Case Report

Spontaneous Bilateral Retinal Pigment Epithelium Rips with Good Visual Acuity

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Retinal pigment epithelium (RPE) rips commonly occur in retinochoroidal disorders including age-related macular degeneration, idiopathic polypoid alchoroidal vasculopathy, central serous chorioretinopathy, high myopia andchoroidal neovascularization. Most patients have unilateral involvement and poor visual prognosis.

A 55-year-old female presented with decreased vision in her right eye for one week. Her best-corrected visual acuity was 6/12 in the right eye and 6/6^{-1} in the left. Fundus examination revealed a large juxtafoveal RPE rip in the right eye and multiple small pigment epithelium detachments in the left. No abnormalhyperfluorescentlesions were detected by fundus angiography. High-doseoral antioxidant was prescribed. A pigment epithelium detachment (PED) in the left eye grew larger over the follow-up period. Ultimately, a RPE rip also occurred in the left eye in the 17th week of follow-up. Her best-corrected visual acuity was 6/9. Although reinvestigation was done, no other choroidal abnormalities were demonstrated by optical coherence tomography (OCT) and fundus angiography. During the observation, RPE tears were reattached spontaneously in both eyes. A considerable amount of RPE proliferation, migration, and repopulation was also demonstrated by OCT and fundus autofluorescence. After 2.5 years of follow-ups, her best-corrected visual acuity was 6/9 in the right eye and 6/6 in the left.We hypothesize that the increased surface tension of RPE is the etiology of RPE rips in this case. Furthermore, the underlying chorioretinal abnormality directly affects the visual prognosis and further studies are needed in prevention, pathogenesis and treatment.

Keywords: *Retinal pigment epithelium rip, Pigment epithelium detachment, Antioxidants, Bilateral*

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Spontaneous retinal pigment epithelium (RPE) tears have been associated with age-related macular degeneration (AMD), idiopathic polypoid alchoroid alvasculopathy (IPCV), central serous chorio retinopathy (CSCR), high myopia with posterior staphyloma and subretinal proliferative vitreo retinopathy[1-5]. The rips also have been developed following laser photo coagulation, laser capsulotomy, photodynamic therapy (PDT), intravitreal anti-vascular endothelial growth factor (anti-VEGF) injection, vitrectomy and blunt ocular trauma[6-14]. Most patients have unilateral involvement and tears often occur at the edge of RPE detachment. Additionally, most RPE rip patients have at least one chorioretinal abnormality before developing the RPE rip. We describe an atypical case of bilateral spontaneous RPE rips in a patient who did not have another chorioretinal disease.

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Case Report

A 55-year-old female presented with decreased vision in her right eye for one week. Her visual acuity (VA) was 6/18^{-2} with pinhole 6/12 in the right eye and 6/6^{-1} in the left. The anterior segment was normal. Fundus examination revealed a juxtafoveal RPE rip in the right eye and multiple, small, pigment epithelium detachments (PEDs) in the left. A sharp-edged pigment epithelial sheath—separated from the neurosensory retina without subretinal fluid—was demonstrated by optical coherence tomography (OCT) (Stratus OCT, Carl Zeiss Meditec, Inc., Dublin, CA, USA). Notwithstanding, no abnormal hyperfluorescent lesion was detected by fluoresceine and indocyanine green fundus angiography (FA) (HRA-2; Heidelberg Engineering, Dossenheim, Germany) (Fig. 1). Oral high-dose

antioxidant was prescribed for prevention of disease progression. One of the PEDs in the left eye grew larger during the follow-up period. Seventeen months later, an extrafoveal RPE rip also occurred in the left eye. VA of the affected eye was 6/12 with pinhole 6/9. Although reinvestigation was performed, no other choroidal abnormalities were demonstrated by OCT and FA. The patient preferred observation and entered a 3-monthly follow-up program. Over the observation period, RPE tears were reattached spontaneously in both eyes; despite not being on the same margin nor any choroidal neovascularizations (CNV) having occurred. Considerable tissue remodelling was also revealed by OCT and fundus autofluorescence (FAF)—including, RPE proliferation, migration, and repopulation at the edge of the lesion (Fig. 2, 3). The left eye remained symptom-free and the VA was 6/6 for at least 1 year. The patient, however, continues to have mild metamorphopsia in the right eye. After 2.5 years of follow-up, her best-corrected VA was 6/9 in the right eye and 6/6 in the left.

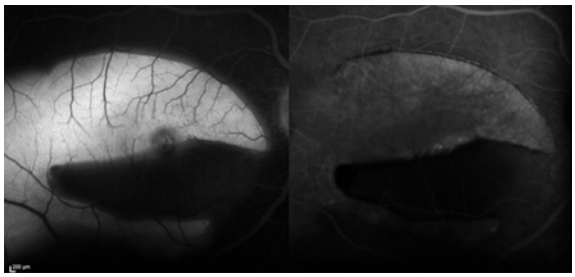

Fig. 1 No abnormal hyperfluorescent lesion was detected by fluoresceine and indocyanine green fundus angiography.

