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Serpentine retinal pigment epithelial tear

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Abstract:

We report a case of retinal pigment epithelial tear in a patient with peripheral exudative hemorrhagic chorioretinopathy (PEHCR). A 60-year-old diabetic female presented with left eye metamorphopsia. Fundus examination showed bilateral peripheral retinal pigment epithelium (RPE) degeneration, and a large serpentine-shaped RPE degeneration tract extending from the superotemporal arcade to the inferior periphery with associated subretinal hemorrhages in her left eye. This tract curved around the fovea, just sparing it. Fundus fluorescein and indocyanine green angiographies showed bilateral polyps in the superotemporal periphery. Optical coherence tomography through the tract showed scrolled up RPE at its edges with bare underlying Bruch's membrane and choroid in the region of the rip. There was no sign of an underlying pigment epithelial detachment. The patients with PEHCR should be prognosticated about such a rare vision-threatening macular complication.

Keywords:

Curvilinear, giant, peripheral exudative haemorrhagic chorioretinopathy, pigment epithelial detachment, retinal pigment epithelial tear, serpentine

Introduction

Retinal pigment epithelial tear or rip was first described in 1981 by Hoskin *et al.*^[1] It has been described as the tearing and retraction of the retinal pigment epithelium (RPE layer), exposing the underlying Bruch's membrane and choroid. This rare manifestation is most commonly seen in diseases associated with pigment epithelial detachment (PED) including exudative age-related macular degeneration (AMD) and polypoidal choroidal vasculopathy (PCV). Atypical tears have been described in pathologies not associated with PED or AMD.^[2]

Peripheral exudative hemorrhagic chorioretinopathy (PEHCR) has been defined as the presence of choroidal and retinal peripheral lesions associated with subretinal or sub-RPE hemorrhages with or

without subretinal exudation.^[3-5] Although it is diagnosed clinically based on careful fundus examination, multimodal imaging may be necessary to avoid misdiagnosis, especially from choroidal melanoma. B-scan ultrasound is the most important modality in differentiating PEHCR from choroidal melanoma. PEHCR is characterized by the presence of a dome- or plateau-shaped heterogeneous lesion with the absence of choroidal excavation and orbital shadowing.^[3-5] Indocyanine green angiography (ICGA) shows the presence of pachyvessels and peripheral polyps. Fundus fluorescein angiography (FFA) shows choroidal blockage due to the presence of subretinal/sub-RPE hemorrhage in addition to peripheral hyperfluorescence secondary to polyps and window defects due to RPE atrophy. Wide-field retinography and angiography allow complete evaluation of both the central as well as peripheral retina.^[3-5] Optical coherence tomography (OCT) may show signs of maculopathy. The various treatment modalities include laser

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photocoagulation and cryotherapy for the peripheral lesions, and intravitreal antivascular endothelial growth factor (VEGF) injections as well as photodynamic therapy for choroidal neovascularization. Rarely, surgical intervention may be necessary to treat massive submacular and/or vitreous hemorrhage.^[3-5]

We describe a case of a peculiar serpentine-shaped giant macular RPE tear secondary to PEHCR with preserved visual acuity in the absence of a pigment epithelial detachment (PED).

Case Report

A 60-year-old female presented with recent-onset metamorphopsia in her left eye. She was on treatment for diabetes mellitus (insulin and metformin), hypertension (telmisartan), dyslipidemia (atorvastatin), and coronary heart disease (aspirin). While her blood sugar was normal, her blood pressure was 200/100 mm Hg. Her bilateral best-corrected visual acuity was 20/20. Intraocular pressures, pupillary reactions, and anterior segment examination were normal in both the eyes. Fundus examination showed multiple microaneurysms and peripheral RPE degeneration in both the eyes; and a large serpentine-shaped RPE degeneration tract extending from the superotemporal arcade to the inferior periphery with associated subretinal hemorrhages in her left eye. This tract curved around the fovea, just sparing it [Figure 1a]. Fundus autofluorescence showed hypoautofluorescence corresponding to the tract with hyperautofluorescence along its edges and hypoautofluorescence corresponding to the subretinal hemorrhage [Figure 1b].

