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CASE REPORT

A Case Report of Retinal Pigmented Epithelium rip

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ABSTRACT:

Tears of the retinal pigment epithelium (RPE) are defined as complication of pigment epithelial detachments (PED) in the setting of wet age-related macular degeneration (AMD). RPE rips, also known as RPE tears, have been described spontaneously in association with central serous retinopathy, angioid streaks, and trauma and as a result of various treatments for neovascular AMD. Clinically, a well-demarcated area of bare choroid is visible immediately adjacent to a hyperpigmented area, which represents the redundant, retracted RPE. The overlying neurosensory retina remains intact and a localized exudative detachment of the neurosensory retina may be present. In this study, we discuss a case of Retinal pigmented epithelium rip reported to the Department of Opthalmology, MGM hospital, Kamothe.

Key words: Macular degeneration, Pigment epithelial detachments, Retinal Pigmented Epithelium.

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INTRODUCTION:

Tears of the retinal pigment epithelium (RPE) are defined as complication of pigment epithelial detachments (PED) in the setting of wet age-related macular degeneration (AMD).[1,2] RPE rips, also known as RPE tears, have been described spontaneously in association with central serous retinopathy, angioid streaks, and trauma and as a result of various treatments for neovascular AMD. Clinically, a well-demarcated area of bare choroid is visible immediately adjacent to a hyperpigmented area, which represents the redundant, retracted RPE. The overlying neurosensory retina remains intact and a localized exudative detachment of the neurosensory retina may be present.[3, 4] Retinal pigment epithelial (RPE) tears can complicate various chorioretinal disorders, including traumatic chorioretinopathy, high myopia, angioid streaks, choroidal tumors, light chain deposition disease, central serous chorioretinopathy, and polypoidal choroidal vasculopathy (PCV).[5] The most common association of RPE tear by far is the neovascular form of age-related macular degeneration (AMD). Hoskin and colleagues were the first to identify RPE tears in patients with PED and neovascular AMD, and they described the clinical and angiographic findings, which included a window defect corresponding to the area of RPE loss and blockage in the region of the retracted RPE.[6] Interestingly, the fundus autofluorescence findings are the reverse with hypoautofluorescence in the region of lost RPE and hyperautofluorescence corresponding to the retracted RPE.[7]

Henceforth, we discuss a case of Retinal pigmented epithelium rip reported to the Department of Opthalmology, MGM Hospital, Kamothe.

CASE REPORT:

A 53 years old woman reported at the outpatient clinic of our department with the chief complaint of distorted vision is her right eye from 1 year. Her visual acuity was: L.E. (left eye) light perception and R.E. (right eye) 0.4. The right fundus showed a large serous detachment of the pigment epithelium located in a large area of hard and soft drusen scattered throughout the posterior pole. There was no evidence of choroidal neovascularization. The fundus picture remained stable for six months when the patient had a sudden onset of blurred vision in his R.E.: her visual acuity was reduced to counting fingers at 20 cm. On opthalmoscopy of right eye, a large retinal pigment epithelial rip with a retracted torn temporal edge was seen.

The tearing was more evident on fluorescein angiography (fig 1).

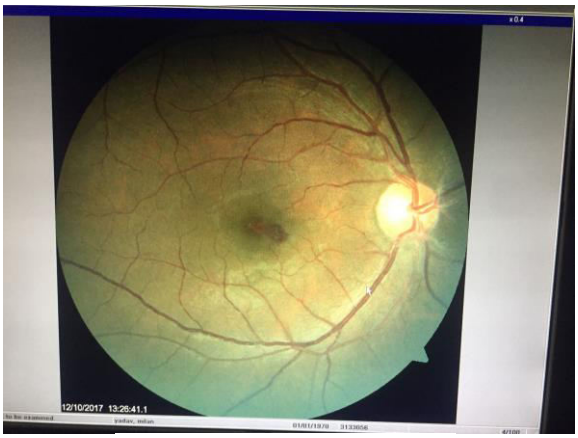

Figure 1: Photograph of Right eye fundus

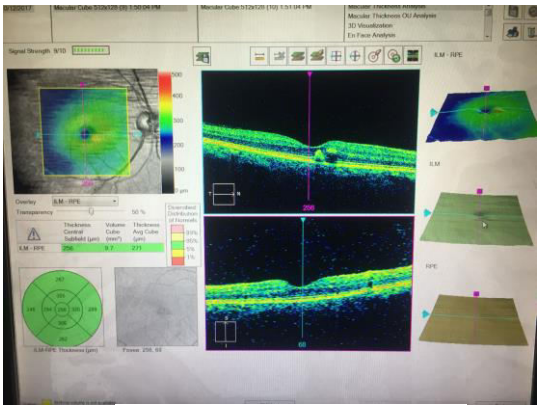

Figure 2: Right eye OCT showing RPE rip

The area of bare choroid was sharply outlined and showed intense early hyperfluorescence with larger choroidal vessels being easily detected and no late sub-retinal leakage of dye. The adjacent folded edge of RPE was intensely hypofluorescent and well demarcated. The remaining detachment showed uniform hyperfluorescence.

