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Case Report

Atypical Presentation of Uveal and Conjunctival Melanoma in the Anterior Segment

Introduction

Ocular melanoma is a rare malignancy arising from melanocytes of the uvea, the conjunctiva, and the orbit. Uveal melanoma is the most common intraocular malignancy in adults, accounting for 85-95% of ocular melanomas [1-3]. The conjunctival form accounts for 5% of ocular melanomas. Uveal and conjunctival melanoma are very distinct from each other biologically. We describe a case of conjunctival melanoma and a uveal melanoma both presenting as large masses of the anterior segment.

Case Report

Patient 1

75 year old female from Haiti presented with spontaneous bleeding from her left eye for 1 day. She had a black mass present on the surface of her left eye which was noted by her family 4 months prior when she arrived from Haiti. The patient did not seek any medical care until bleeding began. She did not complain of pain. She denied past ocular trauma or surgery or a history of anticoagulation therapy. On examination, vision was 20/40 in the right eye, light perception in the left eye. The right pupil was reactive. The mass prevented the visualization of the left pupil. In the left eye, there was 3+ diffuse conjunctival injection, and a large black and somewhat bloody mass measuring 12x12x4mm that precluded visualization of the entire cornea and anterior segment (Figure 1A). Computed Tomography (CT) scan demonstrated a formed and normal sized globe with a solid mass overlying an intact cornea. There was no evidence of intraocular extension radiologically (Figure 1B). Blood tests including liver function tests were in normal

limits. An examination under anesthesia and exploration was performed.

Intraoperatively, the mass was easily removed from the corneal epithelium. It was adherent to the limbal conjunctiva temporally. A small, bleeding limbal blood vessel was cauterized. The corneal epithelium was intact. No other lesions were noted on the anterior aspect of the globe and dilated exam with scleral depression revealed normal anatomy. Histopathology disclosed mixed spindle B and pigmented epithelioid cells (Figure 1C). MHB-45, MART-1, Mel A, and S-100 markers were positive. The final diagnosis was malignant melanoma, clinically arising from the limbal conjunctiva. Postoperatively, the patient's vision in the left eye was 20/100, attributable to a nuclear cataract. There was a 2 mm area of conjunctival melanosis at 12 o'clock and the eye was otherwise grossly anatomically normal (Figure 1D). There was no evidence of tumor in the angle on gonioscopy. The patient underwent a metastatic workup

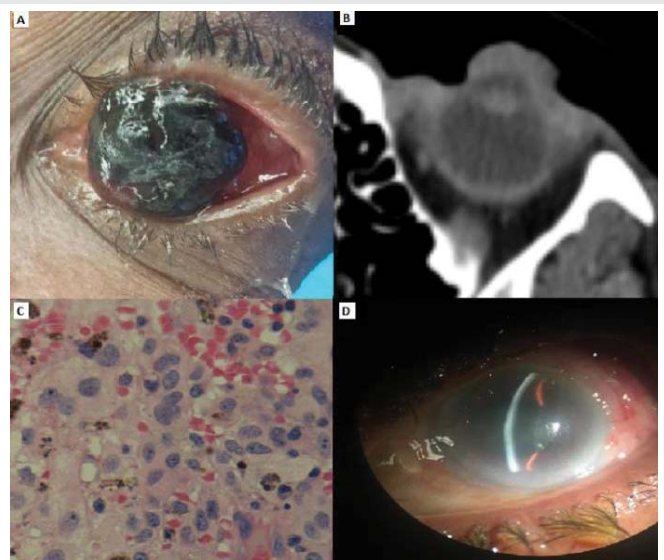


Figure 1: (A) Anterior segment mass (B) CT scan of orbit (C) Histological stain showing mixed spindle B and pigmented epithelioid cells (D) Appearance of eye after excision of anterior segment mass.

which was negative. She underwent further cryotherapy of the conjunctival tissue and treatment with interferon topical drops.

