JAMA Ophthalmology Clinical Challenge

A Suspicious Pigmented Lesion of the Conjunctiva

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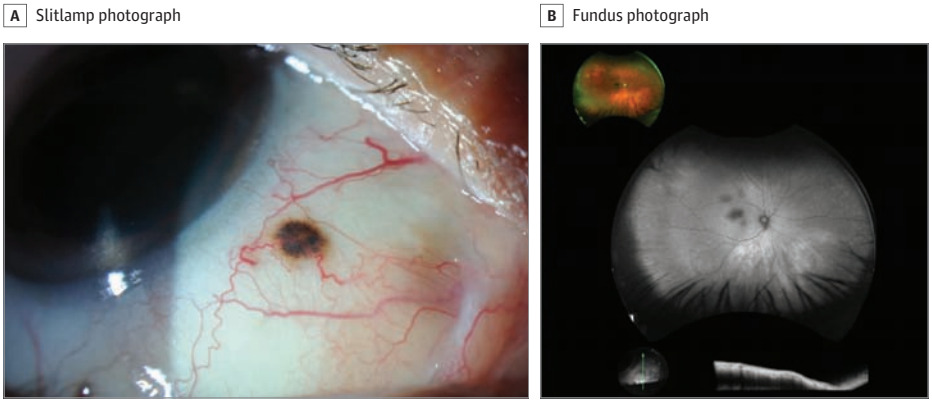

Figure 1. Right eye slitlamp and fundus photographs. A, Slitlamp photograph of the right eye shows a small elevated pigmented lesion at the 4-o'clock position with sentinel vessels. B, Right eye fundus photograph appears normal with no evidence of lesions, pigmentary changes, or abnormal vasculature. Right eye fundus autofluorescence photograph depicting normal foveal hypofluorescence (upper left). Anterior-segment optical coherence tomography of the right eye depicts a small elevated subepithelial lesion at the 4-o'clock position (lower quarter of component B).

A 74-year-old woman with a history of breast cancer, now in remission, presented for evaluation of a conjunctival lesion of the right eye. Per her history, the lesion was noted over 18 years in the past by her optometrist. She reported no noticeable growth or changes to the lesion but was sent for evaluation with ocular oncology. Visual acuity with correction was 20/40 in the right eye and 20/30 in the left eye. Intraocular pressure was 14 mm Hg in the right eye and 16 mm Hg in the left eye. Anterior segment examination results were within normal limits for the left eye, and dilated fundus examination results for both eyes did not reveal concerning pathology. The conjunctiva of the right eye was notable for a small elevated pigmented lesion measuring approximately 2 × 2 mm with sentinel vessels (**Figure 1**A). Fundus photographs and autofluorescence (Figure 1B) as well as optical coherence tomography (OCT) were normal bilaterally. An anterior-segment OCT was performed and revealed a small elevated subepithelial lesion with shadowing vs possible scleral extension (Figure 1B).

WHAT WOULD YOU DO NEXT?

- **A.** Surgical excision of lesion with cryotherapy
- **B.** High-resolution ultrasound
- **C.** Topical chemotherapy/immunotherapy
- **D.** Plaque radiation therapy

CME Quiz at jamacmelookup.com

Diagnosis

Ciliary body melanoma with extrascleral extension masquerading as a conjunctival nevus

What to Do Next

B. High-resolution ultrasound

Discussion

Uveal melanoma is the most common intraocular malignancy in adults and can manifest with a multifaceted initial clinical presentation based on its location and growth.[1-3] Most often, uveal melanoma is found incidentally on a dilated eye examination or when a patient presents with blurry vision.[4] However, there are also reports of patients presenting with hyphema, flashing lights, retinal detachment, spontaneous subretinal bleeding, and glaucoma, among others.[5,6] There are few case reports highlighting extrascleral extension as the primary presentation leading to a diagnosis.

Although the most common melanocytic lesion of the conjunctiva is a conjunctival nevus, melanoma must be considered in the differential diagnosis of any pigmented conjunctival lesion. Though rare, the incidence of melanoma increases with age and peaks in the seventh and eighth decades of life.[7] Additionally, ocular melanoma rates are 8 to 10 times higher in the White population.[4,7] Reasonable treatment options are to excise with cryotherapy, with or without topical chemotherapy/immunotherapy, in the setting of a con-

junctival melanoma. However, surgical excision with cryotherapy (choice A) and topical chemotherapy (choice C) would not be the preferred answers as one must confirm that there is no intraocular tumor present. Plaque radiation therapy (choice D) would not be recommended as the immediate next step until we ensure no underlying intraocular disease. Pursuing choices A and C immediately postexamination would result in inadequate treatment of the patient's underlying pathology. Given the clinical findings above (possible scleral involvement on anterior-segment OCT, sentinel vessels, transpupillary illumination shadowing), it is imperative to first obtain an ultrasound biomicroscopy/high-resolution ultrasound to ensure no underlying pathology before treatment.

In short, the ultrasound biomicroscopy/high-resolution ultrasound of the right eye revealed an intraocular lesion measuring 2.96 mm in depth with low acoustic internal reflectivity and cystic cavities (**Figure 2**). This patient had the conjunctival lesion for 18 years, with no color changes or growth noticed. Upon transillumination of the extrascleral lesion, a 10-mm shadow appeared in the 4-o'clock position. Diagnosis confirmation involved a B-scan ultrasound revealing a 9.5 × 8× 3-mm mass. The diagnosis was a ciliary body melanoma with extrascleral extension masquerading as a conjunctival lesion. This lesion was likely to have gone undetected for years given that it was not visible on fundus examination without depression and did not affect the patient's vision.

Standard of care for ciliary body melanoma of this size is radiotherapy. She was ultimately treated with iodine 125 brachytherapy with fine-needle aspiration biopsy that revealed a melanocytic neoplasm most compatible with malignant uveal melanoma. The gene sequencing panel of the patient's uveal melanoma revealed a missense mutation in GNAQ without a secondary driver mutation. This mutation is tier II, a variant of potential clinic significance, with level C evidence, but has no clinical implications for the patient's future course of management.[8] The patient completed her first follow-up visit 4 months after surgery with expected regression of the ciliary body tumor (now 8 × 7 × 2.5 mm).

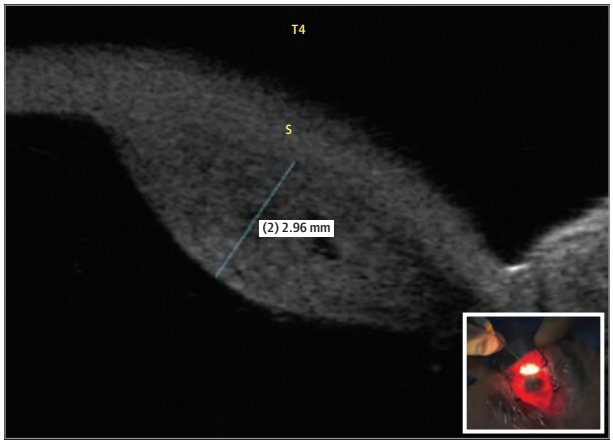

Figure 2. Ultrasound biomicroscopy imaging of the right eye shows an intraocular lesion measuring 2.96 mm in depth with low acoustic internal reflectivity and cystic cavities. Clinical photograph of right globe transillumination reveals a subscleral lesion at the 4-o'clock position (bottom right).

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