

Grand Rounds

July 23, 2009

Introduction: Elmer Tu, MD (*Attending*)

This week's Cornea Grand Rounds presents several interesting cases encountered by our cornea faculty.

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Conjunctival Malignant Melanoma: Brittany Osgood, MD (*Resident*)



A 69 year-old white male was referred to the cornea clinic after biopsy of a conjunctival lesion of the right eye was noted to be malignant melanoma with involvement of the margins. According to the patient, he had noted a "colored spot" on the conjunctiva for many years, but had noted recent growth over the past year. His medical, surgical, social and family histories were all unremarkable.

The best corrected visual acuity was 20/25 in the right eye and 20/20 in the left. His motility and confrontation visual fields were full bilaterally. On slit lamp examination of the right eye, a conjunctival graft was noted temporally with pigment on the graft, and abnormal vessels were noted superotemporally. Examination of the left eye was normal. Gonioscopy was normal in both eyes. No lymph nodes were palpated and the dilated fundus exam was normal.

A wide resection of the main lesion was planned. This was conducted in June 2009 and while in the OR, another small superior pigmented lesion was noted which was also removed, along with 2 map biopsies in the superior and inferior fornix. Cryotherapy was applied to the margins and absolute alcohol to the limbus. An amniotic membrane was used for resurfacing.

On pathology, the main lesion was noted to be malignant melanoma with a thickness of 0.34mm, arising in the context of primary acquired melanosis (PAM) with atypia (figure 1), with extension to the superior margin. The smaller superior lesion was also noted to be melanoma (figure 2) and the 2 map biopsies were negative.

The patient was referred to oncology and a sentinel lymph node biopsy performed in July 2009 was negative.

BACKGROUND

Conjunctival malignant melanomas are rare, occurring 40 times less than choroidal melanomas and 500 times less than cutaneous melanomas. They can arise from PAM in 60-77% of cases, a pre-existing nevus in 20%, or rarely de novo. In the setting of PAM with atypia, 50% give rise to malignant melanoma within 2 ½ years.

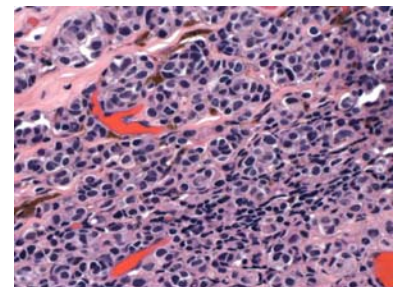
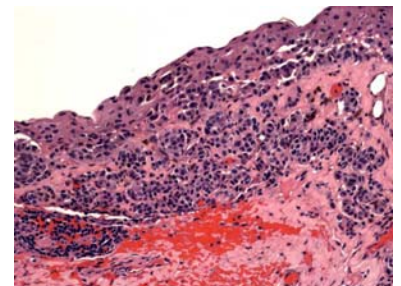


Figure 1 (top) and Figure 2 (bottom)

Courtesy of Amy Lin, MD

DISCUSSION

Surgical treatment of keratoconus can be challenging in a patient with mental retardation. Careful selection of patients who are good candidates is essential, and requires observation of the patient, as well as questioning of the family or caretaker. A combative and noncooperative patient endangers not only the eye and graft, but also makes follow-up examinations extremely difficult. The second case demonstrates some of the difficulties in managing a traumatic hyphema with corneal blood staining in a young child. Although corneal blood staining in an adult can often be observed, in a young child the risk of amblyopia may necessitate earlier intervention. Since our patient was slightly on the older end of those susceptible to amblyopia, it was elected to observe the patient carefully. However, the use of

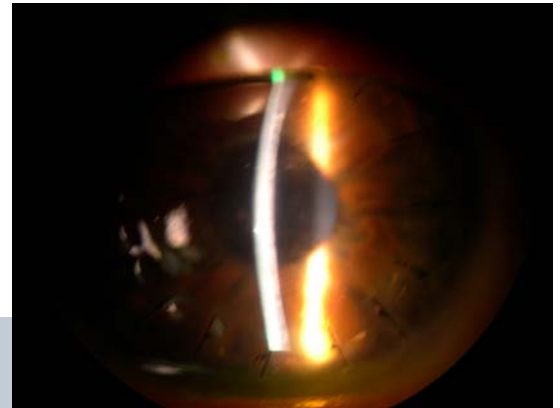
patching in the unaffected eye can prevent amblyopia.

