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Case Report

Peripapillary Retinoschisis as a Manifestation of Ocular Hypotony

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Peripapillary retinoschisis · Ocular hypotony

Abstract
The aim of this case report was to present an unusual case of peripapillary retinoschisis (PPRS) associated with ocular hypotony after glaucoma surgery. It refers to a 78-year-old man with primary open-angle glaucoma who developed PPRS while hypotonous. Optical coherence tomography of the peripapillary and the macular area of the right eye revealed PPRS temporally and nasally to the optic disc, more prominent at the level of the outer nuclear layer and less so at the inner nuclear layer. The PPRS completely regressed after 1 month of treatment and restoration of intraocular pressure to normal levels. This case report highlights the fact that PPRS in glaucoma patients may present in the setting of ocular hypotony and appears to resolve when the hypotony is successfully managed. Hydrostatic pressure gradient across retinal vasculature that allows movement of fluid into the extracellular spaces is a potential mechanism for the development of PPRS in ocular hypotony.

Introduction

Peripapillary retinoschisis (PPRS) is a splitting of the retina's neurosensory layers with accumulation of intraretinal fluid in the area adjacent to the optic nerve head. It has been observed in various ocular conditions like degenerative myopia and in optic disc abnormalities such as optic disc pit and optic disc coloboma, as well as in pachychoroid spectrum diseases [1]. Recently, the occurrence of PPRS has been described in eyes with various types of glaucoma associated with high or fluctuating intraocular pressures (IOPs) [2–7].

The clinical course of glaucoma-associated PPRS is usually favourable and does not manifest any symptoms if the macula is not involved. Most cases remain stable or resolve

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spontaneously several months after diagnosis [2]. However, there is much controversy regarding the clinical significance of PPRS for the glaucoma course. Earlier studies [5] reported that PPRS did not affect the glaucoma course, but recent studies have reported that PPRS is associated with rapid glaucoma progression [8, 9].

Furthermore, the exact pathogenesis of PPRS is not fully understood. It is assumed that alterations in the lamina cribrosa or vitreous traction may play a role in the pathogenesis of PPRS [7, 10]. The exact mechanism, however, remains speculative and requires further evaluation.

We report an interesting case of hypotony-related PPRS in a glaucoma patient and the subsequent resolution of the retinoschisis when the IOP returned to normal. Ocular hypotony is known to affect posterior segment structures. Hypotony maculopathy can manifest as a constellation of signs including optic nerve oedema, vascular tortuosity, choroidal folds, and macular striae. Pathophysiologically, hypotony is believed to cause an inward collapse of the scleral wall, resulting in choroidal and retinal redundancy, which gives the characteristic chorioretinal wrinkling [11]. Another less frequent sequela of ocular hypotony is cystoid macular oedema caused by abnormal retinal capillary permeability [11, 12]. Although our patient did not present with the typical features of hypotony maculopathy, the potential mechanisms for the development of PPRS in combination with ocular hypotony in this case are being discussed. The CARE Checklist has been completed by the authors for this case report and is attached as supplementary material.

Case Report

A 78-year-old male patient presented for routine follow-up in the glaucoma service. He had a history of primary open-angle glaucoma in the right eye managed with trabeculectomy 18 months before, and he had uncomplicated cataract surgery in this eye 4 months prior to presentation. His left eye had a history of ocular hypertension managed with dorzolamide/timolol drops twice daily and bimatoprost once daily, and a history of uncomplicated cataract surgery 2 months prior to presentation. His corrected visual acuity was 6/12 in OD and 6/6 in OS. Slit-lamp examination revealed bilateral pseudophakia, and IOP measurements with Goldmann applanation tonometry were 6 mm Hg in the right eye and 17 mm Hg in the left eye.

Dilated fundus examination revealed asymmetric disc cupping, with the right eye showing a more cupped disc. Peripapillary atrophy was noted around the right optic nerve head.

