

CASE REPORT

Open Access



# Glaucomatous optic nerve damage in the contralateral eye of a patient with peripapillary retinoschisis: a case report

Wenbo Zhang, Tian Tian and Liu Yang\*

## Abstract

**Background** Peripapillary retinoschisis (PPRS) is often associated with glaucomatous eyes. It usually occurs in eyes with a more advanced stage of glaucoma with obvious optic nerve damage. We report a patient who was found to have PPRS in one eye during a routine physical examination without obvious glaucoma symptoms. Further examination revealed glaucomatous visual field loss and retinal nerve fiber layer defects in the contralateral eye.

**Case presentation** A 55-year-old man presented for a routine physical examination. The anterior segment was normal in both eyes. Fundus examination revealed an elevated and red optic disc in the right eye. In addition, scattered patchy red lesions were seen on the retina on the temporal side of the optic disc. The color and boundary of the left optic disc were normal, and the cup-to-disc ratio was 0.6. Optical coherence tomography showed retinoschisis on the optic nerve head of the right eye throughout the entire circumference, extending to the retina on the temporal side of the optic disc. The intraocular pressure was 18 mmHg OD and 19 mmHg OS. The patient was diagnosed with PPRS (OD). However, no optic disc pit or optic disc coloboma was found. Further examination showed that the visual field of the patient's right eye was generally normal, while a glaucomatous visual field defect was found in the left eye, which manifested as a nasal step visual field defect. Moreover, stereophotography and a red-free fundus image revealed two retinal nerve fiber layer defects in the supratemporal and infratemporal regions of the retina of the left eye. Continuous intraocular pressure measurement found that the intraocular pressure fluctuated between 18 and 22 mmHg OD and 19–26 mmHg OS during the daytime. Primary open-angle glaucoma was then diagnosed.

**Conclusions** In this case, we found that PPRS was associated with glaucomatous optic nerve changes and visual field defects in the fellow eye.

**Keywords** Peripapillary retinoschisis, Glaucoma, Retinal nerve fiber layer defect, Case report

## Background

Peripapillary retinoschisis (PPRS) is the splitting of the retina around the optic nerve head (ONH) and often occurs in patients with congenital optic disc

abnormalities, such as optic disc pit and optic disc coloboma [1–3]. Recent evidence indicates that PPRS is more likely to develop in glaucoma patients than in normal people [4–11]. PPRS usually occurs in eyes with a more advanced stage of glaucoma, a higher intraocular pressure, more severe optic nerve damage, a poorer visual field and a thinner retinal nerve fiber layer (RNFL) [4, 6, 9, 10, 12, 13]. However, the patient we reported was found to have PPRS in one eye during a routine physical examination without obvious glaucoma symptoms.

\*Correspondence:

Liu Yang

lucy02114@163.com

Department of Ophthalmology, Peking University First Hospital, No. 8 Xishiku Street, Xicheng District, Beijing 100034, China

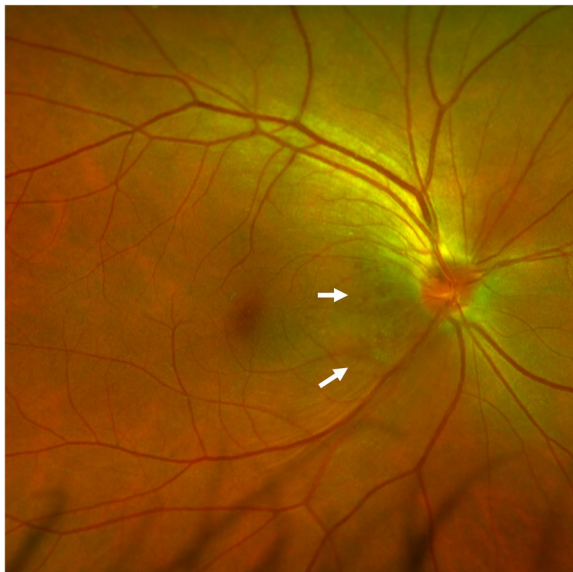


© The Author(s) 2023. **Open Access** This article is licensed under a Creative Commons Attribution 4.0 International License, which permits use, sharing, adaptation, distribution and reproduction in any medium or format, as long as you give appropriate credit to the original author(s) and the source, provide a link to the Creative Commons licence, and indicate if changes were made. The images or other third party material in this article are included in the article's Creative Commons licence, unless indicated otherwise in a credit line to the material. If material is not included in the article's Creative Commons licence and your intended use is not permitted by statutory regulation or exceeds the permitted use, you will need to obtain permission directly from the copyright holder. To view a copy of this licence, visit <http://creativecommons.org/licenses/by/4.0/>. The Creative Commons Public Domain Dedication waiver (<http://creativecommons.org/publicdomain/zero/1.0/>) applies to the data made available in this article, unless otherwise stated in a credit line to the data.

