NANOS 2017
FRANK B. WALSH SESSION

Sunday, April 2, 2017

7:45 a.m. – 5:00 p.m.

Co-Chairs: John Jing-Wei Chen, MD, PhD and Jacqueline A. Leavitt, MD
Neuroradiologist: Jonathan Morris, MD
Neuropathologist: Caterina Giannini, MD, PhD

This symposium is designed to present a wide variety of Neuro-Ophthalmic cases to an audience of physicians with varying neuroscience backgrounds who have a common intellectual interest in the broad range of conditions that impact the human visual pathways and ocular motor systems.

The format is a clinicopathologic conference. Clinical cases will be presented by Neuro-Ophthalmologists with comments by a neuroradiologist, neuropathologist and other selected experts. Necropsy, surgical pathology, and neuroimaging will help illuminate clinical points. Cases will be discussed from clinical, anatomic, radiologic and pathologic aspects with emphasis on diagnosis, pathophysiology and management. Audience participation is encouraged.

Upon completion of this course, participants should be able to: 1) Recognize the varied presentations of Neuro-Ophthalmic disease; 2) Correlate the anatomic localization and histopathologic appearance with the clinical presentations; 3) Effectively use radiologic procedures in diagnosis; 4) Recognize both the value and limitations of neuropathology; and 5) Discuss newly described diseases and their connection to Neuro-Ophthalmology.
Frank B. Walsh Session I
Moderators: Sophia M. Chung, MD & Collin McClelland, MD

8:00 am - 8:20 am  Pseudo-Pseudotumor Cerebri
Valerie Bioussé, MD

8:20 am - 8:40 am  A “Frosty” Altered Level of Consciousness
Alaa Bou Ghannam, MD

8:40 am - 9:00 am  Uncertainty with a Twist of Lyme
Amrita-Amanda Lakraj, MD

9:00 am - 9:20 am  Papilledema Gone Wrong
Ahmara Ross, Fellow in Training

9:20 am - 9:40 am  Occam Rings True
Reid Longmuir, MD

9:40 am - 10:00 am  A Prolonged Path To The Final Diagnosis
Cindy Lam, MD, FRCSC

10:00 am - 10:30 am  Coffee Break

Frank B. Walsh Session II
Moderators: Michael C. Brodsky, MD & Yanjun (Judy) Chen, MD, PhD

10:30 am - 10:50 am  Keep Your Eye On the Ball
Danielle Rudich, MD

10:50 am - 11:10 am  Alcohol is Never the Answer, but it Does Make You Forget the Question
Ali Saber Tehrani, MD

11:10 am - 11:30 am  Nonchalant Midterm-taker Develops Altered Mental Status
Shira Simon, MD, MBA

11:30 am - 11:50 am  It Must Be Voodoo... and he was Far from Spineless
Norah Sydney Lincoff, MD

11:50 am - 12:10 pm  M.I.A.
Julie DeBacker, MD

12:10 pm - 12:30 pm  The Sound of Hoofbeats
Lilangi Ediriwickrema, MD
Frank B. Walsh Session III
Moderators: James A. Garrity, MD & Heather E. Moss, MD, PhD

3:00 pm - 3:20 pm
A Wrong Turn at the Angle
Erica Archer, MD

3:20 pm - 3:40 pm
Strike a Chord
Anastasia Neufeld, MD

3:40 pm - 4:00 pm
Island Fever
Laura Hanson, MD

4:00 pm - 4:20 pm
Lions and Tigers and Bears, Oh My!
Larissa Ghadiali, MD

4:20 pm - 4:40 pm
Not Surprised Surprise
Yu Zhao, MD

4:40 pm - 5:00 pm
A Diagnostic Potpourri
Padmaja Sudhakar, MD

"Pseudo-Pseudotumor Cerebri"

Valerie Biousse1, Jose Velázquez Vega2, Amit Saindane3, Nancy Newman4
1Emory University School of Medicine. Emory Eye Center, Atlanta, Georgia, USA, 2Emory University School of Medicine. Department of Pathology, Atlanta, Georgia, USA, 3Emory University School of Medicine. Department of Radiology, Atlanta, Georgia, USA, 4Emory University School of Medicine, Atlanta, Georgia, USA

