

Peripapillary and macular retinoschisis - A vision-threatening sequelae of advanced glaucomatous cupping

Vijayalakshmi A Senthilkumar, Chitaranjan Mishra¹, Naresh B Kannan¹, Priyanka Raj

Purpose: To present a selected case series of advanced glaucoma-associated peripapillary and macular retinoschisis and response to various treatment strategies with a comprehensive literature review. **Methods:** Retrospective observational case series. Retrospective review of five selected cases of advanced glaucoma with peripapillary and macular retinoschisis. **Results:** All five patients had advanced glaucomatous damage with macular and peripapillary retinoschisis, three (patients 2, 3, and 5) had a neurosensory detachment of the macula. Increased intraocular pressure was managed with maximal antiglaucoma medications and G6 micropulse diode laser treatment in the first patient, transscleral diode laser in the second patient, mitomycin-C augmented trabeculectomy in the third patient, maximal antiglaucoma medications alone in the fourth patient, pars plana vitrectomy followed by trabeculectomy in the fifth patient. **Conclusion:** We speculate that peripapillary and macular retinoschisis may indicate a vision-threatening sequelae of advanced glaucoma. The probable inciting factor for this vision-threatening pathology being elevated intraocular pressure, fluctuations in intraocular pressure, and chronic glaucoma with advanced cupping. We emphasize that meticulous examination of the macula in patients with advanced glaucoma is mandatory. It is imperative to do OCT macula in patients with advanced glaucoma to diagnose this distinct entity at an earlier stage and preserve the existing visual potential.

Key words: Advanced glaucoma, deep cup associated maculopathy, macular retinoschisis, peripapillary retinoschisis, serous retinal detachment

Macular retinoschisis occurs in eyes with optic nerve head structural abnormalities such as optic nerve pit, optic nerve coloboma, tilted disc syndrome, morning glory syndrome, and high myopia.^[1] An association between primary glaucomas (primary open-angle glaucoma and primary angle-closure glaucoma) with peripapillary and macular retinoschisis has been sparsely reported in the literature.^[2,3] However, the exact mechanism of this schisis in patients with advanced optic nerve head cupping without a definite optic disc pit is still inconclusive. The basic underlying pathogenesis of peripapillary and macular retinoschisis in advanced glaucoma need to be explored. This additional retinal pathology may in turn deteriorate the quality of central vision in advanced glaucoma patients.

In this article, we report five patients of different age groups and different types of glaucoma who presented with advanced glaucoma, macular, and peripapillary retinoschisis. None of the patients had vitreous traction or acquired optic disc pit. Management modality was different in all these patients as it was dependent on the discretion of the treating ophthalmologist and there is no unified consensus for the treatment modalities. Though further progression of peripapillary and macular retinoschisis was halted anatomically, the visual outcomes

were suboptimal. To the best of our knowledge, our case series contains the highest number of patients of this relatively uncommon disease from India.

Methods

This was a retrospective observational case series. The study was approved by the Institutional Review Board and adhered to the tenets of the Declaration of Helsinki. Five eyes of five patients with macular and peripapillary retinoschisis were included in the study. Informed consent has been obtained for all the patients. All patients had undergone detailed ophthalmological examination, including best-corrected visual acuity (BCVA) (Snellen's chart), slit-lamp biomicroscopy, color fundus photography, Humphreys visual field analysis, OCT retinal nerve fiber analysis, and high-resolution OCT macula as well as peripapillary retina. The Institutional Review Board approved the study and the IRB number is RET202100359-approval date is 1/9/2021.

Results

Case 1: A 29-year young adult male, diagnosed to have bilateral advanced primary angle-closure glaucoma on

This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

For reprints contact: WKHLRPMedknow_reprints@wolterskluwer.com

Cite this article as: Senthilkumar VA, Mishra C, Kannan NB, Raj P. Peripapillary and macular retinoschisis - A vision-threatening sequelae of advanced glaucomatous cupping. Indian J Ophthalmol 2021;69:3552-8.

Access this article online

Website:
www.ijo.in

DOI:
10.4103/ijo.IJO_668_21

Quick Response Code:



Departments of Glaucoma and ¹Retina and Vitreoretinal Surgery, Aravind Eye Hospital and Postgraduate Institute of Ophthalmology, Madurai, Tamilnadu, India

Correspondence to: Dr. Vijayalakshmi A. Senthilkumar, Department of Glaucoma, Aravind Eye Hospital and Postgraduate Institute of Ophthalmology, Madurai, Tamilnadu, India. E-mail: vijimmc@gmail.com

Received: 23-Mar-2021

Revision: 07-Jul-2021

Accepted: 23-Jul-2021

Published: 26-Nov-2021

irregular treatment presented with a gradual drop in visual acuity in the right eye (OD) for 1 month. His best-corrected visual acuity (BCVA) and intraocular pressure (IOP) in OD and left eye (OS) were 20/120, 38 mmHg, 20/20, and 26 mmHg, respectively. Gonioscopy showed closed angles in both eyes (OU). Fundus examination revealed a cup-disc-ratio (CDR) of 0.9 OU [Fig. 1a and c] with macular edema in OD without any optic disc pit or coloboma. His axial lengths were 22.61 mm OD and 22.82 mm OS. Humphreys visual field analysis (HFA) and optical coherence tomography (OCT - RNFL analysis) OU were remarkable for advanced glaucomatous damage. OCT macula revealed peripapillary retinoschisis and schitic changes in the nasal macula OD [Fig. 1a, b] and normal macula OS [Fig. 1d]. The patient was treated with maximal ocular hypotensive agents OU and subsequently underwent micropulse G6 diode (IRIDEX) laser treatment in OD. Invasive glaucoma filtration procedure was deferred in this patient as he had split fixation field defect OD. The IOP was brought under control with maximal medical therapy and the retinoschisis in OD remained non-progressive at 8 months follow-up, with a BCVA of 20/40.

