



Grand Rounds

9-30-09

Moderator: Allen Putterman, MD (*Attending*)



This week's grand rounds presents several interesting cases encountered by the oculoplastics service.

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Orbital Lymphangioma: Elizabeth Grace, MD (*Resident*)



A 5 year-old female presented with a 1-month history of a nasally-located, enlarging, right eyelid mass. One month prior to when the mass was first noticed, the overlying skin had no discoloration. As the lesion increased in size, she saw a pediatrician who referred her to an ophthalmologist to evaluate the lesion. It was felt the lesion was a cyst and she was taken to the operating room, where upon opening the superior lid the mass was noted to be larger and more extensive than previously thought, possessing bluish hue. The outside ophthalmologist closed the lid, did not take a biopsy of the lesion and sent her to be evaluated at UIC for possible orbital varix (Fig. 1).



FIGURE 1. External photograph of the patient upon presentation at UIC, demonstrating right upper eyelid fullness and bluish discoloration.

At UIC, the patient underwent an exam, at which time the differential diagnosis included orbital lymphangioma, capillary hemangioma, AV malformation, orbital varix, and ruptured dermoid cyst. She received a MRI scan of the brain and orbits, showing an extensive soft tissue mass of dilated and convoluted vessels, filling the superomedial portion of the right orbit, displacing the globe inferolaterally (Figs. 2-3). The mass was also compressing the superior oblique vein causing it to be markedly engorged. The mass was felt to be a orbital lymphangioma and was observed closely, with decreased size noted at the one week follow-up.



FIGURE 2. Axial T2 MRI image demonstrating cystic structures in the right superomedial orbit. Fluid-fluid layering of red blood cells and serum is seen.

BACKGROUND: Orbital lymphangiomas are benign hamartomatous tumors that usually become apparent during the first decade of life. They can be seen on the conjunctiva, eyelids, orbit, oropharynx, and sinuses. Clinically, the behavior of the lesions varies and is reflected by their clinical presentation. The lesion can grow slowly with progressive proptosis, displacement of the globe, ptosis and restriction of the eye movement. Otherwise, a spontaneous intralesional hemorrhage can lead to acute proptosis, compressive optic neuropathy, and possible loss of vision.

Histologically, lymphangiomas represent combined vascular malformation with venous and lymphatic components that rarely are connected with venous circulation and most have no radiologic evidence of vascular connection. The large serum filled spaces are lined by flattened endothelial cells with scattered lymphoid follicles, whereas radiologically it presents as multiple grapelike cystic lesions with fluid-fluid layering of red blood cells and serum.

Treatment varies, as the vascular malformation often insinuates between intraconal and extraconal structures, making it essentially impossible to completely resect. Conservative management and observation is recommended in most cases. However surgery is indicated in cases with optic nerve compression, corneal exposure, anisometropia, or those with significant cosmetic deformity.

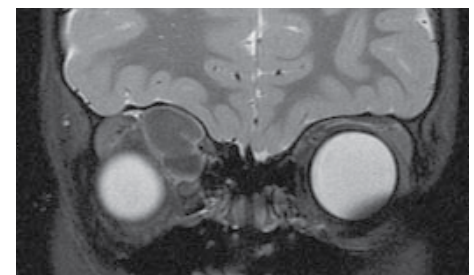


FIGURE 3. Coronal T2 MRI image demonstrating cystic structures in the right superomedial orbit, causing inferolateral displacement of the globe.

Capillary Hemangioma of the Eyelid and Orbit: Shivani Gupta, MD (Resident)



A 46-year-old Caucasian male presented with an enlarging right upper eyelid lesion. The patient noted increased pigmentation over past few months, occasional tearing, and more recently constriction in his peripheral visual field secondary to the eyelid lesion. His past ocular history was significant for a hemangioma since childhood, and past medical history significant for hypertension. The remainder of his history is noncontributory.

External examination demonstrated a raised vascular lesion of the upper eyelid extending laterally and inferiorly, with resultant ptosis (Fig. 5). Visual acuity was 20/20 in both eyes. Pupillary exam and motility was within normal limits in both eyes. Intraocular pressures were 17mmHg and 17.5mmHg, in the right and left eyes respectively. Anterior segment examination and dilated funduscopic examination were within normal limits in both eyes. The patient had an MRI with contrast done prior to presentation (Fig. 6) that demonstrated an enhancing lesion in the eyelid extending into the lateral orbit.

At initial evaluation, the differential diagnosis included capillary hemangioma, capillary hemangioma with malignant transformation, and other vascular tumors such as varix or AV malformation. At this point the discussion arose as to the appropriate diagnostic and/or treatment options. These included serial monitoring, biopsy, or treatment for presumed capillary hemangioma. It was decided to proceed with diagnostic biopsy of the mass lesion prior to initiating treatment of the lesion. Biopsy revealed capillary hemangioma, and treatment with systemic beta blockers was subsequently initiated.

