JAMA Ophthalmology Clinical Challenge

Gravitational Shifting of a Pigmented Iris Lesion in a Young Boy

Kevin R. Card, BS; Matthew R. Barke, MD; Carol L. Shields, MD

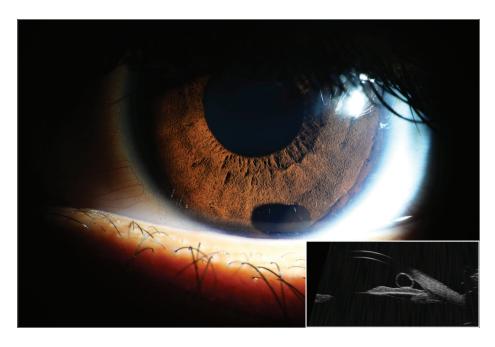


Figure. Slitlamp biomicroscopy of the left eye showing a round, pigmented lesion in the inferior anterior chamber angle abutting the corneal endothelium and resting on the iris stroma measuring 3 × 3 mm in basal dimension. Inset, Ultrasound biomicroscopy scan of the free-floating cyst showing central lucency, no fluid level, and no solid component.

A 13-year-old boy with a history of sports-related blunt trauma to the left eye was referred for evaluation of an asymptomatic, pigmented iris lesion in the left eye. On examination, his best-corrected visual acuity was 20/20 OU, and intraocular pressures were normal in



Multimedia

both eyes. Results of slitlamp examination of the right eye were unremarkable. Slitlamp examination of the left eye revealed a round, pigmented lesion measuring 3 × 3 mm in basal dimension and with gravitational shift-

ing within the anterior chamber fluid with patient head tilt (Video). There was no corneal guttatae or edema. Anterior segment optical coherence tomography depicted the lesion in the anterior chamber angle abutting the corneal endothelium and resting on the iris stroma with no internal fluid level and no solid component. Ultrasound biomicroscopy confirmed the lesion to be cystic with a thickness of 1.6 mm. Dilated fundus examination revealed normal findings in both eyes.

WHAT WOULD YOU DO NEXT?

- **A.** Fine-needle aspiration biopsy with cytology and cytogenetics
- B. Iodine 125 plaque radiotherapy
- C. Observe with no additional testing or intervention
- **D.** Cyst paracentesis with complete drainage
- + CME Quiz at jamacmelookup.com

Diagnosis

Free-floating iris pigment epithelium cyst

What to Do Next

C. Observe with no additional testing or intervention

Discussion

1000

A pigmented mass in the anterior chamber could represent a benign entity but also raises suspicion for malignancy, such as iris melanoma. Iris tumors are predominantly characterized as solid (79% of cases) or cystic (21% of cases). Solid tumors include melanocytic lesions, with the 3 most common being benign iris nevus (60%), iris melanoma (26%), and iris freckle (4%). Cystic iris tumors are subclassified based on their tissue layer of origin, arising from either the iris stroma or iris pigment epithelium (IPE). Iris stromal cysts appear translucent, located on the anterior iris surface and with smooth lobulated surface, typically in young children. By contrast, IPE cysts appear on the posterior iris surface with brown or

JAMA Ophthalmology October 2023 Volume 141, Number 10

jamaophthalmology.com

black color simulating solid melanocytic tumors, such as iris melanoma or IPE adenoma. ^{1,2} For this reason, some IPE cysts have historically been mistaken for melanoma, even leading to enucleation. As iris melanoma is malignant with risk for metastasis, proper treatment for life protection with consideration of the potential for ocular adverse sequelae should be realized. Differentiation of the various iris tumors is paramount to management.

Ultrasound biomicroscopy and anterior-segment optical coherence tomography are useful tools in assessing the internal quality of iris tumors and differentiation of solid from cystic tumors. These technologies are critical for defining tumor basal dimension and thickness to monitor change over time or response to treatment. In this patient, slitlamp biomicroscopy (Figure) demonstrated the gravitational shifting of the mass (Video), and both ultrasound biomicroscopy (Figure, inset) and anterior-segment optical coherence tomography documented the mass to be cystic with no internal fluid or solid component. The Video demonstrates the superior-to-inferior descent of an IPE cyst across the visual axis, coursing purely through the aqueous humor with gravity, offering insight into the dynamic behavior of this lesion.

Free-floating IPE cysts occur when the cyst becomes separated from the IPE, speculated to occur following ocular trauma, as in this patient.^{4,5} These free-floating IPE cysts are benign and rarely cause endothelial decompensation or visual sequelae.^{2,5} Fine-needle aspiration biopsy with cytology and cytogenetics (choice A) was not appropriate because the lesion in question was not solid or within the iris, so iris melanoma was not likely. lodine 125 plaque radiotherapy (choice B) was not appropriate, as this was a cystic, benign mass that was unlikely to be malignant. Cyst paracentesis with complete drainage (choice D) was not necessary in this case because the cyst did not affect the visual axis and was visually asymptomatic. Additionally, drainage of a free-floating cyst without any portion anchored to the iris would likely prove to be technically difficult.

In summary, free-floating IPE cysts are benign epithelial cysts that can dislodge into the aqueous or vitreous following trauma, as in this patient. This form of cyst is benign and rarely leads to adverse sequelae; therefore, conservative observation is advised.

Patient Outcome

This patient was counseled on the benign nature of the free-floating IPE cyst and advised to follow up in 12 months for repeated examination with imaging. There was no expectation that this cyst would grow, rupture, transform into a malignant process, or cause damage to the cornea.

ARTICLE INFORMATION

Author Affiliations: Ocular Oncology Service, Wills Eye Hospital, Thomas Jefferson University, Philadelphia, Pennsylvania.

Corresponding Author: Carol L. Shields, MD, Ocular Oncology Service, Wills Eye Hospital, Thomas Jefferson University, 840 Walnut St, Ste 1440, Philadelphia, PA 19107 (carolshields@gmail.com).

Published Online: September 14, 2023. doi:10.1001/jamaophthalmol.2023.4019

Conflict of Interest Disclosures: Dr Shields is supported in part by the Eye Tumor Research Foundation. No other disclosures were reported.

Additional Contributions: We thank the family of the patient for granting permission to publish this information.

REFERENCES

- 1. Shields CL, Shields PW, Manalac J, Jumroendararasame C, Shields JA. Review of cystic and solid tumors of the iris. *Oman J Ophthalmol*. 2013;6(3):159-164. doi:10.4103/0974-620X.122269
- 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. doi:10.1016/j.ophtha.2011.07.059
- 3. Bianciotto C, Shields CL, Guzman JM, et al. Assessment of anterior segment tumors with ultrasound biomicroscopy versus anterior segment optical coherence tomography in 200 cases. *Ophthalmology*. 2011;118(7):1297-1302. doi:10.1016/j.ophtha.2010.11.011
- 4. Singh G, Narendran K, Saravanan VR, Narendran V. Pigmented free-floating iris cysts. *Br J Ophthalmol*. 2007;91(8):1037. doi:10.1136/bjo.2007.126078
- 5. Rotsos T, Bagikos G, Christou S, et al. Free-floating iris pigmented epithelial cyst in the anterior chamber. *Case Rep Ophthalmol Med*. 2016; 2016:4731037. doi:10.1155/2016/4731037