FFA and ICGA showed normal arm-to-retina and arteriovenous transit time; pachyvessels; blocked fluorescence secondary to subretinal hemorrhage; multiple microaneurysms; cluster of focal hypercyanoscent spots in superotemporal periphery suggestive of polyps in both the eyes; and peripheral window defects secondary to RPE atrophy. In addition, there was a RPE window defect corresponding to the curvilinear RPE tract in the left eye [Figure 2]. OCT through the tract showed

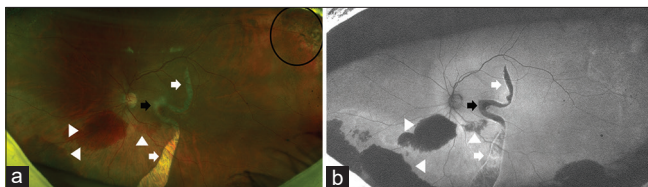


Figure 1: (a) Ultrawide-field fundus image showing a serpentine-shaped retinal pigment epithelial degeneration tract (white arrow), which curved around the fovea (black arrow) and extended from the supero-temporal arcade to the inferior periphery with associated subretinal haemorrhages (white arrow head) and peripheral retinal pigment epithelial degeneration (black circle); and (b) the corresponding ultrawide-field fundus autofluorescence

epiretinal membrane; scrolled up RPE at its edges causing back shadowing masking the underlying choroid; bare underlying Bruch's membrane and choroid in the region of the rip; subretinal fluid overlying the subfoveal RPE defect and pachyvessels [Figure 3]. The subfoveal choroid thickness was 300 μ m. There was no sign of an underlying PED. There was no evidence of choroidal neovascularization on OCT angiography. She was diagnosed to have a bilateral pachychoroid, PEHCR,

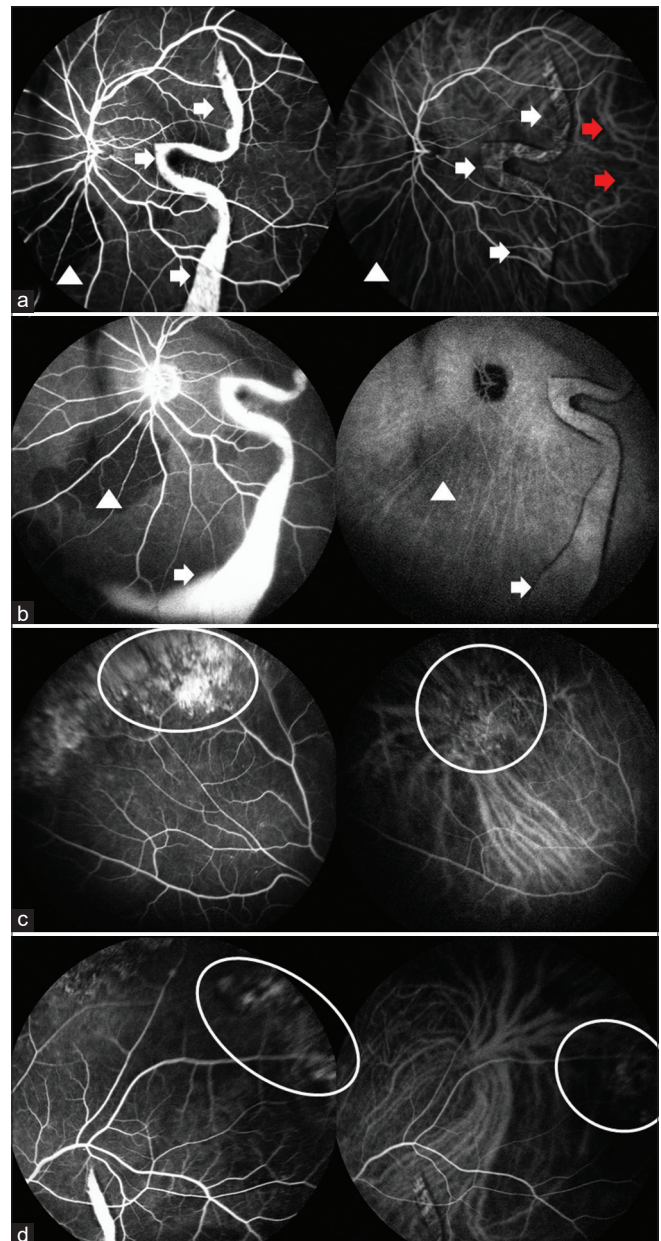


Figure 2: Combined fundus fluorescein angiography (right) and indocyanine green angiography (left) images of (a) left eye recirculation phase showing pachyvessels (red arrow), blocked fluorescence nasal to the disc (white arrow head), multiple microaneurysms and retinal pigment epithelial window defect curving around (sparing) the fovea (white arrow); (b) left eye late phase showing the extent of the retinal pigment epithelial tear (white arrow); (c) right eye superotemporal quadrant showing focal hypercyanoscent spots suggestive of cluster of polyps (white circle); and (d) left eye superotemporal quadrant showing cluster of polyps (white circle)

