



FRANK B. WALSH SESSION

Sunday, February 22, 2015

7:45 a.m. – 5:00 p.m.

Co-Chairs: Benjamin M. Frishberg, MD & Howard R. Krauss, MD

Neuroradiologist: John Rhee, MD

Neuropathologist: Yuki Takasumi, MD

Neurosurgeon: Daniel Kelly, MD

Otorhinolaryngology/Head & Neck Surgeon: Chester Griffiths, MD, FACS

The theme for this year's Walsh Session is 'Some Like It Hot' based on the iconic movie which was filmed at the Hotel del Coronado in 1959. The session 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.

At the conclusion of this program, 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: Laura Bonelli, MD, MDCE & Peter Savino, MD

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***“Some Like It Hot”* Filmed at Hotel del Coronado**

Excerpted from the Hotel del Coronado website

Named #1 Comedy by the American Film Institute

Regarded by critics as one of the finest American movies ever made, *Some Like It Hot* continues to delight audiences 50 years after it debuted in 1959; in fact, the American Film Institute named it No. 1 on their list of the 100 best comedies of all time.

Filmed in 1958, the United Artists movie was shot on location at the Hotel del Coronado. The Del’s iconic Victorian architecture made it the perfect backdrop for the film’s 1929 setting, along with acting icons Marilyn Monroe, Jack Lemmon and Tony Curtis.

Plot

The Prohibition-era story follows the exploits of Lemmon and Curtis, (Joe and Jerry), out-of-work Chicago musicians who accidentally witness a gangland slaying. Making a run for their lives, the men disguise themselves as women and join an all-girl band traveling by train to Florida. Here, a ukulele-strumming singer, played by Monroe, catches the eyes of both men, but it is Curtis’ character who assumes still another identity – an unlucky-in-love millionaire – to successfully woo and win Monroe.

Lemmon’s cross-dressed character, meanwhile, is vigorously pursued by a bona fide millionaire, played by Joe E. Brown. The hilarious gender-shifting romantic romp is played out at California’s famed Hotel del Coronado, which director Billy Wilder found to be the perfect substitute for Florida in the Roaring Twenties.

An “Uproariously Improbable Set”

Like all American resorts, the Hotel del Coronado had endured some tough years during the Depression and World War II, but it was this period of benign neglect that helped preserve the resort, making it the perfect setting for Wilder’s 1929 story, which he co-wrote with I.A. Diamond. Said Wilder, “We looked far and wide, but this was the only place we could find that hadn’t changed in thirty years. People who have never seen this beautiful hotel will never believe we didn’t make these scenes on a movie lot. It’s like the past come to life.”

Although at least one critic didn’t believe the hotel was real, describing The Del as “an uproariously improbable set.” The hotel’s 1888 Queen Anne Revival-style architecture does tend toward the fanciful, with rambling white clapboard, lazy verandas and red-turreted roofs, which an earlier writer had characterized as a cross between an ornate wedding cake and a well-trimmed ship.

Although only exterior scenes were filmed at the hotel, the interior scenes do look very Del-like (right down to the placement of the lobby elevator and stairs). This probably explains why so many *Some Like It Hot* devotees – even after seeing the Hotel del Coronado for themselves – absolutely refuse to believe that the movie’s interior scenes were not filmed at The Del.

Only at The Del: The Stars Align

During filming, Marilyn Monroe was accompanied by her husband, esteemed playwright Arthur Miller (he made two special trips from the East Coast to join her at The Del). Also in Monroe’s entourage was acting coach Paula Strasberg, along with Monroe’s secretary and press agent. Coronado police officers were assigned to guard Monroe throughout her stay.

Meanwhile, Tony Curtis’ wife, Janet Leigh, was also on hand (Leigh was pregnant with their second child, Jamie Lee Curtis, at the time). Jack Lemmon’s future wife, Felicia Farr, also joined the troupe.

By almost everyone’s account, Monroe was very difficult to work with throughout the film’s production – her tardiness and inability to remember lines have become legendary. Interestingly, however, quite a few reports confirm that Monroe was “on her mettle” during the entire Coronado portion of filming.

In fact, in his book *Conversations with Wilder* (1999), writer/director Cameron Crowe addressed Billy Wilder about this aspect of the film, saying, “I grew up in San Diego [and] the legend is that the hotel was the most magical part of the filming ... that Marilyn felt relaxed there.”

To which, Wilder replied, “Yeah, that was fun. We had a good time there. Marilyn remembered her lines ... everything was going according to schedule.” Added Crowe: “Marilyn seems fully engaged in those scenes.”

According to another source, Wilder speculated that Monroe was inspired at The Del, where adoring spectators were plentiful because she preferred a live audience. Wilder later told Crowe that the Coronado fans were “screaming and yelling,” and then added that when he wanted the crowd to quiet down, he had her say, “‘Shhh’ ... they listened to her.” In the end, Wilder probably characterized Monroe the best, calling her “a calendar girl with warmth, with charm.”

