Grand Rounds

A 66-year-old woman with extensive conjunctival melanosis

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History

A 66-year-old white woman was referred to the Orbital Oncology & Ophthalmic Plastic Surgery service at The University of Texas MD Anderson Cancer Center for evaluation and management of new, extensive conjunctival pigmentation of the right eye concerning for a conjunctival neoplasm. The patient had been in her usual state of health until 5 weeks prior to presentation, when she suffered a mechanical fall onto a cement sidewalk, sustaining blunt trauma to the right side of her face and head. She had initially presented to her established local ophthalmologist 10 days following her injury complaining of right eye floaters. Examination by the outside ophthalmologist had revealed near 360-degree conjunctival pigmentation, most dense superiorly, in the right eye that had not been previously noted on routine examination 6 months prior. The patient was subsequently referred to our cancer center for evaluation and management of suspected primary acquired melanosis (PAM). The patient's past medical history was significant for rheumatoid arthritis, coronary artery disease, and heart failure. Her past ocular history was significant for bilateral cataract extraction with intraocular lens placement.

Examination

On examination, visual acuity was 20/40 in the right eye and 20/30 in the left eye. Intraocular pressure was 6 mm Hg in the right eye and 16 mm Hg in the left eye. The right pupil was noted to be irregular prior to dilation; the left pupil was within normal limits. There was no relative afferent pupillary defect. Extraocular motility was

Figure 1. Temporal bulbar conjunctiva with diffuse pigmentation in the patient's right eye at initial presentation.

full and visual fields were full to confrontation bilaterally.

Ocular adnexal examination was unremarkable. Slit-lamp examination revealed diffuse pigmentation of the bulbar conjunctiva nearly 360-degrees, with denser pigment superiorly. There was an area of raised conjunctiva superiorly. Pigment did not extend to the palpebral conjunctiva (Figures 1–2). Lids, lashes, cornea, and anterior chambers were unremarkable bilaterally.

Both pupils were pharmacologically dilated. Posterior chamber intraocular lenses were clear and well-positioned in each eye. Fundus examination revealed unremarkable maculae, vessels, and periphery in both eyes.

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Figure 2. Magnified inferotemporal conjunctiva demonstrating lack of palpebral conjunctival pigment on initial presentation.

Figure 3. Pupil peak at 12 o'clock on repeat evaluation, 2 weeks following initial presentation.

Given the very asymmetric intraocular pressure measurements in the two eyes, the possibility of a trauma-induced, small, self-sealing scleral rupture was considered; nevertheless, given the intact anterior chamber and the extent of abnormal conjunctival pigmentation, a conjunctival biopsy was scheduled.

Two weeks later the patient returned for a scheduled biopsy of the conjunctiva. At this time, examination revealed a corectopic pupil with a peak at 12 o'clock (Figure 3). The conjunctiva was elevated superiorly overlying the area of iris prolapse (Figure 4). Conjunctival pigmentation was present and unchanged from initial examination.

Figure 4. Superior conjunctiva showing iris prolapse with overlying conjunctival pigmentation on repeat evaluation, 2 weeks following initial presentation.

Treatment

The patient's conjunctival biopsy was canceled due to strong suspicion for perforating scleral injury with iris prolapse. The patient's care was immediately transferred to another ophthalmologist for evaluation for urgent surgical exploration and repair of the ruptured cataract wound.

Differential Diagnosis

The differential diagnosis of conjunctival pigmentation includes conjunctival nevus, racial melanosis, primary acquired melanosis, secondary acquired melanosis, and conjunctival melanoma. Secondary acquired melanosis may be due to previous irradiation, hormonal changes, metabolic disorders, chemical exposure, topical glaucoma medications, or chronic inflammatory disorders of the eye.¹ Several cases of secondary acquired melanosis of the conjunctiva with findings similar to our patient's findings attributable to perforating injury or iris trauma have been reported.²⁻⁶

Diagnosis and Discussion

Surgical exploration confirmed our clinical suspicion of a superior scleral laceration in the area of the previous cataract wound with iris prolapse through the wound. Subsequent histologic examination confirmed benign melanosis, with pathology revealing numerous regions of subepithelial and substantia propria accumulations of dotlike pigment consistent with uveal pigmentation. Immunohistochemical stains including HMB-45 and S100 confirmed no abnormal melanocytic cells within the epithelium or elsewhere.

The differential diagnosis for pigmentation of the conjunctiva includes several etiologies, as previously mentioned, however new-onset conjunctival pigmentation that is rapidly progressive narrows the top of the differential to acquired melanosis (primary or secondary) and pigmented malignant neoplasia.

Conjunctival melanomas represent about 2% of all ocular malignancies and typically present as focal areas of raised macules, plaques, or masses in an asymptomatic patient, most commonly in the sixth decade.^{7,8} Clinical features that may suggest conjunctival melanoma include large size, lack of mobility, limbal location, variegation, and presence of feeder vessels.⁸ Conjunctival melanoma may arise from areas of primary acquired melanosis, *de novo*, or from a conjunctival nevus. Usually melanoma arising from a prior nevus or *de novo*, appears as black or grey vascularized nodule, whereas lesions arising from PAM with atypia have areas of nodularity and thickening. Conjunctival melanoma can also present as an amelanotic lesion with a pink, smooth “fish-flesh” appearance.⁹ Whereas our patient demonstrated large areas of new conjunctival pigmentation, the history was significant for recent blunt trauma to the eye. Additionally, histological examination our patient’s conjunctiva confirmed that the deposition of pigment cells was not in the epithelial layer but rather deeper within the subepithelial layer and substantia propria.

Our patient’s medical history may have predisposed her to scleral perforation after blunt trauma. First, she has a history of cataract extraction performed through a superior scleral tunnel incision. Expulsive iridodialysis secondary to trauma has been noted in several case reports as a rare complication of blunt trauma to eyes that have undergone previous cataract surgery.^{10–13} Second, our patient suffers from rheumatoid arthritis. Because there is an association between scleral thinning and perilimbal scleromalacia, she may have additionally been predisposed to scleral perforation after blunt trauma.^{14,15}

Several case reports describe secondary acquired conjunctival melanosis after ocular trauma or surgery including intrableb pigmentation (IBP) after trabeculectomy and phacoemulsification,^{2,3} pigmentation following pars-plana vitrectomy,⁴ and pigmentation attributable to a surgical clamp.⁵

A similar case report by Benson et al⁶ describes centrifugal pigment dispersion diffusely throughout the conjunctiva following traumatic scleral perforation with iris incarceration. Electron microscopy of conjunctival tissue from this case revealed many pigment-laden cells in the conjunctival substantia propria closely resembling

the melanosomes found in the normal posterior iris pigmented epithelium. Presumably the incarcerated iris in both our case and this published case led to dispersion of iris pigment from mechanical forces rapidly into the conjunctiva.

Our patient’s rapid development of secondary acquired conjunctival pigmentation was a direct result of a perforating scleral injury with uveal tissue prolapse, and although uncommon, should be considered in the differential diagnosis for melanosis after trauma.

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