Case 2: A 75-year male patient diagnosed with pseudoexfoliation glaucoma on maximal medical management with poor drug compliance, presented with defective vision in OD since 1 month. His BCVA was OD (hand movements) and OS (20/20), with an IOP of 46 mmHg OD and 22 mmHg OS. The fundus examination showed a CDR of 0.95 with macular edema OD [Fig. 2a] and 0.65 CDR OS [Fig. 2b]. His axial lengths were 23.11 mm OD and 22.86 mm OS. OCT B-scan through the macula showed neurosensory detachment with subretinal fluid with schisis and intraretinal fluid [Fig. 2c]. OCT B-scan through the optic disc showed peripapillary retinoschisis without any optic disc pit in OD [Fig. 2d]. The patient underwent partial diode cyclophotocoagulation (IRIDEX) of the inferior 180 degrees for control of IOP considering the poor visual potential. Two-year follow-up OCT showed a decrease in retinoschisis and decreased retinal thickness [Fig. 2e].

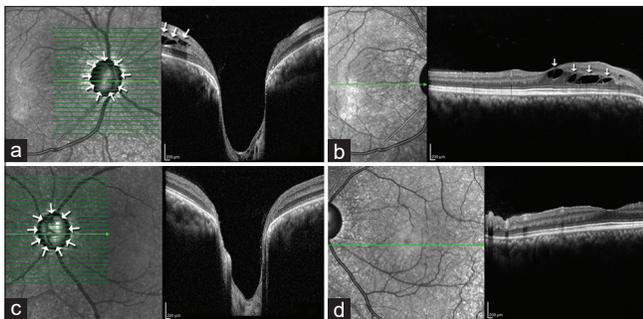


Figure 1: (a) Left panel- Confocal scanning fundus image zoomed over the optic disc of OD showing 0.9 cup disc ratio (white arrows) without any optic disc pit. Right Panel- OCT B-scan through the disc OD showing deep cup and peripapillary retinoschisis (white arrows). (b) Right Panel- OCT B-scan through the center of the fovea of OD showing macular retinoschisis and intraretinal fluid (white arrows). The schitic cavities are located in the inner nuclear layer, outer plexiform layer, and outer nuclear layer. (c) Left panel- Confocal scanning fundus image zoomed over the optic disc of OS showing 0.9 cup disc ratio (white arrows) Right panel- OCT B-scan showing no peripapillary retinoschisis in OS. (d) Right panel- OCT B-scan through the center of fovea in OS showing normal retinal layers. (OCT images taken in HRA Spectralis machine. The scanned OCT lines in Figure a-c are shown as green arrows and in Figure d as a green line.)

Case 3: A 32-year female patient, diagnosed with bilateral pigmentary glaucoma on poor drug compliance, presented to us with high IOP in OU. Her BCVA OD was 20/30, OS was 20/20, and presenting IOP were OD = 52 mmHg and OS = 40 mmHg. Gonioscopy of both eyes showed open angles with grade 4 hyperpigmented trabecular meshwork. Fundus evaluation showed OD 0.9 CDR with macular edema and OS 0.85 CDR [Fig. 3a]. Her axial lengths were 21.14 mm OD and 22.16 mm OS. HFA and OCT-RNFL analysis OU were remarkable for advanced glaucoma [Fig. 3b]. OCT B-scan through the center of fovea revealed subretinal fluid