The final case involves a challenging presentation of malignant melanoma of the conjunctiva. It is important to note that the vast majority of cases arise from PAM, and especially PAM with atypia. Any patient with PAM must be monitored carefully, and malignant transformation suspected if there is a change in elevation or increased vascularity develops. Although not applicable in our patient's case, it is good practice to send pterygia for pathologic examination after they are excised, as they can sometimes harbor a malignant melanoma.

Keratoconus: Genie Bang, MD (*Resident*)



A 16 year-old male was referred to the Cornea Clinic at the IEEI for evaluation and management of keratoconus. He has a history of Down Syndrome, eye rubbing, and newly diagnosed keratoconus. At the time of initial evaluation, visual acuity was 20/400 in the right eye, and 20/60 in the left. Keratoconus was noted in both eyes, however thinning in the right eye was marked, with apparent central scarring. After discussion with the family, it was decided to proceed with an anterior lamellar keratoplasty of the right eye.



BACKGROUND

Down syndrome patients have a much higher incidence of keratoconus than the general population. Reports of the prevalence of keratoconus in Down's Syndrome patients varies from 5% to 30%. Keratoconus is also difficult to manage in these patients, as they are prone to eye rubbing, and may or may not be able to comply with management.

The question of whether surgical treatment is indicated for these patients has been addressed multiple times over the years. Keratoconus is the most common indication for penetrating keratoplasty (PKP). However, PKP requires careful follow up, management, and care of the graft. A few studies have addressed the success of PKP for patients with Down's Syndrome, and indicate that while the 5 year graft survival rate in Down's patients with keratoconus (67%) is less than the general population graft survival rate for keratoconus (90-95%), it is similar to the 5 year graft survival rate of corneal grafts for any reason. Physicians are now reporting that non-penetrating keratoplasty options (Anterior lamellar keratoplasty, epikeratophakia), have similar success rates with fewer serious complications than PKP.

Keratoconus is a disease that is managed in different ways, and surgical decisions vary by practitioner. Keratoconus management in the Down syndrome patient is even more variable. The question of who is a good surgical candidate for PKP requires careful questioning by the physician of not only the patient, but also the caretakers, in order to assure proper postoperative management. Those patients who are not good candidates for PKP, may be candidates for DALK or epikeratophakia, although further studies and outcomes need to be assessed.

Corneal Blood Staining: Jessica Wong, MD (*Resident*)



A 6 year-old African-American female with no significant past medical history presented to an outside hospital with a complete hyphema of the left eye after reportedly falling off of a swing four days earlier. At the time of presentation, a sickle cell prep was negative, reported vision was no light perception (NLP) in the left eye, the presence of a relative afferent papillary defect was noted in the left eye, and intraocular pressure (IOP) in the affected eye was 55 to 60.

The patient was taken to the operating room immediately for an AC washout, but post-operative day #1 a re-bleed was already noted and blood once again completely filled the anterior chamber OS. Over the course of the next few days, the patient's IOP was maintained under 20 on medical therapy alone, so she was not taken for another washout, but on post-operative day #4 she was noted to have corneal blood staining. When she presented to our cornea service, her vision was still NLP, her IOP was 24, and she was noted to have very dense bright red blood staining that involved the entire thickness of the cornea from limbus to limbus. Ultrasound biomicroscopy revealed the presence of a clot in the anterior chamber and B-scan revealed no vitreous debris or retinal detachment.

BACKGROUND

This case was presented to discuss the optimal treatment of corneal blood staining in children. This case presents some treatment dilemmas because of the patient's age. For instance, should deprivation amblyopia be a concern in this child? If this patient were a few years younger or a few years older, would that change our management in this situation? Is this patient truly NLP, and if so, why is this patient NLP, and what is her visual potential? Is surgical intervention indicated, and if so, when would be the optimal time to intervene?

After extensive discussion, it was decided that this patient would be managed conservatively and medically for now since her age makes her high risk for a procedure such as a penetrating keratoplasty. Additionally, at age 6, deprivation amblyopia is less of a concern than if the child were a few years younger.

Our current strategy will be to monitor the IOP and observe for spontaneous clearing of the blood staining; if her exam changes to warrant surgical intervention, it will be undertaken at that time. In conclusion, corneal blood staining in a child presents an interesting and complex therapeutic dilemma.



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 4, 2010

Retina Symposium

April 16, 2010

Uveitis Symposium

May 21-22, 2010

Oculoplastics Symposium

June 25, 2010

34th Annual Alumni Day

September 25, 2010

Pediatric Ophthalmology/Adult Strabismus