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An unusual presentation of peripapillary pachychoroid syndrome

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ABSTRACT

Purpose: Peripapillary pachychoroid syndrome (PPS) is a recently described entity of the pachychoroid disease spectrum and is characterized by thickening of the nasal choroid and peripapillary fluid pockets. This case illustrates the remarkable natural history of this recently described disorder.
Observation: This case report describes a patient with PPS who presented with severe cystoid macular edema (CME) that spontaneously resolved without treatment.
Conclusions and importance: This report indicates that PPS can exhibit significant fluctuation of CME and a favorable natural history in some cases. It also suggests that observation may be a viable option in the initial management of this finding if vision is not significantly unaffected.

1. Introduction

The pachychoroid disease spectrum (PDS) includes pachychoroid pigment epitheliopathy,[1] central serous chorioretinopathy (CSCR),[2] pachychoroid neovasculopathy,[3] polypoidal choroidal vasculopathy,[4] focal choroidal excavation,[5] and peripapillary pachychoroid syndrome (PPS).[6] PDS exhibit the following imaging characteristics: focal or diffuse choroidal thickening associated with reduced fundus tessellation, dilated Haller layer vessels or pachyvessels with thinning of the overlying inner choroid, and choroidal hyperpermeability with indocyanine green angiography (ICGA).[6] These eyes may also display disruption of the retinal pigment epithelium (RPE), choroidal folds and shorter axial lengths.

Of the PDS entities, PPS is the most recently described entity characterized by nasal choroidal thickening, peripapillary fluid pockets and peripapillary RPE disruption with only a few cases reported to date.[6–10] While cases of PDS can be complicated by vision loss due to cystoid macular edema (CME), the natural history and optimal management of this novel disorder are unknown. In this report, we describe an interesting case of PPS with spontaneously resolving CME. To the authors' knowledge, no similar case has yet been reported.

2. Case

An elderly septuagenarian patient with history of hypertension, obstructive sleep apnea, and uncomplicated bilateral cataract surgery (8 years prior) presented with decreased vision and blurriness in the left eye. Anterior segment examination was only remarkable for bilateral posterior chamber intraocular lens implants. There were no signs of inflammation or lens-iris chafing. Dilated funduscopic examination was remarkable for peripapillary RPE mottling in the left eye and oblique choroidal folds in the temporal macula of both eyes. Enhanced depth imaging spectral-domain optical coherence tomography (EDI-OCT) illustrated severe CME, nasal choroidal thickening and peripapillary fluid pockets in the left eye (Fig. 1a). Interestingly, the macula was flat and dry OS with no evidence of CME 8 months prior (Fig. 1b). The CME spontaneously resolved 3 months later without treatment (Fig. 1c). Fundus autofluorescence of the left eye taken after CME resolution showed peripapillary RPE mottling (Fig. 2). FA after resolution of CME displayed late staining of the disc and oblique choroidal folds, but was otherwise unremarkable (Fig. 3). Patient's right eye also showed increased choroidal thickness on OCT (Fig. 4). Patient's axial lengths were 23.55 mm in the right eye and 22.87 mm in the left eye.

3. Discussion

The case described in this report was associated with many of the typical features of PPS including nasal choroidal thickening, and peripapillary fluid pockets, choroidal folds, short axial lengths, and late staining of the optic disc on fluorescein angiography.[10] In addition, we

