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CASE REPORT

Accidentally detected unilateral peripapillary retinoschisis: A case presentation

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Abstract

This study aims to describe an atypical presentation of peripapillary retinoschisis (PPRS) in a young myopic patient. A 14-year-old female with high myopia −10.50 diopters in the right and −12.0 diopters in the left eye and good visual acuity (20/20) in both eyes. She presented with splitting of the inner retinal layers in the superior peripapillary quadrant as an incidental finding on spectral-domain optical coherence tomography (SD-OCT) on her left eye. The macula and outer retinal layers were unaffected and it was not associated with any other ocular pathology except myopia in both eyes. Our patient represents an atypical form of PPRS determined incidentally on SD-OCT with schisis of inner retinal layers without macular involvement.

Keywords: Myopia; optical coherence tomography; peripapillary retinoschisis; young patient.

Peripapillary retinoschisis (PPRS) is characterized by the abnormal splitting of the peripapillary retinal nerve fiber layer and frequently tends to be bilateral with asymmetrical involvement. Macular retinoschisis is mostly found together with PPRS and associated with X-linked retinoschisis,[1] stellate nonhereditary idiopathic foveomacular retinoschisis (SNIFR),[2,3] high myopia,[4] glaucoma,[5] vitreomacular traction syndrome,[6] and congenital optic disc abnormalities such as optic pit[7] and optic disc coloboma.[8] The underlying pathophysiology and the factors associated with PPRS have not been completely understood yet.

In this case report, multimodal imaging in a case with atypical presentation of unilateral PPRS without any sign of macular involvement was presented.

Case Report

A 14-year-old female admitted to our clinic for a routine eye examination without any complaint. Her medical history was unremarkable. She had bilateral high myopia (−10.50 D in OD and −12.0 D in OS). Her best-corrected visual acuities were 20/20 in both eyes. The axial lengths were 27.0 mm OD and 28.0 mm OS. Applanation tonometry revealed intraocular pressures of 13 mmHg OD and 14 mmHg OS. Her anterior segment examination was unremarkable in both eyes. There was no evidence of afferent pupillary defect. The color vision was normal in both eyes. Family history was negative for hereditary eye diseases. In dilated fundus examination, there was a slight elevation of the superotemporal peripapillary retina in the left eye

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(Fig. 1a) and myopic fundus appearance in the right eye. Fundus fluorescein angiography (Heidelberg retinal angiography 2, Spectralis®, Heidelberg Engineering, Heidelberg 2, Germany) did not reveal any sign of leakage in both eyes. In the fundus autofluorescence (Spectralis®, Heidelberg Engineering, Heidelberg 2, Germany), there was a slight hypofluorescence in the superotemporal area adjacent to the left optic disc (Fig.1b and c). The spectral-domain optical coherence tomography (SD-OCT; Spectralis Heidelberg Engineering, Heidelberg 2, Germany) scans demonstrated splitting of various layers of the inner retina in the superotemporal peripapillary region, primarily at the level of the nerve fiber layer, ganglion cell layer, and inner plexiform layer. There was no foveal involvement in the left eye, and the right eye was normal (Fig. 1d and e). The splitting in the left eye corresponded to the area of retinal thickening noted topographically on SD-OCT (Fig. 1f). The swept-source OCT angiography (SS-OCTA; DRI OCT Triton Plus®; Topcon Corporation, Tokyo, Japan) images (12×12 mm) revealed no prominent changes in the superficial capillary plexus. The reflectivity of splitting in superficial retinal layers partially causes dark back shadowing in DCP. En face SS-OCTA images highlighted the areas of retinoschisis as areas of increased reflectivity of the retinal nerve fiber layer (Fig. 2a-d). There was no evidence of vitreoretinal traction and SS-OCT did not reveal any pathology in the optic disc and the fovea. Structural SS-OCT determined normal choroidal thickness and no lamina cribrosa alterations. There was no defect in the visual field testing (Humphrey®, Visual Field Analyzer-3, Zeiss, Germany) of both eyes. During 24 months of follow-up, no changes have been detected in the inner retinoschisis pathology and the patient was scheduled for 6 monthly follow-up visits.