And a last bit of Del trivia: During her stay, a hotel chef reported that Marilyn fancied his cold soufflé vanilla pudding with egg-white decoration, which she requested daily.

Favored by the Fans, Overlooked by the Oscars. The movie was a box office success, grossing over \$8 million initially and earning several million more over the next few years – somewhere between \$10 and \$15 million. Monroe's financial deal – she received between \$100,000 and \$300,000, as well as 10 percent of the film's gross profits – was a very lucrative arrangement in its day, and *Some Like It Hot* turned out to be her most profitable venture.

The movie was also a critical success. *Variety* called it the biggest hit of 1959; Monroe received a Golden Globe for her performance, as did Jack Lemmon. The film itself also won a Golden Globe for "best comedy."

In spite of its financial success and public accolades, the film received only one minor Academy Award for "Best Black and White Costume Design." Today it is thought that *Some Like It Hot* was just too risqué for 1959, when the big winner that year was *Ben-Hur* (also in the running for various Academy Awards were the likes of *Diary of Anne Frank*, *Room at the Top*, *Pillow Talk* and *Porgy and Bess*).

The *Some Like It Hot* story line is racy, and Monroe's costumes are incredibly revealing, even by today's standards (though, according to Wilder, Marilyn was not interested in fashion ... as long as the costumes revealed "something," she was satisfied). Ahead of its time perhaps, present-day reviewers marvel that the movie still comes across as such a wholesome film; this was Monroe's forte: she was sexy, but childlike. Although this is the Monroe film most shown on television today, the actress reportedly never liked her performance.

Fun Film Facts

Writers Wilder and co-author I.A. Diamond were inspired by another cross-dressing comedy, the 1932 German musical *Fanfare of Love*, and they deliberately set the story in the past because, as Diamond put it, "When all the costumes look peculiar to us, a guy in drag looks no more peculiar than anybody else."

Much like *The Del* itself – which was designed as it was being built – the last 15 minutes of *Some Like It Hot* was being written and rewritten as it was being filmed.

The film was shot in black and white because Wilder thought that male actors in female make-up would look too ridiculous in color. The black-and-white format – which also suited the period style of the film – did not appeal at all to Monroe, who contractually insisted that all her films be shot in color. Wilder was able to convince her that the 1920s setting would look more authentic in black-and-white. Interestingly, Wilder (who chose to make many of his movies in black and white) later said that *Some Like It Hot* was the one movie that would have benefited from color.

Although Wilder hired one of the world's most famous female impersonators to teach Lemmon and Curtis how to walk in high heels, Lemmon refused the help – he didn't want his character to be that adept as a woman.

What to Look For

Monroe was displeased at her initial entrance – also at the train station – and Wilder and Diamond concurred. They rewrote the scene so that Monroe's entrance was punctuated by steam blasts from the train.

The film clearly shows *The Del*'s two original front entrances. When the resort opened in 1888, the hotel offered a combined men's and women's entrance and a separate "unaccompanied" women's entrance, which afforded lone women travelers a discreet way to check in. Though the two entrances survived past the 1958 filming of *Some Like It Hot*, only one remains today.

In the role of gangster Spats Colombo, George Raft parodies the gangster role he played in the 1932 film *Scarface*, in which his character repeatedly flipped a coin. In *Some Like It Hot*, Spats Colombo is very irritated when he sees someone else flipping a coin, demanding, "Where did you pick up that cheap trick?" Raft – who didn't accompany the cast to Coronado – was at *The Del* in 1936, during the filming of *Yours for the Asking*.

There were two scenes that supposedly gave Monroe the most trouble: The scene where she knocks on the door and says, "It's me, Sugar" required 47 takes; another scene, where Monroe had to rummage through a dresser drawer for a bottle of bourbon, proved even more challenging, requiring 59 takes. In fact, Wilder claimed that after he put the cue inside one of the dresser drawers, Monroe couldn't remember which drawer it was in.

The last line – uttered by Joe E. Brown when he says to Jack Lemmon, "Nobody's perfect" – was never intended to remain the last line, but Wilder and Diamond couldn't come up with anything they liked better, so it stayed. Ironically, it has become a classic last line.

Some Like It Hot

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History & Exam

A 71 year old Caucasian man with a past medical history of hypertension, hyperlipidemia, Type 2 DM, ESRD status post renal transplant, facial melanoma, currently on ASA for a stable left sided putaminal hemorrhage presented with new right sided ptosis and lower extremity weakness. Brain MRI obtained on admission showed small cortical hemorrhages consistent with prior stroke, without evidence of acute pathologic changes. Repeat fine cut MRI of the brain and orbits with and without contrast showed an enhancing lesion in the right parietal bone, clinoid process, and associated abnormal soft tissue changes extending into the right orbital apex, adjacent superior right sphenoid sinus, and the right anterolateral cavernous sinus.

