

Grand Rounds

8-26-09

Introduction: Jennifer Lim, MD (*Attending*)

This week's retina grand rounds presents several interesting cases encountered by our faculty.

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Ciliary Body Leiomyoma: Mark Krakauer, MD, MPhil (*Resident*)



A 66-year-old Caucasian female with a history of breast cancer in remission, presented to her primary care doctor after a 6-hour bout of vertigo and right earache. CT and MRI of the head incidentally showed a tumor of the right orbit, apparently unrelated to her presenting symptoms. She was then referred to the UIC retina clinic. Her family history was significant for her skin melanoma in her brother and mother. Past medical history, social history, and review of systems were non-contributory.

Dilated fundus exam revealed a tumor located inferonasally, solid-appearing and brown in color. It was located in the pars plana, extending almost to the ora serrata. On ultrasound biomicroscopy, the tumor was found to be 6.4 x 5.5 x 3.2 mm, with low-to-medium internal reflectivity (Fig. 1). It was not cystic, and was relatively uniform and solid. The differential diagnosis included malignant melanoma, metastatic carcinoma, leiomyoma, schwannoma, and adenoma/adenocarcinoma of the RPE or ciliary epithelium. Metastases from breast carcinoma to the eye are not common, and the ultrasound characteristics were not typical for a metastatic lesion, which would have higher internal reflectivity.

Partial lamellar sclerouvectomy was performed to excise the tumor (Fig. 2); this procedure would be both diagnostic and therapeutic. Another option, needle biopsy, was not performed as it has a false negative rate, would be anatomically difficult to perform, and would necessitate a second procedure if malignancy had been found. Diathermy, vitrectomy, and endolaser barricade were performed during removal of the tumor in order to reduce the risk of retinal detachment.

Based on histology and immunohistochemistry, a diagnosis of ciliary body leiomyoma was made. The tumor was composed of well-circumscribed spindle cells in the pars plana of the ciliary body (Fig. 3). It stained positive for desmin and actin, which is consistent with leiomyoma, and negative for cytokeratin AE 1/3 (epithelium), S100 (schwannoma, melanoma), and melan A (melanoma).

The post-operative course was remarkable for a vitreous hemorrhage, the most common complication of partial lamellar sclerouvectomy.

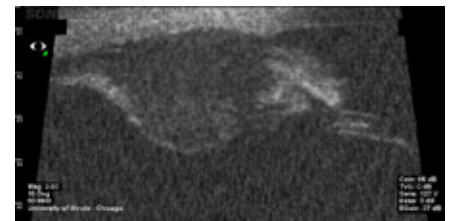


FIGURE 1
Ultrasound biomicroscopy demonstrating a mass at the pars plana with low-to-medium internal reflectivity.

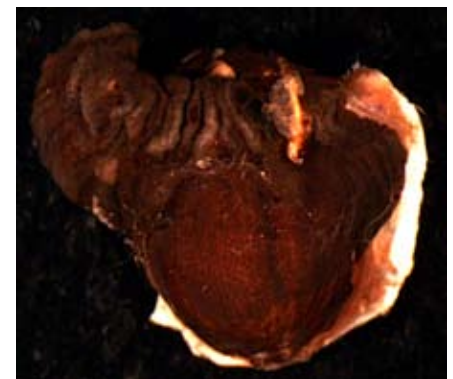


FIGURE 2
Pathology specimen. Tumor after excision, located at the pars plana.

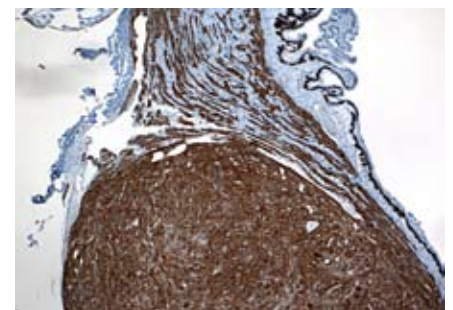


FIGURE 3
Photomicrograph of the tumor, demonstrating a well-circumscribed ciliary body mass consisting of numerous spindle cells.