Optical coherence tomography (OCT) of the peripapillary and the macular area of the right eye (Fig. 1a) revealed schisis of the peripapillary retina, more prominent at the level of the outer nuclear layer and less so at the inner nuclear layer. There was no extension of the schisis to the parafoveal region. Peripapillary atrophy of the retina was noted in the OCT. Thickness map of the OCT showed significant retinal thickening in the temporal peripapillary area, corresponding to the peripapillary schisis. No signs of optic disc abnormality were noted, neither any vitreoretinal traction nor any pachychoroid features in the area affected by the retinoschisis, except for the presence of a subtle epiretinal membrane in the papillomacular area. A review of previous OCT scans of the patient indicated the presence of this subtle epiretinal membrane in previous visits but without any associated PPRS. OCT of the retinal nerve fibre layer (RNFL) of the right eye showed thinning in the superotemporal, superonasal, and inferotemporal sectors. OCT RNFL of the left eye showed only borderline thinning in the superotemporal and inferonasal sectors.

OCT angiography of the right optic disc and the right macula confirmed the absence of peripapillary or macular neovascularization in the retina or the choroid. It was also noted that the PPRS involved the retinal area nasally to the optic disc, with the presence of schitic fluid in the outer retinal layers. (Fig. 1b).

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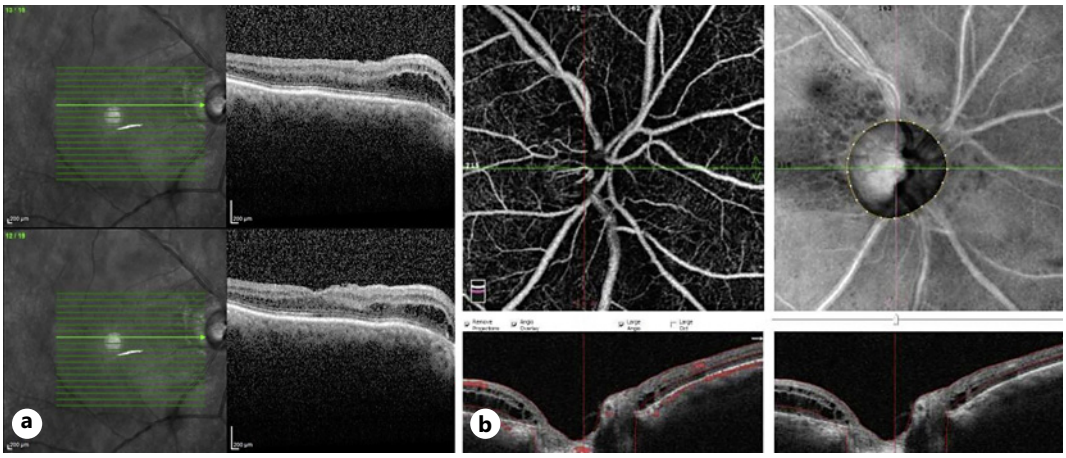

Fig. 1. a OCT of the peripapillary area (top) shows retinoschisis at the level of the outer nuclear layer and, to a smaller degree, the inner nuclear layer. The retinoschisis does not extend to involve the fovea (bottom). **b** OCT angiography of the optic disc confirms the absence of peripapillary neovascularization in the retina or the choroid. Structural OCT B-scan shows peripapillary schisis involving both the temporal and nasal retina around the optic disc.

The patient was managed with topical dexamethasone 0.1% b.i.d. and topical nepafenac 1 mg/mL t.i.d. Four weeks later, OCT showed complete resolution of the PPRS in the right eye (Fig. 2) and IOP measured 16 mm Hg. Best corrected visual acuity improved to 6/7.5. Topical treatment with dexamethasone and nepafenac was discontinued. During follow-up 2 months after initial presentation, OCT was normal with no PPRS and IOP measured 13 mm Hg.

Discussion

Various theories for the development of PPRS have been proposed. Lee JH et al. [1] postulated that defects of the lamina cribrosa were associated with PPRS. The location of the lamina cribrosa defects (central vs. peripheral) appeared to be associated with the involved retinal layers. Inner layer retinoschisis developed in eyes with central focal lamina cribrosa defects. Retinoschisis ranging from the inner nuclear layer to the outer nuclear layer developed in eyes with peripheral focal lamina cribrosa defects. It is proposed that the lamina cribrosa defects provide a conduit for cerebrospinal fluid to enter the peripapillary retina.