EMG showed no evidence of a neuromuscular disorder. Lung and parietal bone biopsy revealed CD10 positive atypical lymphoid cells. On laboratory evaluation, a CBC revealed a low red blood cell count. A lumbar puncture revealed an opening pressure of 12.5 mmHg with an Epstein Barr Virus load of 115 with normal cell counts otherwise. Lymphoma panel showed no abnormal lymphatic cells. Additional CT of the chest, abdomen, and pelvis revealed multiple well-circumscribed pulmonary nodules suspicious for pan-lobar metastatic disease involving both the right and left lungs in addition to profound mesenteric lymphadenopathy. Bone scan showed evidence of multiple areas of involvement in the skull, right humerus and left tibia.

Vital signs were normal. Visual acuity was 20/60 OD and 20/30 OS, color vision was intact, visual fields were normal and pupils were anisocoric (R 4mm, L 3mm both reactive) with near complete ptosis on the right with severely depressed levator function. His right eye was unable to adduct past the midline while the left eye had normal range of motion. There was an associated right-sided facial droop. Motor testing showed normal bulk and tone with normal reflexes. There were no sensory deficits.

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A Shot in the Dark

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History & Exam

A 74 year-old man with a chief complaint of difficulty with night time driving for several years as well as difficulty going from a lighted room to a dark room. PMH of high cholesterol, BPH, hypothyroidism, GERD, glottic squamous cell cancer without recurrence, and a prior history of alcoholism. POHx: cataract extraction right eye and AMD. Medications include Multivitamin, Aspirin, Synthroid, Tamsulosin, Omeprazole, Crestor. Drinks wine intermittently and stopped drinking after possible DT's. He owns a bar. His initial exam VA: 20/25 OU, normal IHCP, CFVF: OD constricted, OS normal. Pupils with no APD but were sluggish. IOP's 13 mm Hg OU. Motility showed some slight decreased abduction OU: 5ET primary, 10ET left, 5ET right gazes. SLE: PCIOL OD, NS cataract OS. Optic nerves had 0.7 cups with difficulty in appreciating any pallor. He had a few RPE macular changes OU. The HVF appeared reliable with severe constriction OD>OS (MD -29 and -17 respectively). RNFL OCT showed a mean thickness of 69 and 61 microns respectively. Testing was later done.

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Speakeasy

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History & Exam

A 64 year-old Caucasian gentleman presented with an unusual complaint. He stated that if he touched the inside of his right cheek with his tongue, he felt a tingly sensation in his R eyebrow. This had started one month ago. He also complained of foreign body sensation and discomfort in the right eye for six months. Additionally, he had recently noted a droopy R eyelid for which his local ophthalmologist had placed a stitch in his R upper lid to lift it. He also complained of recent binocular diagonal double vision. Past medical history included hypertension, hyperlipidemia, emphysema, and coronary artery disease requiring 4 stents (2002 and 2011). Social history was significant for 50 pack-years of smoking, and he drank 6 beers per week. His medication list included simvastatin, prasugrel, fluticasone-salmeterol, tiotropium bromide, aspirin, metoprolol, and hydrochlorothiazide. Exam revealed best-corrected visual acuity of 20/40 and 20/25. BP was 125/70. Pupils were 5.5 mm and 5 mm, with no RAPD. IOP was 19 and 21. Eye movements revealed -1 adduction, -3 supraduction, and -1 abduction in the R eye only. Visual fields were full to confrontation OU. External exam revealed ptosis with an MRD of -2 OD and +2 OS. Facial sensation to cotton tip was WNL bilaterally. Slit lamp revealed a decreased tear film OD and mild corneal scarring OD. There were 2+ nuclear sclerotic cataracts OU. Dilated fundus exam revealed pink, sharp optic discs with normal cups and spontaneous venous pulsation OS. A diagnostic procedure was then performed.

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Shady Double Crosser

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History & Exam

A 62-year-old woman presented with a one-month history of severe light sensitivity and headache, and a two week history of diplopia. Her photophobia was severe enough for her to wear sunglasses indoors. Her past medical history was significant for a cholecystectomy and Crohn's disease, for which she had undergone a partial colectomy. Her only medication was ranitidine. Review of systems was unremarkable. Neuro-ophthalmic examination revealed visual acuity of 20/20 OU. She had normal anterior segments, pupillary reactions, and fundi OU. Motility assessment showed an isolated left abduction deficit. The fifth and seventh cranial nerves were intact. There was no ptosis, proptosis, or nystagmus. CT scan of the brain revealed a 3.6 x 2.4 cm lesion centered on the left skull base. It extended to and filled most of the sphenoid sinus. There was bony destruction, with complete destruction of the petrous apex. The mass also broke through the clivus to extend into the premedullary cistern. There were areas of hyperdensity within the mass. MRI showed a large lesion of the left clivus and petrous bone. It was hyperintense on T1 and enhanced homogeneously with gadolinium. The mass was shown to extend to the lower aspect of the sella. The patient underwent a metastatic workup. Her blood work was significant for an elevated alkaline phosphatase of 155 and a carcinoembryonic antigen of 61.6. CT of the chest was unremarkable. Abdominal CT showed several hypodense liver lesions, suggesting metastases. A 2.5 x 2.1 cm hypodense mass was present in the pancreatic tail. CA19-9, a serum marker for pancreatic cancer, was less than one. A diagnostic study was performed.