Further examination revealed glaucomatous visual field defects and RNFL defects in the contralateral eye.

### Case presentation

A 55-year-old man presented for a routine physical examination. His best-corrected visual acuity was 20/20 in the right eye (0 Diopters Sphere) and 20/20 in the left eye (-2.00 Diopters Sphere), and intraocular pressure was 18 mmHg OD and 19 mmHg OS (central corneal thickness: 539  $\mu\text{m}$  OD and 546  $\mu\text{m}$  OS). In slit-lamp biomicroscopy examination, the cornea, anterior chamber, iris and lens were normal. However, fundus examination (Optos p200t, Optos PLC, Dunfermline, UK) revealed an elevated, red, boundary-blurred ONH in the right eye. In addition, scattered patchy red lesions were seen on the retina on the temporal side of the optic disc, similar to retinal hemorrhage (Fig. 1, pseudocolor). The color and boundary of the left optic disc were normal, and the cup-to-disc ratio was 0.6. Spectral-domain optical coherence tomography (OCT, Spectralis HRA-OCT, Heidelberg Engineering, Heidelberg, Germany) was performed and showed retinoschisis on the ONH of the right eye throughout the entire circumference, extending to the retina on the temporal side of the optic disc. The splitting was mainly located in the RNFL layer. The outer nuclear layer, inner nuclear layer and ganglion cell layer were also involved in the temporal side of the optic disc. The red elevated ONH was caused by the splitting and towering of the neuroretina rim. On infrared fundus imaging, the



**Fig. 1** Fundus photography of the right eye. The optic nerve head is elevated, red and boundary-blurred. Scattered patchy red lesions are seen on the retina on the temporal side of the optic disc (marked with arrows)

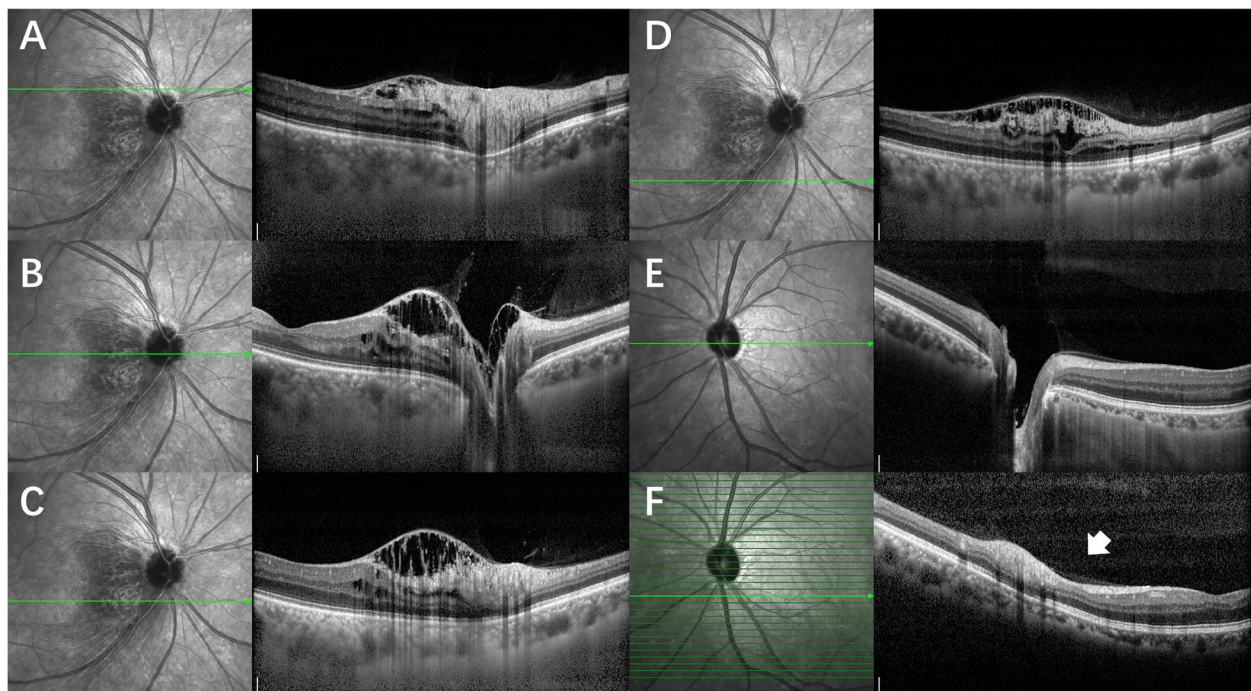
lesion on the temporal side of the optic disc presented as a wedge-shaped dark area with the base attached to the edge of the optic disc, scattered along the nerve fiber bundles (Fig. 2A-D). The OCT and infrared fundus image of the left eye was normal (Fig. 2E). The patient was diagnosed with PPRS (OD). However, no optic disc pit or optic disc coloboma was found. Since PPRS is often associated with glaucoma, visual field examination was subsequently performed. Visual field examination (Humphrey visual field analyzer 750i, Carl Zeiss Meditec, Dublin, USA) showed that the right side was generally normal, while a nasal step visual field defect was found in the left eye (Fig. 3). Furthermore, stereophotography and a red-free fundus image revealed two RNFL defects in the supratemporal and infratemporal regions of the retina of the left eye (Fig. 4). In line scan OCT, obvious RNFL thinning was found through the areas of RNFL defects (Fig. 2F). Intraocular pressure was continuously measured and fluctuations between 18 and 22 mmHg OD and 19–26 mmHg OS were found during the daytime. Because no other factors leading to secondary glaucoma were found and the anterior chamber angle was open, the patient was diagnosed with primary open-angle glaucoma and treated with local intraocular pressure lowering drugs.