Another theory about the pathogenesis of PPRS suggests that vitreous traction on peripapillary areas of glaucomatous RNFL thinning causes micropores in the internal limiting membrane and RNFL complex. These micropores may provide a conduit for vitreous fluid to enter the peripapillary retina [2, 7].

In our case, multimodal imaging did not reveal any defect in lamina cribrosa or any tractional elements from the vitreous. Furthermore, in our opinion, the presence of the subtle epiretinal membrane in the schitic area could not justify the development of the retinoschisis, as the membrane was present in previous scans of this eye without causing any retinoschitic features. Also, it is noteworthy that the PPRS completely disappeared within a month after normalization of IOP. We propose the hypothesis that ciliary body inflammation caused hypotony in the involved eye, which subsequently resulted in shortening of the antero-posterior diameter [13]. We believe that this change in vitreoretinal interface enabled the movement of fluid from the vitreous tracks through a potential hole in the thin tissue of the optic cup and into the retina. A similar mechanism has been proposed by Zumbro et al. [14] for

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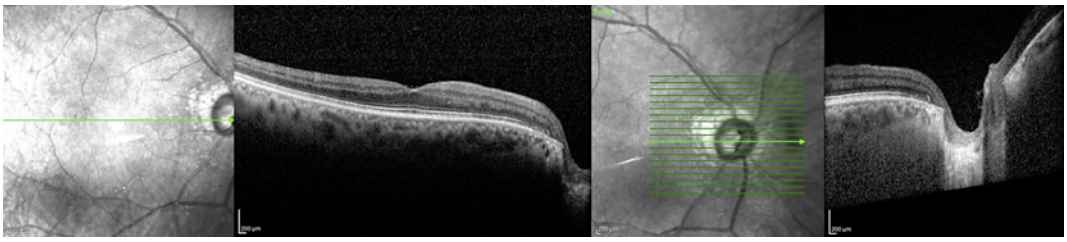

Fig. 2. One month after presentation, OCT (left) shows complete resolution of PPRS and EDI-OCT (right) confirms the absence of any structural defect in the lamina cribrosa. EDI-OCT, enhanced depth imaging OCT.

the development of macular schisis in these eyes. This theory seems to be supported by a recent detection of vitreous proteins in the macular schisis fluid associated with advanced glaucomatous excavation [15].

Another potential mechanism for the development of the schisis is based on the hypothesis that low IOP causes a higher hydrostatic pressure gradient across retinal vasculature that allows movement of fluid into the extracellular spaces. Kokame GT et al. [12] reported a case of hypotony maculopathy where the IOP normalized and the macular oedema subsequently resolved on OCT. As described in his case, low IOP represents low tissue hydrostatic pressure, resulting in a higher hydrostatic pressure gradient across retinal capillaries, which promotes a net movement of fluid into the extracellular spaces. Restoring the IOP by removal of a Baerveldt glaucoma implant in that case resulted in prompt resolution of the cystic changes and serous macular detachment, showing that restoration of a more normal IOP and, thus, higher hydrostatic tissue pressure in the Starling law resulted in less outflow of fluid from retinal capillaries. Less fluid outflow allowed reabsorption of extracellular fluid in the retina and an improvement in vision.

Similarly, in our case, anti-inflammatory treatment resulted in normalization of IOP and disappearance of the intraretinal peripapillary fluid. The mechanism for the development of hypotony is not clear to us. No inflammatory reaction was present in the anterior chamber at the time of examination; however, we cannot exclude the possibility that inflammation had occurred prior to presentation. Although the patient denied using anti-glaucomatous medication in the right eye, we cannot totally exclude this possibility as a cause of hypotony.