History & Exam
A 54-yo man presented with a 10-month history of daily headaches and bilateral disc edema. PMHx was remarkable for uncomplicated type-2 diabetes mellitus, and hypothyroidism. He developed headaches in 07/2014 after being stung by wasps. He also had fluctuating blurry vision. An ophthalmologist found normal VA and disc edema OU. MRI brain/orbits with contrast (12/19/2014) was normal except for some "volume loss". RPR, Lyme, TSH, ANA, ESR were normal/negative. A neurologist (03/03/2015) found normal neurologic and general examinations (BMI 30kg/m2). Brain MRI with contrast (03/11/2015) showed no abnormal enhancement and normal parenchyma. The subarachnoid spaces along the bilateral perirolandic and posterior frontal parenchyma were prominent. Repeat ophthalmologic evaluation (April 23, 2015) showed normal VA and persistent bilateral disc edema. HVF showed enlarged blind spots and nasal steps OU. Headaches and visual changes persisted with pulsatile tinnitus. Brain MRV/MRA with contrast (04/28/2015) showed a large left arachnoid granulation in distal left transverse sinus. LP (04/29/2015) was traumatic and painful and showed CSF -OP 12cm, pink hazy CSF, 124/ul nucleated cells (32% neutrophils, 66% lymphocytes), 22599 RBC/ul, glucose 104mg/dl, protein >300mg/dl, CSF cytology negative; there were a few plasmacytoid/plasmablasts felt to result from blood contamination. SPEP showed moderate hypogammaglobulinemia. Headaches and tinnitus improved dramatically after the LP. The diagnosis of pseudotumor cerebri was made and he was given acetazolamide 500 mg bid, which was not tolerated and was stopped. Ophthalmologic examination (05/22/2015) was unchanged with persistent bilateral disc edema and improved VF. Neurologic evaluation (06/02/2015) continued to be unremarkable. LP under fluoroscopy (06/03/2015) was again difficult and painful and showed CSF-OP 25.8cm, clear CSF with xanthochromia, 90/ul nucleated cells (90% lymphocytes), 288 RBC/ul, glucose 100mg/dl, protein <6mg/dl, CSF cytology negative. His symptoms improved for 10 days. Ophthalmologic examination (06/25/2015) was unchanged with persistent bilateral disc edema and stable VF. Another test was recommended.

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None.
"Pseudo-Pseudotumor Cerebri"

Valerie Biousse¹, Jose Velázquez Vega², Amit Saindane³, Nancy Newman⁴

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History & Exam
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"A "Frosty" Altered Level of Consciousness"

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History & Exam
15-year-old previously healthy male, transferred because of 9-day history of headache, difficulty walking, diplopia, blurry vision and new onset altered level of consciousness. Neurologic examination was remarkable for delirium with semi-coherent speech, VA of CF 2ft and 20/400, no APD, and bilateral abduction deficits. He had bilateral hip flexor weakness(3/5), 2+ DTR, flexor plantar responses, bilateral upper extremity dysmetria, and ataxic gait requiring two-person assist. Prior MRI imaging showed bilateral optic sheaths’ complex enlargement and leptomeningeal enhancement. Lumbar puncture revealed opening pressure of 46 cmH2O with 298WBCs (46% PMN, 36% L, 14% Mono), protein 266, and glucose 20. Cytology was negative for atypical cells. Patient was started on vancomycin, ceftriaxone and levofloxacin. Extensive workup for infections was negative (Quant gold, EBV, Bartonella, RPR, HIV, Coxieilla, Brucella, VZV, Enterovirus, HSV, WNV and Lyme). CSF cultures were negative for bacterial and fungal growth. ESR(47) and CRP(7.7) were both elevated. ANA, c-ANCA, p-ANCA, ACE, C3, C4 were all negative. The patient’s mental status declined and he developed bilateral lower extremity weakness; after 10 days, he became difficult to arouse with moaning verbal responses, 3+ DTR, and extensor plantar responses. Serial LPs showed rising WBC. Repeat MRI brain and spine showed increased diffuse leptomeningeal enhancement, T2 hyperintensity extending from lower cervical spine to conus involving ventral gray matter; bilateral optic nerve, chiasm and optic tract enhancement, and irregularities in posterior globes on T2 weighted images. Ophthalmologic exam revealed visual acuity of CF at 1ft in right and BTL in left eye with normal intraocular pressures. Pupils were small and minimally reactive with no APD; EOM assessment was difficult due to impaired mental status. Dilated fundus exam showed bilateral disc edema with diffuse, perivascular sheathing of the major retinal veins, and scattered intraretinal hemorrhages. Several studies were performed and a diagnostic test was completed.

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"Uncertainty with a Twist of Lyme"