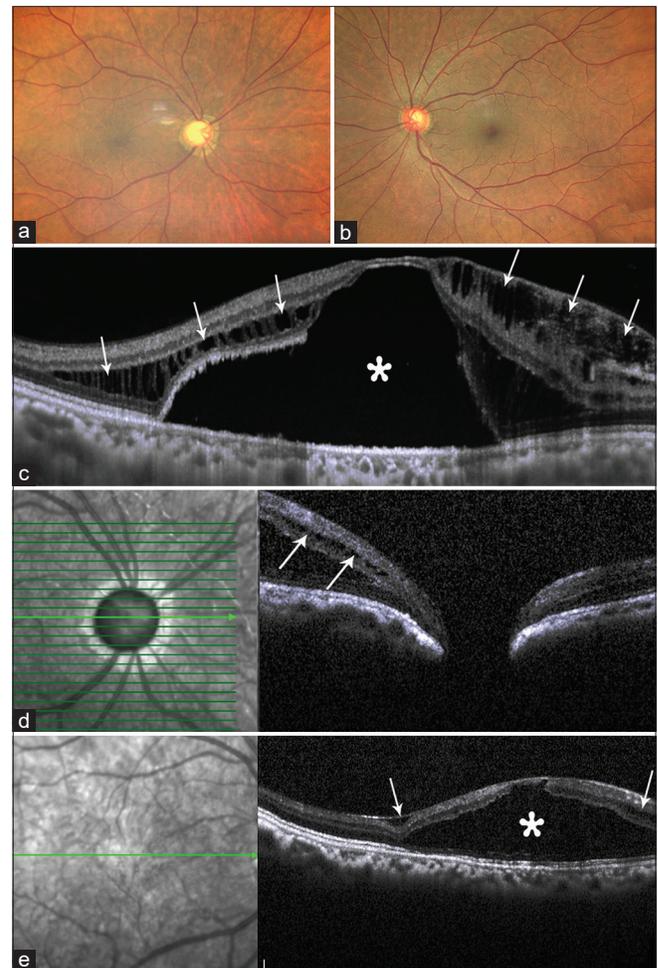


Figure 2: (a) Color fundus photo OD showing large cup disc ratio of 0.95 with near total cupping (white arrows) (b) Color fundus photo OS showing cup disc ratio of 0.65 (c) OCT B-scan through the center of fovea OD showing intraretinal schisis (white arrows). Also note the outer retinal hole with large neurosensory detachment (white asterix). The schitic cavities are located in the retinal layers, from the nerve fiber layer to the outer nuclear layer. (d) Left panel- Confocal scanning fundus image zoomed over the optic disc of OD showing 0.95 cup disc ratio without any optic disc pit. Right panel- OCT B-scan showing peripapillary retinoschisis (white arrows). (e) Right panel- OCT B-scan through the center of fovea OD showing reduced neurosensory detachment (white Asterix) and intraretinal schisis (white arrows) in comparison to Fig. 2c. The schitic cavities are limited to the inner nuclear layer and the outer plexiform layer. (Fundus image taken in Clarus (Zeiss) and OCT images taken in HRA Spectralis machine. The scanned OCT lines in Figures d-e are shown as green arrows.)

and OCT B-scan through the disc showing peripapillary neurosensory detachment (NSD) and no obvious optic disc pit in OD [Fig. 3c, d]. We excluded central serous retinopathy clinically as the female patient does not belong to any of the risk categories involved in CSR (nonsmoker, not a Type- A personality, no h/o steroid intake, antianxiety/antidepressants for stress disorders, no h/o insomnia) as well as by Fundus fluorescein angiography. She underwent Mitomycin-C augmented trabeculectomy OU for control of IOP. At six months follow-up, OCT macula showed non-progressive persistent NSD with favorable IOP control.

Case 4: A 72-year-old male patient diagnosed as primary open-angle glaucoma on maximal medical management with good drug compliance, presented with defective

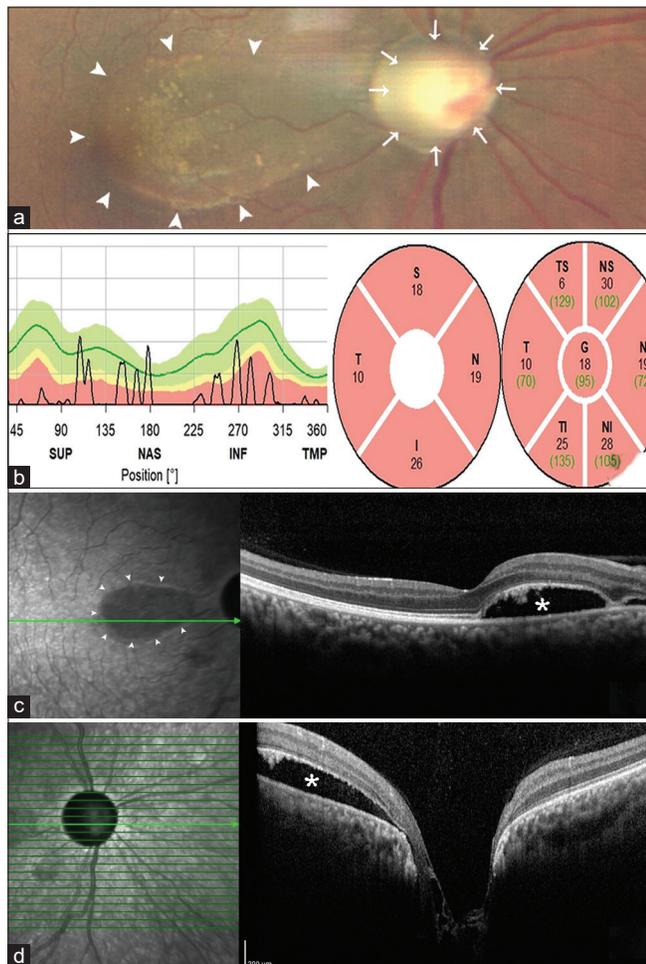


Figure 3: (a) Color fundus photo OD showing 0.9 cup disc ratio (white arrows) without any optic disc pit. Also note the macular elevation (white arrowheads) (b) OCT-RNFL analysis of the right eye showing significant RNFL thinning in all the quadrants c. Left panel- Confocal scanning fundus image zoomed over the macula of OD showing fluid collection in the macular region (white arrowheads). Right panel- OCT B-scan through the center of fovea showing neurosensory detachment with subretinal fluid. d. Left panel- Confocal scanning fundus image zoomed over the optic disc of OD showing 0.9 cup disc ratio. Right panel- OCT B-scan through the disc showing peripapillary neurosensory detachment (white Asterix) and no obvious optic disc pit. (OCT images taken in HRA Spectralis machine. The scanned OCT lines in Figures c-d are shown as green arrows.)

