Photo Essay

Foveolar simple retinal pigment epithelial hamartoma

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Key words: Optical coherence tomography angiography, retinal pigment epithelial hamartoma, Swept source optical coherence tomography

Case Report

A 28-year-old male patient underwent routine ophthalmic evaluation. Visual acuity was 20/20, and the anterior segment was unremarkable in both the eyes. Right eye retinal examination was normal whereas the left eye showed a healthy optic disc along with a well-defined circular pigmented lesion at the foveola [Fig. 1a and b]. The rest of the retinal examination was within normal limits. Swept-source optical coherence tomography (OCT) revealed a hyper-reflective lesion just inferior to the umbo [Fig. 1c]. The hyperreflective lesion was seen just adjacent to the foveal dip in the inner retinal layers. The outer retinal and choroidal layers below the lesion were obscured because of the shadowing. However, the outer retinal and pigment epithelial layers surrounding the lesion appeared intact without any discontinuity or neurosensory detachment [Fig. 1d]. OCT angiography revealed a mildly distorted foveal avascular zone at the location of the lesion. However, the adjacent superficial retinal plexus was intact and well preserved. No abnormal or intrinsic vasculature was noted in the lesion [Fig. 1e]. The patient was counseled about the nature of the retinal lesion and advised for regular follow-up.

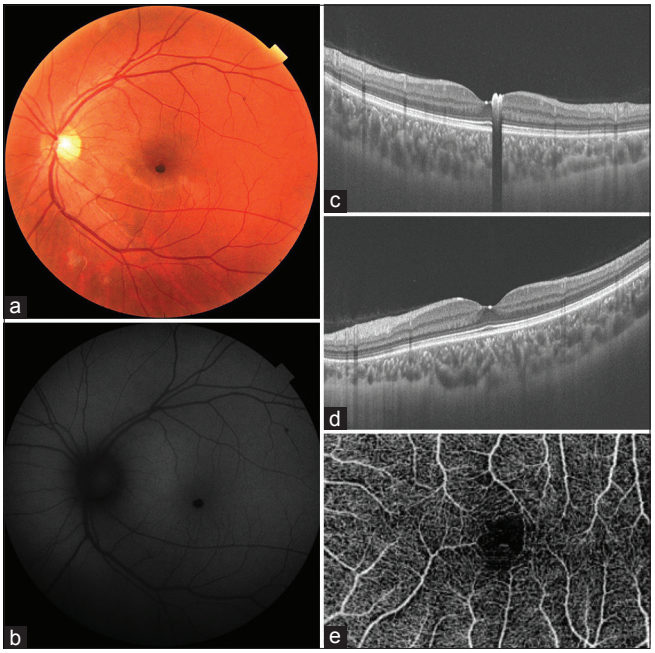

Figure 1: (a and b) Left eye color and red-free fundus photographs showing a well-defined pigmented lesion along the inferior part of the foveola. (c) Vertical scans of swept-source optical coherence tomography showing a hyperreflective lesion just inferior to the umbo. (d) Horizontal scans passing through the foveola showing a normal foveal dip in the absence of any retinal anatomical distortion. (e) Optical coherence tomography angiography showing a mildly distorted foveal avascular zone with a normal superficial retinal plexus anatomy

Discussion

Gass described three patterns of retinal involvement in congenital simple retinal pigment epithelial (RPE) hamartoma, (1) superficial retinal involvement, (2) full thickness retinal involvement with a preretinal extension, or (3) full thickness retinal involvement with a preretinal extension having intrinsic vascularization.[1] Similarly, shields elaborated five such cases focusing mainly on the dimensions of the tumor, distance from the foveola, and the disc and associated retinal changes. They described all the cases as involving the full thickness of retina however OCT features were not described.[2] The noted superficial retinal changes include dilated feeder retinal vessel, surrounding retinal traction, pigmented vitreous cells, and retinal exudations.[2]

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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.

Conclusion

OCT-A is a better noninvasive tool in the evaluation of changes along the retinal vascular plexus in cases of RPE hamartoma.

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Nil.

Conflicts of interest

There are no conflicts of interest.

References

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2. Shields CL, Shields JA, Marr BP, Sperber DE, Gass JD. Congenital simple hamartoma of the retinal pigment epithelium: A study of five cases. Ophthalmology 2003;110:1005-11.