Amrita-Amanda Lakraj¹, Sang Hong², Jennifer Connelly³, Elizabeth Cochran⁴

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History & Exam
The patient is a 59-year-old woman who presented with vision loss. Her ophthalmologic history is significant for myopic astigmatism and presbyopia. Her past medical history is notable for left breast cancer (T2N0 invasive lobular carcinoma, ER/PR positive, HER2/neu negative) s/p left mastectomy, chemotherapy, and radiation (2006); Celiac disease; Grave’s disease s/p partial thyroidectomy (1977) and radioactive iodine (2000); Lyme disease (2013); and progressive neuropathy of unclear etiology (onset 1998). Extensive testing for her neuropathy from 1998-2014 was significant for positive serum Lyme IgM Western Blot 10/2013 (treated with IV antibiotics). In January 2015, the patient developed blurred vision. By April 2015 she presented to her optometrist with double vision and was found to have mild fullness of bilateral optic discs. MRI brain was reportedly unremarkable. Recent MRI of the C/T spine demonstrated abnormal leptomeningeal enhancement extending along the conus medullaris and lumbar nerve roots as well as mild cord edema. Lumbar puncture revealed elevated white count (12), elevated protein (>600), normal glucose, positive Lyme IgM western blot, and negative culture and cytology. She was referred for neuro-ophthalmologic evaluation. Her initial neuro-ophthalmologic exam 5/2015 was notable for visual acuities with correction of 20/50 OD (ph 20/50+2) and count fingers at 6 inches OS; 1+ APD OS; 11/11 Ishihara color plates OD and 4/11 OS; superotemporal constriction OD on confrontation visual fields, and generalized constriction OS; mild limitation of upgaze OU, mild limitation of adduction and abduction OD and mild limitation of abduction OS. On fundus exam she had mild optic disc swelling OU. The presence of bilateral optic neuropathy with multifocal cranial nerve involvement on exam raised concern for a leptomeningeal process, particularly given recent leptomeningeal enhancement on spinal MRI. The patient was admitted for further workup and management.

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"Papilledema Gone Wrong"

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History & Exam
A 12 year old obese female with a history of enterovirus meningitis presented with throbbing, constant, 10/10, occipital and positional headaches, associated with diplopia, photophobia, nausea, and vomiting for one week in the setting of a recent 20 lb. weight gain. Her vision, pupils, fields, and color vision were all unremarkable. Her fundus exam was significant for bilateral papilledema, and her neurologic exam was otherwise normal. MRI with and without contrast and MRV were unremarkable. Her lumbar puncture opening pressure was 60 cmH2O with a normal CSF cell count, protein and glucose. She was started on Diamox with a presumptive diagnosis of IIH and discharged home. Two weeks later she re-presented with similar symptoms and a change in mental status. Neurologic exam was remarkable for posturing, mutism, poor food intake, and regressive behaviors such as urinary incontinence. Her fundus exam showed improved papilledema but her neurologic exam was significant for delirium and slowed reaction time. Routine EEG, MRI brain, MRA head and CSF studies were obtained and all were unremarkable, except for a repeat but bland lumbar puncture with an elevated opening pressure of 48 mmHg. Six weeks later her exam was significant for obtundation, extensive bulbar dysfunction requiring intubation for airway protection, and flaccid quadriparesis resulting in admission to the Neuro-NICU. Extensive tests showed abnormal laboratory findings that were non-specific including a persistently elevated ESR, normocytic anemia, mild transaminitis. Repeat MRI showed diffuse white matter changes involving the periventricular regions extending into the centrum semiovale, corpus callosum, brainstem and cerebral peduncles. MR spectroscopy showed non-specific findings of diffuse elevation of lactate throughout the brain parenchyma, especially in the CSF. N-acetylaspartate was decreased diffusely with a pronounced decrease of N-acetylasparate in the right basal ganglia compared to the left basal ganglia. Choline and myoinositol was diffusely elevated.

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"Occam Rings True"

Reid Longmuir1, Michael Bradshaw1, Taylor Davis1, Woon Chow1, Laura Craig-Owens1, Kim Ely1, Katherine McDonell1

1Vanderbilt University Medical Center, Nashville, Tennessee, USA

History & Exam
A 57-year-old, previously healthy man was referred for vision loss in both eyes. Six weeks before our evaluation, he presented to an outside ophthalmologist with "greying" of the central vision in his right eye that progressed to blindness over a week. After another week, the same symptoms affected his left eye and likewise progressed over two weeks, although he remained able to see hand motion with the left eye. There was a new holoccephalic headache that was worse in the mornings and evenings but was not positional. He also reported burning, electric pain over the left shoulder that radiated down the spine with neck movements. Review of systems was otherwise unremarkable. His ophthalmologist noted severe bilateral papilledema and a brain MRI reportedly revealed a 2-3 cm presumed olfactory groove meningioma. An outside neurosurgeon felt there was no indication for intervention on the olfactory groove lesion and the patient was referred to neuro-ophthalmology 2 weeks later. He was a prison guard but never had positive purified protein derivative testing. He neither drank alcohol nor smoked and was current on preventative cancer screening. His mother died of a brain tumor in her 70s but there was otherwise no significant family history. He was transferred to the emergency department and admitted to the neurology service the day of presentation to clinic, based on poor vision and need for further diagnostic testing. He had normal vital signs, left-sided anosmia, a relative afferent pupillary defect on the right, and severe bilateral papilledema. Visual acuity was counting fingers at 1 foot OD and at 4 feet OS. He had a large central scotoma in each eye with confrontation visual field testing. He had normal ocular motility. He reported Lhermitte's phenomenon and had subtle nuchal rigidity. The remaining general and neurologic examinations were normal.

