Validating the pachychoroid disease spectrum using multimodal imaging

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Pachychoroid disease spectrum has garnered considerable interest and has been discussed in detail in recent literature. It refers to a group of retinal and choroidal disorders, namely pigment epitheliopathy, central serous chorioretinopathy, neovasculopathy, and polypoid choroidal vasculopathy, all arising from a thickened choroid and hyperpermeable large choroidal vessels. We describe a case which had simultaneous presentation of multiple disorders on the pachychoroid spectrum. Multimodal findings in this patient have been described in this report. The presence of pachychoroid should prompt thorough imaging since coexistence of multiple disorders can potentially change the management and follow-up schedule of these patients.

Key words: Central serous chorioretinopathy, optical coherence tomography angiography, pachychoroid, pachychoroid pigment epitheliopathy, polypoidal choroidal vasculopathy

Pachychoroid spectrum is being increasingly diagnosed, with its characteristic features being elucidated on spectral domain optical coherence tomography (SD-OCT), indocyanine green angiography (ICGA), and optical coherence tomography angiography (OCTA). We present a case that had simultaneous presentation of polypoidal choroidal vasculopathy, central serous chorioretinopathy (CSC), and pigment epitheliopathy, thus validating the pachychoroid spectrum of diseases.

Case Report

A 37-year-old male patient presented to us with complaints of decreased vision in the right eye (OD) for 1 week. Vision in the OD was 20/60 N36 and the left eye (OS) was 20/20 N6. Anterior segment evaluation was unremarkable. OD fundus revealed a large submacular hemorrhage with a diameter of 4 disc areas, with drusen superotemporal to the fovea [Fig. 1 top]. OS showed few drusen inferotemporal to the fovea [Fig. 1 bottom right].

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SD-OCT of the OD showed subretinal hemorrhage with underlying notched pigment epithelial detachment (PED) with a "double-layer sign" [Fig. 2 top].[1] The OS showed small PEDs corresponding to drusen [Fig. 3 bottom right]. Pneumatic displacement was done in the OD using 0.3 cc of C3F8 (100%) gas, following which the submacular hemorrhage was displaced [Fig. 1 bottom left]. One week later, there was a persistent PED with subretinal altered blood settled inferiorly [Fig. 1 bottom left]. At this point, fundus fluorescein angiography (FFA) and ICGA were carried out. FFA in the OD showed blocked fluorescence corresponding to the thin film of submacular hemorrhage and inkblot leaks of CSC temporal to the blocked fluorescence [Fig. 4 bottom left - inset]. ICGA in the OD revealed hypofluorescence in the area of PED associated with branched vascular network of around 2 disc areas nasal and superior to the PED along with two discrete polyps. There were dilated large choroidal vessels around the area of inkblot leaks and near the superior part of branch vascular network [Fig. 4 bottom right]. Branching vascular network with polyps was also demonstrable on OCTA in the OD [Fig. 4 top].

In the OS on FFA, there was pooling due to PED, and ICGA showed choroidal hyperpermeability, large choroidal vessels, and a quiescent polyp inferotemporal to the fovea [Fig. 3 bottom left]. Enface OCTA in the OS showed enlarged choroidal vessels focally (focal pachychoroid) underlying the area of the drusen (pachychoroid pigment epitheliopathy [PPE]) [Fig. 3 top]. Enhanced depth imaging-OCT revealed increased choroidal thickness of 550 microns in OD and 543 microns in the OS (focally corresponding to the area of abnormality seen on enface OCTA) with large outer choroidal vessels compressing the Sattler's layer and choriocapillaris [Fig. 3 bottom right].

A diagnosis of OD diffuse pachychoroid with polypoidal choroidal vasculopathy (PCV) and CSC was made. OS was diagnosed as a focal pachychoroid with PPE and PCV. Intravitreal aflibercept 2 mg/0.05 ml (Eylea, Regeneron Pharmaceuticals) was administered to the OD, following which there was resolution of subretinal hemorrhage and the height of the PED. The patient regained vision of 20/20, N6 in the OD at 3 months. He continues to be under 3-monthly follow-up and has maintained 20/20, N6 in the OS without any other complications due to pachychoroid.

