Multimodal imaging in circumpapillary congenital hypertrophy of retinal pigment epithelium

Mahesh Shanmugam, Devashish Dubey, Rajesh Ramanjulu, Divyansh Mishra

Key words: OCT angiography, peripapillary congenital hypertrophy of retinal pigment epithelium, peripapillary pigmentary lesion

A 34-year-old gentleman came to the clinic for a regular check-up. His best-corrected visual acuity was 6/6, N6 in both eyes. Anterior segment examination was unremarkable. On fundus evaluation, the right eye showed a flat, darkly pigmented, circumpapillary lesion with distinct margins and lacunae, without any alteration in the vascular architecture [Fig. 1a]. The left eye fundus was within normal limits. Autofluorecence imaging of the right eye revealed a uniformly hypo-autofluorescent well-demarcated lesion [Fig. 1b]. SS-OCT (Swept source optical coherence tomography) scan showed thickened irregular retinal pigment epithelium (RPE) along with loss of outer retinal layers and a large sub-retinal cleft [Fig. 1c]. OCT-angiography at the level of the choroidal vessels showed vascular attenuation over the lesion along with signal void areas [Fig. 2].

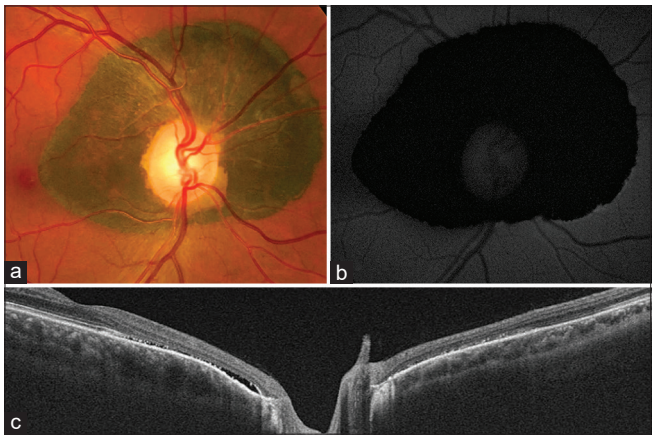

Figure 1: Fundus picture showing the flat, pigmented, peripapillary lesion with lacunae (a). Autofluorescence imaging showing a uniformly hypoautofluorescent well-demarcated lesion (b). SS-OCT scan showing the presence of thickened irregular RPE along with loss of outer retinal layers and a large sub-retinal cleft in the circumpapillary region (c)

Access this article online	
Quick Response Code:	Website: www.ijo.in
	DOI: 10.4103/ijo.IJO_196_20

Department of Vitreoretina, Sankara Eye Hospital, Bengaluru, Karnataka, India

Correspondence to: Dr. Devashish Dubey, Department of Vitreoretina and Ocular Oncology, Sankara Eye Hospital, Kundalahalli, Bengaluru - 560 037, Karnataka, India. E-mail: devashishdubey17@gmail.com

Received: 29-Jan-2020 Revision: 15-May-2020
Accepted: 21-May-2020 Published: 23-Sep-2020

Discussion

Congenital hypertrophy of retinal pigment epithelium (CHRPE) is a benign retinal lesion with an indolent course usually located in the equatorial region.[1] The term was coined in 1975 by Buettner.[1] It has a prevalence of 1.2% in the normal population.[2] Solitary peripapillary CHRPE is an extremely rare entity and has been reported to comprise <1% cases of CHRPE.[3] Unilateral solitary CHRPE is not associated with any other ocular or extraocular findings.[4] CHRPE usually does not affect the vision unless it involves the fovea or a choroidal neovascular membrane develops at its margin. OCT-A features of peripheral CHRPE have been described recently, it is a useful noninvasive modality to assess the vascularity of CHRPE as it can distinctly image the choroidal vascularity.[5]

Combined hamartoma of retina and retinal pigment epithelium (CHRRPE) and choroidal nevus are main differentials for such a lesion that may have more drastic visual consequences requiring close monitoring and treatment. In this photo essay, we highlight the different imaging signatures of this rare lesion, which will help to correctly diagnose and differentiate it from other circumpapillary pigmentary lesions.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have

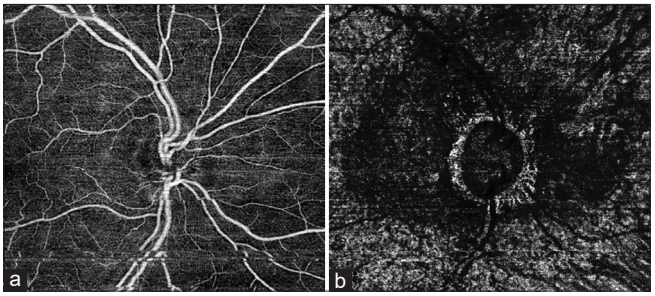

Figure 2: OCT-A superficial slab showing normal vascularity (a). OCTA through the level of choroidal vessels after manual segmentation showing vascular attenuation and signal void areas corresponding to the area of CHRPE (b). Projection artifact not affecting the image details were noted

Cite this article as: Shanmugam M, Dubey D, Ramanjulu R, Mishra D. Multimodal imaging in circumpapillary congenital hypertrophy of retinal pigment epithelium. Indian J Ophthalmol 2020;68:2232-3.

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.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Buettner H. Congenital hypertrophy of the retinal pigment epithelium. Am J Ophthalmol 1975;79:177-89.
2. Coleman P, Barnard NA. Congenital hypertrophy of the retinal pigment epithelium: Prevalence and ocular features in the optometric population. Ophthalmic Physiol Opt 2007;27:547-55.
3. Shields CL, Mashayekhi A, Ho T, Cater J, Shields JA. Solitary congenital hypertrophy of the retinal pigment epithelium: Clinical features and frequency of enlargement in 330 patients. Ophthalmology 2003;110:1968-76.
4. Shields JA, Shields CL, Shah PG, Pastore DJ, Imperiale SM Jr. Lack of association among typical congenital hypertrophy of the retinal pigment epithelium, adenomatous polyposis, and Gardner syndrome. Ophthalmology 1992;99:1709-13.
5. Shanmugam PM, Konana VK, Ramanjulu R, Mishra KD, Sagar P, Simakurthy S. Ocular coherence tomography angiography features of congenital hypertrophy of retinal pigment epithelium. Indian J Ophthalmol 2019;67:563-6.