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"A Prolonged Path To The Final Diagnosis"

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History & Exam
Previously healthy 25-year-old male presented with a 3-week history of frontal headache, right 3rd nerve palsy with ptosis, and areflexia of the left leg. MRI brain was normal. Two days later new symptoms developed: lower back pain, paresthesias, left leg weakness, right facial weakness. On neuro-ophtalmic exam pertinent findings included bilateral upper lid ptosis, right adduction deficit, pupil-sparing left 3rd nerve palsy, and right 7th nerve palsy. MRI brain and spine demonstrated enhancement and thickening of the cauda equina nerve roots, enhancement within the right internal auditory canal with a nodularity of the right facial nerve, and enhancement in left Meckel’s cave. Two LPs demonstrated very elevated protein (3.3-5.5 g/L), oligoclonal banding, and pleocytosis (100 cells/um, 98% lymphocytes). Extensive infectious and inflammatory work-up of the serum and CSF was negative. CSF flow cytometry, lymphoma panel, and SPEP were negative. A provisional diagnosis of atypical GBS (Miller Fischer variant) was made and IVIG was administered without symptom improvement. Methylprednisolone was administered intravenously (1 g x 5 days), leading to improvement in lower back pain, paresthesias and left leg weakness. Two months later all symptoms had resolved. However, eleven months after initial presentation, new progressive right sided weakness developed. MRI of the brain and spine demonstrated a large enhancing lesion in the left centrum semiovale with a surrounding halo of restricted diffusion. No cauda equina enhancement was seen. Four expert neuroradiologists debated whether diagnosis was Balo’s concentric sclerosis or CNS lymphoma. Three LPs were performed: cytology was negative for malignancy and flow cytometry was inconclusive. CT thorax, abdomen, and pelvis was negative. A new left facial palsy developed. Repeat MRI showed increase in the left frontal mass, enhancement of left facial nerve and internal auditory canal and resolution of enhancement in right IAC and left Meckel’s cave. A diagnostic procedure was performed.

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In March 2014, an 82-year-old male developed left sided facial pain and numbness over his left temple and periorbital region. He was treated for a presumed dental infection with no response. MRI brain in July 2014 showed only microvascular white matter changes. Because his symptoms persisted, MRI brain was repeated in February 2015, and the left sided muscles of mastication were relatively hyperintense on FLAIR/T1 post contrast weighted images and there was abnormal enhancement of the third branch of the left fifth cranial nerve, just beyond Meckel’s cave. MRI orbits showed questionable asymmetric enhancement of the second branch of the left trigeminal nerve within the left infraorbital foramen. Neuro-oncology work up including spinal tap and laboratory workup were negative. A neurologist diagnosed Tolosa Hunt syndrome but there was no response to Prednisone. The patient gradually developed binocular horizontal diplopia, blurred vision and redness of the left eye, and was referred for neuro-ophthalmology consultation in May 2015. Visual acuity was 20/20 OD and 5/200 OS with left fifth and sixth cranial nerve palsies. The left eye was injected with a central corneal ulcer and small hypopyon. MRI from June 2015 showed subtle enhancement of the left fifth and sixth cranial nerves at the brainstem exit, atrophic left pterygoid and temporalis muscles and thinning of the left lateral rectus muscle. Prior dermatology records/pathology slides showed no malignancy. ENT felt there was no lesion to biopsy for diagnosis. Perineural tumor spread was suspected but there were no available biopsy sites. MRA/V brain was normal and PET scan showed no evidence of malignancy or metastases. The corneal ulcer grew Candida albicans and worsened, leading to a corneal perforation, requiring penetrating keratoplasty. A procedure was performed.

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"Alcohol is Never the Answer, but it Does Make you Forget the Question"

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History & Exam
A 63-year old male with a history of alcohol abuse presented with two weeks of confusion and imbalance. On exam, he had left beating nystagmus on left gaze, and right beating and torsional nystagmus on right gaze. Head impulse test was abnormal in all canals bilaterally with video head impulse test showing left horizontal vestibulo-ocular reflex gain: 0.3, right: 0.4. Finger to nose and heel to shin were abnormal bilaterally. Gait was abnormal and he was falling towards his right side. Neuropsychiatric evaluation revealed impairment in the following domains; intellectual skills, attention/visual sequencing and memory. MRI did not show any acute abnormalities. Infusion of 500 mg of thiamine did not improve head impulse test gain.