Discussion

Pachychoroid describes a set of choroidal characteristics seen on SD-OCT as focal or diffuse choroidal thickening with larger outer choroidal vessels compressing the inner choroidal layers. On ICGA, large caliber vessels have been described that typically do not taper as they approach the posterior pole.[2]

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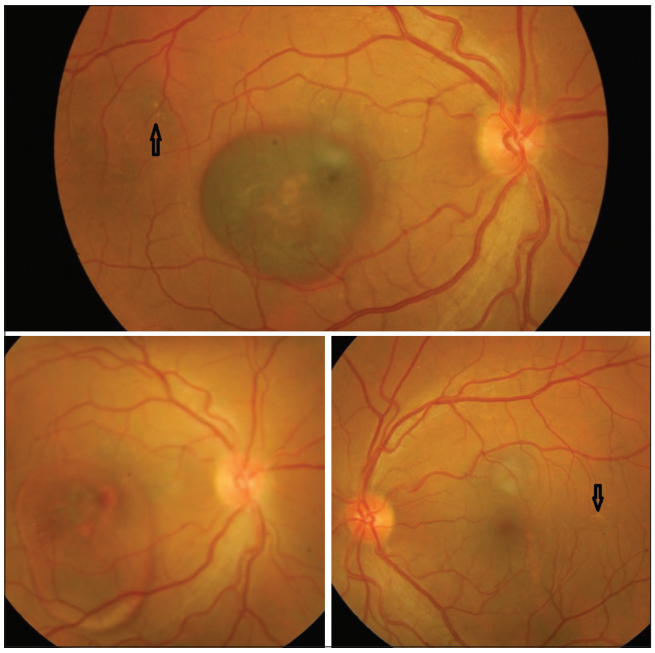

Figure 1: Top – Fundus photograph of the right eye at presentation showing a large submacular hemorrhage with drusen (black arrow). Bottom left – After pneumatic displacement of subretinal hemorrhage. Bottom right – pachydrusen seen temporal to fovea (black arrow)

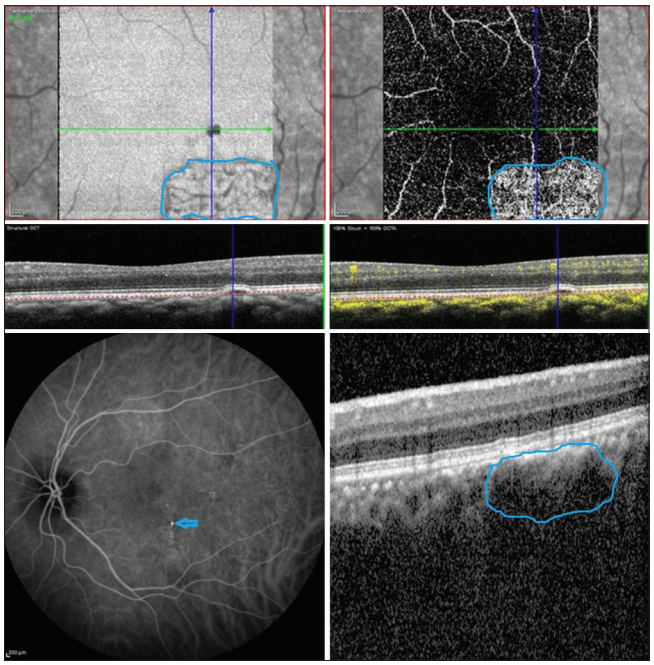

Figure 3: Top – Enface optical coherence tomography image showing large choroidal vessels in the choriocapillaris slab (blue outlined area). Bottom left – Indocyanine green angiography (4 min 55 s – showing enlarged choroidal vessels and a polyp (blue arrow)). Bottom right – Spectral domain optical coherence tomography image showing enlarged choroidal vessels compressing the choriocapillaris at the location corresponding to the enface optical coherence tomography image

Pachychoroid is generally bilateral and can be incidentally detected with or without the disease manifestations. Genetic inheritance of pachychoroid was suggested by Lehmann *et al*.[3]