Further evaluation of RPE detachment was done with fluorescein angiography of the right eye. The flourescein angiography revealed that the hyperfluorescence was early and irregular on the temporal side of the detachment, while late hyperfluorescence was relatively uniform in the remaining area. At the posterior pole, hard and soft drusen were detected around the detached RPE. Larger than hard drusen, soft drusen fluoresced early and showed mild late staining. Present at the edge of the detachment, they spread temporally mixed with nodular drusen, over to the border of the wide drusen area at the posterior pole.

DISCUSSION:

Tears of the RPE may be spontaneous or following laser photocoagulation. They may be relatively frequent, occuring in about 10% of pigment epithelial detachments, according to Hoskin and co-workers. Misdiagnosis may have lowered actual figures of this condition until recently. In all cases reported, tearing of the RPE has always been shown to occur along the border of the serous detachment. This location led some authors to suggest a mechanical cause of the pigment epithelial rip.[8-10]

Battaglia Parodi M et al reported a case showing adult-onset foveomacular vitelliform dystrophy (AOFVD), associated with vascularized pigment epithelial detachment. A 72-year-old female affected by AOFVD complained with blurred vision and metamorphopsia in her right eye, seven months after a routine clinical examination. Visual acuity in right eye dropped from 0.6 to 0.3, and biomicroscopic fundus examination revealed a serous pigment epithelial detachment arising from the temporal margin of the pseudo vitelliform lesion. Fluorescein angiography showed an uneven filling of the pigment epithelial detachment, suggesting the presence of a subfoveal choroidal neovascularisation, which was confirmed by indocyanine green angiography. The authors concluded that the association between AOFVD and vascularized pigment epithelial detachment supports the hypothesis that AOFVD may be a different subgroup of age-related macular degeneration with specific genetic predisposition. Bottoni FG et al reported a case of a 71-year-old man presenting with a large disciform scar in the right eye and hard (nodular) as well as soft (granular) drusen scattered throughout the posterior pole of the left eye. A large serous pigment epithelial detachment was evident in the drusen area of the left fundus. Six months later a tear of the retinal pigment epithelium (RPE) occurred in the left eye. It was located exactly along the peripheral border of the drusen area instead of the border of the RPE detachment where it would have been expected. It is considered that reduced adhesion between the RPE and Bruch's membrane as well as the sharp change in tissue cohesiveness at the border of the drusen area and surrounding healthy tissue were relevant in the pathogenesis of this case.[11, 12]

Fryczkowski P et al presented a case of a spontaneous retinal pigment epithelial tear in high myopia. A 56 years old woman with high myopia (refractive error -10.0D) in both eyes and amblyopia in right eye since childhood, complained of a decrease in left eye vision for one and a half month. Full ophthalmic examination, fundus photography, ultrasound examination and indocyanine green angiography were performed. At first visit BCVA was 5/25 in her left eye. A subretinal hemorrhage accompanied by retinal pigment epithelial tear inferotemporal to the fovea was found in ocular examination. B-scan ultrasound did not revealed any traction at the fovea. 3 months later BCVA was 5/16 OS. The area of the atrophy of RPE--Bruch's membrane--choriocapillaris complex expanded and hyperpigmentation occured above the RPE tear. 3 months later BCVA was 2/50 OS, we observed a dark brown pigment spot surrounded with a subretinal hemorrhage somewhat above the RPE tear. Indocyanine green

angiography demonstrated an initial choroidal neovascularization at superotemporal edge of hypertrophic pigment epithelium (Fuch's spot). The authors concluded that a spontaneous RPE tear might be the beginning of Fuchs' spot. A mechanical stress associated with eye movement is transmitted through the thin sclera and Bruch's membrane onto RPE and may cause a tear in its most vulnerable place i.e. at the margin of posterior staphyloma. Sinawat S et al reported that 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 6/6-2 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 abnormal hyperfluorescent lesions were detected by fundus angiography. 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. The authors hypothesized 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.[13,14]

CONCLUSION:

This can be concluded that the Retinal pigmented epithelium is a common occurring clinical entity and should be efficiently diagnosed and further management to prevent further any deliberating effects.

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