During the ensuing month, the patient received 3 FOS injections at the OSH, but then “lost vision” OD abruptly and saw OSH retina who recommended fluorescein angiography (showing profound peripheral ischemia extending to the fovea, vasculitis, arterial occlusions and shunt vessels). The patient returned to see the UIC uveitis service, at which time vision OD was hand motions. Anterior segment exam OD was notable for 4+ pigmented AC cells, 3+ flare, rubeosis at the pupil border with gonioscopy revealing fine rubeosis temporally (but open to ciliary body for 360 degrees). Dilated exam OD was obscured by a dense vitreous hemorrhage. At that point the differential diagnosis included ocular ischemic syndrome, given the profound ischemia OD with rubeosis. An intravitreal injection of bevacizumab was administered that day, and carotid dopplers were ordered, demonstrating mild obstruction. The patient was advised to sleep with his head elevated, and return for panretinal photocoagulation once there was sufficient clearing.

BACKGROUND Capillary hemangiomas are common, benign orbital tumors in children. They are often seen in the first few months of life, and commonly resolve within the first decade of life. They have 3 distinct stages, including a proliferative phase, plateau phase, and involutinal phase. Ocular complications related to capillary hemangiomas include astigmatism, amblyopia, strabismus, ptosis, proptosis, optic atrophy and glaucoma. Lesions that are small and do not pose a risk for the aforementioned complications may be observed. Alternatively, treatment may be initiated with systemic or intralesional steroids, systemic propranolol, interferon, irradiation, or intralesional laser. Excision may also be attempted but is frequently associated with hemorrhage and recurrence.



FIGURE 5. External photograph of the patient demonstrating a raised vascular mass of the right upper eyelid.



FIGURE 6. MRI image demonstrating an enhancing lesion in the eyelid extending into the lateral orbit.

Mohs Reconstruction: Genie Bang, MD (Resident)



A 68 year-old male presented with a basal cell carcinoma found on his upper nose, near his left medial canthus. He was scheduled for a Mohs surgery with dermatology, and same-day oculoplastic repair of the defect. Presurgical evaluation showed a defect 17x17mm, involving orbicularis, located on the left upper nasal skin, near, but not involving the left medial canthus (Fig. 4).

Surgical repair includes skin graft or local flap options. The scope of this discussion is limited to local flaps, as that is what was chosen for this case. Local flaps have the advantage of an integrated blood supply, similar skin type to the defect, and similar coloration to the skin defect area.

Important terms when describing reconstruction are: 1) primary defect – the original defect the surgeon is repairing; 2) secondary defect – the new defect created by closing the primary defect. The art of plastic surgery and reconstruction lies in the hiding of the secondary defect; 3) primary movement – the movement of the flap into the primary defect; 4) secondary movement – the movement of the surround tissue into the primary defect

Also important in making flap incisions and in the movement of the flap is remembering Relaxed Skin Tension Lines (Langer's Lines). These imaginary lines note the natural tendency of the skin and underlying collagen to take certain orientation. Incisions are well hidden in these Langer's Lines.

Local flaps are classified in different ways. Random flaps rely on mucocutaneous blood vessels, whereas axial flaps rely on an anatomically identified septocutaneous artery (ex. paramedian forehead flap using the supratrochlear artery). Another classification of flaps is whether they are advancement flaps, translocation/pivotal flaps, or a combination of both. Example of advancement flaps are single pedicle flaps, bipedicle flaps, and the V-Y advancement flap. Pivotal flaps include rotational flaps, interpolation flaps, and transposition flaps. Transposition flaps are further classified into rhombic flaps, bilobed flaps, and Z-plasty.

Postoperatively, the flap goes through histologic changes, including an inflammatory phase, and also neovascularization of the flap. Flaps must also be monitored carefully for signs of complications, including dehiscence, hematoma, and failure of the flap. Failure of the flap can be multifactorial, with factors including: patient health, too small of a flap, damaged blood supply, inadequate blood supply for size of flap, or too much tension on the flap.

The patient underwent a V-Y plasty with rotational advancement for closure of the wound. A good cosmetic result is expected. Postoperatively, the incisions are well apposed, and the flap is healthy without signs of failure.



FIGURE 4. Photograph of defect following excision of basal cell carcinoma, prior to reconstruction.

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.

UPCOMING CME COURSES

March 13-19, 2010	Illinois Eye Review
April 10, 2010	Retina Symposium
May 12, 2010	Glaucoma Meeting
May 21, 2010	Oculofacial Plastic Surgery Conference
June 25, 2010	34th Annual Alumni Day