BACKGROUND Ciliary body leiomyoma is extremely rare. UBM is the most useful imaging modality for the diagnosis and follow-up of anterior segment tumors. Preoperatively, it is difficult to distinguish leiomyoma from malignant melanoma. Partial lamellar sclerouvectomy can be both diagnostic and therapeutic, as in this case. The most common complications are vitreous hemorrhage, subretinal hemorrhage, retinal detachment, and cataract. Histology and immunohistochemistry are necessary for definitive diagnosis, and the prognosis is favorable for this benign tumor.

Ocular Histoplasmosis: Neema Nayeb-Hashemi, MD (Resident)



A 44 year old female with a 1-week-history of blurred vision and metamorphopsia in her left eye was referred from a retina specialist in St. Louis. She had a history of bilateral chorioretinopathy secondary to ocular histoplasmosis syndrome (OHS) diagnosed in 1996 necessitating multiple vitrectomies as well as focal laser and intravitreal steroid injection for complications related to extrafoveal choroidal neovascularization of her right eye.

Examination of the patient demonstrated 20/30 vision in her newly affected eye, with metamorphopsia paracentrally on Amsler grid. Fundus examination of the left eye revealed peripapillary atrophy with focal areas of hyperpigmentation, small peripheral chorioretinal scars, macular edema, as well as two extrafoveal areas of subretinal hemorrhage adjacent to a fibrotic scar (Fig. 4). Fluorescein angiograms revealed late leakage in the region of the scar extending centrally into the fovea owing to choroidal neovascularization (CNV) (Fig. 5). OCT performed on the initial visit revealed significant retinal thickening temporal to the fovea.

Ranibizumab was injected intravitreally in an effort to avoid scarring and possible vision loss from focal laser photocoagulation. One month later, the patient's vision was stable with some improvement in the metamorphopsia. Fluorescein angiogram demonstrated less prominent late phase leakage as well as partial regression of subretinal fluid on OCT (Fig. 6). She was offered a second injection of ranibizumab, and on return to the clinic one month later, OCT demonstrated further regression of subretinal fluid with a slight improvement in visual acuity to 20/25.



FIGURE 4
Fundus photo of left eye demonstrating peripapillary atrophic pigmentary changes, a 1 DD area of extrafoveal macular edema, small regions of subretinal hemorrhage, and a chronic fibrotic scar.

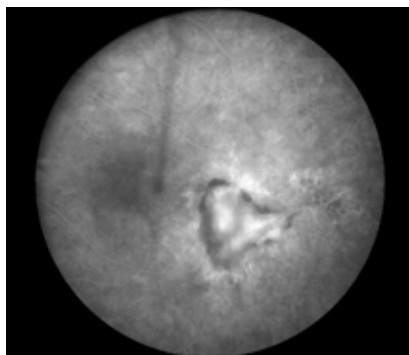


FIGURE 5
Late phase FA showing leakage from CNV in the area of fibrotic scarring.

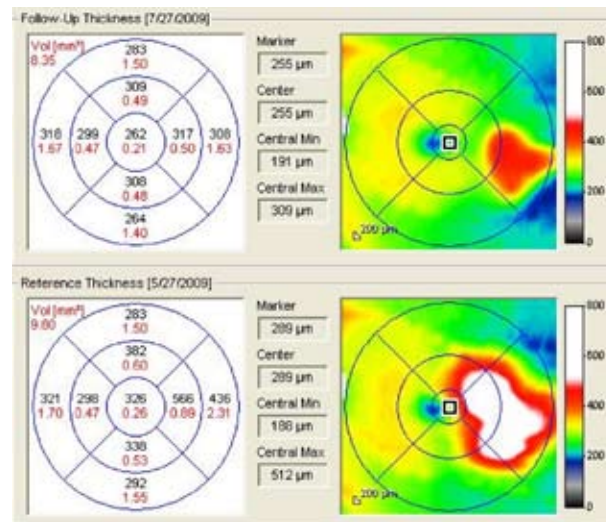


FIGURE 6
Comparison of the initial (inferior) and final (superior) OCT thickness maps after 2 injections of ranibizumab in the left eye. There is significant regression of subretinal fluid.

