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An Iris Tumor

Rajesh C. Rao, M.D.^{1,2,3,4} and J. William Harbour, M.D.^{5,6}

¹Department of Ophthalmology and Visual Sciences, W.K. Kellogg Eye Center, University of Michigan Medical School, Ann Arbor, MI

²Department of Pathology, University of Michigan Medical School, Ann Arbor, MI

³University of Michigan Comprehensive Cancer Center, Ann Arbor, MI

⁴Division of Ophthalmology, Surgical Service, Veterans Administration Ann Arbor Healthcare System, Ann Arbor, MI

⁵Ocular Oncology Service, Bascom Palmer Eye Institute, University of Miami Miller School of Medicine, Miami, FL

⁶Sylvester Comprehensive Cancer Center, University of Miami Miller School of Medicine, Miami, FL

A 58-year-old woman noticed a gradually enlarging, white-brown, circular lesion on her right iris over the last several years, but denied visual changes. She did not report trauma, changes in vision, foreign travel, and did not take any topical ophthalmic medications. Her past medical history was unremarkable. Her best corrected visual acuity was 20/25 in the right eye and 20/20 in the left. Her pupils were round with no afferent pupillary defect, although the circular lesion did move in concert with the pupillary constriction and dilation. The intraocular pressure was 14 mm Hg in both eyes. Extraocular movements were normal. Slit lamp examination of the anterior segment showed a 2mm × 2mm circular iris lesion at the inferotemporal border of the iris. The mass appeared to have a fluid-debris level (Figure 1, A,B, arrowheads). When the anterior segment visualized with a gonioscope (Figure 1, C, D) the iridocorneal angle was open, no neovascularization was present, and a spherical mass about 2 mm in height arose from the pupillary margin (arrowheads). Its nasal surface appeared white with speckled brown pigmentation, while the remainder of the lesion had brown pigment (Figure 1D). The anterior segment of the left eye, and posterior segment of both eyes were unremarkable.

What would you do next?

- A. Observe at yearly intervals
- B. Obtain high-frequency ultrasound biomicroscopy
- C. Perform transcorneal biopsy of the lesion

Corresponding author: Rajesh C. Rao, MD, W.K. Kellogg Eye Center, 1000 Wall St., Brehm Rm 8326, Ann Arbor, MI 48105, rajeshr@med.umich.edu.

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- D.** Completely excise the lesion with pathological analysis

Diagnosis

Iris stromal cyst of the right eye.

What to do next

Obtain high-frequency ultrasound biomicroscopy (UBM).

The key feature in this case is that the patient's iris mass contained a fluid-debris level (Figure 1A) and arose anterior to the posterior iris surface, confirmed by ultrasonography (Figure 2), which is consistent with an iris stromal cyst.¹

Discussion

Iris tumors can be benign or malignant lesions. It is important to differentiate iris cysts from solid lesions, as the latter class is more often associated with a variety of malignancies.

Cystic and solid lesions constitute 20% and 80%, respectively, of iris tumors.² Cystic iris tumors can be further classified into stromal or pigmented epithelial cysts (IPE). Derived from iris posterior pigmented epithelium, IPE cysts are typically opaque, grow on the back surface of the iris, and may not be apparent until large enough to bow the anterior iris. Stromal cysts arise from the iris stroma, are often lucent, have a smooth surface, contain a fluid-debris level, and are generally unilateral (Figures 1 and 2).¹

Stromal cysts can be acquired, as in our patient, or congenital.¹ These tumors can grow, rupture, and hemorrhage, leading to hyphema, amblyopia (in children), glaucoma, lens dislocation, corneal touch, cataract, uveitis, photophobia, and discomfort.¹ Stromal cysts diagnosed under 10 years of age tend to have a more aggressive course, often requiring multiple surgeries and leading to vision loss, whereas those in later years tend to be more benign, with only one-quarter requiring intervention.¹

Ocular oncologists may encounter iris tumors distinct from stromal cysts. The differential of iris stromal cysts include primary cysts such as IPE, and secondary iris cysts due to trauma, surgery, paraneoplastic syndromes, malignancies, parasites (e.g. cysticercosis), inflammation (e.g. Fuchs heterochromic iridocyclitis), systemic disorders (e.g. mucopolysaccharidoses, hypertension), as well as medications such as anticholinesterases and prostaglandin analogues.^{1,3-7} Solid lesions can masquerade as iris cysts, and these include: iris nevi, melanoma of the iris, ciliary body, and choroid, ciliary body medulloepithelioma, metastases (e.g. breast, lung), juvenile xanthogranuloma, iris pigment adenoma and adenocarcinoma.² Other non-neoplastic lesions that can simulate iris cysts include iridocorneal endothelial syndrome, iris atrophy, coloboma, foreign body, varices, and hemangiomas.² Lisch nodules and Brushfield spots of the iris are multifocal and occur in patients with neurofibromatosis type 1, and Down syndrome, respectively.²