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Under Pressure

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History & Exam

A 29-year-old woman was referred to neuro-ophthalmology clinic for 1 year of headaches and papilledema discovered 2 months earlier. She complained of recently blurred vision but no positional headache, pulsatile tinnitus, transient visual obscurations, or other neurologic issues. She used no medications and denied any recent illnesses except for thrombocytopenia (59k) on recent blood work, and had no significant past medical history. Her initial examination in 4/2009 revealed normal acuity with 20/20 VA OU, equal pupils without RAPD, and severe grade 4 papilledema OU with several choroidal folds through both maculae. No hemorrhages were noted. All other portions of the examination were normal. Her BMI was 27. Humphrey SITA Standard visual fields were performed and showed slight enlargement of the blind spot OU with a nasal step in the L eye. An MRI of the brain and orbits with and without gadolinium was normal with normal venous flow voids. CSF analysis showed an opening pressure of 26 cm H₂O with 1 RBC, 1 WBC, Protein 75, Glucose 69, and no abnormalities on cytology. Although she had atypical features, a preliminary diagnosis of Probable Pseudotumor Cerebri was made and she was initiated on acetazolamide, with improved papilledema over the next several months. Over the next three years, however, she developed worsening vision with multiple additional symptoms.

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Requiem for a Cabinet Maker (*Numb of the Above*)

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History & Exam

A 54-year-old white male presents with 3 weeks of painless horizontal nystagmus and 6 months of left sided forehead numbness. He reports a 20 lbs weight loss. Medical history includes a renal transplant 30 years prior. He takes prednisone 30 mg QOD and azathioprine. Visual acuity is 20/20 OU, color vision is 8 of 8 OU and confrontational fields are full OU. Pupils are equal and reactive without an R APD. Ductions are full but with end gaze nystagmus. There is no proptosis or ptosis but there is decreased V1 sensation on the left. Fundus examination is normal. A contrasted cranial and orbital MRI showed an enhancing mass in left superior orbit. He was treated with a Medrol dose pack and had mild improvement in symptoms. Left orbital biopsy was read as orbital fibrotic histiocytoma. One month later his examination showed NLP vision OS associated with an amaurotic pupil. He had decreased abduction OS. He had 4 mm of proptosis OS. Left V1 sensation was still diminished and fundus remained normal OU. He was admitted to the hospital and prescribed IV methylprednisone. Repeat MRI showed increased in size of left orbital mass, now involving the optic nerve. CT orbits showed adjacent bone demineralization. He underwent left orbital radiation. Five months later, patient presents to ED with shortness of breath and transient visual loss OD. Vision remained 20/20 OD and NLP OS. Ductions now show limited superior gaze OD and limited all directions OS. Decreased left sided V1 and V2. No optic disc edema OD and mild optic nerve edema OS. MRI shows enlarging left sided orbital mass with a new right-sided retro-orbital mass. Further work up reveals new bilateral pulmonary nodules and metastatic appearing hepatic lesion. A diagnostic procedure was performed.

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Three Weeks in Florida

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History & Exam

A 36 year-old man presented with severe headaches, bilateral leg numbness, and bilateral decreased vision. He was born in Ecuador where he received BCG vaccination and immigrated to US at age 19. In 2005 he enrolled in nursing school and volunteered in homeless shelters. PPD was positive with a negative chest x-ray. In August 2011 he developed pleuritic chest pain; chest x-ray showed a 2 cm cavitory lesion in the right upper lobe. He was diagnosed with active TB and completed treatment in August 2013. In December 2013 he developed headaches associated with neck stiffness and intermittent blurry vision which progressed over 6 months. In May 2014, while on vacation in Ecuador, he had a prolonged seizure requiring intubation. Upon recovery he returned to Florida and presented to the emergency department with visual loss and drowsiness, but arousable to verbal stimuli. Vision was light perception OU with sluggish pupils. Extraocular movements were full but demonstrated exotropia. Fundus examination showed papilledema. Strength was reduced in all four limbs with apraxia in the upper right. Sensation was decreased from groin to foot, left worse than right. Serum white count was elevated to 25,300. Brain MRI demonstrated hydrocephalus and diffuse leptomenigeal enhancement including bilateral intracranial optic nerves and chiasm. CSF analysis showed 110 leukocytes with 66% monocytes. Lumbar spine MRI was negative. The patient was diagnosed with recurrent TB with meningitis and was restarted on TB medications. CSF smears, cultures, and PCR were negative for infection including TB. Lumbar drain was placed and ICP was 40 cm water. Repeat brain MRI showed progressive leptomenigeal enhancement and hydrocephalus. Repeat CSF analysis demonstrated cellular atypia with equivocal immuno-histochemical staining. Flow cytometry revealed a mixture of immune cells but no B lymphocytes. A diagnostic procedure was performed.