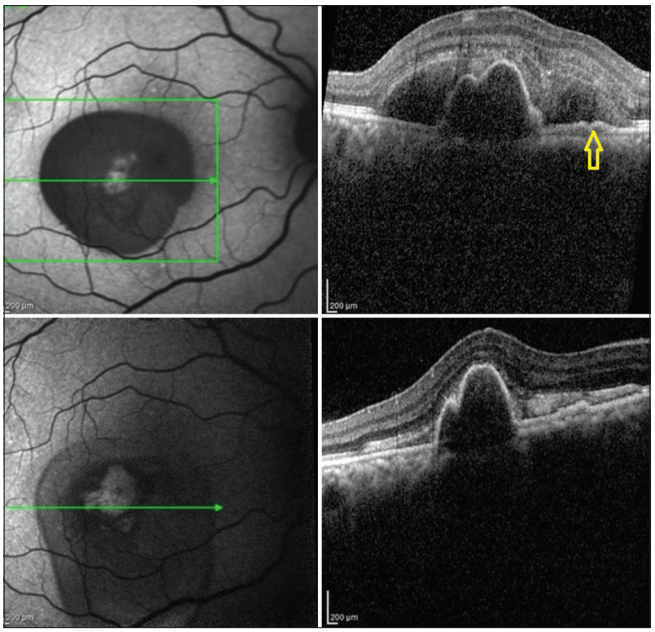

Figure 2: Top – Spectral domain optical coherence tomography of the right eye showing subretinal hyperreflectivity due to hemorrhage and an underlying notched pigment epithelial detachment. Double-layer sign noted (yellow arrow). Bottom – Postpneumatic displacement. Resolution of subretinal hemorrhage and persisting pigment epithelial detachment

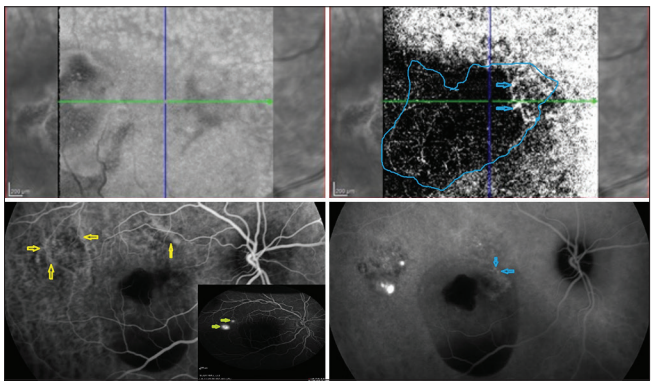

Figure 4: Top left – Enface optical coherence tomography image. Top right – A slab segmented between retinal pigment epithelium and Bruch's membrane delineating the branch vascular network (blue outlined area) and polyps (blue arrows). Bottom left – Indocyanine green angiography early frames (43 s) – showing enlarged choroidal vessels (yellow arrowheads). Inset shows fundus fluorescein angiography with inkblot leaks of central serous chorioretinopathy corresponding to the area of the enlarged choroidal vessels (green arrows). Bottom right – Late frames (11 min 45 s) showing branch vascular network and 2 discrete polyps (Blue arrows)

Dansingani *et al*. studied 66 eyes of 33 patients and showed that pachychoroid leads to a spectrum of choroidal diseases, namely PPE, chronic serous chorioretinopathy (CSCR), pachychoroid neovasculopathy (PNV), and PCV; usually in chorological order.[4]

In an eye with a hyperpermeable pachychoroid, PPE may result, if the retinal pigment epithelium (RPE) is able to overcome the choroidal congestion. If RPE fails to overcome

the stress, there might be choriocapillaris damage and CSC as a result of RPE damage. Microrips in the Bruch's membrane may be seen along with the ischemic changes, leading to neovascular network, i.e., PNV. PCV finally develops in cases of long-standing PNV where there is aneurysmal dilation of the ends of the neovascular network.[5] It is hypothesized that the choroidal hyperpermeability and vascular congestion lead to the development of PCV and CSCR.

Manayath *et al.* demonstrate simultaneous PCV and CSC in 6 eyes associated with a dilated choroidal vessel on ICGA and hypothesized that both PCV and CSC are offshoots of the pachychoroid spectrum.[5] The mean age in their series was 62.6 years. Our report however is of a much younger patient with bilateral simultaneous presentation of all the disease manifestations of pachychoroid except PNV. We also utilized the enface feature of OCTA and were able to demonstrate a focal abnormality in the choroidal slab in the OS. Branching vascular network with polyps was also demonstrable in the OD on OCTA in our case.

Conclusion

Our findings add to the existing scant literature about the presence of the entire spectrum of pachychoroid in a single patient. We recommend regular follow-up of such patients to look for future recurrences of CSC, PNV, and PCV. These patients need to be forewarned about symptoms arising from the pachychoroid disease spectrum.

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Conflicts of interest

There are no conflicts of interest.

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