

# Grand Rounds

8-05-09

## Introduction: James Goodwin, MD (*Attending*)

This week's neuro-ophthalmology grand rounds reviews three interesting cases encountered by our faculty.

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## Optic Nerve Enlargement: Trucian Ostheimer, MD (*Resident*)



An 18 year-old Caucasian male initially presented at an outside hospital with a chief complaint of blurry vision and pain affecting his right eye. The patient stated that his symptoms began with pain and stiffness in his neck lasting two days, followed by right eye pain exacerbated by movement of the eye. The patient also noted blurring of his nasal visual field and altered color vision in his right eye one week later. The patient's history is unremarkable except for a family history of scleroderma. Initial ophthalmologic evaluation revealed a relative afferent pupillary defect in the right eye, marked disc swelling with hemorrhage, and vision reduced to finger counting. The general medical workup was reportedly normal but MR imaging of the brain and orbits displayed thickening and enhancement of the intraconal and prechiasmatic portions of the right optic nerve with irregularity of the nerve surface. The patient was treated with IV methylprednisolone pulses for four days followed by oral prednisone and experienced symptomatic improvement.

The patient was referred to Dr. Goodwin and evaluated approximately one month after the onset of symptoms. At the time of this exam, the patient's manifest refraction was 20/20 and 20/15 with normal color vision, pupils, motility, and intraocular pressure. The anterior segment examination was unremarkable but the dilated fundus examination revealed 1+ cells in the anterior vitreous, a minimally swollen disc with a resolving flame hemorrhage, and an epiretinal membrane involving the macula in the right eye. Visual field examination (Octopus) displayed depression in the inferior quadrants primarily on the 10-degree and 30-degree static threshold exam.



**FIGURE 1**  
Orbital MRI with contrast demonstrating enhancement and irregular thickening of the intraconal and prechiasmatic portions of the right optic nerve.

**BACKGROUND** The differential diagnosis of optic nerve enlargement is vast, but considering this patient's presentation and findings, particular consideration was given to the possibility of sarcoidosis, optic neuritis (demyelinating, infectious, parainfectious, autoimmune, radiation-induced), Wegener's granulomatosis, and idiopathic orbital inflammatory syndrome. Dr. Goodwin felt that the combination of the patient's visual symptoms, disc swelling, and MRI findings were most likely suggestive of optic neuropathy secondary to granulomatous inflammation giving lumpy enlargement of the nerve, the most common cause of which is sarcoidosis in our region. Visual system abnormalities are the most common extrathoracic manifestation of this disease, and virtually any part of the globe or orbit can be involved. Ideally, diagnosis is confirmed by biopsy which must be interpreted in the context of the patient's clinical presentation and correlated with other laboratory or imaging tests. If sarcoidosis is strongly suspected as in this patient, the treatment is typically corticosteroids.

## Palinopsia: Javaneh Abbasian, MD (Resident)



A 38 year-old male came to neuro-ophthalmology clinic for evaluation after falling 15 feet from a scaffold to the ground. The patient recalls bleeding from his right ear and suffered a temporal bone fracture, several broken ribs, and a broken right clavicle. He denied visual symptoms immediately after the accident but reports that six months following the accident, he experienced episodic visual disturbances. He described seeing multiple images displaced horizontally and vertically, although he denied true diplopia. He also described moving objects leaving a trail of images behind the object.

