

Glaucoma Secondary to Iris Melanoma

BY EIYASS ALBEIRUTI, MD

CASE PRESENTATION

A 63-year-old white woman was referred for advanced unilateral glaucoma. She presented to a local ophthalmologist urgently due to pain and blurred vision in her right eye. Upon examination, her IOP measured 58 mm Hg OD, with advanced unilateral glaucomatous cupping and a suspicious-appearing iris lesion. The physician started the patient on latanoprost 0.005% and a fixed combination of brimonidine tartrate 0.2% and timolol maleate 0.5% in her right eye. Her IOP soon decreased to 22 mm Hg OD, and she was referred to our service the following week for further management.

On presentation at our clinic, the patient's eye was comfortable, her visual acuity was 20/40 OD and 20/30 OS, and her IOP measured 25 mm Hg OD and 14 mm Hg OS (with average corneal thickness) using the aforementioned drops. An examination of her right eye revealed a clear cornea and conjunctiva and a deep, quiet anterior chamber. A lightly pigmented inferotemporal iris mass appeared to have internal vascularity and was associated with ectropion uveae (Figure 1). The surface of the iris showed lightly scattered pigmentary deposition, especially near the lesion. There was mild nuclear sclerosis without focal opacification. The poste-

rior segment was unremarkable except for nearly total cupping and excavation of the optic disc. Gonioscopy showed increased pigmentation of the angle in the patient's right eye compared with mild pigmentation in her left eye. The view to the inferotemporal angle was partially obscured by the mass, but no direct involvement was discernible. Ultrasound biomicroscopy (UBM) showed an iris mass that radially measured 3 mm X 1.4 mm, but no ciliary body involvement was noted (Figure 2). Initial visual field testing of the patient's right eye (24-2 Swedish interactive threshold algorithm-standard) showed a nearly total blackout. The examination of her left eye was unremarkable.

A review of the patient's records revealed that she had initially been seen 5 1/2 years earlier for a raised iris mass (Figure 3), with radial dimensions of 2.5 mm X 1 mm on UBM. At that time, the mass had somewhat distorted the pupil, but the IOP measured in the midteens with no evidence of glaucoma. Gonioscopy had revealed no involvement of the angle. A systemic workup at that time (including computed tomography and bone scans) was negative for any signs of malignancy. Notably, the patient had a medical history of breast cancer, and she had undergone a mastectomy with chemotherapy 12 years earlier, followed by a lumpectomy with chemotherapy

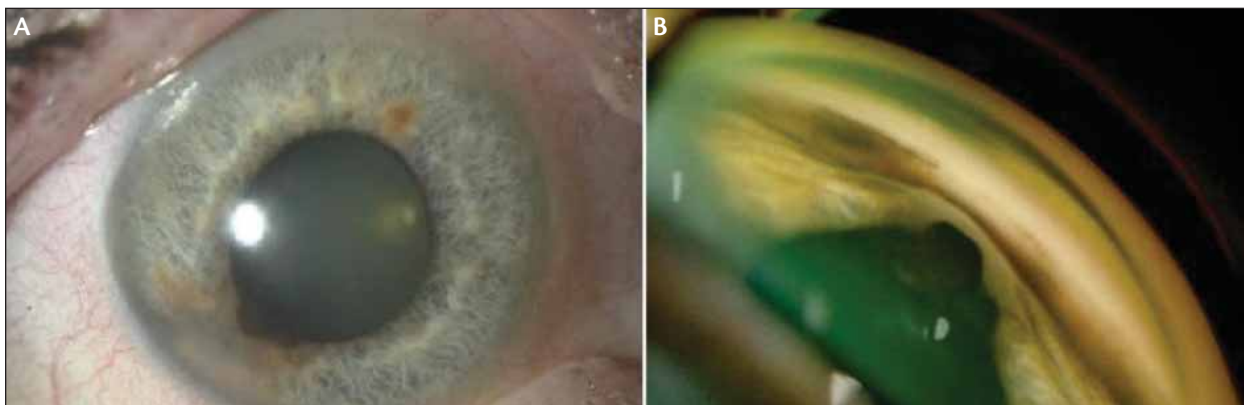


Figure 1. Pigmented iris lesion causing corectopia and ectropion uveae (A). Gonioscopy shows a raised iris mass blocking the view to the angle with dispersed angle pigmentation (B).