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"Nonchalant Midterm-taker Develops Altered Mental Status"

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History & Exam
A 19 year-old female presented in April 2016 with two weeks of worsening headaches, malaise, left-hand weakness, and confusion. Symptoms started during a trip to Cancun and one week after visiting a friend with mumps. Although she was becoming increasingly lethargic, she refused to seek medical attention during midterm exams. En route to an exam, she bent over to tie her shoelaces and collapsed, prompting her hospital presentation. Past medical history included optic neuritis of the right eye in 2013 with good response following ONTT protocol steroid therapy and headaches since junior high, which responded to NSAIDs. During her examination, she became increasingly confused, paranoid, and combative. She required sedation and sitters to obtain further investigations. Due to concern for meningoencephalitis, she was empirically started on IV vancomycin, ceftriaxone, and acyclovir. MRI brain and spine with contrast demonstrated enhancement of both optic nerves as well as a 6 mm focus of enhancement in the anterior aspect of the C4 vertebral body. CSF gram stain was negative, but there was an elevated protein concentration (132 mg/dL) and white cell count (195 cells per milliliter) with lymphocytic predominance. EEG demonstrated generalized slowing. Visual acuity was 20/40 in each eye, with no RAPD. Ocular motility was full without pain, internuclear ophthalmoplegia, or nystagmus. The anterior segment examination was normal; posterior segment showed optic disc pallor in the right eye and mild diffuse optic disc edema with no associated pallor in the left eye. Formal perimetry could not be obtained secondary to her confusion. Serologic and CSF testing demonstrated no infectious etiology (negative HSV, VZV, EBV, HHV5, West Nile, enterovirus, Lyme, mycoplasma, syphilis, HIV, TB, histoplasmosis, cryptococcus, bartonella, Q fever, and mumps). She was started on IVIG (400 mg/kg/day) for presumed aseptic meningitis. Her mental status continued to worsen. A diagnostic test was performed.

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Grant Support: None.
Nonchalant Midterm-taker Develops Altered Mental Status

Answer

Final Diagnosis
Tumor-negative anti-N-methyl-D-aspartate receptor (NMDA-R) encephalitis with bilateral optic neuritis

Summary of Case
Repeat MRI and LP were performed, along with a complete autoimmune and paraneoplastic panel. MRI demonstrated persistent mild patchy bilateral optic nerve enhancement as well as subtle leptomeningeal enhancement. ANA titer was positive; ESR, CRP, and anti-thyroid peroxidase antibody were elevated. CSF testing revealed negative aquaporin-4, with positive oligoclonal bands, elevated IgG index, and positive NMDA receptor antibody. NMDA receptor antibody was positive in serum, consistent with anti-NMDA receptor encephalitis with optic neuritis. Repeat PET and ultrasound examinations showed no teratoma or other malignancy. Anti-NMDA receptor encephalitis was first described in 2007, in association with ovarian teratoma. The classic presentation is with a prodromal illness, psychiatric symptoms, and decreased responsiveness. Since its description, it has become the second most common immune-mediated cause of encephalitis after acute disseminated encephalomyelitis. 80% of patients are women, 60% of whom have an associated tumor (5% of men). 75% of patients with anti-NMDA receptor encephalitis have a good recovery with treatment. Brain MRI is normal in up to 67% of patients, and EEG is abnormal in 90% of patients. Brain biopsy is unhelpful in diagnosis. Tumor-negative anti-NMDA receptor encephalitis with optic neuritis is very rare. There have been two adult cases reported since 2007: a 29 year-old male with history of presumed lymphocytic meningitis and a 32 year-old female with chronic depression. No standard of care exists, but case studies report promising results with concurrent IVIG (0.4 g/kg per day for 5 days) and methylprednisolone (1 g/day for 5 days) as first line therapy, and then second-line therapy of rituximab and cyclophosphamide. Our patient continued to deteriorate despite receiving IVIG with methylprednisolone. She was subsequently treated with rituximab and methylprednisolone with taper, and has returned to her baseline, with no recurrences to date.

Struggle/Dilemma of the Clinical Presentation Description
The differential diagnosis was originally infectious given her history of recent travel and exposures. However, clinical evaluation was challenging due to suboptimal patient cooperation with the precipitous decline in mental status necessitating quick action. Diagnostic and management dilemmas arose given a largely unrevealing infectious and inflammatory work-up and lack of clinical response to IVIG therapy.

Keywords:
Optic neuritis, Meningo-encephalitis

References

It must be Voodoo... and he was Far from Spineless

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History & Exam
A 35 yo legally blind gentleman was referred for evaluation because of neck and new lower back pain. His visual loss began 4 years prior, following a trip to “N’Orleans”. He had been on a “Boys Trip” for 10 days, and had been cursed with Voodoo for not paying a cemetery tour guide as promised. He also was cursed with hearing loss and imbalance. He reported that no trial of medicine saved his sight, but that he was now at least less off balance, and felt his hearing loss was 50% improved. He was diagnosed with aseptic meningitis. He had been treated with antibiotics, steroids, IVIG, Methotrexate, and Cyclophosphamide. To further convince himself of the curse, he found himself in Tower One on 9/11, barely making it out alive. Neuro-ophthalmologic exam revealed vision of HM right eye and CF left eye. His pupils were poorly reactive. Motility exam was normal. Slit lamp examination was normal. Applanation Tonometry was normal. Funduscopic examination revealed bilateral severe optic atrophy, cupping, and vascular narrowing. He had markedly constricted visual fields of less than 20 degrees both eyes. An MRI of his brain revealed bilateral optic atrophy. An MRI of his spine revealed a diffuse "moth eaten" appearance to his vertebrae. A biopsy was performed.

