Optical coherence tomography and angiography features of pigmented ocular fundus lesions in hereditary colonic polyposis syndrome

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Key words: Hereditary Colonic Polyposis Syndrome, optical coherence tomography angiography, pigmented ocular fundus lesions

A 32-year-old gentleman with family history of colonic polyposis was detected to have multiple slate gray-pigmented, oval-shaped, flat lesions on fundus examination. Colonoscopy showed numerous polyps more than100 in number, suggestive of colonic polyposis syndrome.

Three lesions were seen in the right eye and two were seen in the left eye [Fig. 1a and b]. Swept-source optical coherence tomography (OCT) showed attenuation of outer retinal layers and thickening and hyperreflectivity of retinal pigment epithelium (RPE). The superonasal lesion in the right eye showed intraretinal hyperreflective foci [Fig. 1c and d]. En face OCT of the lesion at the outer retinal slab showed hyperreflectivity [Fig. 2a and d] On OCT angiography (OCTA), the vascular density was reduced in the superficial capillary plexus (SCP) and increased in the deep capillary plexus (DCP) [Fig. 2b, c, e, f].

Discussion

Pigmented ocular fundus lesions (POFL) in familial adenomatosis polyposis (FAP) were initially described as multiple congenital hypertrophy of RPE (CHRPE).[1] But it is recognized that these lesions are distinct from solitary CHRPE.[2] Solitary CHRPE shows RPE hypertrophy, whereas POFL shows RPE hyperplasia and hamartomatous changes.[3] Tzu *et al*. described that the intraretinal RPE migration on OCT is unique to POFL.[4] Intraretinal hyperreflective deposits seen in our case could represent intraretinal RPE migration.

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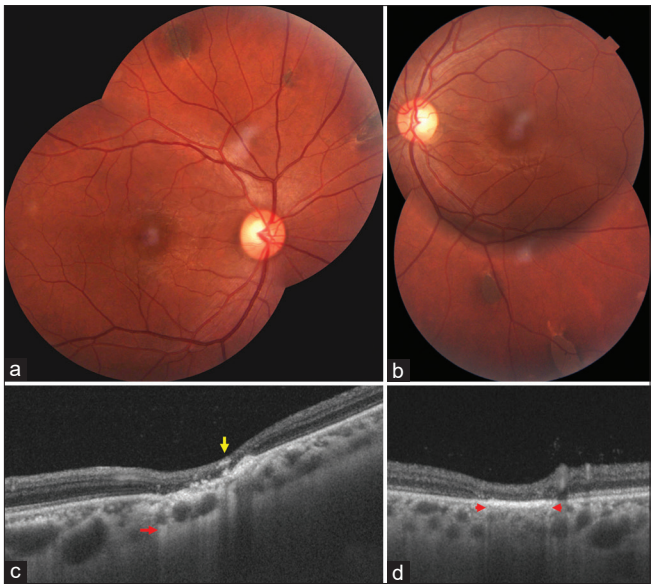

Figure 1: (a) Color fundus photo showing multiple pigmented oval flat lesions with well-defined borders with marginal halo in the right eye. (b) Color fundus photo showing multiple pigmented oval flat lesions with tailing in the left eye. (c) OCT showing intraretinal hyperreflectivity possibly due to pigment migration (yellow arrow). Hyper-transmission is seen due to loss of RPE at the site of lacunae (red arrow). (d) OCT showing hyperreflective thickened RPE (between red arrows) with loss of outer retinal layers. OCT = optical coherence tomography, RPE = retinal pigment epithelium

The reduced vascular density in SCP on OCTA would be a true reduction in vascularity, but the increase in DCP would be an artifact. In slab subtraction-based projection artifact removal (PAR), the SCP is subtracted from DCP to generate final DCP. Due to the lesser vascular density in the SCP in this lesion, the vascular density in DCP is probably overestimated after PAR. OCTA findings of POFL are not described in literature. But in a study of OCTA in solitary CHRPE, the retinal vasculature was noted to be normal.[5] This report highlights the reduction in vascular density in SCP on OCTA in POFL.