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Growing Suspicion

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History & Exam

A 44 year-old man presented to the emergency department in July 2011 for progressive painless vision loss in the right eye for 3 months. On presentation, vision was 4/200 in the right and 20/20 in the left with an afferent pupillary defect on the right. His visual field was full to confrontation but automated perimetry revealed a central scotoma. The remainder of the exam was normal with the exception of slight elevation of the optic nerve head along with mild perivascular sheathing. He received IV solumedrol for 3 days followed by an oral steroid taper. Fat saturated MRI did not show enhancement of the optic nerve nor any brain abnormalities or mass lesions. He had a normal lab workup including CBC, BMP, quantiferon gold, B12, RPR, and ACE and a normal CXR. On follow up one month later, he experienced no vision improvement. At this point, testing for Leber's hereditary optic neuropathy (LHON) was performed and was negative. He returned 6 months later with subjective worsening of vision in the right eye, however acuity was stable at 4/200. He was started on IVIg therapy and autoimmune and NMO antibodies were drawn. Antibody testing was negative and on follow up one month later, his acuity remained unchanged and his scotoma was larger and denser. He was again lost to follow up for 3 years until he began losing vision in the left eye. Exam revealed counting fingers vision in the right eye and 20/40 with a temporal visual field defect in the left eye. MRI showed an enhancing mass extending from the planum tuberculum and suprasellar area to the right temporal lobe and into both orbits. A procedure was performed.

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Hiding and out of Sight

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History & Exam

A 75-year-old white woman presented with a history of biopsy-proven giant cell arteritis (GCA) presented with recurrence of severe left sided headaches and left global ophthalmoparesis for 4 days. GCA had been diagnosed 4 months prior by biopsy. Left eye vision loss occurred when an outside physician tapered corticosteroid therapy 5 weeks into her illness. MRI 5 weeks into her illness showed enhancement of the left orbital apex, adjacent temporal dura and neighboring cavernous sinus. Dural biopsy at that time was normal, and a second temporal artery biopsy performed then confirmed GCA. Examination was remarkable for left eye vision loss to no light perception with an amaurotic pupil and global left ophthalmoparesis including ptosis and pupil involvement with a normal fundus. Repeat MRI showed progression of the prior lesions. A diagnostic procedure was performed.

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It's Not Just a FAD (*EHR Fatigue Syndrome*)

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History & Exam

A 29 year-old female nurse, nine months postpartum, presented with an inability to see her computer well for the past two months. She denied eye pain, diplopia, numbness, tingling or weakness. There were no changes in vision in bright vs. dim lighting. She also had a headache at the back of her head for two months that was relieved with OTC medications. She denied any events immediately preceding the blurred vision. She also complained of shortness of breath, unexplained weight loss and extreme fatigue, sleeping 10 hours per night and taking naps over her lunch break at work. Workup by her primary care doctor revealed a normal CXR, ECG with sinus bradycardia and anemia (Hgb 9.6, Hct 28.7). Towards the end of her recent pregnancy she was evaluated for polydipsia (drinking up to 9 L /d) and nocturia (6-7 x /night). Water deprivation testing during pregnancy was not possible but sodium of 133 made the diagnosis of diabetes insipidus (DI) unlikely. Symptoms improved after delivery and therefore the polyuria and polydipsia was attributed to pregnancy. Postpartum she also developed fairly severe anxiety and depression that was managed with sertraline and clonazepam. On examination, best corrected visual acuity was 20/30 OU, Ishihara color plates were 11/13 OD, 13/13 OS. Pupils reacted briskly without an RAPD. Visual fields were full to confrontation. Slit lamp and dilated fundus examination was unremarkable, including absence of macular abnormalities or optic disc pallor. Goldmann fields showed a suggestion of a homonymous field defect, but had variable responses and the perimetrist noted that she was often falling asleep during the test. Optical coherence tomography showed a normal average retinal nerve fiber layer thickness OU. Lab workup showed an elevated ESR of 70 and an elevated ACE of 62. Tests were performed.

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Now you see it. Now you don't.