Figure 2. UBM gives a radial view of a stromal iris mass, measuring 3 mm long and 1.4 mm thick. There is no apparent involvement of the iris root and ciliary body.

and radiation 8 years later for a recurrence. She has been in remission and has been following up regularly with her oncologist for surveillance. At the time of her initial ophthalmic evaluation, the possibility of an iris melanoma was raised, and close observation was advised. The patient, however, was lost to ophthalmic follow-up until recently.

DISCUSSION

This uncommon case of secondary glaucoma from an iris lesion raises concern about melanoma. The differential diagnosis of an iris melanoma includes iris nevus, melanocytoma, iris cyst, and metastasis to the iris. Although nevi are common, iris melanomas are rare and account for only about 3% of all uveal melanomas.¹ Both nevi and melanomas usually occur in the inferior half of lightly colored irides among white patients and can be difficult to tell apart clinically. The presence of findings such as corectopia, ectropion uveae, internal vascularity, cataract formation, and even lesion growth may not be diagnostic. Ophthalmologists, however,

must consider excising lesions that demonstrate growth as suspected melanoma.² A careful examination and documentation of the tumor's size and location using slit-lamp photography and UBM, if available, are critical in this regard. Other signs of concern include a large tumor (> 3 mm in diameter), prominent vascularity, pigment dispersion or glaucoma, and signs of extension into adjacent structures.³

Iris melanoma is not very aggressive, and the overall chance of metastasis of a biopsy-proven melanoma is only 5% after 10 years.¹ Some risk factors for metastasis are older age, elevated IOP, involvement of the iris insertion and angle, and extraocular extension. Iris melanomas can be circumscribed lesions or diffuse infiltrating tumors that are seen as hyperpigmentation of the iris and/or angle. The standard options for treatment are excision by iridectomy or iridocyclectomy if the tumor is not too extensive, or enucleation if the tumor is not resectable. Plaque radiotherapy is also available as an adjunct or alternative to excision or enucleation in some cases.

Iris melanoma can be associated with elevated IOP in 7% of clinically suspected cases but in up to 30% of microscopically confirmed cases.⁴ The mechanism by which IOP elevation typically occurs is either direct infiltration of tumor cells into the angle or obstruction of the trabecular meshwork by dispersed pigment (and any engulfing macrophages). Less common mechanisms are angle closure and neovascularization. Treating glaucoma due to iris melanoma can be challenging in light of the contraindication to surgeries such as trabeculectomy, which create a pathway to potential extraocular extension.⁵ Even laser trabeculoplasty may pose a theoretical risk of promoting the tumor's spread; no evidence supports this concern, however, and a cautious attempt at laser trabeculoplasty away from the areas of tumor involvement may be reasonable if medical thera-