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"M.I.A."

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History & Exam
A 69-year-old man presented with a several year history of vertical double vision. At age 46, he presented with a two year history of imbalance, four year history of urinary urgency/frequency, and a six-month history of leg numbness and tingling. His history was significant for tuberculosis, treated (age 12). He took a multivitamin. He had no known toxic exposures. Family history revealed two brothers, ages 45 and 39 with dysuria. Prior exams revealed gynecomastia and high feet arches with hammertoe deformity, decreased right hearing, normal strength, lower extremity hyperreflexia, and progressive impaired pinprick and vibration sensation and tandem gait. Brain MRI was initially unremarkable, though two years later showed cerebellar vermian atrophy and 4 years later atrophy and T2 hyperintensities in the periventricular, deep, and subcortical white matter of both cerebral hemispheres. MR spectroscopy showed decreased parieto-occipital NAA. C-T spine MRI showed thoracic cord atrophy without abnormal signal. EMGs showed severe axonal polyneuropathy. Serum and CSF were unremarkable for vitamins B12 and E, ANA, heavy metals, Lyme, leukocyte Aroylsulfatase A, CPK, Anti-glycolipid antibodies, very long chain fatty acids, phytic acid, HTLV-1 and 2, peripheral blood smear for monocyte/lymphocyte vacuoles, and a peroxisomal panel. A debranching enzyme assay was inconclusive twice. Urine porphyrins were unremarkable. Skin punch biopsy electron microscopy demonstrated a demyelinating neuropathy. On our exam, mental status was normal. Visual acuities were 20/30 OU. Visual fields were intact to confrontation. He perceived Ishihara control plate only OU. There was no APD. He had a right 4th palsy, saccadic pursuits. Optic discs were atrophic OU. OCT showed RNFL thinning. Muscle bulk in his legs was reduced with 0/5 lower and normal upper extremity strength. Sensation was diminished to knees B/L. Patient was in a wheelchair with trunk support. Reflexes were absent.

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"The Sound of Hoofbeats"
Lilangi Ediriwickrema¹, Neil Miller¹, Thomas Bosley¹
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History & Exam
A 42-year-old woman with a recurrent cerebral malignant glioma in the left frontal lobe on clinical trial presented with blurry vision in her right eye 6 days after undergoing sinus surgery for paranasal inflammatory changes. She presented approximately 1 month after the sixth cycle of systemic steroids, Durvalumab (PD-L1 inhibitor), and Bevacizumab. Her initial examination revealed best-corrected visual acuity of 20/25 OU, a right relative afferent pupillary defect, and an unremarkable anterior segment exam. Fundus exam was notable for mild right optic disc swelling. Automated static perimetry showed no deficits in either eye. Magnetic resonance imaging (MRI) of the brain showed parenchymal disease progression. In addition, the orbital portion of right optic nerve was enlarged, hyperintense on T2-weighted imaging, and enhanced after intravenous injection of paramagnetic contrast material. Ten days later, the patient developed sudden, complete vision loss in her right eye. Examination revealed no light perception vision OD and 20/20 OS. The anterior segment was unchanged; however, she now had marked optic disc swelling, a peripapillary retinal hemorrhage, and a superior branch artery occlusion. The patient underwent a lumbar puncture that showed no abnormalities. Examination two days later showed interval development of a central retinal artery and vein occlusion, as well as a new collection of layered, whitish material suspended anterior to the internal limiting membrane, forming a pocket in the posterior vitreous just inferior to the optic disc. A diagnostic procedure was performed.

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"A Wrong Turn at the Angle"

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History & Exam
A 54-year-old woman developed numbness of the right jaw and right side of the tongue. She attributed her symptomatology to recent dental work, but when dental examination, including dental x-rays, failed to reveal an answer, she underwent brain MRI. Imaging disclosed an enhancing right cerebellopontine angle (CPA) mass that appeared to originate in the tentorial dura (Slides 1,3). There was also a small mass on the right medullary surface (Slides 2,3). Neurosurgical examination found decreased sensation in the right V2 and V3 distributions and diminished hearing in the right ear. The lesion was interpreted as a meningioma. Over the next few weeks, she developed vertical diplopia. Our neuro-ophthalmologic examination diagnosed a small comitant right hypertropia interpreted as skew deviation, together with mild right appendicular and gait ataxia. The diplopia was palliated with a Fresnel prism. Because of increasing symptoms, the patient underwent right retrosigmoid craniotomy with “90% resection of the tumor” (Slide 4). Post-operatively, the ataxia worsened and right hearing loss became complete. Neuro-ophthalmologic evaluation disclosed no change in the skew deviation, but now development of sidebeat nystagmus to both sides and saccadic pursuit, as well as profound ataxia. The pathology report on the brain lesion came next...