Prem S. Subramanian

Wilmer Eye Institute Baltimore, MD, USA

History & Exam

A 70 yo RH African American female presented to our institution with sequential, severe vision loss in the preceding 4 months. Her past medical history included well-controlled hypertension and hyperlipidemia, CAD s/p coronary stent placement x 2 after MI, and stroke. She had no prior history of visual loss or eye surgeries. She was well until January 2014 when she was hospitalized for severe cholangitis complicated by septic shock requiring vasopressor support in the ICU. She does not recall exactly when she lost vision in her left eye but suspects it was during the hospitalization. In mid-April 2014 she was noted with Va 20/70 OD and NLP OS, had a left RAPD, and possible left optic disc pallor. Decreased vision OD was consistent with cataract, and uncomplicated surgery was performed on 5/2/14. She noted poor vision postop, and on 5/5/14 was HM OD and NLP OS with possible pallor of the right optic disc. ESR reported 3 days later was 102, and she was prescribed prednisone 60 mg po qd. Bilateral temporal artery biopsies were performed and were read as negative. Prednisone was tapered over the next month to 10 mg daily with some vision improvement OD. She then presented to our institution on 7/10/14 with Va 20/400 OD and NLP OS. The left pupil was amaurotic. Fundus exam showed mild pallor of both optic discs. OCT of the RNFL and macular GCC demonstrated diffuse loss. She remained on 10 mg prednisone daily at that time and felt her vision was slowly improving. A diagnostic procedure was performed.

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A Weak Presentation

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History & Exam

An 82-year-old right-handed man with myasthenia gravis presented in May 2014 with double vision and right facial numbness and weakness. He was first seen in 1998 with horizontal diplopia. He had an abduction deficit of the right eye, and right nasolabial fold flattening. He was diagnosed with myasthenia based on a positive acetylcholine receptor blocking antibody. His chest CT scan was negative for thymoma. His diplopia and facial weakness resolved with azathioprine and prednisone. He had central retinal vein occlusion (CRVO) in the right in 2002, with resultant optic neuropathy and central vision loss. He had removal of innumerable squamous and basal cell carcinomas, coronary artery disease, prostate cancer with prostatectomy and laryngoplasty. In 2009, he first noticed right brow numbness. He had surgery for ectropion OD in April 2013. In May 2013, he developed dysesthesia of his right brow. A basal cell carcinoma was removed without benefit. In July 2013, his myasthenia was stable, but his azathioprine was decreased due to reduced platelets and hematocrit. In September 2013, he developed stabbing pain of his right cheek, with right cheek sensory loss and right facial weakness. A MRI in November 2013 showed a small enhancing intraconal mass, not in an MRI from 2006. In April 2014, he had Mohs excision of a poorly differentiated scalp squamous cell carcinoma. In May 2014, his afferent examination was stable. Eye movements showed new -2 limitation of abduction OD. He had sensory loss of his right cheek, and right facial weakness. Increasing his prednisone dose did not improve his eye movements. Repeat brain MRI in May 2014 showed increase in size of the orbital mass. The third, fourth, fifth, sixth, and seventh cranial nerves appeared normal. A diagnostic procedure was performed.

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The Man with No Face (*About Face*)

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History & Exam

A 61-year-old man presented with one week history of decreased vision OS. Past medical history includes asthma and amblyopia OD, but with complete visual loss OD 18 years prior. He takes no medications. He is retired but worked for 21 years in social science research. The remarkable thing upon meeting this articulate intelligent individual is he had no face. On exam his visual acuity was NLP OD and HM OS. There was no view of the pupil OD and a poor view OS but there is minimal reactivity. Color vision was non-recordable. Confrontational visual field showed marked constriction OS. Extraocular muscle exam showed complete restriction OD and a small amount of movement in all cardinal directions OS. External exam OD revealed absence of eyelids, exposed globe, and dense corneal scar with no view into the anterior chamber. External exam OS showed thick, keratinized upper and lower lids temporally and absence of lids medially, and 360 degrees of conjunctival chemosis, diffuse corneal edema with white lesion inferonasally, and a formed AC with poor view. Intraocular pressure was non-measurable OD and 40 mm Hg OS. No view of the fundus OU. Cranial and orbital CT and MRI revealed extensive destruction of soft tissue of the face that abuts and likely invades the dura. The right globe was involved, the left globe appeared intact. The intracranial and prechiasmatic portion of the optic nerves appeared normal. There was no definite evidence of involvement of the cavernous sinuses. However, there was disease surrounding several intracranial nerve roots, which likely represents perineural invasion of these structures. A biopsy was obtained.

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Who Deserves a Second Chance?

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History & Exam

An 81-year-old woman presents with a one-month history of blurred vision OS, acutely worse in the past 5 days. She reports pain with left gaze, left sided forehead tenderness and some weight loss. Medical history includes hypertension, borderline diabetes, cerebral vascular accident and basal cell carcinoma of the face. Her medications include rivaroxaban, metoprolol, atorvastatin and omeprazole. Visual acuity is 20/20 OD and light perception OS. Color vision is 11/11 OD and 0/11 OS. Confrontational fields are full in the right eye and non-recordable in the left eye. Pupils are reactive and equal OU, with a greater than 1.2 log unit APD OS. Anterior segment exam reveals bilateral intraocular lenses. Dilated exam: OD is unremarkable, and OS is noted to have pallid disc edema with several hemorrhages. Initial blood work reveals an ESR 36mm/hr and CRP 5.4 mg/L (normal <3.0). MRI head obtained at outside facility showed abnormal enhancement along the orbital portion of the left optic nerve. Prednisone 60mg/day is initiated for presumed GCA and temporal artery biopsy scheduled. Contact is also made with the patient's primary care physician to stop rivaroxaban. A left temporal artery biopsy showed calcific atherosclerosis, without evidence of active or treated arteritis. Vision in left eye progressed to no light perception. Dedicated orbital MRI study with and without gadolinium showed mild enlargement and enhancement of the left optic nerve, possibly worse than prior. Patient admitted to hospital for workup. CT Chest – no evidence of sarcoidosis. LP – glucose 113 (serum 276), protein 31, WBC <1, cytology negative for malignant cells. Repeat dilated exam showed increased vitreous cells and debris in the left eye. A procedure was performed...