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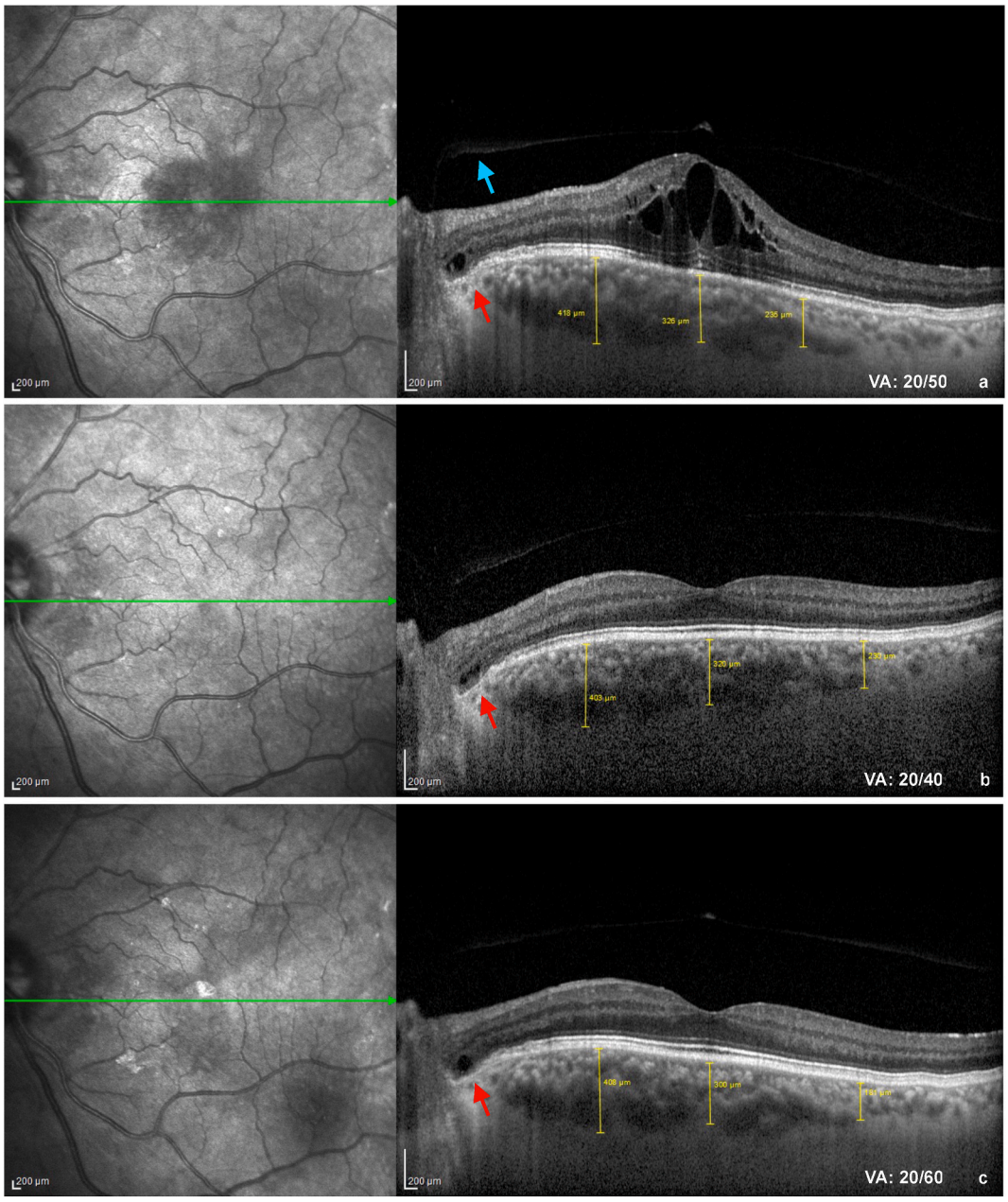

Fig. 1. EDI-OCT B scans of patient's left eye acquired at baseline presentation (a), at 8 months prior (b) and 3 months after baseline presentation (c). Note the presence of severe CME at the baseline visit (a) with no evidence of fluid before or after. Note the nasal choroidal thickening as well as the presence of peripapillary fluid pockets (red arrows). Measurements are shown at the foveal center and 1500 µm on either side. In normal eyes, choroidal thickness ranges from 142 to 194 µm, 196–363 µm, and 218–268 µm at the nasal, central, and temporal choroid, respectively.[13–16] Grade 3 posterior vitreous detachment is identified in all photographs (blue arrow) with no evidence of vitreomacular traction or epiretinal membrane formation. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

demonstrate self-resolving CME, non-contiguous with peripapillary fluid pockets, and thinning of the temporal choroid associated with thickening of the nasal choroid. We speculate that thickening in both PDS and PPS may be attributed to venous outflow congestion due to anomalous inter-vortex venous anastomoses. Whereas other PDS eyes demonstrate anastomoses in all four quadrants, PPS eyes may have a predilection for anastomoses in the inferonasal quadrant.[11,12] We speculate that as the nasal choroid becomes thicker and more congested, it may be diverting blood from the temporal choroid thus resulting in temporal choroidal thinning.