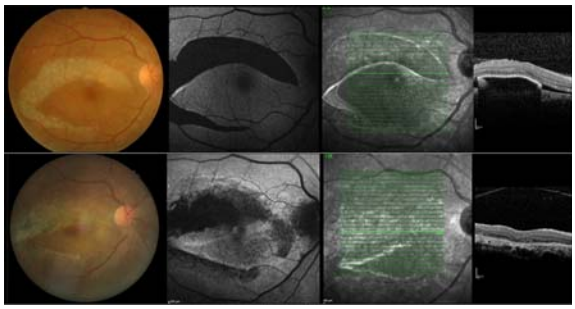

Fig. 2 Fundus photography, fundus autofluorescence and optical coherent tomography of the right eye at the initial clinical presentation were presented on the top row and the last follow-up on the bottom row.

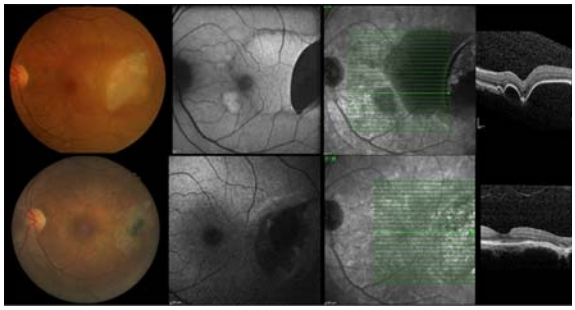

Fig. 3 Fundus photography, fundus autofluorescence and optical coherent tomography of the left eye at the initial clinical presentation were presented on the top row and the last follow-up on the bottom row.

Discussion

Tears in the RPE were first described in 1981 by Hoskins et al[15]. They are usually associated with progressive serous pigment epithelium detachments in AMD and polypoidal choroid alvasculopathy, or with intravitreal anti-VEGF injection[1-12,15]. Based on previous studies, the pathogenesis of RPE rip is not exactly known. Tears may be associated with increased hydrostatic pressure generated by damaged chorio-capillaris. Gass[16] countered that choroidal neovascularization directly separates the RPE from Bruch's membrane and contractile forces of the choroidal neovascular membrane tears the RPE. Two studies reported that by using angiographic and histologic examination, CNV was observed in the bed of RPE rips as well as at the site of the scrolled PRE[17,18]. Two otheraetiologies include traction from a proliferative vitreoretinopathy subretinal membrane or fibrin, and acute tangential force from blunt ocular trauma[5,14]. Since no chorioretinal lesions were found, we hypothesize that increased surface tension of RPE detachment is the etiology of RPE rips in this patient. Most patients with RPE rips have unilateral involvement. The visual outcome mainly depends on the underlying chorioretinal disease(s), size and location of the tear. Patients with RPE tears have a poor visual prognosis, even if the tear does not involve the subfoveal region[19,20], but some case reports document stable visual acuity (VA) after a RPE tear involving the fovea[21,22]. Most patients with a RPE rip have chorioretinal diseases (i.e., AMD and PCV), but after a full evaluation none was detected in our patient. The RPE rips we report here also had bilateral involvement; viz., a juxtafoveal RPE rip in the right eye and aextrafoveal RPE rip in the left. Lack of any chorioretinal

pathology and non-subfoveal involvement could explain why the patient continued to have good visual outcome.

A clinical trial on preventing RPE tears is difficult to achieve. Mones et al[23] suggested that a bimonthly half-dose ranibizumab—for large pigment epithelial detachment associated with retinal angiomatous proliferation—may be able to prevent RPE tears. We tried to prescribe oral high dose anti-oxidant for rip prevention, but the tears still occurred in the other eye. The treatment of RPE tears depends on the etiology and severity of the rip.

Assessment of the areal extent of RPE tears from fundus autofluorescence images is more accurate and reproducible than near-infrared reflectance images[24]. In a RPE tear, the pigment epithelial sheath separates from the neurosensory retina. Although photoreceptor cells are not able to function during this separation, they can survive up to 325 days after the tear[25]. This knowledge is essential for planning treatment such as macular translocation or autologous pigment epithelium and choroidal transplantation. After discussion with the patient, we decided to observe as no chorioretinal disease had been detected.

Advances in technology, FAF and spectral-domain OCT can demonstrate both the predictive signs and tissue remodelling[26-28]. Recovery of RPE correlates with restoration of retinal sensitivity in eyes with RPE tear[29]. We were able to observe RPE proliferation, migration, and repopulation at the edge of the lesion using FAF and OCT. Further research, however, are needed into prevention, pathogenesis and treatment of RPE rips.

Conclusion

We hypothesize that increased surface tension of RPE is the etiology of RPE rip in this case report. Consequently, the length of the lesion will increase with the degree of tightness. The underlying chorioretinal abnormality and rip location will also directly affect visual prognosis.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

Acknowledgement

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What is already known on this topic?