### Discussion and Conclusions

At present, more studies support that PPRS tends to occur in glaucoma patients. The incidence of PPRS in glaucoma patients is approximately 3.1-6.0%, which is significantly higher than the incidence of approximately 0.5% in normal people [4–6]. PPRS can be found in various types of glaucoma, such as open-angle glaucoma [7, 8], narrow-angle glaucoma [9], secondary glaucoma [10], and even normal-tension glaucoma [11].

The specific mechanism of PPRS in glaucoma is still controversial. However, there are several possibilities. Kahook et al. suggested that the fluctuation in the intraocular pressure accompanied by small changes in the axial length is the main cause of PPRS [9]. Lee et al. reported that the liquid flows into the ONH from the subarachnoid space and vitreous cavity through the lamina cribrosa defect, thus causing PPRS. Glaucoma patients are more likely to have lamina cribrosa defects coupled with the destruction of glial barrier function caused by atrophy of inner nerve retinal tissue, which aggravates the entry of liquid into the retina [3]. Other possible mechanisms include lateral mechanical forces on the ONH caused by reactive gliosis of Müller cells [1, 6] and vitreous traction [11, 14].

PPRS usually resolves spontaneously [8], but it is still worthy of attention. First, spontaneous recovery of PPRS can lead to a sudden decrease in RNFL thickness