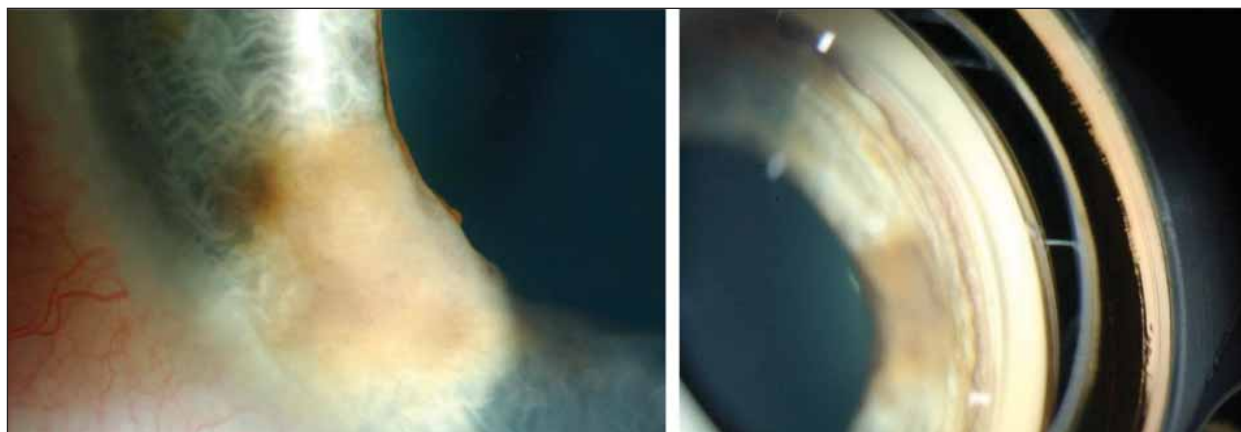


Figure 3. The same lesion several years ago. Early pupillary distortion is seen. The angle is wide open with little pigmentation.

py fails. Likewise, transscleral cyclophotocoagulation can be performed, preferably in quadrants away from the tumor.

It is worth reiterating the possibility that iris melanoma will present in its diffuse infiltrative form, leading to unilateral glaucoma. If its spread is confined to the angle, creating a rare so-called ring melanoma, it may be mistaken for primary open-angle glaucoma if careful gonioscopy is not performed. Alternatively, if the melanoma also diffusely involves the entire iris, it can be mistaken for a unilateral pigmentary glaucoma and inappropriately treated with surgical filtration.⁶ A hyperchromic heterochromia of the iris with ipsilateral glaucoma should raise this possibility in the clinician's mind and trigger a careful investigation.

CLINICAL COURSE

This case was highly suspicious for a primary iris melanoma given the size of the lesion, its documented growth, and the secondary glaucoma induced by the tumor. A metastatic lesion from breast cancer was also a consideration, but it was unlikely, particularly since the systemic workup was normal and typical features of metastatic carcinoma were absent. Because there did not appear to be any direct extension or seeding into the angle, pigment dispersion was the presumed mechanism of glaucoma. The patient began therapy with dorzolamide hydrochloride 2% in addition to her other eye drops, and her IOP decreased to the upper teens, which was acceptable. She was referred to a regional ocular oncologist. Because there was no evidence of extension of the tumor into the ciliary body, the patient did not require enucleation. Rather than undergo extensive iris resection, she elected to receive plaque radiotherapy. □

Section Editor Eiyass Albeiruti, MD, is an assistant professor of ophthalmology at the University of Pittsburgh in Pennsylvania. Dr. Albeiruti may be reached at (412) 647-8199; albeirutie2@upmc.edu.



1. Shields CL, Shields JA, Materin M, et al. Iris melanoma, risk factors for metastasis in 169 consecutive patients. *Ophthalmology*. 2001;108:172-178.
2. Territo C, Shields CL, Shields JA, et al. Natural course of melanocytic tumors of the iris. *Ophthalmology*. 1988;95:1251-1255.
3. Singh AD. Tumors of the uvea: benign melanocytic tumors. In: Singh AD, Damato BE, Pe'er J, et al, eds. *Clinical Ophthalmic Oncology*. W.B. Saunders: Philadelphia, PA: 2007;185-186.
4. Shields CL, Materin MA, Shields JA, et al. Factors associated with elevated intraocular pressure in eyes with iris melanoma. *Br J Ophthalmol*. 2001;85:666-669.
5. Grossniklaus EI, Brown RH, Stulting RD, et al. Iris melanoma seeding through a trabeculectomy site. *Arch Ophthalmol*. 1990;108:1287-1290.
6. Demirci H, Shields CL, Shields JA, et al. Diffuse iris melanoma: a report of 25 cases. *Ophthalmology*. 2002;109:1553-1560.