Photo Essay

An unusual case of congenital hypertrophy of retinal pigment epithelium with overlying hemorrhages

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Key words: Congenital hypertrophy of retinal pigment epithelium, intraocular tumors, pigmented lesions, retinal pigment epithelium, retinal tumors, retinal vein occlusion

A 24-year-old asymptomatic female was referred to the vitreoretina services in view of the presence of a pigmented lesion in the right eye. Fundus examination showed the presence of a solitary, hyperpigmented flat lesion with scalloped margins in the inferotemporal quadrant suggestive of congenital hypertrophy of retinal pigment epithelium (CHRPE). Scattered retinal hemorrhages were evident over the lesion [Fig. 1]. Fundus fluorescein angiography (FFA) showed blocked fluorescence due to pigment epithelial hypertrophy. A small retinal vein within the lesion showed segmented filling suggestive of sluggish flow. Areas of capillary dropout with leakage from smaller venules adjacent to the vein described above were seen [Fig. 2]. Spectral domain optical coherence tomography (SD-OCT) of the lesion showed thickened hyper-reflective retinal pigment epithelium (RPE) with shadowing, attenuation of the overlying retinal layers, abruptly terminating outer layers at the edge of the lesion and irregular focal hyper-reflective areas in the inner retinal layers corresponding to intraretinal hemorrhages [Fig. 3]. The above features suggested CHRPE with small vein occlusion.

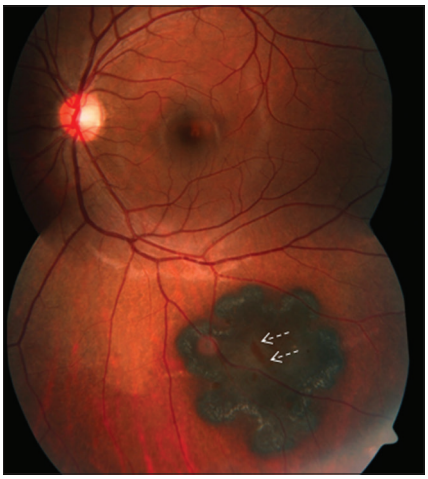

Figure 1: Fundus color image showing a solitary congenital hypertrophy of retinal pigment epithelium lesion with overlying retinal hemorrhages (dashed arrow)

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Discussion

Retinal vascular abnormalities overlying CHRPE have been documented in literature.[1,2] The most common vascular abnormality noted in CHRPE is capillary nonperfusion,

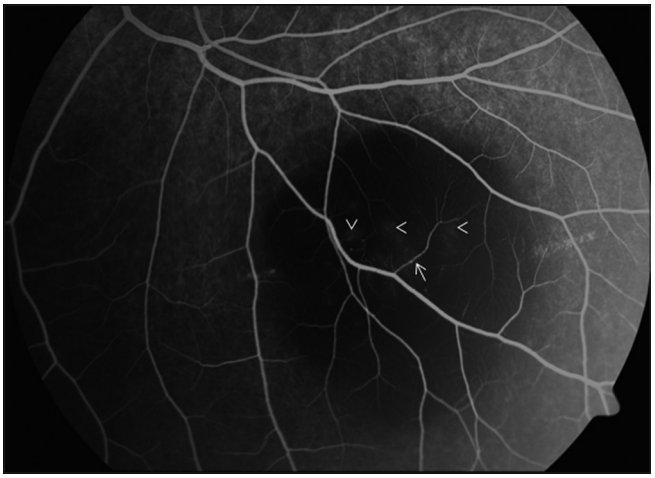

Figure 2: Fundus fluorescein image of the congenital hypertrophy of retinal pigment epithelium lesion showing blocked fluorescence from pigment epithelial hypertrophy, segmented filling within a small retinal vein suggestive of sluggish flow (arrow) and areas of capillary dropout with leakage from smaller venules (arrowheads)

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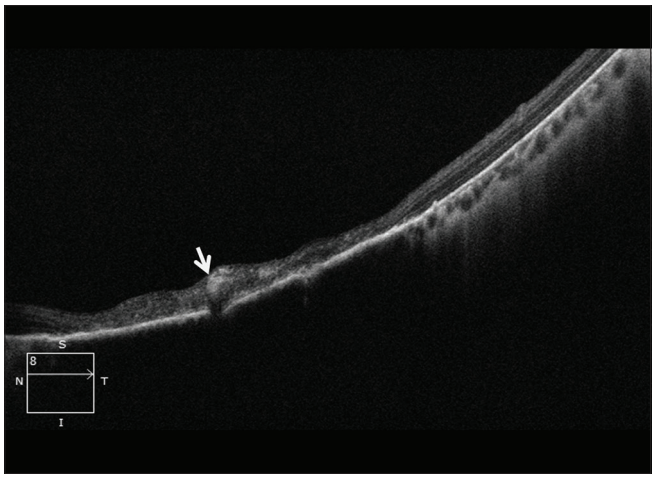

Figure 3: Spectral domain optical coherence tomography showing thickened hyper-reflective retinal pigment epithelium with shadowing, attenuation of the overlying retinal layers with abruptly terminating outer layers at the edges of the lesion and irregular focal hyper-reflective areas in the inner retinal layers corresponding to the intraretinal hemorrhages (thick arrow)

which was seen in our case. Other vascular abnormalities include microaneurysms, chorioretinal anastomosis, leak from abnormal capillaries, and focal attenuation of vessels crossing the lesion leading to partial vascular blockage.[1,2] Intraretinal hemorrhages have not been documented as a spectrum of vascular abnormalities in patients with CHRPE but were a predominant feature in our case supported by SD-OCT. Hemorrhages within the area of CHRPE can also be seen as a feature of choroidal neovascular membrane (CNVM) complicating CHRPE or in adenocarcinoma arising from CHRPE.[3,4] Our case did not show evidence of CNVM on FFA.[3] There was also no evidence of any nodular mass arising within the lesion excluding adenocarcinoma.[4]

SD-OCT showed attenuation of retinal layers with the loss of retinal architecture and thickened RPE similar to the previous observations.[5] However, choroidal structures were not visible in SD-OCT due to shadowing by hypertrophic RPE as the axial scan was captured through the pigmented area and not the lacunae. Focal hyper-reflective intraretinal lesions corresponding to hemorrhages in fundus photograph were visible. Careful clinical examination along with SD-OCT and FFA identified the benign nature of the lesion in our case.

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Conflicts of interest

There are no conflicts of interest.

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