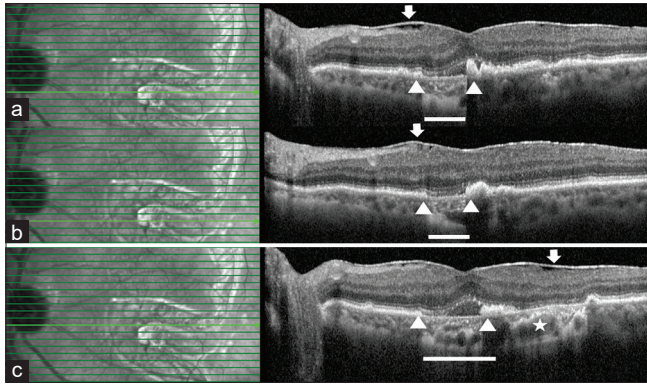


Figure 3: Optical coherence tomography line scans through the curvilinear tract in the macula at different levels (a-c) showing epiretinal membrane (white arrow); scrolled retinal pigment epithelium at the edges of the tract (white arrow head) leaving underlying Bruch's membrane and choroid bare (white line); and pachyvessel (star)

and mild nonproliferative diabetic retinopathy, with a giant RPE tear in the left eye.

She was advised a close follow-up and a strict metabolic control. However, she was lost to follow-up.

Discussion

RPE tear is most commonly associated with a vascularized PED, especially after anti-VEGF treatment.^[2] It can also be associated with serous PEDs, especially those with more than 400 μm height.^[6] Causes not associated with PED include choroidal neovascular membrane (CNVM), RPE thinning, choroidal swelling, vitreoretinal traction, and blunt trauma. These usually present with giant RPE tears.^[2]

The RPE rip is usually crescent or concentric shaped.^[2] However, the RPE rip in our patient had a very peculiar serpentine shape. Although no PED was noted, the presence of an earlier PED cannot be completely ruled out as a large hyperautofluorescent area was noted nasal to the RPE rip. However, the shape of the tear is unlikely to be caused by an adjacent PED. Giant RPE rip has also been reported in patients with hypertensive choroidopathy.^[7,8] The blood pressure of our patient was also high. However, unilateral presentation as well as the absence of other typical features such as retinal hemorrhages, hard exudates, disc edema, and exudative retinal detachment makes hypertensive choroidopathy in this patient unlikely.^[9] The rip in our patient was probably secondary to pachychoroid and PEHCR. The widespread subretinal hemorrhages, peripheral polyps, and surrounding atrophic RPE changes suggest the diagnosis of PEHCR. Mantel *et al.* proposed that atrophic changes in PEHCR occur as a sequela to the previous exudative changes, while widespread subretinal hemorrhages occur secondary to the RPE rupture.^[3] We believe that a complex interplay between the increased hydrostatic pressure (secondary to pachychoroid) and peripheral polyps may be the reason for the peculiar

serpentine shape of the RPE tear in our patient. Although the choroidal thickness was not very high, the presence of pachyvessels seen on OCT and ICGA can contribute to the increased hydrostatic pressure. However, the real cause of such a peculiar RPE rip in the absence of PED remains inconclusive.

PEHCR is an asymmetric bilateral pathology presenting with variable severity, typically affecting the older population. It has been postulated to be a peripheral variant of the PCV.^[10,11] As noted in our case, it is present more often in patients with hypertension and those treated with anticoagulants.^[5,12,13] Annesley reported anticoagulant therapy to be a risk factor for the extended involvement.^[12] The common macular complications include drusens, RPE alterations, macular edema, and epiretinal fibrosis. RPE rip has been rarely reported in a case of PEHCR.^[4,5] Kumar and Tewari reported a crescentic RPE rip temporal to the fovea.^[14] They believed the tear occurred secondary to a preexisting large serous PED. However, there was no evidence of a PED in our case.