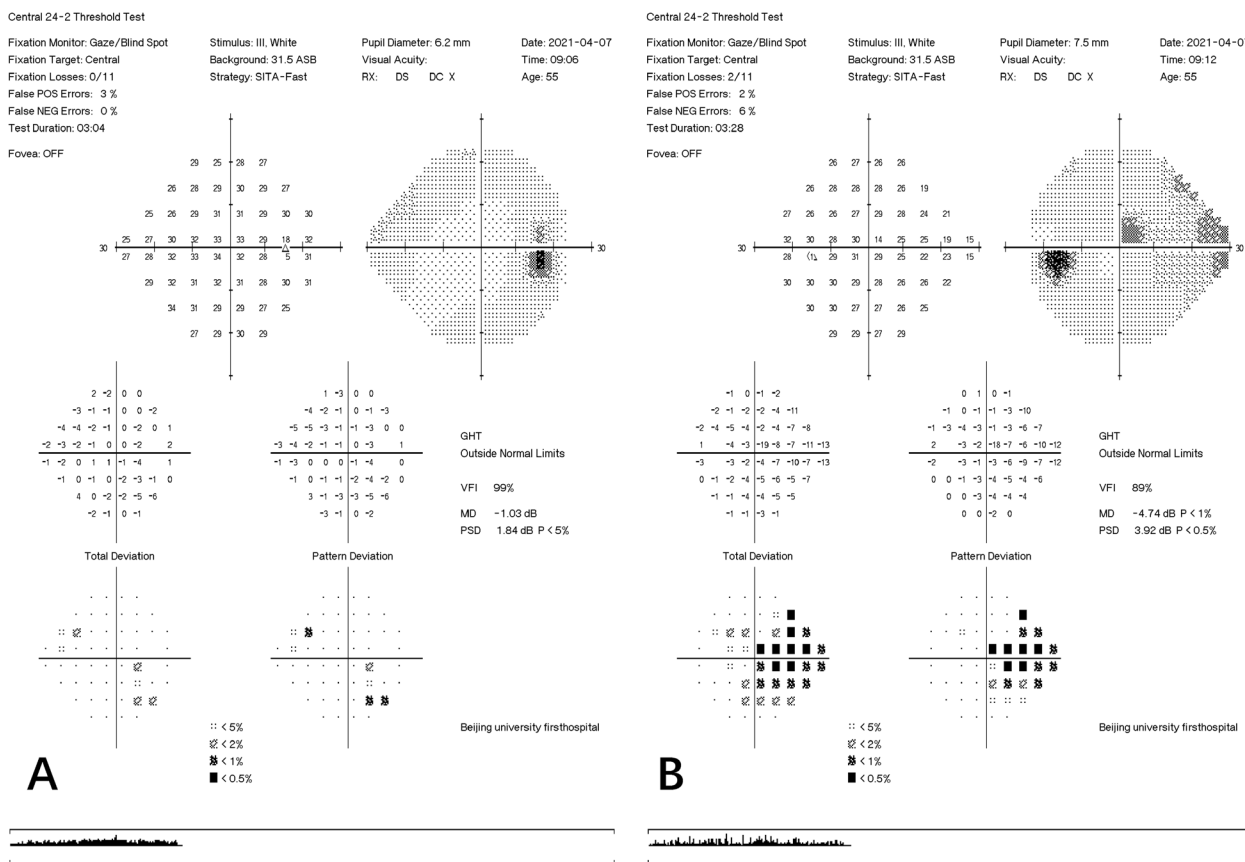
**Fig. 2** Optical coherence tomography (OCT) and infrared fundus image of the optic disc of both eyes. OCT shows retinoschisis on the optic nerve head of the right eye throughout the entire circumference, extending to the temporal retina. The splitting is located mainly in the retinal nerve fiber layer, ganglion cell layer, inner nuclear layer and outer nuclear layer. Infrared fundus image shows the lesion on the temporal side of the optic nerve head of the right eye presents as a wedge-shaped dark area with the base attached to the edge of the optic disc, scattered along the nerve fiber bundles (**A, B, C, D**). The OCT image and infrared fundus image of the left eye is normal (**E**). OCT shows obvious retinal nerve fiber layer thinning through the areas of retinal nerve fiber layer defects in the left eye (marked with an arrow, **F**)

measurements, resulting in misinterpretation of the rapid progression of glaucoma [7, 8]. Second, recent studies have shown that the emergence of PPRS is indeed related to the rapid aggravation of glaucoma [1, 6]. Third, based on this case, we found that PPRS can sometimes indicate the presence of glaucoma.

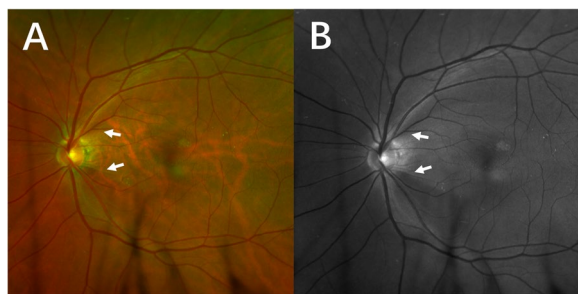
This patient was found to have PPRS in his right eye during a routine physical examination. No history of glaucoma or obvious symptoms was revealed. OCT showed no signs of optic disc pit or lamina cribrosa defect. Due to the close relationship between PPRS and glaucoma, we performed relevant examinations. Further examinations found that the patient actually had high intraocular pressure and glaucomatous optic nerve damage. This suggests that PPRS may indicate the existence of glaucoma, and additional examination should be performed for diagnosis. Interestingly, in this patient, glaucomatous optic nerve damage was found in the contralateral eye rather than the eye with PPRS. In fact, PPRS often occurs in a more advanced stage of glaucoma, and the affected eye is often accompanied by higher intraocular pressure, larger intraocular pressure fluctuation, higher cup-to-disc ratio, thinner RNFL thickness, and poorer visual