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Lights Out

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History & Exam

A 78 year-old man presented with a one-month history of progressive painless binocular vision loss. He had sustained head trauma without loss of consciousness three days prior to the onset of vision loss. On the morning of his presentation to us, he had awoken with complete binocular vision loss and had been started on oral prednisone (70 mg daily) by his local eye care provider. He denied symptoms of giant cell arteritis. His past medical history was remarkable for hypertension, diabetes, and a distant history of prostate cancer that was thought to be in remission. Neurologic review of systems was unremarkable. Examination revealed no light perception OU. The pupils were dilated and minimally reactive to light. There was no RAPD. Intraocular pressures were within normal limits OU. Extraocular movements were full OU. Anterior segment examination revealed pseudophakia OU. Dilated funduscopic examination revealed diffuse optic atrophy OU. Neurologic examination was unremarkable.

There were no choroidal filling defects or perfusion abnormalities on fundus fluorescein angiography. ESR, CRP, and PSA were all within normal limits. MRI brain revealed enlargement and heterogeneous enhancement of the optic chiasm. An enhancing lesion was seen adjacent to the anterior horn of the right lateral ventricle. CSF examination revealed markedly elevated protein and glucose levels, with negative cultures, cytology, and flow cytometry. Bone scan was negative for bony metastases. In an attempt to improve his vision, intravenous methylprednisolone was given for five days, followed by a taper of oral prednisone over seven days. The right frontal lesion decreased in size dramatically with the steroid treatment, making biopsy difficult, but his vision did not improve. Repeat MRI brain obtained two weeks after completion of the steroids revealed interval growth of the right frontal lesion, with surrounding vasogenic edema, and growth of the chiasmal lesion. A diagnostic procedure was performed.

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Star Spangled Banner

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History & Exam

A 12-year-old girl with a history of bilateral optic nerve enlargement, enterovirus meningitis, seizures, and bilateral hygromas, presented with acute onset chronic vision loss in her left eye. Two years prior, she presented to an outside hospital with headaches, intermittent speech arrest and right-sided weakness, and was diagnosed with enterovirus meningitis and seizures. Incidentally, she was noted to have bilateral optic nerve enlargement. She had no visual symptoms until four months prior to presentation when she was again admitted with headaches and transient neurologic deficits. Neuroimaging was notable for new bilateral hygromas, further enlargement of the optic nerves, and septations of the subarachnoid space thought to be related to prior meningitis. Lumbar puncture was unremarkable, and PET scan revealed a paraspinal mass near the left L4-L5 nerve root. Biopsy was consistent with neuroblastic tumor, thought to be isolated, and managed with partial resection. Additionally, extensive dural ectasia of the spine was observed. An attempt to drain the hygromas revealed high CSF output that required subgaleal-peritoneal shunt placement, later removed due to infection. During her admission, she reported blurred vision. Initial ophthalmological evaluation revealed subnormal acuities in the left more than right eye and dyschromatopsia in both eyes, with normal pupils and optic discs. The etiology of vision loss was postulated to be optic neuropathy secondary to high intracranial pressure from her hygromas. After discharge she remained stable for another three months until she noted complete vision loss in her left eye and was referred to our facility for neuro-ophthalmological consultation. Her examination was significant for visual acuity of 20/60 in the right eye and light perception in the left eye, a left afferent pupillary defect, and bilateral optic disc temporal pallor with nasal edema in the right eye. There were no Lisch nodules.

