JAMA Ophthalmology Clinical Challenge

Amelanotic Fundus Lesion in an Older White Woman

Sanjana Davuluri, MHS; Vijitha S. Vempuluru, MD; Carol L. Shields, MD

A Fundus photograph of the left eye

B Magnetic resonance imaging of the orbits

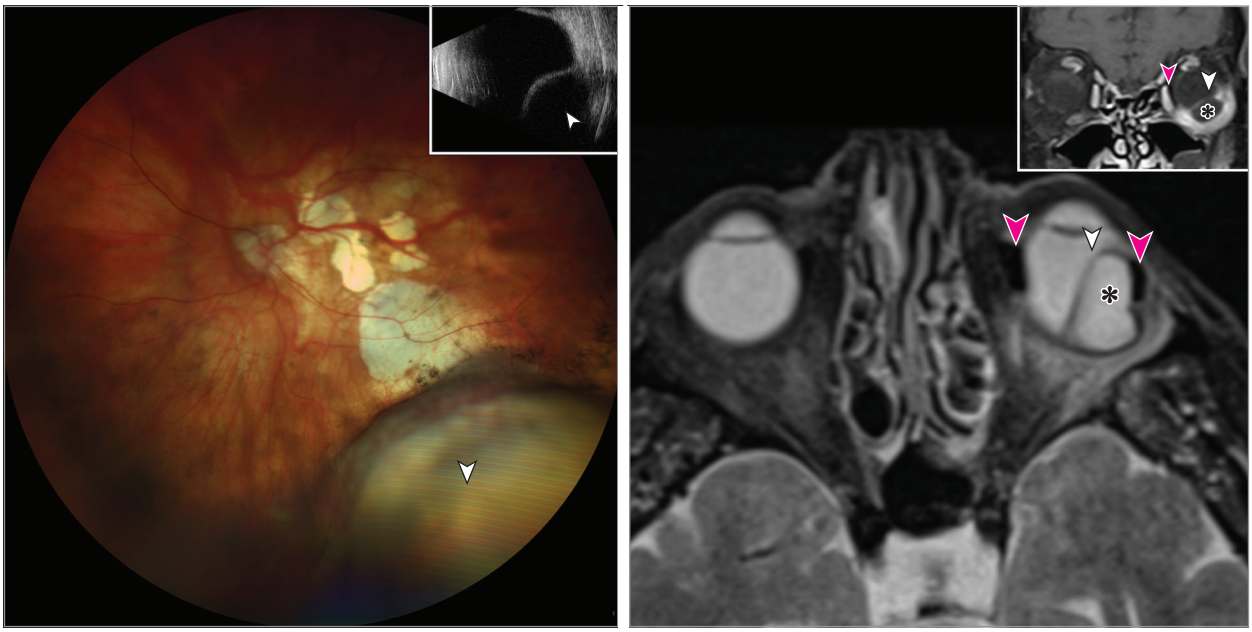

Figure. Fundus photograph and magnetic resonance imaging of the left eye. A, Fundus photograph of left eye showed a pale optic disc, geographic macular atrophy, and an inferotemporal amelanotic mass (white arrowhead); inset shows ultrasonography revealing an echolucent mass (white arrowhead). B, Magnetic resonance imaging (MRI) (T2-weighted axial orientation) revealed a shallow hypointense circumferential band in the left eye (pink arrowheads), inferotemporal hyperintense nodular mass (asterisk), and inward compressed sclera (white arrowhead); inset (T1-weighted coronal orientation with gadolinium) shows hypointense encircling band (pink arrowhead), subtle scleral indentation (white arrowhead), and no gadolinium enhancement of mass (asterisk).

An 81-year-old White woman noted decreased vision in her left eye for 6 months. She was referred to the Ocular Oncology Service, Wills Eye Hospital, Philadelphia, Pennsylvania, for suspected choroidal melanoma. She disclosed a history of macular degeneration in both eyes and retinal detachment in the left eye that was treated 35 years previously. Medical history revealed cutaneous basal cell carcinoma and squamous cell carcinoma, both treated surgically.

On examination, visual acuity was 20/50 OD and 20/400 OS. External examination showed posterior-chamber intraocular lenses in both eyes and conjunctival scarring in the left eye from circumferential scleral buckle surgery with no visible extraocular tumor. Fundus evaluation showed macular drusen in the right eye and a pale optic disc with geographic macular atrophy in the left eye, explaining her visual acuity of 20/400 OS. In addition, a shallow circumferential buckle effect and an inferotemporal mass measuring 15.0 × 10.0 mm in basal dimension and 9.1 mm in thickness (**Figure**, A) were seen in the left eye. The mass appeared elevated, amelanotic with overlying retinal vasculature, and with chorioretinal atrophy. No retinal detachment or breaks were seen. By ultrasonography the mass was echolucent (Figure, A inset). Magnetic resonance imaging (MRI) revealed a T1, T2-hypointense shallow circumferential band in the left eye. In addition, there was an inferotemporal nodular mass in the left eye showing T1 (gadolinium) hypointense, T2-hyperintense features, underlying the encircling buckle, and with adjacent subtle delineation. There was no enhancement with gadolinium (Figure, B).