field [4, 6, 9, 10, 12, 13]. In addition, PPRS often appears at the site of RNFL defects, and the range and location of PPRS often overlap with those of RNFL defects [1, 4–6, 15, 16]. However, we found a high intraocular pressure, a nasal step visual field defect and two RNFL defects in the contralateral eye with PPRS. Therefore, although PPRS is associated with glaucomatous eyes, it can appear in the eye without obvious glaucomatous optic nerve damage. Visual field loss and RNFL defects can be found in the contralateral eye.

However, the above view is still speculative and requires more evidence to be confirmed. In fact, even if no visible disc pit was found in this case, occult disc pits may still exist. In Hedels C's study, small hyporeflexive cavitations were found adjacent to the ONH in a patient with PPRS without an obvious optic pit using OCT with an enhanced depth imaging model [17]. Nagesha CK et al. also found cavitation in the disc stroma in OCT images of a glaucoma patient with PPRS without visible disc pits, demonstrating an occult disc pit [18]. Using a narrow band raster OCT scan, Banerjee M et al. detected cavitations along the slope of the ONH in a patient with a circumscribed,



**Fig. 3** Automated perimetry examination of both eyes. The right visual field is generally normal (A), and the left visual field shows nasal step visual field defects (B)



**Fig. 4** Stereophotography and red-free fundus image of the left eye. The color and boundary of the left optic disc are normal, and the cup-to-disc ratio is 0.6. Retinal nerve fiber layer defects are found in the supratemporal and infratemporal regions of the retina (marked with arrows, A). Red-free fundus image shows obvious retinal nerve fiber layer defects in the supratemporal and infratemporal regions of the retina (marked with arrows, B)

hypopigmented macular lesion with peripapillary extension and no clinical evidence of disc pits [19]. In our study, we did not find such a sign of cavitation in the ONH on OCT images. Nevertheless, because there are gaps between lines in the OCT line scan, some

lesions may be missed, so the possibility of occult disc pits cannot be completely ruled out.

In general, PPRS does not require additional treatment and can resolve spontaneously within an average of 9 months [8]. Controlling intraocular pressure can also alleviate PPRS, whether through trabeculectomy or drug administration [4, 14]. Unfortunately, this patient was not followed up long enough to observe the outcome of PPRS.

This case report has some limitations in regards to the clinical data collection. First, we did not repeat the visual field examination for the patient, which may affect the reliability of the results. Second, ONH scan OCT, which can display RNFL thickness in different areas around the ONH, has not been performed. However, in line scan OCT, we found obvious RNFL thinning through the areas of RNFL defects. Third, we did not measure the diurnal variation in intraocular pressure for this patient.

In conclusion, we report a patient who was found to have PPRS in one eye without obvious glaucoma symptoms. Further examination revealed glaucomatous

optic nerve damage in the contralateral eye. PPRS may indicate the existence of glaucoma, further examination and follow-up is required. In this case, we found that PPRS was associated with glaucomatous optic nerve changes and visual field defects in the fellow eye.

#### Abbreviations

OCT	Optical coherence tomography
ONH	Optic nerve head
PPRS	Peripapillary retinoschisis
RNFL	Retinal nerve fiber layer.

#### Acknowledgements

Not applicable.

#### Authors' contributions

ZWB collected case information and wrote the majority of the manuscript with support from TT. YL reviewed the manuscript. All the authors have read and approved the manuscript.

#### Funding

Not applicable.

#### Availability of data and materials

Not applicable.

#### Declarations

#### Ethics approval and consent to participate

Not applicable.

#### Consent for publication

Written informed consent to publish this information was obtained from the participant.

#### Competing interests

The authors declare that they have no competing interests.