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**"Strike a Chord"**

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**History & Exam**

A 26 year old woman presented in August 2016 with migraine over the last 4 years, getting worse over the last 2 weeks. She reported occasional “glittering” in her vision in both eyes. She had no constitutional symptoms. Her past medical history included migraine, idiopathic leukocytosis, and nephrolithiasis. There was no family history of neurocutaneous disease. Due to worsening of her headache, her primary care provider ordered an MRI Brain, which showed an enhancing solid mass, measuring 2.3 cm in the suprasellar region, thought to be originating from the optic chiasm with involvement of both pre-chiasmatic optic nerves as well as the optic tracts. There was no hydrocephalus or any other lesions in the brain parenchyma. Primary diagnosis of concern based on imaging characteristics was optic pathway glioma. Endocrine work up was normal. Best-corrected visual acuity was 20/20 OD and 20/30 OS. There were no pupillary abnormalities. Intraocular pressure was normal in both eyes. Optic nerve function tests were normal, including color, critical flicker fusion frequency, red desaturation, and Humphrey visual field. Dilated fundus examination showed mild temporal pallor of the optic nerves. OCT of the retinal nerve fiber layer showed superior and inferior elevation and mild temporal atrophy on the right, and superior elevation and mild temporal atrophy on the left. A diagnostic procedure was performed.

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"Island Fever"

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**History & Exam**

A 22 year-old male paralegal in the military presented with double and blurred vision. One month ago, he had traveled to Hawai‘i for a wedding. Two weeks ago, he had been evaluated for an acute onset of fever, headache, emesis, light sensitivity and neck stiffness. He was diagnosed with viral meningitis after a lumbar puncture revealed 270 WBC with 85% mononuclear cells, 12% eosinophils and 3% PMNs, and sent home on convalescent leave to recuperate with his family. Shortly after arriving home, his headache, neck stiffness and malaise had improved, but he developed new horizontal, binocular diplopia and blurred vision in both eyes. Ophthalmologic examination revealed bilateral 6th nerve palsies and Frisen stage 3-4 optic disc edema. Humphrey visual field testing showed enlarged blind spots. A second lumbar puncture was performed, which revealed 588 WBC with an eosinophilic predominance of 61%, protein 343 mg/dL, and glucose 36 mg/dL. The opening pressure was 28 cm of water. A contrasted CT of the chest revealed multiple pulmonary nodules with surrounding ground-glass appearance. A gadolinium-enhanced MRI of the brain revealed scattered cortical-based nodular foci of enhancement with areas of T2 signal abnormality and sheet-like enhancement. An MRV showed no venous sinus thrombosis. A diagnostic procedure was performed.

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"Island Fever"

Answer

Final Diagnosis

Eosinophilic Meningitis secondary to Angiostrongylus cantonensis.  Angiostrongylus cantonensis is also known as the "Rat Lungworm" and was first found in the lungs of rats in South China. The nematode larvae live in the pulmonary veins of their definitive host, the rat, and are perpetuated by snails, which serve as a secondary host. It is endemic in Southeast Asia, the Pacific Basin, the Caribbean, and Hawai'i. Humans become infected by eating undercooked snails, freshwater shrimp, frogs, or unwashed produce contaminated by one of these hosts. The larvae migrate to the central nervous system and the induced inflammatory response creates the manifestations typically seen in humans, including meningitis, cranial nerve abnormalities, ataxia, encephalitis, and increased intracranial pressure. The larvae are presumed to die before reaching the lungs. Treatment generally includes high-dose systemic corticosteroids. Albendazole can be used in combination with steroids, but there is some concern that the rapid death of the larvae from the anthelmintic can worsen the inflammatory response.

Summary of Case

The infectious workup for this patient included testing stool for ova and parasites, CSF PCR for VZV, HSV, and EBV, and CSF evaluation for Syphilis, Rickettsia, Cryptococcus and Bartonella, all of which were normal. Evaluation for coccidioides and HIV was also normal. Cytology of the CSF revealed the absence of abnormal cells, but did show an abundance of eosinophils.  CSF and serum were sent to the CDC for ELISA testing of parasites, which confirmed the presence of Angiostrongylus cantonensis.  Repeat lumbar punctures revealed persistently elevated intracranial pressures. The patient was treated with oral corticosteroids for three weeks, as well as acetazolamide.

Struggle/Dilemma of the Clinical Presentation Description

Appropriate treatment for this condition was delayed, as it was initially misdiagnosed as a viral meningitis. Increased intracranial pressure is common in Angiostrongylus Eosinophilic Meningitis, and may have been detected earlier, if appropriate clinical suspicion was maintained.  The imaging was another clue to the diagnosis, but was not obtained during his initial hospitalization.