WHAT WOULD YOU DO NEXT?

- **A.** Whole-body positron emission tomography scan
- **B.** Fine-needle aspiration biopsy
- **C.** Plaque radiotherapy
- **D.** Observation

CME Quiz at jamacmelookup.com

Diagnosis

Expanded hydrogel scleral sponge

What to Do Next

D. Observation

Discussion

In this case, based on MRI and the lack of gadolinium enhancement, the final diagnosis was expanded hydrogel sponge from previous retinal detachment repair. This mass did not show MRI features of choroidal melanoma. The mass showed complete ultrasonographic acoustic hollowness, potentially suggestive of choroidal melanoma, but MRI confirmed a subtle delineation on the inner portion of the mass (Figure, B) suggestive of thinned sclera, indicating that the mass was episcleral and indenting the globe. The mass was hypointense on T1-weighted (gadolinium-enhanced) imaging suggesting a nonvascular mass and hyperintense on T2-weighted imaging suggesting a hydrophilic mass, possibly a hydrogel sponge. We suspect that the sponge, over time, had slowly expanded underneath the circumferential buckle, leading to inward indentation of the globe and the appearance of an intraocular tumor (Figure).

Scleral buckle is one of the highly effective options for retinal detachment repair with success rates ranging from 63% to 99% between various studies.[1] The hydrogel scleral buckle was introduced in the 1980s and was deemed safe with low risk of infection when soaked in antibiotics, owing to the hydrophilic nature. In addition, the buckle slowly expanded over time, a presumed beneficial effect to further buckle the retina.[2] However, delayed complications were later recognized as this implant continued expansion disproportionately, leading to extreme scleral buckle effect, resulting in diplopia, dysmotility, pseudotumor formation in the orbit, eyelid, conjunctiva, and globe, occasionally with inflammation and pain.[2,3]

Our group has previously described expanding hydrogel (MIRAgel) scleral sponge mimicking orbital cysts and tumors.[4] This patient had no symptom of an enlarging orbital mass but was clinically found to have an asymptomatic intraocular mass, suspicious for uveal melanoma. However, the clinical features of the episcleral mass combined with imaging that demonstrated the nonvascular mass overlying markedly thinned sclera, confirmed our suspicion of a hydrogel sponge that presumably expanded over 35 years.

Swollen hydrogel implants can be managed by careful surgical excision or close observation.[3,4] Despite various techniques described for excision of this discohesive, distended material, complete excision is challenging and risks include continued swelling of remnant implant, retinal detachment recurrence, and thinned scleral wall leading to perforation.[3] In the absence of intervention, continued enlargement of the buckle effect can lead to complications; hence, long-term surveillance is advised. In view of this patient's age and absence of symptoms, observation (option D) was recommended. On subsequent follow-up, if enlargement, extrusion, or any signs of infection are noted, implant removal may be considered. Since the absence of tumor or malignancy was suggested by MRI, there was no need for positron emission tomography scan (option A), biopsy (option B), or radiotherapy (option C).

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 28, 2023. doi:10.1001/jamaophthalmol.2023.4325

Conflict of Interest Disclosures: Dr Vempuluru reported grants from the Victoria Cohen Eye Cancer Charitable Trust Scholarship for pursuing fellowship outside the submitted work. Dr Shields reported fees from the Eye Tumor Research Foundation and Aura Biosciences outside the submitted work. No other disclosures were reported.

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

REFERENCES

1. Fallico M, Alosi P, Reibaldi M, et al. Scleral buckling: A review of clinical aspects and current concepts. *J Clin Med*. 2022;11(2):314. doi:10.3390/jcm11020314

2. Thompson JT, Chambers WA. Good ideas gone bad: the MIRAgel saga. *Ophthalmology*. 2016;123(1):5-6. doi:10.1016/j.ophtha.2015.09.038

3. Crama N, Klevering BJ. The removal of hydrogel explants: An analysis of 467 consecutive cases. *Ophthalmology*. 2016;123(1):32-38. doi:10.1016/j.ophtha.2015.08.018

4. Shields CL, Demirci H, Marr BP, Mashayekhi A, Materin MA, Shields JA. Expanding MIRAgel scleral buckle simulating an orbital tumor in four cases. *Ophthalmic Plast Reconstr Surg*. 2005;21(1):32-38. doi:10.1097/01.IOP.0000148409.31127.F5