vision in OD since 1 month. His BCVA was OD = 20/25 and OS = 20/20, with an IOP of 18 mmHg OD and 16 mmHg OS. The fundus examination showed O.85 CDR with macular edema OD [Fig. 4a] and 0.75 CDR OS. OCT-RNFL analysis showed superior and inferior RNFL thinning. His axial lengths were 21.16 mm OD and 21.18 mm OS. OCT B-scan through the macula showed cystic cavities at the center [Fig. 4b], peripapillary retinoschisis and a thin epiretinal membrane. B-scan of the macula taken at a level just superior to the optic disc revealed intraretinal schisis and fluid collection at multiple

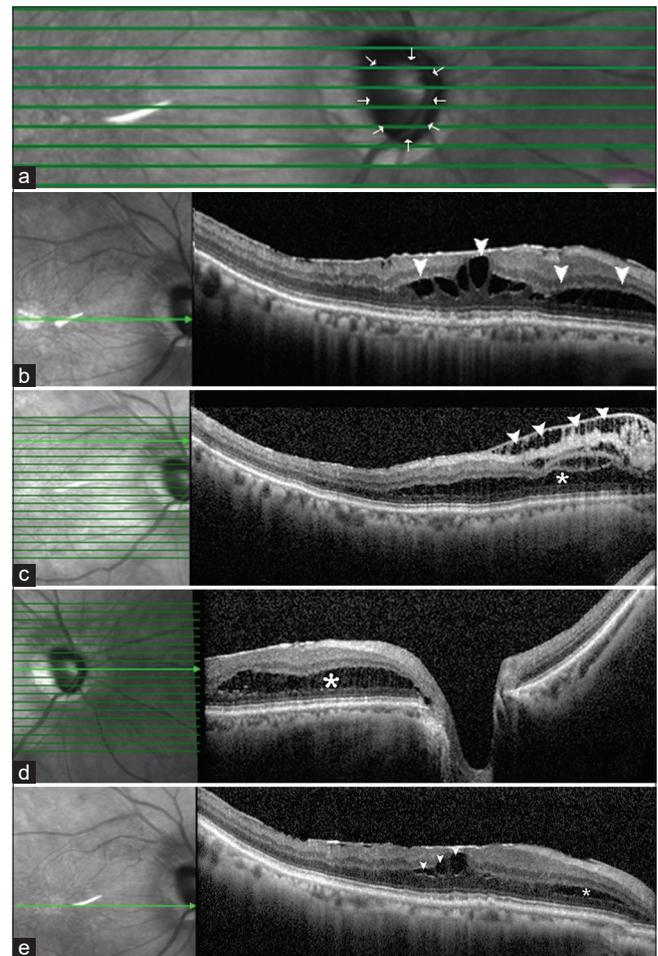


Figure 4: (a) Confocal scanning fundus image zoomed over the optic disc of OD showing 0.85 cup disc ratio (white arrows) without any optic disc pit. (b) Right panel- OCT B-scan through the macula suggestive of cystic cavities at the center and peripapillary retinoschisis (white arrowheads). Also note the thin epiretinal membrane. (c) Right panel- OCT B-scan of the macula taken at a level just superior to the disc suggestive of intraretinal schisis and fluid collection at multiple layers (white arrowheads and asterix). The schitic cavities are present in the nerve fiber layer, ganglion cell layer, inner nuclear layer, outer plexiform layer, and outer nuclear layer. (d) Right panel- OCT B-scan through the disc showing peripapillary retinoschisis (white Asterix) and no obvious optic disc pit. The schitic cavities are present in the outer plexiform layer. (e) Right panel- OCT B-scan through the disc and macula of OD showing decreased peripapillary retinoschisis limited to the outer plexiform layer only (white asterix) and intraretinal cystic cavities limited to the sub-foveal inner retinal layers (white arrowheads). (OCT images taken in HRA Spectralis machine. The scanned OCT lines in Figures b-e are shown as green arrows.)

layers, and B-scan through the optic disc showing peripapillary retinoschisis and no obvious optic disc pit in OD [Fig. 4c, d]. Surgical intervention was deferred in this patient considering the age and BCVA. The patient was observed only with maximum AGM. Six months follow-up OCT OD revealed decreased peripapillary and macular retinoschisis [Fig. 4e].