Keywords:

Meningitis, Increased intracranial pressure, 6th nerve palsy

References


"Lions and Tigers and Bears, Oh My!"

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History & Exam

A 79 year-old female s/p resection of a right CPA meningioma in 1998 noted sudden painless visual loss OD on 6-12-2016. Two days later examination revealed 20/150 vision OD, a right RAPD, and an otherwise normal ophthalmic exam. Her vision had been 20/40 OD on 5-31-2016. An MRI of the brain and orbits was interpreted as an intracranial meningioma of the right optic nerve. She was started on prednisone with subjective improvement and referred for radiation. Prior to radiation, she presented to us for a second opinion. Seen on 6-21-2016, she reported one month of generalized fatigue, a 10 pound weight loss and having a tick removed from her right upper lid in May 2016. Her visual acuity was 20/300 OD and 20/25 OS. She had a right RAPD and mild right temporal disc pallor. Visual fields on the right revealed a large cecocentral scotoma and were full on the left. The MRI was reviewed and revealed a thickened right prechiasmal optic nerve and chiasm with heterogeneous areas of both solid and peripheral enhancement, a previously unrecognized left temporal lobe infiltrating cortical T2 FLAIR hyperintensity, and a right frontal contrast-enhancing dural-based lesion. Postsurgical gliosis was noted in the right temporal lobe and pons secondary to prior meningioma resection. Bloodwork and lumbar puncture were unremarkable. She was tapered off prednisone and referred to neuro-oncology. Full body PET CT revealed a subpleural ground glass opacity in the left lower lobe with low-grade FDG uptake measuring 2.4 x 1.3 cm. A trial of azithromycin was initiated for the lung lesion with no change. What would you do next?

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History & Exam
A 68-year-old Hispanic woman presented to our emergency department with significant vision loss OS for 1 week. A month ago, she was evaluated locally for mild blurry vision OS, and corrected acuity was 20/30 OD, 20/40 OS. She was diagnosed with cataracts and epiretinal membrane OD. Medical history included smoking, hypertension, hypothyroidism, hyperlipidemia, transient ischemic attack, and aortic/mitral valve placement. Family history was positive for lymphoma and prostate cancer in father. Best-corrected visual acuity was 20/30 OD and 3/800 OS with RAPD OS and mild bilateral cataracts. Extraocular movements were normal. The right optic disc was small and crowded, and the left disc had diffuse edema. There were no other cranial nerve dysfunction. She had no headaches and no giant cell arteritis symptoms. ESR was 20, CRP was 1.3 (normal <1.0), and CBC was normal. Non-arteritic anterior ischemic optic neuropathy OS was suspected given the risk factors and crowded disc on the right, and the patient was placed on prednisone 60 mg daily. Three days later, her corrected acuity OS had improved to 20/150. The left disc edema persisted with a peripapillary hemorrhage nasally to the disc. Humphrey visual field showed temporal and inferior defects OS. Prednisone was continued at 60mg. Two days later, the patient presented with difficulty falling asleep, fatigue, severe stabbing left eye pain, scalp tenderness and neck pain. The corrected vision OS dropped to light perception with projection with an amaurotic pupil OS. A procedure was performed.

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"A diagnostic potpourri"

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History & Exam
A 63 yr. old incarcerated man with history of diabetes, hypertension, rheumatoid arthritis (RA) (on hydroxychloroquine, sulfasalazine and prednisone 7 mg), cutaneous lupus and Hepatitis C presented with sudden painless vision loss of the left eye without headache or symptoms of temporal arteritis. Visual acuity was 20/40 OD and count finger OS. He had left RAPD, quiet anterior segment OU, normal fundus on the right and optic disc edema on the left. Contrast enhanced brain and orbital MRI revealed an abnormal enhancing soft tissue involving left> right retrobulbar space, facial soft tissues, right temporal fossa, and bilateral infratemporal fossae.

Differentials included lymphoma, lupus panniculitis and idiopathic orbital inflammatory syndrome. His labs showed ESR of 102 mm/hr, CRP 3 mg/dl, ACE 32, normal CBC and BMP, negative serology for HIV and syphilis. Lumbar puncture revealed opening pressure of 18 cm H2O, normal CSF contents except mild elevation of protein (53). CSF and serum IgG levels were also increased without CSF oligoclonal bands. CT chest suggested interstitial lung disease involving bilateral apices. CT abdomen and pelvis was normal. Left orbital biopsy showed sclerosing fibro-proliferative process. Oral prednisone was increased to 60 mg daily. Six weeks later his visual acuity improved to 20/30 OD and 20/50 OS. Fundus exam remained normal on the right and showed disc pallor on the left. Prednisone was slowly tapered with a plan to add another immunosuppressive agent (Rituximab). A follow-up MR orbit was recommended. While waiting for initiating treatment with Rituximab, he presented 8 weeks later with painful complete vision loss in the left eye concerning for ophthalmic artery occlusion. Did he have another disease?

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