Intraretinal, and more uncommonly subretinal, fluid can complicate PDS.[1–3] However, in eyes with PPS, fluid pockets are usually located in a nasal or peripapillary distribution.[6,10] In our patient with CME not contiguous with peripapillary fluid pockets, we speculate that fluid leaked from thickened nasal choroid, entered the retina through a peripapillary external limiting membrane defect, and subsequently diffused to the central retina. All other causes of CME were excluded, including inflammatory, tractional, and vascular etiologies. OCT B scans before and after baseline presentation showed a grade 3 posterior vitreous detachment with no evidence of vitreomacular traction or epiretinal membrane formation. In addition, there were no signs of inflammation or retinal degeneration with only a very remote history of cataract extraction, and the patient denied use of any eyedrops or medications associated with CME. FA taken after resolution of CME failed to demonstrate any evidence of retinal vascular disease in either eye.

Interestingly, the CME in our case completely and spontaneously

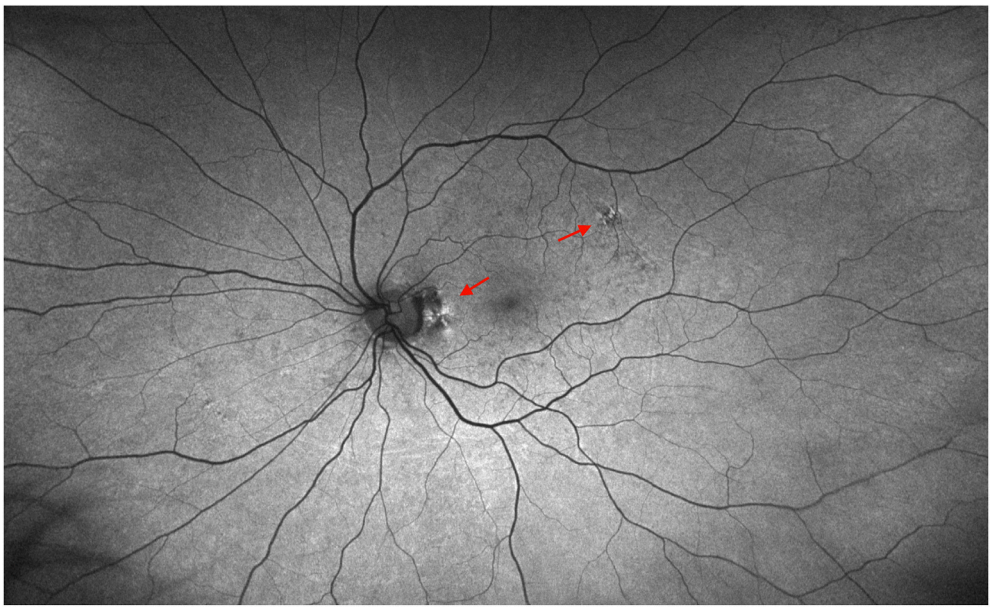

Fig. 2. Fundus autofluorescence image of patient's left eye after resolution of CME showed peripapillary and macula RPE disruption (red arrows). (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

