

# Grand Rounds

8-12-09

## Introduction: Amjad Ahmad, MD (*Attending*)



This week's oculoplastics grand rounds discusses interesting cases encountered by our faculty.

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## Cavernous Hemangioma: Jennifer Hu, MD (*Resident*)



A 44 year-old Asian woman presented with chronic dry eyes and "eyes looking wide for many years." She was seen by an outside ophthalmologist who, given question of lid retraction, ordered a CT scan of the orbits to rule out Grave's disease. Incidentally, on CT she was noted to have an orbital mass in the right eye, presumptively a cavernous hemangioma. She was then seen at the IEEI oculoplastics clinic for a second opinion, without any visual complaints or ocular pain. On exam, her vision, pupils, motility, confrontation visual fields, and Ishiharas were normal. She had 3 mm of right proptosis on Hertel's and bilateral lid retraction. Anterior segment and fundus exam were normal. On CT with and without contrast, there was a well-defined soft tissue density in the right retro-orbital space between the medial rectus and optic nerve, with the

optic nerve being stretched along the lateral aspect of the mass (fig 1). The mass enhanced with contrast, and was most likely a cavernous hemangioma.

Though the differential diagnosis of a well-circumscribed orbital mass is extensive, and in addition to cavernous hemangioma includes schwannoma, fibrous histiocytoma, hemangiopericytoma, lymphomas, lymphoid hyperplasia, optic nerve meningioma, lymphangioma, neurofibroma, and orbital metastasis; it was felt that this mass, by radiographic appearance as well as clinical judgment, was a cavernous hemangioma. Thus conservative management with observation of the mass and any symptoms was recommended, given the risks of surgical excision. The patient elected to pursue 3rd and 4th opinions on treatment. The plan, should the patient return, would be to obtain an MRI for further evaluation of the mass.



**FIGURE 1**  
CT scan of the orbits demonstrating a mass in the intracanal space of the right orbit. The mass measured 18.6 mm x 12.9 mm.

**BACKGROUND** Cavernous hemangiomas are the most common primary orbital tumor in adults and occur mostly in middle-aged women. Because of their slow growth, the most common presentation is painless proptosis, though it can cause optic nerve compression with visual loss and visual field defects. They are usually solitary lesions occurring in the lateral aspect of the retrobulbar, intracanal space and are well-circumscribed because of a distinct fibrous pseudocapsule and have minimal neoplastic potential. CT and MRI are the primary imaging modalities used to evaluate these lesions, with a higher accuracy of diagnosis when using multi-phase contrast studies. Ultrasound is also sometimes used in conjunction to corroborate the diagnosis. In terms of management, surgical removal is reserved only for those cases with severe proptosis or optic nerve compression.

## Thyroid Ophthalmopathy: Donny Hoang, MD (Resident)



A 79 year-old African American male presented to the IEEI oculoplastics service in May 2009 with a 6-week history of proptosis of the left eye, followed by gradually increasing proptosis of the right eye over the last 2 weeks. He also complained of blurring, tearing and slight irritation. He had a history of known Graves disease, diagnosed and treated with radioactive iodine in 2007. He was currently hypothyroid and taking levothyroxine. Of note, his past medical history also included diabetes, hypertension, COPD, chronic kidney disease, and refractory anemia. He was taking lubricating drops and ointment, as well as prednisone 2.5mg daily, levothyroxine, insulin, amlodipine, lasix and a statin.

On presentation, best corrected visual acuity was 20/200 in the right eye, and 20/100 in the left. Intraocular pressure was 24 bilaterally. Extraocular movements were limited (-2 in all directions) bilaterally. Color vision was 6/11 and 5/11 on Ishihara plates. Hertels measured 30mm bilaterally

(base 114). He had 13 and 13+ mm lag with 4+ conjunctival chemosis in both eyes, and there was corneal punctate staining (Fig 2). Additionally, the patient brought with him CT images of the orbits taken a month prior at an outside hospital that showed bilateral proptosis, severe enlargement of the superior and inferior recti, as well as moderate enlargement of the medial rectus. There was apical crowding noted.

The patient was therefore diagnosed with Grave's eye disease with resultant bilateral proptosis, lagophthalmos and compressive optic neuropathy. He was presented with three treatment options: oral prednisone, radiation therapy, and orbital decompression surgery. Since the patient adamantly refused surgical intervention and also turned down the notion of pulse steroid therapy, he was started on oral prednisone 60 mg daily, and returned 1 week later with significant subjective improvement in vision and comfort, but much more erratic blood sugars. Best corrected vision had improved to 20/100 in the right eye and 20/70 in the left. Color vision was stable at 6/11 and 5/11 plates, and there was no relative afferent pupillary defect (Fig 3).

Given the patient's diabetic history, the prednisone was tapered and the radiation oncology service was consulted. The radiation oncology service recommended radiation (20 Gy in 2 Gy fractions). On his 4 week follow-up visit, he had completed 6 out of 10 radiation sessions, and was on prednisone 30mg daily. Best corrected vision was 20/100 in the right eye and 20/40 in the left. Color plates had slightly improved to 9/11 in the right and 7/11 in the left. Hertels had improved to 29, 31 (base 105). EOMs remained decreased in all directions (-2). His lag had significantly improved, now 3-4 mm in the right and 4-5 mm in the left. There was trace conjunctival chemosis and inferior corneal staining bilaterally (Fig 4).



**FIGURE 2**  
External photograph at presentation demonstrating proptosis and significant conjunctival chemosis.



**FIGURE 3**  
Follow-up external photograph following oral prednisone, demonstrating dramatically reduced proptosis and conjunctival chemosis.



**FIGURE 4**  
External photograph following orbital irradiation with further improvement in proptosis and minimal conjunctival chemosis.

**BACKGROUND** Thyroid ophthalmopathy is the most common cause of unilateral or bilateral proptosis in adults, with females affected more frequently than males (8:1). Although patients are often asymptomatic, they may present with eyelid retraction (90%), proptosis (63%), diplopia (restrictive myopathy), tearing, redness, foreign body sensation, lagophthalmos, lid lag on downgaze (von Graefe's sign), reduced blinking, superficial keratopathy, conjunctival injection and chemosis, decreased motility (supraduction most common), increased intraocular pressure on upgaze, and resistance to retropulsion. Compressive optic neuropathy may also be present in less than 5%, with decreased vision, color, visual field defects, and a relative afferent pupillary defect.

Irreversible intervention is usually deferred until the disease is stable for 6 months, except if optic neuropathy or extreme proptosis causing severe exposure is present. Surgical reconstruction typically proceeds from posterior to anterior with orbital bony decompression first, followed by strabismus surgery, with eyelid surgery performed last.

The management of optic neuropathy includes immediate treatment with systemic steroids, consideration of external beam irradiation, and orbital decompression for compressive optic neuropathy via either a Caldwell-Luc or transcutaneous/transconjunctival approach.

## Discussion:

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Cavernous hemangiomas are the most common type of primary orbital tumor in adults. Cavernous hemangiomas behave indolently with slow growth; however can cause compressive optic neuropathy. Surgical intervention should be avoided unless necessary.

The final case underscores the role of thyroid ophthalmopathy as the leading cause of unilateral or bilateral proptosis in adults. Additionally, it stresses the need for urgent treatment in the cases of compressive optic neuropathy. Our patient responded well to prednisone, however needed radiation therapy for long-term treatment.

## 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.