Case 5: A 34-year male patient, diagnosed to have bilateral juvenile glaucoma on irregular follow-up and poor drug compliance presented to us with a gradual drop in visual acuity in OS for 3 months. His BCVA and IOP in OD and OS were 20/20, 42 mmHg, 20/200, and 36 mmHg, respectively. Gonioscopy revealed open angles with prominent iris process. Fundus examination revealed an OD CDR of 0.8 and an OS CDR of 0.9 with macular elevation without any optic disc pit or coloboma [Fig. 5a]. HFA and OCT - RNFL analysis OU were remarkable for advanced glaucomatous damage [Fig. 5b]. The OCT B-scan through the center of the macula in OS showed schisis with neurosensory detachment [Fig. 5c], OCT B-scan through the disc showed peripapillary retinoschisis without any obvious optic disc pit [Fig. 5d]. His axial lengths were 22.12 mm OD and 22.85 mm OS. The IOP was brought under control by topical ocular hypotensive medications for 3 weeks as well as systemic carbonic anhydrase inhibitors for one week and the IOP dropped to 26 mmHg in RE and 22 mmHg in LE. After stabilizing the IOP, we proceeded with vitrectomy surgery with ILM peeling and gas tamponade (SF6) for the maculopathy in OS first. However, in the postoperative period, as we targeted the IOP to be further lower and it could not be achieved with antiglaucoma drugs, we performed sequential trabeculectomy OU. After 2 months of vitreoretinal surgery, there was a full-thickness macular hole (FTMH) in OS [Fig. 5e, f]. The patient was observed only with the possibility of spontaneous closure of FTMH over a longer follow-up. The final BCVA of the patient at six months follow-up was OD 20/20 and OS 20/200 with an IOP OD of 18 mmHg and an OS of 16 mmHg. The FTMH remained the same at the 6-month follow-up visit [Fig. 5g].

Discussion

Macular retinoschisis, a term that describes large separations of the retinal layers in the posterior pole, is often associated with congenital pits of the optic nerve head. The typical finding of macular retinoschisis in patients with advanced optic disc cupping without a visible optic disc pit has been reported sparsely in the recent past.^[1-3] In our case series, we could document the variable morphology of macular retinoschisis and peripapillary retinoschisis with the help of OCT and documented the follow-up images appropriately. In addition, we found the constant association of high IOP, advanced cupping, and poor visual outcome in these eyes [Table 1]. This is the first case series highlighting the association between retinoschisis and advanced glaucoma in Indian ethnic eyes.

The probable pathogenesis of this condition was earlier proposed as an acute rise in intraocular pressure (IOP). Hollander *et al.*^[3] first described the association of macular retinoschisis and serous retinal detachment in a patient with advanced glaucomatous cupping resulting from repeated attacks of acute angle-closure glaucoma. The author presumed the mechanism to be acute rise in IOP and increased optic disc