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Joe & Jerry Flew the Coop

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History & Exam

A previously healthy 13-year-old girl presented to a local hospital with fever and myalgia, followed one day later by lethargy and vision loss. Past medical history was significant only for acne, for which she had been treated with doxycycline 40 mg/day intermittently starting 2 months prior to symptom onset. In the emergency department, the patient was difficult to arouse. Within 24 hours of onset, arousal level spontaneously returned to normal, but vision was light perception in both eyes. Based on fundus examination, she was given a presumptive diagnosis of neuroretinitis and transferred to our hospital. On our examination, visual acuity was light perception only in both eyes. Pupils measured 7mm in dim illumination and constricted moderately to direct light. Ophthalmoscopy in both eyes revealed nearly confluent, sharp-bordered ischemic retinal white patches in the posterior pole. Optical coherence tomography (OCT) showed inner retinal thickening and hyperreflectivity. Fluorescein angiography (FA) revealed occlusion of multiple small arterioles in the areas of retinal whitening. A wide-field FA confirmed multifocal arteriolar occlusions posteriorly with minimal late leakage and no retinal vascular abnormalities in the periphery. Brain MRI demonstrated symmetric T2 hyperintensities on FLAIR images in the region in both LGBs, and in the cerebellar vermis and dorsal midbrain. T2-weighted gradient echo images showed hypointensities with blooming in the LGBs, indicative of hemorrhage. These lesions showed restricted diffusion. ESR was 46, CRP was 0.1 and the following labs were negative: cardiolipin antibody, anti-DsDNA, anti-SSb/anti-La, anti-Sm, anti-RNP, anti-scleroderma, anti-Jol, chromatin, ribosomal protein and centromere B. An additional test was performed.

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Using Muscle

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History & Exam

A 30- year-old Saudi female student with no past medical history presented an eight month history of progressive blurred vision primarily at near, anisocoria OS, and periocular discomfort with eye movements. Neuro-Ophthalmologic examination revealed best corrected visual acuity of OD 20/20 and OS 20/30. Color vision was OD 12/12 and OS 12/12. Automated threshold perimetry was normal. The pupils were 4 mm and normally reactive OD and 5 mm and minimally reactive OS. There was no relative afferent pupil defect (RAPD). The pupil was characteristic of absent sympathetic and parasympathetic input as may be seen in cavernous sinus lesions. The eyes were quiet. IOP was normal. Ocular motility examination was normal at initial evaluation but over the next 3 months she developed limited adduction and mild proptosis; other eye movements were normal throughout her 10 month course. Funduscopy was normal. Magnetic resonance imaging (MRI) of the brain and orbit with contrast revealed an enlarged left medial rectus and a linear enhancing structure running from the medial rectus muscle back to the superior orbital fissure and superior lateral wall of the cavernous sinus, believed to be an enlarged, enhancing third nerve. The following laboratory studies were performed and were negative or normal: Angiotensin-converting enzyme, lysozyme, T4, T3, TSH, thyroid stimulating immunoglobulin, thyrotropin binding inhibitory immunoglobulins, thyroid peroxidase antibody, serum protein electrophoresis, cysticercosis antibodies, trichinella immunoglobulin G, and IgG4 immunoglobulin. A Quantiferon Gold test was sent three times before receiving a report that it was positive after all the above evaluation and the diagnostic procedure were complete. A diagnostic procedure was done

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I Can't See Straight

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History & Exam

In May of 2014 this 30 year old right handed patient was referred for consultation regarding diplopia and dizziness. The patient relates that she had been told that she had “tired eyes” as a child. Two and a half years ago she began to have intermittent exodeviation. She was seen locally and diagnosed as having a IV nerve palsy and treated with 2 diopters of base in prism and 2 diopters of base up prism. Six months before her referral she began to complain of double vision. The patient attributed the onset of double vision to a car accident in 2007. On examination visual acuity was correctable to 20/20 and 20/25. Color was intact. Visual fields demonstrated an inferior Seidel scotoma on the right side. There was no afferent pupillary defect. Motility examination revealed severe limitation in up gaze OD > OS with 45 diopters of right exotropia and 12 diopters of right hypotropia. Counter clockwise torsional nystagmus was noted. Slit lamp examination and applanation tensions were within normal limits. Funduscopy examination revealed no evidence of disc edema or optic atrophy. OCT revealed some epiretinal surface changes, but no obvious epiretinal membrane. Imaging studies were obtained.

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Nobody's Perfect

Alexander Ksendzovsky, Steven A. Newman

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History & Exam

In June of 2006 an 8 year old patient was referred for evaluation. Apparently at age 1 ½ she had developed headaches and was found to have a posterior fossa tumor. In El Salvador she was treated with shunting and chemotherapy plus radiation therapy for presumed medulloblastoma. She underwent a shunt revision at age 2 ½. Near vision was 20/20 OU with arcuate visual field defects, no afferent pupillary defect, normal motility, and normal discs including OCT. She did have an MRI which showed some likely post radiation changes, but a negative spinal MRI. On June 19, 2007 a repeat MRI scan, however, demonstrated a lesion at the L1 level. In March of 2006, a 29 year old patient was referred for a 6 month history of problems with his vision, and two weeks of headaches characterized as steady, and unassociated with nausea or vomiting. Visual acuity was correctible to 20/20 and 20/25. Visual fields demonstrated a bitemporal superior visual field defect. He had evidence of a 0.6 log unit left afferent pupillary defect. There were full ductions and versions and 100 seconds of stereopsis. His past medical history was remarkable for an incomplete form of osteogenesis imperfecta. MRI scan showed evidence of a sellar and suprasellar enhancing mass. A meningioma was suspected. A spinal MRI scan, however, demonstrated evidence of lumbar enhancement. CSF analysis was unremarkable.

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