Received: 21 February 2022 Accepted: 27 March 2023

Published online: 03 April 2023

#### References

- 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. <https://doi.org/10.1167/iov.62.2.16>.
- Agorogiannis EI, Parkes C. Optic disc pit with peripapillary retinoschisis misdiagnosed as glaucoma. *JAMA Ophthalmol*. 2019;137(1):e183851. <https://doi.org/10.1001/jamaophthalmol.2018.3851>.
- Lee JH, Park HY, 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. <https://doi.org/10.1016/j.ophtha.2016.06.033>.
- Lee EJ, Kim TW, Kim M, Choi YJ. Peripapillary retinoschisis in glaucomatous eyes. *PLoS ONE*. 2014;9(2):e90129. <https://doi.org/10.1371/journal.pone.0090129>.
- Bayraktar S, Cebeci Z, Kabaalioglu M, Ciloglu S, Kir N, Izgi B. Peripapillary retinoschisis in glaucomatous eyes. *J Ophthalmol*. 2016;2016:1612720. <https://doi.org/10.1155/2016/1612720>.
- Fortune B, Ma KN, Gardiner SK, Demirel S, Mansberger SL. Peripapillary retinoschisis in glaucoma: association with progression and OCT signs of muller cell involvement. *Invest Ophthalmol Vis Sci*. 2018;59(7):2818–27. <https://doi.org/10.1167/iov.18-24160>.
- Dhingra N, Manoharan R, Gill S, Nagar M. Peripapillary schisis in open-angle glaucoma. *Eye (Lond)*. 2017;31(3):499–502. <https://doi.org/10.1038/eye.2016.235>.
- Hwang YH, Kim YY, Kim HK, Sohn YH. Effect of peripapillary retinoschisis on retinal nerve fibre layer thickness measurement in glaucomatous eyes. *Br J Ophthalmol*. 2014;98(5):669–74. <https://doi.org/10.1136/bjophthalmol-2013-303931>.
- 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. <https://doi.org/10.1016/j.ajo.2006.10.054>.
- Vila N, Chen JC. Peripapillary retinoschisis and serous retinal elevation secondary to steroid-induced glaucoma. *Can J Ophthalmol*. 2016;51(4):e136–8. <https://doi.org/10.1016/j.cjco.2016.03.018>.
- Inoue M, Itoh Y, Rii T, Kita Y, Hirota K, Kunita D, et al. Spontaneous resolution of peripapillary retinoschisis associated with glaucomatous optic neuropathy. *Acta Ophthalmol*. 2015;93(4):e317–318. <https://doi.org/10.1111/aos.12568>.
- Ornek N, Buyuktortop N, Ornek K. Peripapillary and macular retinoschisis in a patient with pseudoexfoliation glaucoma. *BMJ Case Rep*. 2013;2013:bcr2013009469. <https://doi.org/10.1136/bcr-2013-009469>.
- Farjad H, Besada E, Frauens BJ. Peripapillary schisis with serous detachment in advanced glaucoma. *Optom Vis Sci*. 2010;87(3):E205–217. <https://doi.org/10.1097/OPX.0b013e3181d1dad5>.
- 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. <https://doi.org/10.1016/j.ajo.2007.03.027>.
- van der Schoot J, Vermeer KA, Lemij HG. Transient peripapillary retinoschisis in glaucomatous eyes. *J Ophthalmol*. 2017;2017:1536030. <https://doi.org/10.1155/2017/1536030>.
- Lee WJ, Seong M. Disc hemorrhage following peripapillary retinoschisis in glaucoma: a case report. *BMC Ophthalmol*. 2021;21(1):253. <https://doi.org/10.1186/s12886-021-02010-5>.
- Hedels C, Krohn J. Enhanced depth imaging optical coherence tomography of optic disc maculopathy without a visible optic pit. *Clin Exp Ophthalmol*. 2013;41(9):894–6. <https://doi.org/10.1111/ceo.12090>.
- Nagesha CK, Ganne P. Occult optic disc pit maculopathy in a glaucomatous disc. *Middle East Afr J Ophthalmol*. 2017;24(3):165–6. [https://doi.org/10.4103/meajo.MEAJO\\_79\\_17](https://doi.org/10.4103/meajo.MEAJO_79_17).
- Banerjee M, Khandelwal H, Azad SV, Venkatesh P. Utility of narrow band raster scan for detection of occult disc pit. *BMJ Case Rep*. 2023;16(1). <https://doi.org/10.1136/bcr-2022-253164>.

Ready to submit your research? Choose BMC and benefit from:

- fast, convenient online submission
- thorough peer review by experienced researchers in your field
- rapid publication on acceptance
- support for research data, including large and complex data types
- gold Open Access which fosters wider collaboration and increased citations
- maximum visibility for your research: over 100M website views per year

At BMC, research is always in progress.

Learn more [biomedcentral.com/submissions](https://biomedcentral.com/submissions)