Spontaneous RPE tears have been associated with age-related macular degeneration (AMD), idiopathic polypoid alchoroidal vasculopathy (IPCV), central serous chorioretinopathy (CSCR), high myopia with posterior staphyloma and subretinal proliferative vitreoretinopathy. The rips also have been developed following laser photocoagulation, laser capsulotomy, photodynamic therapy (PDT), intravitreal anti-vascular endothelial growth factor (anti-VEGF) injection, vitrectomy and blunt ocular trauma. Additionally, most of the RPE rip patient has at least one of the chorioretinal abnormalities before developing RPE rip. Most patients with RPE rip usually had unilateral involvement. The visual outcome mainly depends on the underlying chorioretinal diseases, size and location of tear. Patients with RPE tears have a poor visual prognosis, even if the tear does not involve the subfoveal region.

What this study adds?

We describe an atypical case of bilateral spontaneous RPE rips in a patient who did not have anychorioretinal diseases and also have good stable visual acuity.

Potential conflicts of interests

None.

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รายงานผู้ป่วยชั้นเซลล์เม็ดสีจอตาฉีกขาดที่เกิดขึ้นเองในตาทั้งสองข้างร่วมกับมีระดับการมองเห็นที่ดี

สุธาสินี สีนะวัฒน์, ชวกิจ ภูมิบุญชู, ยศอนันต์ ยศไพบูลย์, สุพัชญ์ สีนะวัฒน์

ชั้นเซลล์เม็ดสีจอตาฉีกขาดมักเกิดในโรคของจอตาและคอรอยด์ ได้แก่ภาวะจุดภาพชัดเสื่อมในคนสูงอายุ ภาวะหลอดเลือดใต้จอตาโป่งพอง โรคจุดภาพชัดบวมน้ำ ภาวะสายตาสั้นมาก และภาวะหลอดเลือดคอรอยด์งอกผิดปกติ ผู้ป่วยส่วนใหญ่มักเป็นโรคนี้ในตาเพียงข้างเดียว และมีการพยากรณ์โรคด้านการมองเห็นที่ไม่ดี

ผู้ป่วยหญิงไทยอายุ 55 ปี มาพบแพทย์ด้วยอาการตาขวามัวลงเป็นระยะเวลา 1 สัปดาห์ ระดับการมองเห็นเป็น 6/12 ในตาขวา และ 6/6[1] ในตาซ้ายการตรวจจอตาพบชั้นเซลล์เม็ดสีจอตาฉีกขาดขนาดใหญ่ในบริเวณ juxtafovea ของตาขวาและ pigment epithelial detachments (PED) ขนาดเล็กเป็นจำนวนมากในตาซ้ายโดยไม่พบความผิดปกติแบบ hyperfluorescence จากการฉีดสีตรวจหลอดเลือดจอตาและคอรอยด์ ผู้ป่วยได้รับการรักษาด้วยการรับประทานสารต้านอนุมูลอิสระขนาดสูงแต่ PED อันหนึ่งในตาซ้ายมีขนาดใหญ่ขึ้นเรื่อย ๆ และเกิดชั้นเซลล์เม็ดสีจอตาฉีกขาดในอีก 17 สัปดาห์ถัดมา โดยมีระดับการมองเห็นเป็น 6/9 แต่ก็ไม่พบความผิดปกติอื่นเมื่อตรวจเพิ่มเติมด้วย optical coherence tomography (OCT) การฉีดสีตรวจหลอดเลือดจอตาและคอรอยด์ระหว่างการสังเกตอาการพบว่า ชั้นเซลล์เม็ดสีจอตาที่ฉีกขาดสามารถติดกลับได้เองในตาทั้งสองข้างการตรวจ OCT และ fundus autofluorescence พบมีทั้ง proliferation, migration และ repopulation ของชั้นเซลล์เม็ดสีจอตาภายหลังการตรวจติดตามผู้ป่วยเป็นระยะเวลา 2 ปีครึ่ง ผู้ป่วยมีระดับการมองเห็นเป็น 6/9 ในตาขวาและ 6/6 ในตาซ้าย คณะผู้นิพนธ์ตั้งสมมติฐานว่าชั้นเซลล์เม็ดสีจอตาที่ฉีกขาดในผู้ป่วยรายนี้เกิดจากแรงดึงผิวที่เพิ่มมากขึ้นของชั้นเซลล์เม็ดสีจอตาอย่างไรก็ตาม พยาธิสภาพของชั้นคอรอยด์และจอตามีผลโดยตรงต่อการพยากรณ์โรคด้านการมองเห็น การศึกษาเกี่ยวกับวิธีการป้องกัน พยาธิกำเนิดและการรักษาชั้นเซลล์เม็ดสีจอตาฉีกขาดต่อไปในอนาคตมีความจำเป็นยิ่ง