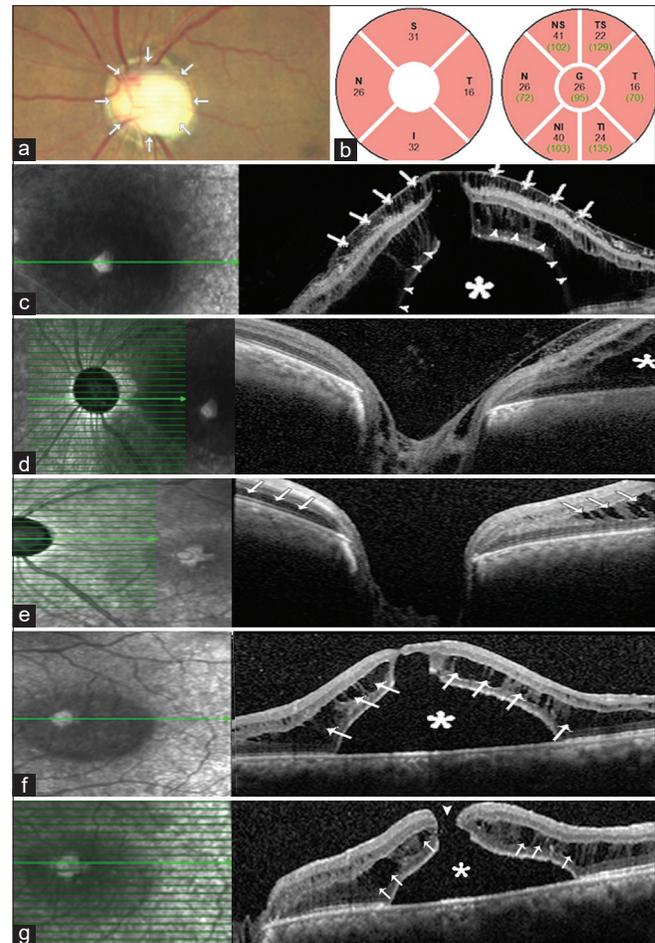


Figure 5: (a) Color fundus photo OS showing 0.9 cup disc ratio (white arrows) without any optic disc pit. (b) OCT-RNFL OS showing significant RNFL thinning in all the quadrants (c) Right panel- OCT B-scan through the center of the macula of the left eye showing macular retinoschisis with intraretinal fluid (white arrows and arrowheads). Also note the outer retinal hole with large neurosensory detachment (white asterisk). The schitic cavities are present in the retinal nerve fiber layer, ganglion cell layer, inner nuclear layer, and outer plexiform layer, and outer nuclear layer (d) Left panel- OCT B-scan through the disc showing peripapillary retinoschisis and neurosensory detachment (white Asterix). There was no obvious optic disc pit. (e) Left panel- OCT B-scan through the disc at 2 months post vitreoretinal surgery showing decreasing peripapillary retinoschisis (white arrows) (f) OCT B-scan through the macula at 2 months post vitreoretinal surgery showing decreasing retinoschisis (white arrows) in comparison to Fig. 5c. Also note the subretinal fluid with full-thickness macular hole (white asterix). The schitic cavities were predominantly present in the inner and outer plexiform layers (g) Right panel- Five months postoperative OCT macula showing a full-thickness macular hole in the left eye (white arrowhead), subretinal fluid (white Asterix), and retinoschisis (white arrows). The schitic cavities were predominantly present in the inner and outer plexiform layers. (Fundus image taken in Optos Daytona and OCT images taken in HRA Spectralis machine). The scanned OCT lines in Figures c-g are shown as green arrows.)

cupping could have led to the liquified vitreous entering the retina resulting in schisis detachment. Subsequently, some case series have been reported showing an indubitable association between large glaucomatous cupping and retinoschisis [Table 2].^[3-6]

Table 1: Demographic and clinical findings of study patients

Pat No	Age	Sex	Type of glaucoma	Duration of glaucoma (in years)	Severity of glaucoma	Treatment given for IOP control	IOP At which the diagnosis of maculopathy was done	Laterality	OCT findings	Treatment given	Final follow-up
1.	29	M	PACG	1	Advanced CDR-0.9	AGM-4, YAG PI	38	RE	Peripapillary schisis and schitic changes in the nasal macula	G6 IRIDEX micropulse	8 months-schisis remained the same
2.	75	M	PXFG	5	Advanced 0.95	AGM-3	46	RE	Peripapillary schisis and neurosensory detachment with macular schisis and intraretinal fluid	IRIDEX Transcleral diode cyclophotocoagulation	24 months - Decrease in schisis and retinal thickness
3.	32	F	Pigmentary glaucoma	3	Advanced CDR-0.9	AGM-3	52	RE	Subretinal fluid in the macula and peripapillary neurosensory detachment	MMC augmented trabeculectomy	6 months Remained the same
4.	72	M	POAG	8	Severe CDR-0.75	AGM-3	18	RE	Cystic cavities in macula and peripapillary and intraretinal schisis in the macula	Maximum AGM	6 months- Decreased retinal and macular schisis
5.	34	M	JOAG	4	Advanced CDR-0.9	AGM-3	36	LE	schitic changes with neurosensory detachment in the macula and peripapillary schisis	PPV + SF6 followed by 5-FU augmented trabeculectomy	6 months- Full-thickness macular hole

The second mechanism suggested for this distinct entity being fluctuations in IOP, which can aggravate the fluid accumulation as were seen in four of our patients.^[3] The fluid from the vitreous tracks through a microhole in the thin tissue of the glaucomatous deep optic nerve cup into the retina and causes edema of the retinal layers, and when it is severe leads to a schisis appearance and ultimately serous retinal detachment.^[7] This mechanism was supported by non-progression of schisis after IOP control as reported by Kahook *et al.* and Zumbro *et al.* [Table 2]. Aribas *et al.*^[5] described a 14-year child with primary congenital glaucoma with high IOP developing acute middle macular retinopathy, and controlling the IOP alone resulted in resolution of macular retinopathy. Pedruzzi *et al.*^[8] in 2018 reported a glaucoma patient developing peripapillary retinoschisis with NSD following a non-penetrating deep sclerectomy, where the patient presented with ocular hypotony in the initial postoperative days. Two of our patients showed a decrease in retinal and macular schisis after controlling IOP alone without any vitreoretinal intervention.

The third mechanism being the role of vitreous traction associated with the tiny holes in the neuroectoderm tissue of ONH, similar to that described in patients with optic disc pits.^[9] Small changes in axial length following IOP fluctuations can lead to vitreous traction in the inner retina resulting in schisis formation. This hypothesis was very well supported by previously published reports by Mavrikakis *et al.*,^[10] Moreno *et al.*,^[11] Takashina *et al.*,^[12] and Zumbro *et al.*, where PPV with gas tamponade resulted in resolution of schisis in patients with glaucoma-associated schisis without any optic

disc pits. [Table 2] All our five patients had no optic disc pits, which was confirmed by OCT through the disc area. One of our patients underwent pars plana vitrectomy for retinal schisis, followed by trabeculectomy, which ultimately resulted in a FTMH.

The role of IOP in causing peripapillary and macular retinoschisis may indicate an ongoing glaucoma progression as this entity has been reported in a patient with normal-tension glaucoma also.^[13] This hypothesis could be assumed to be the underlying pathogenesis of our fourth patient developing peripapillary and macular retinoschisis despite well-controlled IOP. Moreover, he was a chronic glaucoma patient on AGM for more than eight years and chronicity of glaucoma with advanced cupping may be an inciting factor.