Since the slit-lamp examination suggested a fluid-filled cyst, high-frequency UBM is the most appropriate diagnostic step following a history and ophthalmic evaluation.¹ UBM

confirmed (Figure 2) presence of iris stromal cyst, and since there was no obstruction of the visual axis, glaucoma, or other negative effect, the iris stromal cyst can be monitored, and no biopsy was needed. A fluid-debris level was noted (Figure 1A), which is commonly found in these cysts, though this finding is not pathognomonic.¹ The debris material was whitish in color and could be seen on gonioscopy through the epithelial lining of the cyst (Figure 1B), and the fluid-debris level may represent serous material with free iris epithelial cells.⁸ Occasionally, iris stromal cysts have been known to regress spontaneously.⁹ Diathermy, electrolysis, aspiration followed by injection of absolute alcohol, and surgical excision with or without adjunctive cryotherapy and laser photocoagulation are therapeutic approaches that have been described should an iris stromal cyst become symptomatic.¹⁰ Following treatment, iris stromal cysts rarely recur.¹⁰

Patient outcome

The patient was reassured that the condition was an iris stromal cyst that is generally benign. She has been monitored yearly for several years without significant growth of the iris stromal cyst and remains asymptomatic.

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References

1. Lois N, Shields CL, Shields JA, Mercado G, De Potter P. Primary iris stromal cysts. A report of 17 cases. *Ophthalmology*. 1998; 105(7):1317–1322. [PubMed: 9663240]
2. Shields CL, Kancherla S, Patel J, et al. Clinical survey of 3680 iris tumors based on patient age at presentation. *Ophthalmology*. 2012; 119(2):407–414. [PubMed: 22035581]
3. Lai IC, Kuo MT, Teng LM. Iris pigment epithelial cyst induced by topical administration of latanoprost. *Br J Ophthalmol*. 2003; 87(3):366. [PubMed: 12598462]
4. Chin NB, Gold AA, Breinin GM. Iris Cysts and Miotics. *Arch Ophthalmol*. 1964; 71:611–616. [PubMed: 14120647]
5. Joseph A, Rahimy E, Sarraf D. Bilateral diffuse uveal melanocytic proliferation with multiple iris cysts. *JAMA Ophthalmol*. 2014; 132(6):756–760. [PubMed: 24763691]
6. Sato S, Maeda N, Watanabe H, et al. Multiple iridociliary cysts in patients with mucopolysaccharidoses. *Br J Ophthalmol*. 2002; 86(8):933–934. [PubMed: 12140222]
7. Phowthongkum P, Burapasubkajorn P, Intarabeth P, Buddhari W, Kowitoonkij W. Familial aortic dissection and congenital iris flocculi with hypertension. *Ophthalmic Genet*. 2008; 29(3):126–127. [PubMed: 18766992]
8. Boruchoff SA, Kenyon KR, Foulks GN, Green WR. Epithelial cyst of the iris following penetrating keratoplasty. *Br J Ophthalmol*. 1980; 64(6):440–445. [PubMed: 6992858]
9. Brent GJ, Meisler DM, Krishna R, Baerveldt G. Spontaneous collapse of primary acquired iris stromal cysts. *Am J Ophthalmol*. 1996; 122(6):886–887. [PubMed: 8956645]
10. Shields CL, Arepalli S, Lally EB, Lally SE, Shields JA. Iris stromal cyst management with absolute alcohol-induced sclerosis in 16 patients. *JAMA Ophthalmol*. 2014; 132(6):703–708. [PubMed: 24723076]

Figure 1.

(A) Slit-lamp photograph of anterior segment showed an iris lesion with a fluid-debris level (black arrowhead) inferotemporal to the pupillary border (white arrowhead). (B) Gonioscopic views a showed iris lesion (black arrowhead) emerging from pupillary border (white arrowhead).

Figure 2.
(A) High-frequency ultrabiomicroscopy showed an iris stromal cyst emerging anterior to the iris plane (white arrowhead). A debris-fluid level (black arrowhead) correlated with the color photo (Figure 1A, black arrowhead).