In conclusion, this case report highlights the fact that PPRS in glaucoma patients may present not only in the setting of acute elevation of IOP, as previously described, but also in ocular hypotony, and it appears to resolve when the hypotony is successfully managed. In addition, resolution of the PPRS in our case occurred in a much shorter period of time than the average time of spontaneous resolution of PPRS previously described.

Statement of Ethics

The present work was conducted in accordance with the Declaration of Helsinki. Ethical approval was not required for this study in accordance with local or national guidelines. Written informed consent was obtained from the patient for publication of the details of their medical case and any accompanying images.

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

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Author Contributions

Ioannis Markopoulos: writing and editing. Michail Tzakos: supervision and reviewing. Vasilios Tzimis: conceptualization and reviewing. Ioannis Halkiadakis: conceptualization, writing and reviewing.

Data Availability Statement

All data generated or analysed during this study are included in this article. Further enquiries can be directed to the corresponding author.

References

1 Lee JH, Park HYL, Baek J, Lee WK. Alterations of the lamina cribrosa are associated with peripapillary retinoschisis in glaucoma and pachychoroid spectrum disease. Ophthalmology. 2016;123(10):2066–76.
2 Kahook MY, Noecker RJ, Ishikawa H, Wollstein G, Kagemann L, Wojtkowski M, et al. Peripapillary schisis in glaucoma patients with narrow angles and increased intraocular pressure. Am J Ophthalmol. 2007;143(4):697–9.
3 Lee EJ, Kim TW, Kim M, Choi YJ. Peripapillary retinoschisis in glaucomatous eyes. PLoS One. 2014;9(2):e90129.
4 Bayraktar S, Cebeci Z, Kabaalioglu M, Ciloglu S, Kir N, Izgi B. Peripapillary retinoschisis in glaucoma patients. J Ophthalmol. 2016;2016:1612720.
5 van der Schoot J, Vermeer KA, Lemij HG. Transient peripapillary retinoschisis in glaucomatous eyes. J Ophthalmol. 2017;2017:1536030.
6 Grewal DS, Merlau DJ, Giri P, Munk MR, Fawzi AA, Jampol LM. Peripapillary retinal splitting visualized on OCT in glaucoma and glaucoma suspect patients. PLoS One. 2017;12(8):e0182816.
7 Dhingra N, Manoharan R, Gill S, Nagar M. Peripapillary schisis in open-angle glaucoma. Eye. 2017;31(3):499–502.
8 Fortune B, Ma KN, Gardiner SK, Demirel S, Mansberger SL. Peripapillary retinoschisis in glaucoma: association with progression and OCT signs of müller cell involvement. Invest Ophthalmol Vis Sci. 2018;59(7):2818–27.
9 Lee EJ, Kee HJ, Han JC, Kee C. The progression of peripapillary retinoschisis may indicate the progression of glaucoma. Invest Ophthalmol Vis Sci. 2021;62(2):16.
10 Farjad H, Besada E, Frauens BJ. Peripapillary schisis with serous detachment in advanced glaucoma. Optom Vis Sci. 2010;87(3):e205–17.
11 Costa VP, Arcieri ES. Hypotony maculopathy. Acta Ophthalmol Scand. 2007;85(6):586–97.
12 Kokame GT, de Leon MD, Tanji T. Serous retinal detachment and cystoid macular edema in hypotony maculopathy. Am J Ophthalmol. 2001;131(3):384–6.
13 Saeedi O, Pillar A, Jefferys J, Arora K, Friedman D, Quigley H. Change in choroidal thickness and axial length with change in intraocular pressure after trabeculectomy. Br J Ophthalmol. 2014;98(7):976–9.
14 Zumbro DS, Jampol LM, Folk JC, Olivier MM, Anderson-Nelson S. Macular schisis and detachment associated with presumed acquired enlarged optic nerve head cups. Am J Ophthalmol. 2007;144(1):70–4.
15 Patel S, Ling J, Kim SJ, Schey KL, Rose K, Kuchtey RW. Proteomic analysis of macular fluid associated with advanced glaucomatous excavation. JAMA Ophthalmol. 2016 Jan;134(1):108–10.