Discussion

PPRS frequently occurs bilaterally with macular involvement. Most of the reported cases are asymptomatic and incidentally detected on OCT. Data on PPRS are limited, but several retrospective studies reported its association with

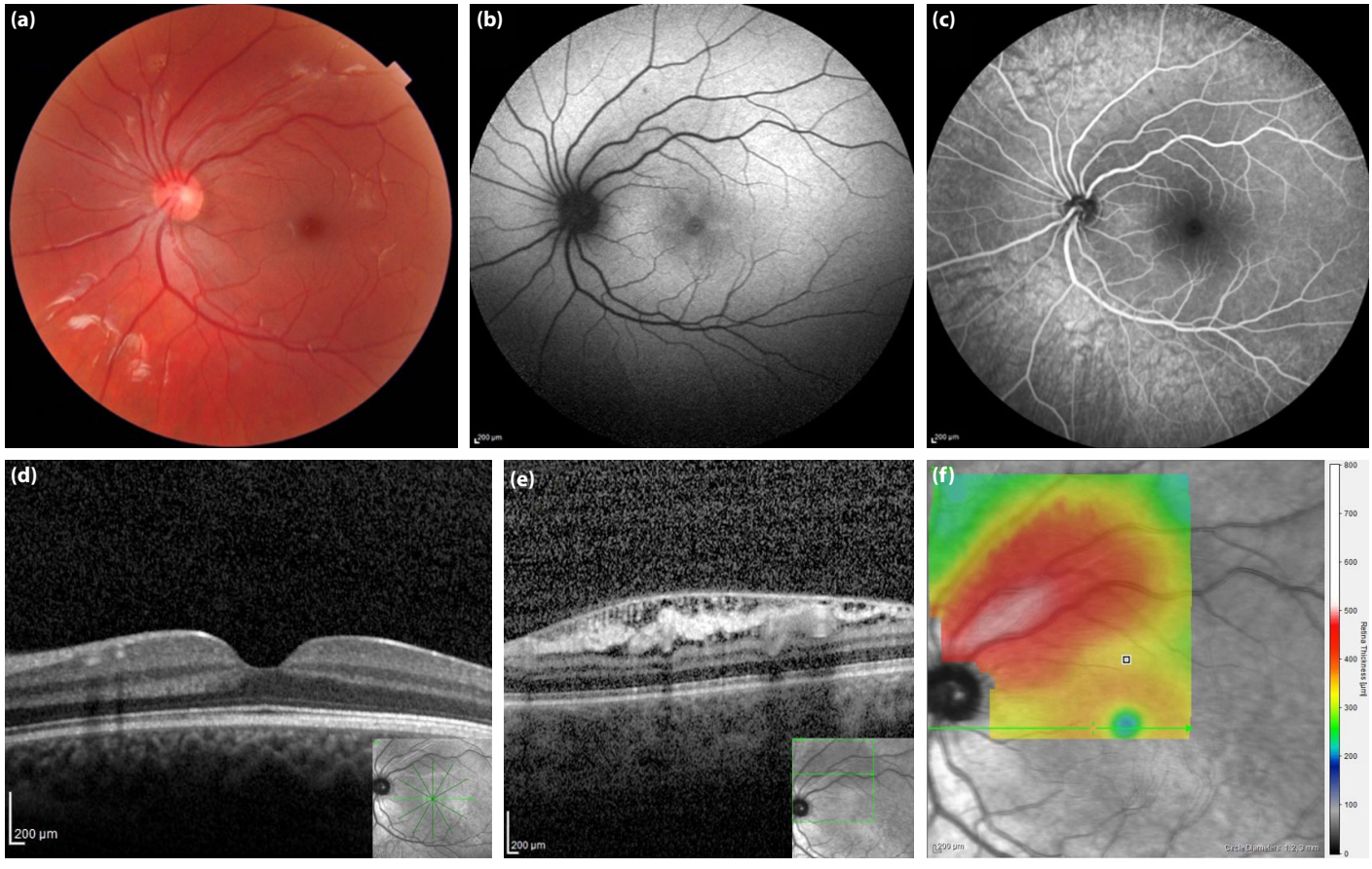

Fig. 1. Peripapillary retinoschisis is typically difficult to discriminate in color fundus photography **(a)**. A slight hypofluorescence in the superotemporal quadrant adjacent to the left optic disc (fundus autofluorescence) **(b)**. Normal fluorescein angiographic appearance in retinoschisis area **(c)**. Spectral-domain optical coherence tomography (SD-OCT) demonstrated a normal foveal contour in the left eye **(d)**. SD-OCT B-scan revealed splitting of various layers of the inner retina in the left eye **(e)**. Retinal thickness map showed significant thickening at the superotemporal peripapillary retina.

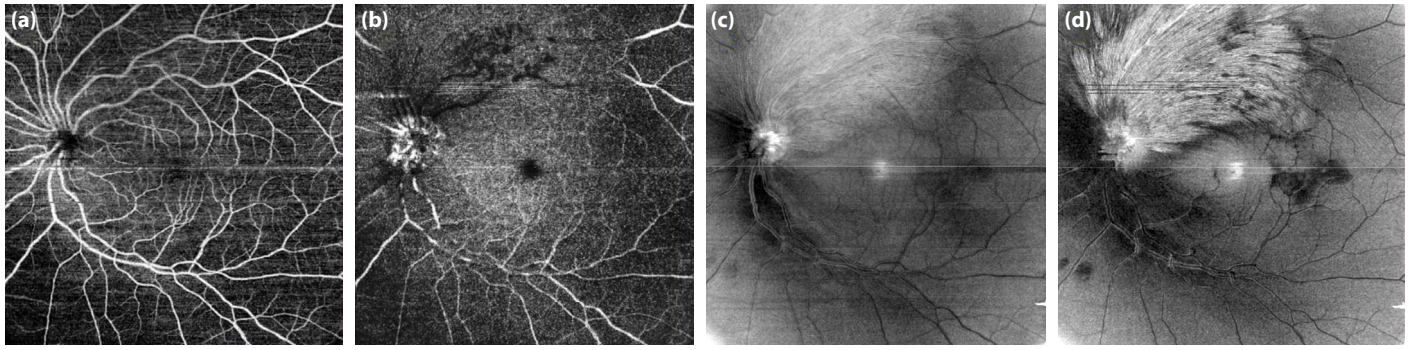

Fig. 2. In swept-source optical coherence tomography angiography (SS-OCTA) images; superficial capillary plexus revealed no prominent changes **(a)**. Splitting in superficial retinal layers partially causes dark back shadowing in DCP **(b)**. En face SS-OCTA images revealed a markedly increased reflectivity in the areas of retinoschisis **(c** and **d)**.

X-linked retinoschisis, primary acquired retinoschisis, SINFR, degenerative myopia, glaucoma, and congenital optic disc abnormalities.[1–8]

Congenital juvenile X-linked retinoschisis is a rare disorder and all affected individuals have typical foveal schisis with approximately half also exhibiting some degree of peripheral schisis. It almost exclusively occurs in males because of the X-linked inheritance pattern and is mostly seen bilaterally.[1,9] Primary acquired retinoschisis has been reported in patients within the third decade of life (20–30 years), commonly involves the inferior temporal retina bilaterally with minimal pigment alterations. It is characterized by splitting of the neurosensory retina at the outer plexiform layer and foveal affection is hardly present, even though rare cases of progression with retinal detachment including the macula were reported.