A large case series (372 patients) by Lee *et al.*^[14] reported peripapillary retinoschisis owing to glaucomatous ONH structural changes without any detectable optic disc pit, optic nerve coloboma, or high myopia. In this observational study, OCT analysis of both the RNFL and macula was performed in 372 patients, of which 25 areas of retinoschisis were identified in 22 patients. Proteomic analysis revealed that the source of fluid filling the schisis cavity in a case of advanced glaucoma was most likely vitreous humor.^[15] Bayraktar *et al.*^[16] compared the RNFL measurements and OCT B-scans of 940 glaucoma patients with 801 glaucoma suspects and concluded that increased RNFL thickness measurements were observed at the time of retinoschisis. In addition, resolution of the retinoschisis can affect the presentation of glaucoma stability

Table 2: Case studies of macular schisis and retinoschisis associated with glaucomatous cupping (As reported in English literature)

Case reports	No of patients	Age	Sex	Type of glaucoma	Cup disc ratio	Investigation	Treatment
Hollander <i>et al.</i> ^[3] (2005)	1	54	M	Repeated episodes of angle-closure glaucoma	0.9	OCT-area of retinoschisis continuous with the ON + NSD extending through the macula	Control of IOP by YAG PI + AGM
Kahook <i>et al.</i> ^[2] (2007)	2	48 64	F F	PACG	N/D	OCT- multiple peripapillary schisis cavities extending into the macula	IOP control (AGM)
Zumbro <i>et al.</i> ^[19] (2007)	5	14 62 73 63 65	F M M F M	JOAG POAG POAG Glau. Suspect Traumatic glaucoma	1 0.6 0.8 Large cup 1	All 5 had enlarged cupping with macular schisis without any vitreous traction	1 patient - trabeculectomy 2 patients -PPV
Mavrikakis <i>et al.</i> ^[10] (2011)	1	65	F	Chronic glaucoma	0.7	OCT- macular schisis with macular detachment	PPV + gas tamponade
Moreno <i>et al.</i> ^[11] (2012)	1	64	F	Chronic glaucoma	0.8	OCT- communication between enlarged optic disc cup with macular retinoschisis and neurosensory detachment at presentation	PPV + gas tamponade0
Takashina <i>et al.</i> ^[12] (2013)	1	78	M	POAG	0.9	OCT- membrane tissue on the optic disc and a tunnellike hyporeflective lesion connecting the schisis cavity and a site near the tissue, but no obvious optic disc pit. Membrane tissue with a sheet-like appearance on the optic nerve head	PPV + membrane removal on the disc and ILM
Inonue <i>et al.</i> ^[23] (2015)	11	60-81	3 F 8M	Chronic glaucoma	>0.7	All 11 patients had macular schisis and glaucomatous optic neuropathy	All 11 patients underwent PPV for macular schisis and glaucomatous optic neuropathy
Oztaş <i>et al.</i> ^[4] (2017)	1	30	M	PACG	0.9-1.0	3D SD-OCT - retinoschisis in the papillomacular and macular areas and focal defects of at the outer ON margins in prelaminar and laminar regions of the ONH	Thermal laser photocoagulation to the peripapillary areas
Peduzzi <i>et al.</i> ^[8] (2018)	1	46	M	Juvenile glaucoma	0.9	Post NPDS -patient developed peripapillary schisis and NSD	IOP control
Aribas <i>et al.</i> ^[5] (2020)	1	14	F	Primary congenital glaucoma	N/D	Paracentral acute middle maculopathy	IOP control

or progression.^[17] Later, Brazerol *et al.*^[18] studied the macular OCT scans of 218 glaucomatous eyes and concluded that glaucomatous damage leads to gradual thickening of the inner nuclear layer, known as microcystic macular edema and in more severe glaucoma cases leads to retrograde maculopathy. All our patients described here had diffuse peripapillary RNFL thinning.

The treatment modality of this distinct entity remains inconclusive. Spontaneous resolution of schisis has also been reported, which makes observation a reasonable option as was observed in the two patients.^[1] Öztas *et al.*^[4] performed thermal laser photocoagulation adjacent to the peripapillary retinoschisis area in a 30-year male with advanced PACG to contain the macular schisis. Few studies have demonstrated that a prolonged increase in IOP may lead to breaks in the

inner limiting membrane with penetration of the vitreous into the retrolaminar space and controlling IOP remains the primary treatment modality.^[13,14,19] Few studies presumed the presence of lamina cribrosa defects in patients with macular schisis, where fluid from the subarachnoid space may also be responsible.^[20-22]

Although the underlying pathophysiology may not be completely understood, it is important to be aware of this atypical finding when managing glaucoma patients with peripapillary and macular retinoschisis because treatment options may differ from other causes of secondary retinoschisis. In the presented case series, the first patient underwent micropulse G6 and the second patient underwent partial diode CPC, whereas the third patient underwent glaucoma filtration surgery only as a treatment modality for glaucoma-associated

schisis. The fourth patient was only observed whereas the last patient underwent PPV and trabeculectomy for the treatment of macular retinoschisis.

Macular retinoschisis. Treatment modality was different in our case series; this may be because of the different clinical modalities of presentation, different treating physicians, and absence of a specific prescribed modality of treatment due to the rarity of the disease. We suggest the underlying pathogenesis as the flow of vitreous fluid into the retina via a microhole in the thin tissue of the advanced excavated glaucomatous disc triggered by fluctuations in IOP or vitreoretinal traction.