The visual acuity of RPE tear depends on location of the tear. Tears passing through the macula are usually associated with poor vision.^[2] Although the giant RPE tear in our patient passed through the macula, it fortunately curved around the fovea just sparing it. Hence, she maintained a good visual acuity. Since the vision was preserved and there was no evidence of CNVM, no intervention was sought. Cases with RPE tears through the fovea but good visual acuity have been previously reported.^[15,16]

To the best of our knowledge, such a peculiar shaped RPE tear has never been reported. The patients with PEHCR should be prognosticated about such a rare vision-threatening macular complication.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Conflicts of interest

The authors declare that there are no conflicts of interests of this paper.

References

- Hoskin A, Bird AC, Sehmi K. Tears of detached retinal pigment epithelium. *Br J Ophthalmol* 1981;65:417-22.

2. Ersoz MG, Karacorlu M, Arf S, Sayman Muslubas I, Hocaoglu M. Retinal pigment epithelium tears: Classification, pathogenesis, predictors, and management. *Surv Ophthalmol* 2017;62:493-505.
3. Mantel I, Uffer S, Zografos L. Peripheral exudative hemorrhagic chorioretinopathy: A clinical, angiographic, and histologic study. *Am J Ophthalmol* 2009;148:932-8.e1.
4. Badawi AH, Semidey VA, Magliyah M, Al-Dhibi H. Updated systematic review and clinical spectrum of peripheral exudative hemorrhagic chorioretinopathy. *Middle East Afr J Ophthalmol* 2020;27:4-9.
5. Vandefonteyne S, Caujolle JP, Rosier L, Conrath J, Quentel G, Tadayoni R, *et al.* Diagnosis and treatment of peripheral exudative haemorrhagic chorioretinopathy. *Br J Ophthalmol* 2020;104:874-8.
6. Chan CK, Abraham P, Meyer CH, Kokame GT, Kaiser PK, Rauser ME, *et al.* Optical coherence tomography-measured pigment epithelial detachment height as a predictor for retinal pigment epithelial tears associated with intravitreal bevacizumab injections. *Retina* 2010;30:203-11.
7. Tai YC, Huang JC, Sun CC, Yeung L. Bilateral retinal pigment epithelial tears in hypertensive choroidopathy. *Taiwan J Ophthalmol* 2016;6:150-4.
8. Matsubara N, Kato A, Kominami A, Nozaki M, Yasukawa T, Yoshida M, *et al.* Bilateral giant retinal pigment epithelial tears in hypertensive choroidopathy. *Am J Ophthalmol Case Rep* 2019;15:100525.
9. Fraser-Bell S, Symes R, Vaze A. Hypertensive eye disease: A review. *Clin Exp Ophthalmol* 2017;45:45-53.
10. Goldman DR, Freund KB, McCannel CA, Sarraf D. Peripheral polypoidal choroidal vasculopathy as a cause of peripheral exudative hemorrhagic chorioretinopathy: A report of 10 eyes. *Retina* 2013;33:48-55.
11. Mantel I, Schalenbourg A, Zografos L. Peripheral exudative hemorrhagic chorioretinopathy: Polypoidal choroidal vasculopathy and hemodynamic modifications. *Am J Ophthalmol* 2012;153:910-22.e2.
12. Annesley WH Jr. Peripheral exudative hemorrhagic chorioretinopathy. *Trans Am Ophthalmol Soc* 1980;78:321-64.
13. Cebeci Z, Dere Y, Bayraktar Ş, Tuncer S, Kır N. Clinical features and course of patients with peripheral exudative hemorrhagic chorioretinopathy. *Turk J Ophthalmol* 2016;46:215-20.
14. Kumar V, Tewari R. Giant retinal pigment epithelium rip in a patient with peripheral exudative hemorrhagic chorioretinopathy. *Indian J Ophthalmol* 2019;67:1164-5.
15. Bressler NM, Finklestein D, Sunness JS, Maguire AM, Yarian D. Retinal pigment epithelial tears through the fovea with preservation of good visual acuity. *Arch Ophthalmol* 1990;108:1694-7.
16. Yoshitani S, Katsura M, Minamoto A, Tsumura K, Tamura H, Hasebe H, *et al.* Retinal pigment epithelial tear involving the fovea with preserved visual function. *Ophthalmic Surg Lasers Imaging* 2003;34:217-20.