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Suspicious Retinal Lesion With a Distinctive Appearance

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Case

A 47-year-old asymptomatic female presented for routine examination. Visual acuity was 20/20 in both eyes (OU). On slit-lamp examination, she was found to have normal anterior segment OU. Fundus examination of the right eye (OD) revealed an amelanotic, noncalcified, intraretinal lesion measuring 5 × 5 mm in basal dimensions [Fig. 1a] with a thickness of 2.2 mm on ultrasonography. Fundus examination of the left eye (OS) showed flat retina with intact macula. The patient had no history of systemic disease. Her family history was insignificant.

What is your next step?

A. Fine needle aspiration biopsy (FNAB)
B. Transpupillary thermotherapy (TTT)
C. Observation
D. Application of radioactive plaque

Findings

Optical coherence tomography (OCT) OD demonstrated a retinal mass within the nerve fiber layer with a "moth-eaten" appearance [Fig. 1b]. This distinct OCT feature was the result of numerous intralesional cavities. No treatment was rendered at this time, and the mass was observed with annual follow-up. The patient has remained asymptomatic, and the tumor has remained stable with no change or growth over 7 years.

Diagnosis: Retinal astrocytic hamartoma (RAH)

Correct answer: C, Observation

Discussion

RAH is a congenital benign tumor composed of well-differentiated astrocytes.[1,2] The mean age at presentation was reported at 32 years.[1] RAH presents with a variety of clinical features on ophthalmoscopy. This lesion can be calcified, noncalcified, or both.[1,2] Differential diagnosis includes retinoblastoma, retinocytoma/retinoma, retinal astrocytoma, granuloma, and pseudoneoplastic gliosis.[2,3] Diagnosis is aided by retinal OCT, with >90% showing the characteristic moth-eaten appearance and 100% arising within the nerve fiber layer.[1] RAH is often associated with tuberous sclerosis complex (TSC) and is considered a diagnostic marker for TSC.[1,2,4] As such, discovery of RAH on routine exam may warrant further systemic workup.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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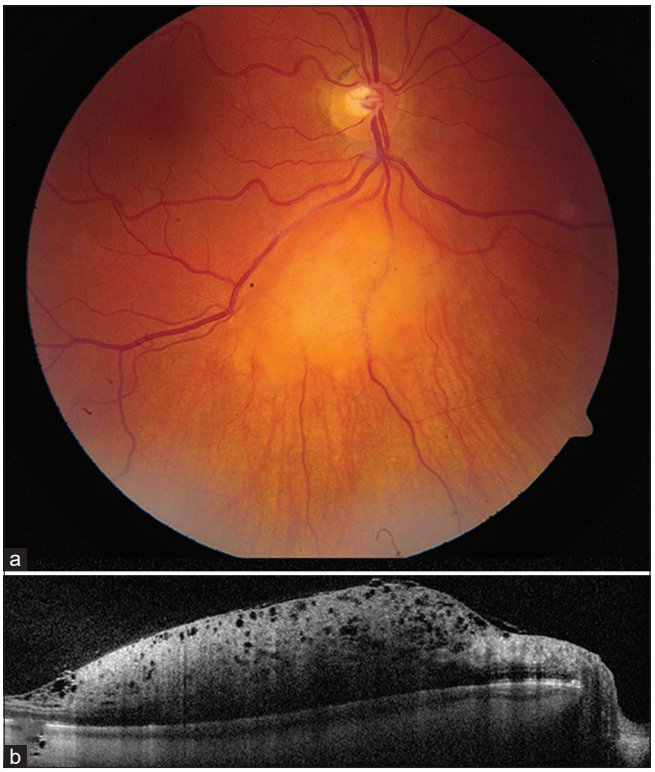

Figure 1: (a) Fundus photograph OD showing an amelanotic, noncalcified, intraretinal lesion in the inferior quadrant. (b) Retinal OCT of the intraretinal lesion showing numerous intralesional cavities contributing to a distinct "moth-eaten" appearance. OCT = optical coherence tomography

Conflicts of interest

There are no conflicts of interest.

References

1. Shields CL, Say EAT, Fuller T, Arora S, Samara WA, Shields JA. Retinal astrocytic hamartoma arises in nerve fiber layer and shows "Moth-Eaten" Optically empty spaces on optical coherence tomography. Ophthalmology 2016;123:1809-16.
2. Shields JA, Shields CL. Glial tumors of the retina. The 2009 King Khaled Memorial Lecture. Saudi J Ophthalmol 2009;23:197-201.
3. Shields CL, Benevides R, Materin MA, Shields JA. Optical coherence tomography of retinal astrocytic hamartoma in 15 cases. Ophthalmology 2006;113:1553-7.
4. Pichi F, Massaro D, Serafino M, Carrai P, Giuliari GP, Shields CL, *et al*. RETINAL ASTROCYTIC HAMARTOMA: Optical coherence tomography classification and correlation with tuberous sclerosis complex. Retina 2016;36:1199-208.

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