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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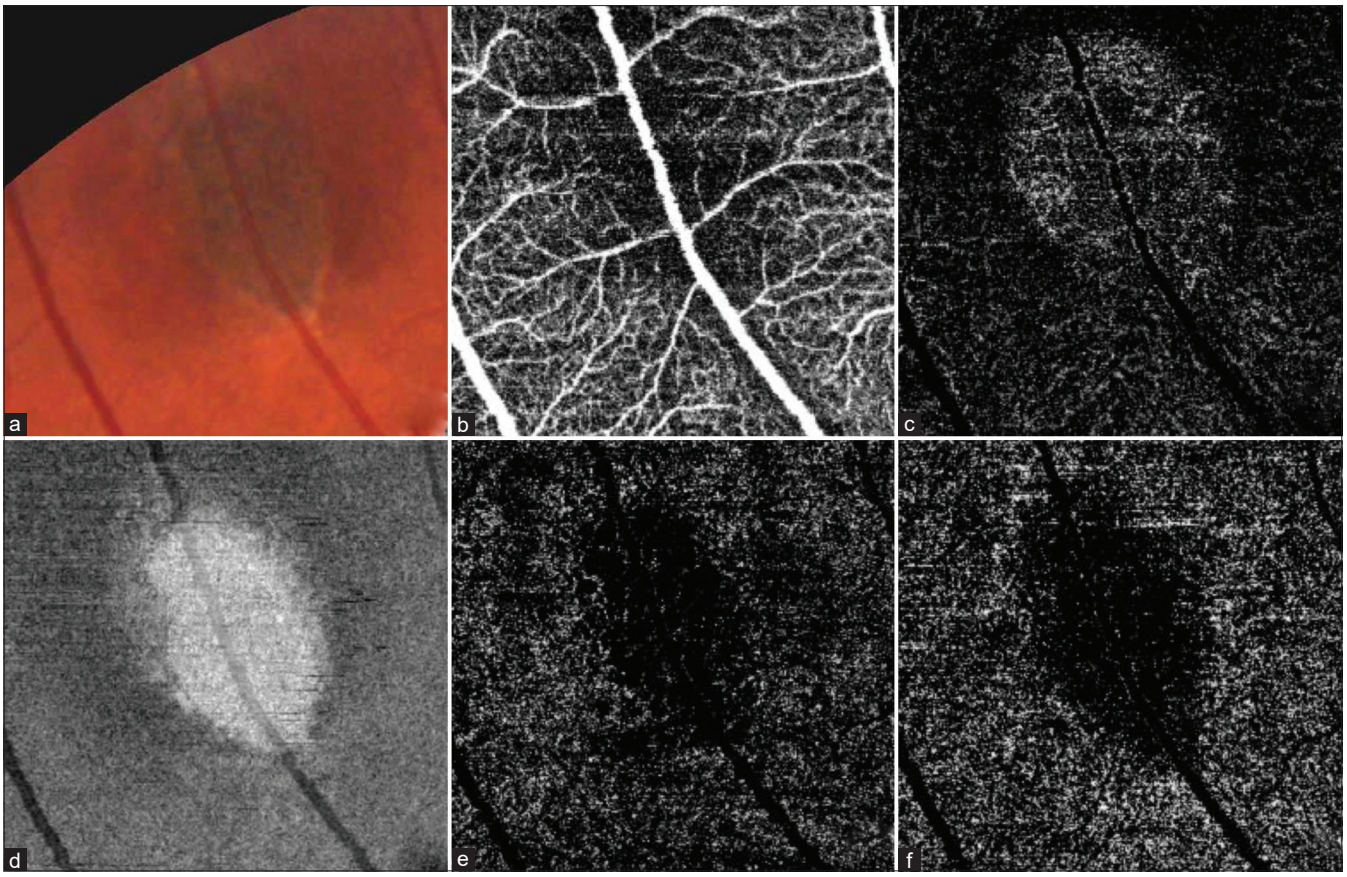

Figure 2: (a) Enlarged view of the right eye superotemporal lesion showing pigmented lesion with marginal halo. (b) OCTA of superficial capillary plexus slab showing decreased vascularity at the site of lesion. (c) OCTA of deep capillary plexus showing increase in vascularity. (d) En face OCT of outer retina showing hyperreflectivity. (e) OCTA of outer retina showing flow void area at the lesion site. (f) OCTA of choriocapillaries showing flow void area at the lesion site due to hypo-transmission. OCTA = optical coherence tomography angiography

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Conflicts of interest

There are no conflicts of interest.

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