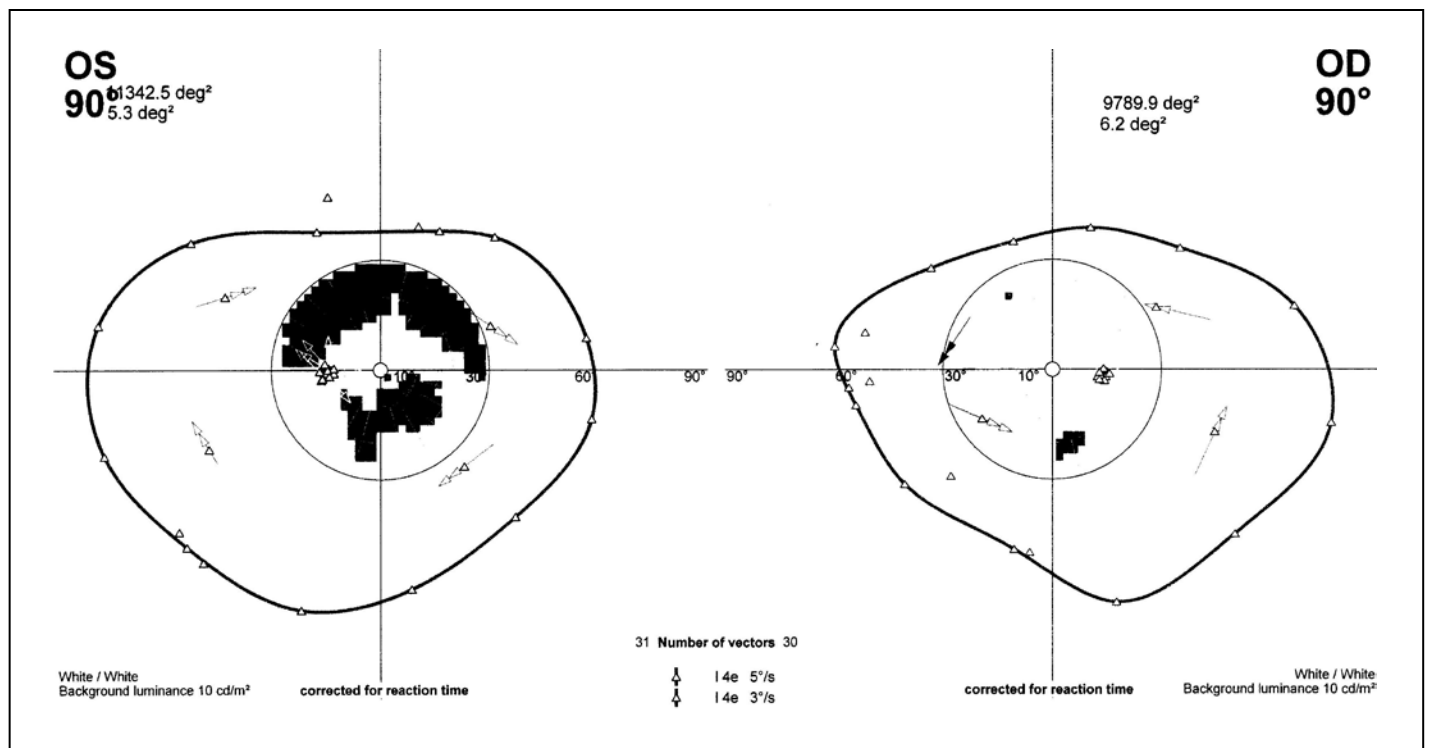
His past medical and surgical history was non-contributory. The patient was taking medications for psychiatric problems after the accident, namely Effexor, Abilify and Lorazepam. The best corrected visual acuity OU was 20/20, color vision was full in both eyes and motility was full. He had a 1 prism diopter esophoria and less than 1 prism diopter left hyperphoria in primary gaze. His pupil exam did not demonstrate a relative afferent pupillary defect and his intraocular pressure was within normal limits. The slit lamp exam and dilated fundus exams were normal in both eyes. Octopus

visual field testing are shown below. (fig 2)

The differential diagnosis for the symptoms described included phosphenes, photopsia, palinopsia, visual hallucinations and seizures. This patient exhibited palinopsia defined as a trail of images after a moving object. He also experienced polyopsia, multiple overlapping images of a stationary object. These findings were consistent with damage to the parieto-occipital convexity where higher cortical visual processing is carried out. In correspondence with the patient's neurologist, it was suggested that seizure mechanism for the paroxysmal polyopsia also be ruled out.

**BACKGROUND** Palinopsia is a symptom distinct from the physiologic after-image in which images of an object persist or reappear after the patient has stopped looking at the object. This symptom was first described by Critchley in 1951 as a type of visual perseveration. In some cases patients see a series of "echoes" of the object, separate from but in proximity and identical to the actual object, giving the impression of polyopia. As in this patient, object movement may make the original object appear to leave a trail of images along its path.

Palinopsia usually is associated with large destructive lesions (infarction, tumor, trauma, AV malformation) of the parieto-occipital cortex. Other causes include seizure discharge, migraine, anoxia, toxic-metabolic disturbances and many medications. Most patients have right hemisphere lesions producing an incomplete left homonymous hemianopia although it has been suggested that left sided lesions may be underrepresented secondary to aphasia.