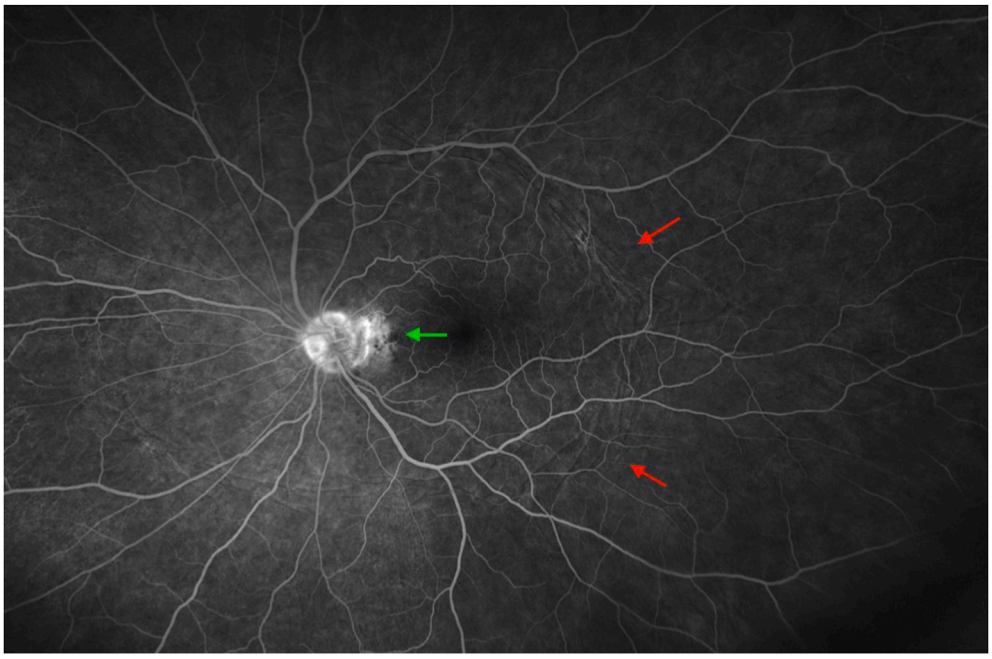

Fig. 3. Fluorescein angiography of patient's left eye after resolution of CME showed late staining of the disc and juxtapapillary region of RPE disruption (green arrow) and oblique choroidal folds (red arrows). (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

resolved at the 3 months follow-up visit without any treatment. In Phasukkijwatana et al.'s study of 31 eyes with PPS, macular fluid associated with peripapillary fluid pockets spontaneously resolved with observation in only one eye. The other eyes in that study exhibited variable responses to different treatment modalities such as intravitreal anti-VEGF injections, verteporfin photodynamic therapy, and topical dorzolamide.[6] Xu et al. recently described the outcomes of 35 patients (56 eyes) with PPS and a mean follow up of 27 months and noted that mean central and nasal retinal thickness significantly improved, although the majority of these eyes received some form of treatment such as anti-VEGF injection or photodynamic therapy.[10] A case report of PPS described fluctuating macular edema that did not correlate with anti-VEGF injections,[7] while another illustrated spontaneous resolution of subretinal and intraretinal fluid within one day.[9]

Peripapillary choroidal congestion associated with high hydrostatic pressure in the choroid may lead to fluid leakage in the retina in PPS. It is possible that degeneration of the peripapillary outer retinal layers may facilitate this process and provide a pathway into the retina from the hyperpermeable choroid.[6] Intraretinal and subretinal fluid in PPS may exhibit significant fluctuation, and even complete spontaneous resolution, due to systemic and local mechanisms and therefore observation may be a viable approach to treatment in the initial management of this