Outcomes

Two patients had decreased retinal schisis and macular schisis while the schisis remained stable in two patients, one patient developed a FTMH during follow-up visits.

Limitation

As it is a retrospective study, all five patients had different treatment modalities for their IOP control. Follow-up periods were also different for each patient. The reason for normal macula in two patients who had advanced glaucoma with high IOP in the fellow eye could not be explained.

Conclusion

Macular and peripapillary retinoschisis associated with advanced glaucomatous cupping without optic disc cavitory lesions is a less reported clinical entity and it signifies an ongoing glaucoma progression. Both primary and secondary glaucomas with advanced glaucomatous cupping can be associated with this rare entity. Detailed and careful evaluation of macula of patients with advanced disc cupping can help in timely intervention and prognostication. Though control of IOP and vitreoretinal intervention in selected cases are the described management modalities, there is no generalized consensus in the management of this clinical entity.

Acknowledgements

Dr. Krishin Krishnakumar, MS- Retina fellow, Aravind eye hospital, Madurai.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

- Farjad H, Besada E, Frauens BJ. Peripapillary schisis with serous detachment in advanced glaucoma. *Optom Vis Sci* 2010;87:E205-17. doi: 10.1097/OPX.0b013e3181d1dad5.
- 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;143:697-9.
- Hollander DA, Barricks ME, Duncan JL, Irvine AR. Macular schisis detachment associated with angle-closure glaucoma. *Arch Ophthalmol* 2005;123:270-2.
- Öztas Z, Mentis J, Ates H, Nalçacı S. Three-dimensional optical coherence tomography imaging and treatment of glaucomatous optic nerve head defects associated with schisis-like maculopathy. *Turk J Ophthalmol* 2017;47:119-22.
- Aribas YK, Aktas Z, Bayrakceken K, Atalay T, Ozdek S. Paracentral acute middle maculopathy in primary congenital glaucoma. *Retin Cases Brief Rep* 2020;14:163-5.
- Nagesha CK, Ganne P. Occult optic disc pit maculopathy in a glaucomatous disc. *Middle East Afr J Ophthalmol* 2017;24:165-6.
- Van der Schoot J, Vermeer KA, Lemij HG. Transient peripapillary retinoschisis in glaucomatous eyes. *J Ophthalmol* 2017;2017:1536030.
- Pedruzzi NG, Ghirelli W, Cvintal V. Peripapillary retinoschisis and serous detachment of neurosensory retina after a non-penetrating deep sclerectomy. *Rev Bras Oftalmol* 2018;77:356-9.
- Brown GC, Shields JA, Goldberg RE. Congenital pits of the optic nerve head. II. Clinical studies in humans. *Ophthalmology* 1980;87:51-65.
- Mavrikakis E, Lam WC. Macular schisis and detachment secondary to large optic nerve head cup: a newly recognized syndrome amenable to vitrectomy. *Acta Ophthalmol* 2011;89:95-6.
- Moreno-López M, González-López JJ, Jarrín E, Bertrand J. Retinoschisis and macular detachment associated with acquired enlarged optic disc cup. *Clin Ophthalmol*. 2012;6:433-6.
- Takashina S, Saito W, Noda K, Katai M, Ishida S. Membrane tissue on the optic disc may cause macular schisis associated with a glaucomatous optic disc without optic disc pits. *Clin Ophthalmol* 2013;7:883-7.
- Zhao M, Li X. Macular retinoschisis associated with normal tension glaucoma. *Graefes Arch Clin Exp Ophthalmol* 2011;249:1255-8.
- Lee EJ, Kim TW, Kim M, Choi YJ. Peripapillary retinoschisis in glaucomatous eyes. *PLoS One* 2014;9:e90129. doi: 10.1371/journal.pone.0090129.
- 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;134:108-10.
- Bayraktar S, Cebeci Z, Kabaalioglu M, Ciloglu S, Kir N, Izgi B. Peripapillary retinoschisis in glaucoma patients. *J Ophthalmol* 2016;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;59:2818-27.
- Brazerol J, Iliev ME, Höhn R, Fränkl S, Grabe H, Abegg M. Retrograde maculopathy in patients with glaucoma. *J Glaucoma* 2017;26:423-9.
- 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;144:70-4.
- 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;123:2066-76.
- Kiumehr S, Park SC, Syril D, Teng CC, Tello C, Liebmann JM, *et al.* *In vivo* evaluation of focal lamina cribrosa defects in glaucoma. *Arch Ophthalmol* 2012;130:552-9.
- Park SC, De Moraes CG, Teng CC, Tello C, Liebmann JM, Ritch R. Enhanced depth imaging optical coherence tomography of deep optic nerve complex structures in glaucoma. *Ophthalmology* 2012;119:3-9.
- Inoue M, Itoh Y, Rii T, Kita Y, Hirota K, Kunita D, Hirakata A. Macular retinoschisis associated with glaucomatous optic neuropathy in eyes with normal intraocular pressure. *Graefes Arch Clin Exp Ophthalmol*. 2015;253:1447-56.