/ Lymphoma with Extraocular Muscle Infiltration

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Introduction:
To report a case of Adult T-cell leukemia/ lymphoma (ATLL) with confirmed infiltration into the extraocular muscles.

Methods:
Case report and review of literature

Results:
A 60-year- old man presented with complaints of diplopia and discomfort around the right eye since February 2015. He visited an eye clinic in his neighborhood, and the ophthalmologist referred him to our clinic. His past medical history was remarkable for adult T-cell leukemia with complete remission for 2 years, biopsied goiter, diabetes, and hypertension. Examination showed visual acuities of 20/20 OU. The pupils appeared round and measured 5 mm OD and 6 mm OS in darkness and 3 mm OU with exposure to light; both pupils constricted briskly to light. There was no afferent pupillary defect. After instillation of 1% apraclonidine, the pupils measured 7 mm OD and 6 mm OS in darkness. Visual field examination revealed normal OU. Proptosis was observed in the right eye, along with 80% supraduction, 100% adduction, 40% infraduction, and 60% abduction. The left eye was normal. Corneal sensation and slit -lamp and funduscopic examination findings were normal OU. The rest of the neurological examination results were normal. Orbital MRI showed swelling of the right lateral rectus, medial rectus, and inferior muscle. The swelling appeared as low intensity on precontrast T1 MRI, homogeneous enhancement on postcontrast T1 MRI, and high intensity on T2 MRI. The partially resected inferior rectus muscle disclosed diffuse proliferation of small- to medium-sized lymphoid cells with irregularly shaped nuclei in the soft tissue. Immunohistochemically, these lymphoid cells were positive for CD3, CD4, but negative for CD79a, CD8. These findings are consistent with ATLL.

Conclusions:
The clinician should consider extraocular muscle invasive ATLL as a potential cause of an orbitopathy. Extraocular muscle biopsy is crucial for an accurate diagnosis.

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Keywords: Orbit/Ocular Pathology, Orbit, Neuro-Ophth & Infectious Disease, Tumors

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