BACKGROUND Ocular histoplasmosis syndrome is most commonly found in the Mississippi and Ohio River valley areas. The fungus responsible is carried by chickens, pigeons, and bats. Patients affected often present years after initial systemic infection. Classic fundus findings include punched out chorioretinal lesions, juxtapapillary atrophic pigmentary changes, and as a late sequelae of infection, vision threatening choroidal neovascularization. Vitritis is notably absent, unlike other diseases in the differential diagnosis such as multifocal choroiditis and birdshot choroidopathy.

Workup of patients with OHS typically includes fluorescein angiography to evaluate for CNV, and more recently OCT. Treatment should be initiated soon after confirmation of CNV, and includes focal laser photocoagulation for extrafoveal CNV, as well as PDT or anti-VEGF injections for subfoveal lesions. A recent retrospective review of bevacizumab use in patients with CNV secondary to OHS by Schadlu et al. has yielded promising results, as well as the possibility that PDT in conjunction with bevacizumab could yield superior results to bevacizumab alone. Prospective studies are currently underway. Submacular surgery has also been found helpful in individuals with vision worse than 20/100. Anti-fungal medications, however, are of no benefit in the treatment of this syndrome.

Tuberous Sclerosis: Brittany Osgood, MD (Resident)



A 27 year old Hispanic female presented to the retina clinic for abnormal findings noted on a previous eye exam. Her past medical history was significant for tuberous sclerosis, along with mental retardation and a history of seizures (a prior MRI demonstrated multiple tubers throughout the white/gray matter of both hemispheres). At her initial exam her vision was noted to be 20/20 in both eyes, and her extraocular movements,

confrontation visual fields, and pupil exam were all within normal limits. Her external exam was significant for adenoma sebaceum, and her anterior segment exam was normal. Her dilated fundus exam was significant for multiple flat, semitransparent lesions (Fig. 7), identified as astrocytic hamartomas.

BACKGROUND Tuberous sclerosis is an autosomal dominant disease, although new mutations account for about 2/3 of cases. Retinal astrocytic hamartomas are found in 50% of patients with TS. There are three types of astrocytic hamartomas. Type 1 lesions are the most common and were noted in our patient. They are flat, semi-transparent, circular/ or oval, ill-defined, and average around a half disc diameter in size. They are found in the retinal nerve fiber layer without calcification. Type 2 lesions are raised, multinodular lesions described as “mulberry-like,” and are often calcified. Type 3 lesions have characteristics of both type 1 and 2 lesions.

The hamartomas tend to remain stable over time and seldom affect vision, however eight cases of subretinal exudation and three cases of vitreous hemorrhage caused by these lesions have been reported in the literature. Most of these cases demonstrated spontaneous improvement after 1 month but a few were treated successfully with laser or PDT, along with one vitrectomy for a case of recurrent vitreous hemorrhage. There was one extreme case of an atypical astrocytic hamartoma that caused a retinal detachment, vitreous hemorrhage, neovascular glaucoma, and spontaneous scleral perforation. This eye was eventually enucleated. Contrastingly, there have been rare reports of hamartomas that have spontaneously regressed.



FIGURE 7
Color fundus photograph of the right eye demonstrating several lesions (superior arcade, inferior arcade, and inferonasal to disc) consistent with Type 1 astrocytic hamartomas.

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 th 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 and click on Grand Rounds under the Education drop down menu. Or, call 312-996-6590.