Simultaneous multiple pachychoroid spectrum entities coexisting in the same eye

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Key words: Central serous chorioretinopathy, choroidal neovascular membrane, focal choroidal excavation, pachychoroid, peripapillary pachychoroid syndrome

A 45-year-old female presented with complaints of decreased vision in the right eye (OD) for the past 2 years with her vision in OD being 20/400. OD anterior segment was unremarkable and fundus showed the presence of a pigmented lesion superior to the fovea with a yellowish grey lesion at the nasal edge [Fig. 1a]. OD ocular coherence tomography (OCT) macula showed non-conforming focal choroidal excavation (FCE) with choroidal neovascular membrane (CNVM), foveal thinning with shallow sub-retinal fluid (SRF), schitic retinal edema temporally and pachychoroid [Fig. 1b-d]. OCT segmentation through the optic disc showed sub-retinal fluid nasal to the disc [Fig. 1e]. In OD, fundus fluorescein angiography (FFA) revealed point leak suggestive of central serous chorioretinopathy (CSCR) and Indocyanine green angiography (ICGA) revealed CNVM with increasing late phase leakage and pachy-vessels [Fig. 2]. A diagnosis of peripapillary pachychoroid syndrome (PPS) with FCE, CNVM, and CSCR was made in OD. The patient was treated with OD intravitreal injection of Bevacizumab along with FFA-guided focal laser to the point leaks [Fig. 3].

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Discussion

PPS is characterized by morphological pachychoroid features preferentially involving the peripapillary retina and nasal macula rather than the fovea along with overlapping findings of chronic CSCR, however they are differentiated by the preferential choroidal thickening in the nasal region in PPS, the presence of late disc leakage/staining and older age.[1] In our case multimodal imaging (MMI) showed the presence of nasal choroidal thickening with intra-retinal schitic fluid, peripapillary stippled fluorescence, extra-foveal leaks, and peripapillary dilated choroidal vessels (pachyvessels). In

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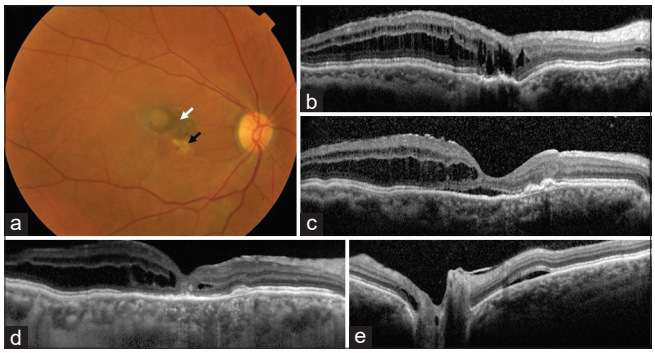

Figure 1: Baseline color fundus photo of the right eye showing a pigmented lesion superior to fovea along with a yellowish grey lesion at its nasal edge (a); OCT RE showing non-conforming FCE (b); foveal thinning with schitic retinal edema temporally (c); thick choroid (480 microns) (d); OCT segmentation through the disc showing outer retinal layer schisis temporal to disc and SRF nasal to it (e)

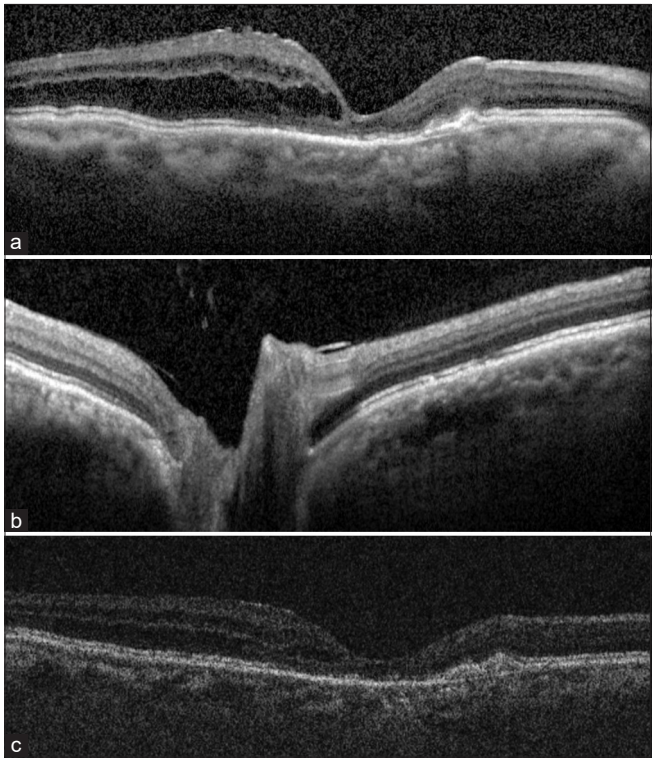

Figure 3: Repeat OCT at 3 months confirming complete SRF resolution under the fovea 3 months' post-bevazizumab (a); nasal resolution of SRF post focal laser (b); follow-up OCT at 1 year with foveal thinning and no recurrence of SRF (c)

addition, focal leaks of CSCR were identified on FFA in our case, which were treated with focal laser resulting in SRF resolution.

FCE, known to coexist along with CNVM, CSCR, and PCV, has recently been described as part of the pachychoroid disease spectrum (PDS).[2] In our case, OCT confirmed the presence of localized choroidal thinning with underlying hyper-reflective tissue suggestive of FCE. FCE-associated CNVM usually develops on the edge of excavation and shows favorable response to anti-VEGF injections[3] as was seen in our case.

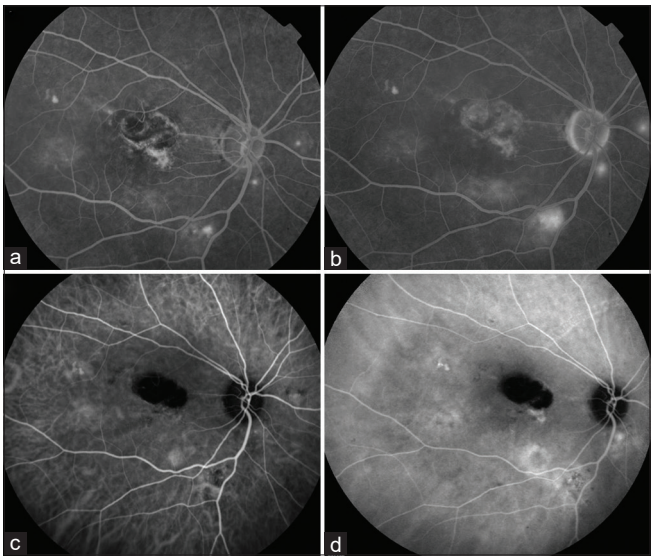

Figure 2: Early phase of FFA showing point leaks nasally suggestive of CSR (a); late phase of FFA showing leak increasing in intensity and size (b); early phase of ICG revealing CNVM (c); late phase hyper fluorescence seen on ICG (d)

As various manifestations of PDS result from the same disease process of choroidal congestion and thickening, these eyes are at high risk of developing different PDS entities at the same time or at different stages. Therefore, routine follow-up with MMI is a must in PDS patients due to heterogeneity. We report a previously unreported combination of various manifestations of PDS, namely PPS, a non-confirming FCE, acute CSCR leak, and type 1 CNVM, in the same eye highlighting their association with common disease process.

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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Nil.

Conflicts of interest

There are no conflicts of interest.

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