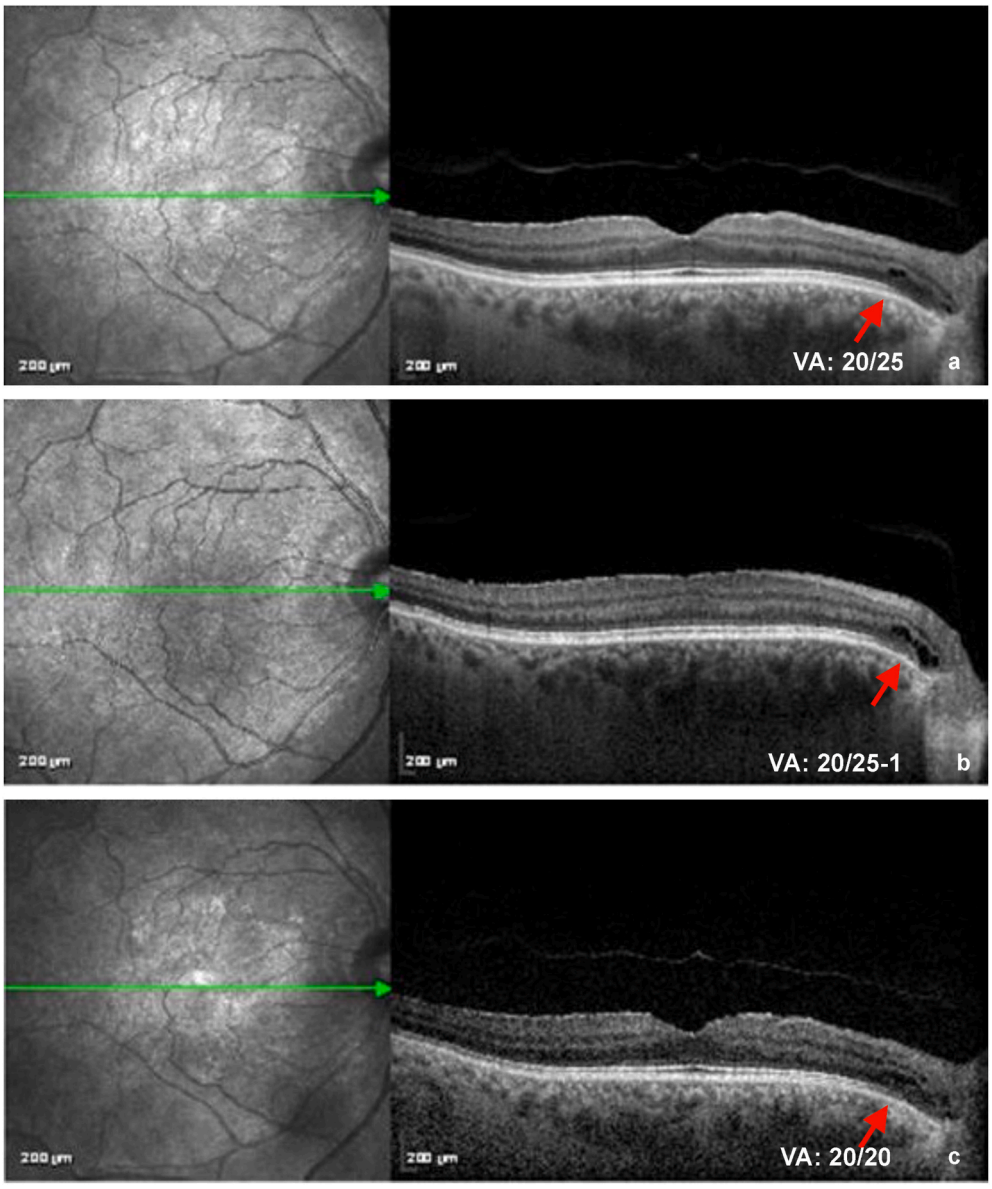

Fig. 4. EDI-OCT B scans of patient's right eye acquired at baseline presentation (a), 8 months prior (b), and 3 months after baseline presentation (c), showing peripapillary cysts and fluid pockets (red arrows). (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

disease.

4. Conclusion

There are no clear guidelines for the management of PPS but it is evident that fluid may significantly fluctuate and even resolve in this disease, and therefore observation may be an initially viable option even with central CME and subretinal fluid if there is no significant change in vision.

Patient consent

Consent was not obtained from the patient mentioned in the study as no identifying information was disclosed.

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Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

Declaration of competing interest

D. Sarraf is a consultant for Amgen, Bayer Healthcare, Genentech, Iveric Bio, Novartis, and Optovue and receives research support from Allergan, Boehringer Genentech, Heidelberg, Optovue, Regeneron and Topcon. The remaining authors have no conflicting interests to disclose.

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