**FIGURE 2**  
Octopus visual field demonstrating superior arcuate and inferior paracentral defects in the left eye. The right eye is essentially full.

## Traumatic Chiasmal Syndrome: Kimberly Truax, MD (Resident)



A 22 year-old white male was referred to the neuro-ophthalmology clinic at UIC by neurosurgery. On February 1, 2009 he had a severe head injury when he was thrown from a moped into a metal fence in Key West, FL. He was immediately flown by helicopter to Jackson Memorial Hospital in Miami. Per the patient's family, he had multiple fractures including the frontal bone, both orbits, the maxilla, and the nose. He underwent surgical repair of the frontal bone, the left orbit, and the maxilla. Prior to surgical

repair of the orbit, it was apparently known that he had lost vision in the left eye.

The patient was intubated for a long period of time, requiring a tracheostomy. He was nonverbal until March 5. On March 9 he was transferred to a rehabilitation center in Chicago. Finally he began breathing through his nose, and it was noted that air was entering the CSF through a defect in his nasal cavity. He underwent tissue grafting at Northwestern University Hospital to repair the defect.

The patient was currently residing with his father in the Chicago suburbs and noticing significant problems with memory. Current medications included gabapentin and trazadone. On examination, his right eye was correctable to 20/30 with -2.50 sphere; his left eye was light perception. Color vision was full in the right eye. He had a 2.1 log left eye relative afferent pupillary defect. His motility was restricted to 50% of expected range in upgaze bilaterally; the left eye only had trace downward movement. Hertels, IOP, and anterior segment exam were within normal limits. Dilated fundoscopic exam was significant for a pale left optic nerve. Goldmann visual field testing demonstrated total field loss in the left eye and a temporal hemianopsia in the right eye (fig 3). He was diagnosed with traumatic chiasmal syndrome.

**BACKGROUND** Traumatic chiasmal syndrome is very rare because most people do not survive trauma severe enough to damage the anatomically-privileged chiasm. There is a strong association with frontal bone fracture. A study by Hasan et al reviewed 19 cases of traumatic chiasmal syndrome, and they found that 9/19 patients had a bitemporal hemianopsia and 10/19 had total field loss in one eye and a temporal hemianopsia in the fellow eye.

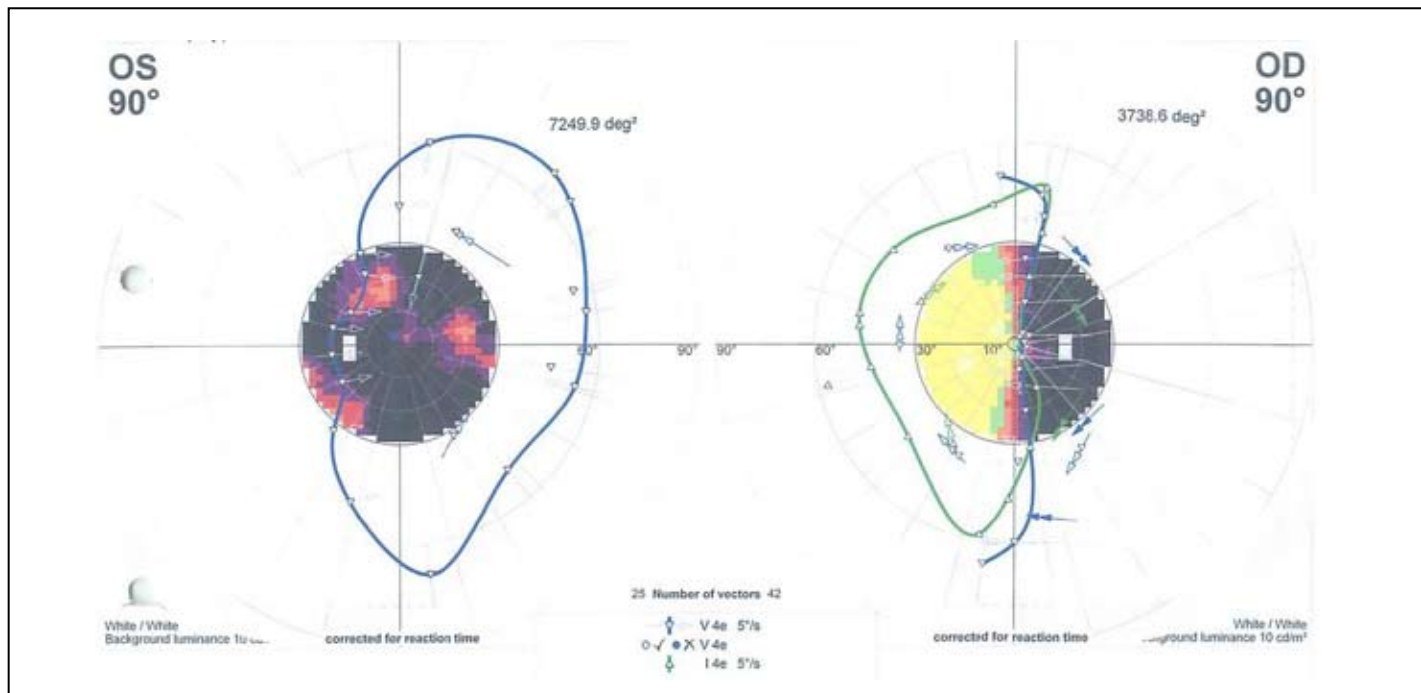
Adjacent structures are rarely spared in traumatic chiasmal syndrome. CSF rhinorrhea, carotid-cavernous fistula, panhypopituitarism, and pneumatocele have been reported. Cranial nerve deficits occur, including anosmia, ocular motility deficits, and hearing loss. Approximately 30% of patients develop diabetes insipidus.