Our case had no associated ocular conditions such as X-linked retinoschisis, primary acquired retinoschisis, glaucoma, and congenital optic disc abnormalities. We speculated two theories regarding the development of PPRS in our case. One hypothesis is that high myopia is responsible for the peripapillary inner retinoschisis. High myopia is characterized by abnormal axial elongation with retinal microstructural degenerative changes such as retinoschisis, especially at the posterior pole. Sherman et al.[10] described that PPRS seems to be a clinical entity more prevalent in high myopia. In their study including 600 eyes, 19 exhibited retinoschisis around the optic disc. The splits were usually bilateral, variable in location and often appeared to exist in several layers, most often found in the inner and outer plexiform layers. Sixteen of them had normal or near-normal visual acuity and none had a macular involvement. However, most eyes demonstrated visual field defects as the enlargement of the blind spot. Eight eyes had one or more zones of vitreoretinal traction that might be the etiology of the schisis. They concluded that PPRS without macula schisis appears to be a new entity not previously reported but easily documented with SD-OCT images around the optic disc. Scans through the macula will miss the PPRS unless the peripapillary area is included in the OCT scan.[10]

Pathologic myopia with staphyloma is another cause of foveomacular retinoschisis due to a tractional maculopathy most likely arising from residual cortical vitreous after posterior vitreous detachment.[4] In their study, Shimada et al.[4] also reported that nearly in 48% of high myopic eyes with myopic conus, the peripapillary retinal vessels with tractional microfolds on OCT scans are associated with retinoschisis mostly showing an extension toward the macular area. Although our patient is bilaterally high myopic with long axial lengths, she did not exhibit any signs of degenerative myopia with myopic conus, staphyloma, or traction maculopathy on radial SD-OCT scans. The retinoschisis was unilateral and only involving the inner retinal layers rather than outer plexiform layer.

Second hypothesis is associated with PPRS, is SNIFR. SNIFR is an uncommon cause of foveomacular retinoschisis. Most cases are unilateral and highly myopic women with good visual acuity.[2,3] Recent evidence suggests that apart from foveomacular retinoschisis, peripheral imaging is key in identifying the other findings of SINFR, including mid-peripheral peripapillary inner retinoschisis. Although the clinical manifestation of SNIFR is based on OCT examination and defined as a stellate foveal splitting of the outer plexiform layer, the latest reports revealed coexisting peripapillary inner retinal changes on OCT.[2,3]

Our case is very similar to patients reported in the SNIFR series of Ober et al.[2] in which most of them were female with relatively good visual acuity, myopia, and unilateral involvement. Javaheri and Sadda[9] reported a 36-year-old woman with the diagnosis of SNIFR. She had mild myopia with good visual acuity (20/20) and exhibited macular split-

ting of the outer plexiform layer with peripapillary inner retinoschisis, involving the outer plexiform layer and inner retina in her left eye. Ahmed et al.[3] described an atypical case of bilateral SNIFR with a petaloid foveomacular splitting of the outer plexiform layer extending to the temporal periphery on the right eye on OCT, whereas on the left eye, there was only the cleavage of the outer retina started at the peripheral posterior pole, approximately 3.5 mm temporal to the umbo of the fovea. No pathology could be detected in FA and OCTA. They also claimed that there might be an early stage of SNIFR without foveal involvement. A possible expansion of the mid-peripheral splitting of the outer plexiform layer toward the center could lead to a secondary affection of the foveomacular zone which develops over a certain time and manifests as slight visual symptoms, once the fovea is chronically damaged.

Our case differs from these case series because she had only unilateral peripapillary inner retinoschisis instead of OPL and macular involvement. Our patient may also represent an early stage of SNIFR without foveal involvement as Ahmed et al.[3] described in their case report. With the standard use of SD-OCT in routine cases, PPRS will likely be diagnosed more frequently in the future as it can easily be recognized with its characteristic pattern. Longer follow-up and larger case series should be maintained to clarify this entity.

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