Patient 2

72 year old African-American man presented with spontaneous bleeding from his left eye for 3 days. He had a black mass present on the surface of the left eye for unknown duration of time. He did not complain of pain but reported a foreign body sensation. His past ocular history included enucleation of the right eye in 1970 following an automobile accident. His left eye was severely injured in a fight in 1979 which had left him with no light perception vision. He denied any recent trauma. He denied any medical history and was not taking any medications. On examination, the right orbit was anophthalmic. His left eye was blind and phthisical. There was a black and bloody mass protruding from and occupying the corneal surface; measuring 15x15x5mm (Figure 2A). CT scan demonstrated a phthisical eye with bone and soft tissue within the globe (Figure 2B). The patient underwent evisceration as the surgeon clinically believed the pigmented lesion did not originate from the uvea. There was no gross intraocular extension seen intraoperatively. An Acrylic implant was inserted into the scleral shell. Histopathology revealed the presence of both intraocular and extraocular melanoma. The tumor was composed of large and small cells with epithelioid features (Figure 2C). Many of the cells stained for melanin. Fragments of bone and degenerated uveal tissue were present as well. HMB-45, MEP IP and XIAP stains were positive. The final diagnosis was malignant melanoma epithelioid cell type arising from the uveal tract. The patient underwent metastatic work up and no metastases were found. He then underwent a definitive exenteration of the left orbit with histopathology showing no evidence of primary or residual tumor (Figure 2D).

Discussion

Primary ocular melanoma can occur in the uvea, conjunctiva, or orbit. These melanomas are distinct entities

although they share a common origin from melanocytes. The annual incidence of uveal melanoma is 5.1 per million [2]. The most common location is of the choroid posterior to the equator in 85% of cases. 15% arise anterior to the equator and involve the iris, ciliary body or the choroid anterior to the equator [4]. Conjunctival melanomas account for only 2–5% of ocular malignancies. Tumor spread occurs through lymphatics and blood stream resulting in metastasis [5,6]. Poor prognostic factors include tumor thickness greater than 2mm, high mitotic rate, and epithelioid cell type. With uveal melanoma, poor prognostic factors have been associated with involvement of the ciliary body, extrascleral extension, pathologic features of epithelioid cytology, and monosomy 3 [7–11]. In both uveal and conjunctival melanomas, involvement of the entire corneal surface is extremely rare. The two cases presented share several striking characteristics. Both individuals had moderate to dark skin pigmentation. Both cases presented as large masses involving the cornea, precluding anterior segment visualization. They appeared to be free of significant intraocular or conjunctival pigmentation elsewhere, at least on clinical evaluation at presentation and on radiologic imaging. The first case was striking as the mass easily peeled off the corneal epithelium with moist cotton tip applicator sticks; there was bleeding at the lateral limbus where the lesion was clinically minimally adherent to the limbal conjunctiva. The differential diagnosis in each case included coagulated blood on the surface of the cornea, ocular surface squamous neoplasia, and a ruptured globe with exposed uvea. The corneal epithelium can be involved when conjunctival melanoma involves the limbus.

Bowman's layer may provide a barrier to deeper invasion [12].

Conclusion

Uveal and conjunctival melanoma presenting as a pigmented large mass overlying the cornea is rare. The mass may resemble exposed uveal tissue from a ruptured globe, large hematoma or squamous neoplasia. The management can be complex. Wide margins should be taken from the visible lesion since positive margins can be associated with higher recurrence rates. Tumor cells should not be irrigated. Care must be taken to not penetrate Bowman's layer during excision to prevent further tumor spread. Post-operatively, patients must be monitored closely since new pigmentation may be a sign of recurrence. In summary, melanoma must be considered for any pigmented lesion obscuring the cornea and prompt management should be undertaken, including a work-up to exclude metastatic disease.

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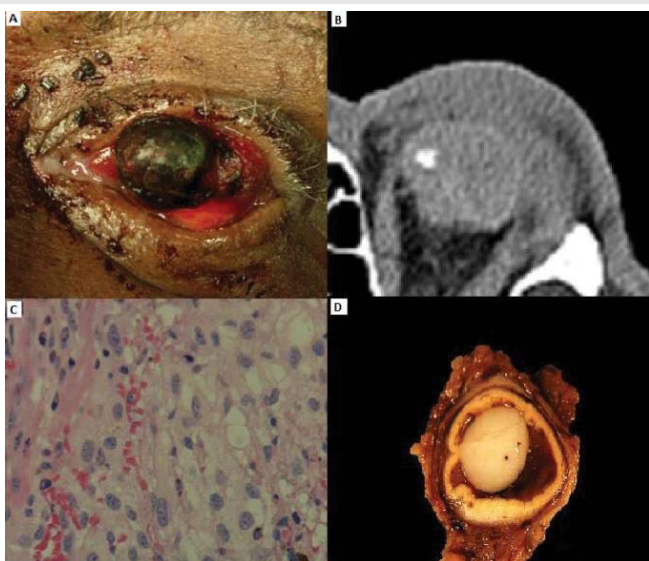


Figure 2: (A) Anterior segment mass. (B) CT scan of orbit. (C) Histological stain showing cells with epithelioid features. (D) Exenterated specimen.

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