The mechanism of traumatic chiasmal syndrome is poorly understood. It may result from a mechanical stretching/tearing of the nerve fibers, compression by surrounding hemorrhage, or compression by cerebral edema. Likely, the mechanism varies in each case and/or results from a combination of these insults.

**REFERENCES:** 1) Hassan et al. Traumatic chiasmal syndrome: a series of 19 patients. Clin Experiment Ophthalmol. 2002 Aug; 30(4): 273-80.

2) Kawai, et al. Traumatic chiasmal syndrome presenting with bitemporal hemianopsia. J Trauma. 1998 Jan; 44(1): 224-9.

3) Foroozan, R. Chiasmal Syndromes. Current Opin Ophthalmol. 2003 Dec; 14(6) 325-31.



**FIGURE 3** Octopus Goldmann visual fields demonstrating temporal hemianopsia in the right eye, and nearly complete visual field loss in the left eye.

## Discussion:

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The differential diagnosis of optic nerve enlargement is wide, and includes etiologies such as inflammatory conditions, tumor/mass, and infections. In this patient, the findings and presentation led us to the diagnosis of sarcoidosis, which was supported by imaging. Sarcoidosis is a condition that can have manifestations in virtually any segment of the eye. Although anterior granulomatous uveitis is more commonly associated with sarcoid, posterior manifestations can include optic neuritis, periphlebitis, retinal vascular disease, CME, and granulomas in choroid, retina, vitreous, and optic nerve.

The second case had cerebral polyopia and palinopsia as an interesting presentation in a patient who presumably suffered right parieto-occipital cortical injury after a fall. A careful history to rule out symptoms caused by medications, toxic-metabolic etiologies must be performed. Work-up should include EEG to look for possible seizure mechanism for this usually paroxysmal visual symptom.

Traumatic chiasmal syndrome, as mentioned above, is exceedingly rare. Traumatic injury to the chiasm often occurs in severe head trauma, and often is associated with damage to several intracranial structures, including the pituitary and cavernous sinus.

## UPCOMING CME COURSES

March 13-19, 2010	Illinois Eye Review
April 4, 2010	Retina Symposium
April 16, 2010	Uveitis Symposium
May 21, 2010	Oculoplastics Symposium
June 25, 2010	34 <sup>th</sup> Annual Alumni Day
September 25, 2010	Pediatric Ophthalmology/Adult Strabismus

## Upcoming Grand Rounds

Illinois Eye and Ear Infirmary Ophthalmology Grand Rounds are held Wednesdays at 5:00 pm on the UIC campus at 909 S. Wolcott in the College of Medicine Research Building. For a complete schedule go to [www.uic.edu/com/eye](http://www.uic.edu/com/eye) and click on Grand Rounds under the Education drop down menu. Or, call 312-996-6590.