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Acquired Retinoschisis Resolved after 23Gage Pars Plana Vitrectomy in Posterior Microphthalmos

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Abstract

Background: Posterior microphthalmos combined with acquired retinoschisis is rare in clinic. Only three similar cases in nanophthalmos were reported in 2009. This report presents a case of acquired retinoschisis in a patient with posterior microphthalmos and discusses the proper treatment for such disease. The patient exhibited the acquired peripheral retinal schisis in both eyes. The schisis in right eye was limited in peripheral retina and the schisis in the left eye presented with a bullous-like type. The schisis was formed from peripheral retina extending to the posterior macular region and nearly contact to the lens in the left eye. The treatment for patients under this condition was very challenging.

Case presentation: The patient presented with a fix scotoma and visual acuity decrease for 2 weeks in his left eye. Ocular examination found that his best corrected visual acuity were 0.6 in his right eye and 0.2 in left eye. He has amblyopia because of hyperopia with spherical +11.75 diopters in the right eye and+12.00 diopters in the left. Slip lamp examination found normal anterior segments. The total axial lengths were 18.41mm in right and 18.43mm in left, relatively. Funduscopy showed bilateral retinoschisis in inferotemporal retina. The schisis present a limited form in peripheral retina in the right eye and a bullous type in the
left eye. The schisis was from peripheral extended to the posterior macular region in left eye and without resolved during 2 weeks’ follow up. The management was to observe the condition changes in the right eye and to perform pars plana vitrectomy on the left eye. Visual acuity restored to 0.6 in left eye and kept stable in right eye after one-year follow up.

**Conclusion:** Posterior microphthalmos combined with retinoschisis is rare. When it appears in peripheral retina, the schisis remains stable. While the schisis extends to posterior pole and affects macula, surgery could be the option.

**Key Words:** microphthalmos, acquired retinoschisis, pars plana vitrectomy

**Background**

Posterior microphthalmos (PM) is a developmental defect in which affected eyes display vitreous chamber foreshortening, normal or nearly normal anterior chamber depth, and papillomacular retinal folds\(^1\). PM is relatively rare in clinic. It usually coexists with high hyperopia, glaucoma, uveal effusion syndrome, and exudative retinal detachment\(^2-3\). PM combined with foveoschisis has been reported in some cases\(^4-5\) and has been proved to be related to gene mutation\(^4,6\). Only three similar cases in nanophthalmos were reported in 2009\(^7\), in which the schisis limited in peripheral retina without treatment. In our case, it presents PM but not
nanophthalmos. And the schisis in one eye presents a bullous type, which extended from peripheral retina to posterior region. The following reports the bullous acquired retinoschisis in a posterior microphthalmos and the management.

**Case presentation**

A 35 year-old Chinese man complained of a fix scotoma and visual acuity decreased for 2 weeks in his left eye. He received amblyopia therapy when he was 4, because of amblyopia resulted from high hyperopia. There was no related family history. The best correct visual acuity (BCVA) was 0.6 in his right eye (OD) and 0.2 in left eye (OS) (spherical equivalent +11.75D OD, +12.00D OS). The corneal diameter was 10.5mm in both eyes (OU), anterior chamber depths were 2.87mm OD and 2.81mm OS (Figure1D), total axial lengths were 18.41mm OD and 18.43mm OS, relatively. The diagnosis of posterior microphthalmos was clear. The morphology of schisis in two eyes varied. While it was limited in right eye (Figure1B), it presented as bullous, smooth elevation with thin surface in the left eye (Figure1C). And it didn’t move under position changing. The bulla in left eye elevated to the posterior surface of the lens (Figure1A) and compressed the macular region resulting in macular folds and edema. Ultrasound biomicroscopy revealed peripheral cystic degeneration OU, without ciliary body or choroid detachment. Fundus fluorescence angiography (FFA) showed peripheral vessels
leakage OU, without background fluorescence changes. Spectralis optical coherence tomography (OCT) scan showed a thick retina neurosensory layer and a thick choroid layer (533 μm) in right eye(Figure1F). In left eye, the schisis was too high for us to get the image and compressed the macular region resulting in macular folds and edema(Figure1G).

In order to exclude inflammation causing retinoschisis, short term systemic corticosteroid was used. Methylprednisolone started from 1.6mg/kg/day for 3 days, reduced to 0.8mg/kg/day for 3 days then 0.4mg/kg/day for 5 days, then discontinued. The bulla showed no change. Considering recently rapid visual acuity decrease, we performed 23 Gage pars plana vitrectomy for him. Before surgery, a carefully scleral depression examination was performed, and there was no outer or inner layer breaks of retina. Triamcinolone acetonide associated posterior vitreous detachment was performed. After an internal drainage hole was made, the bulla collapsed. This was followed by photocoagulation in schisis region under the perfluorocarbon liquid then with C3F8 tamponade.

During follow up, the macular edema and folds disappeared. And the BCVA restored to 0.5 one month after surgery. BCVA stayed at 0.5 at 3 months after surgery, and increased to 0.6 at one year after surgery. The bulla collapsed and macular region was flat after surgery(Figure 2C) but the retinoschisis cavity in inferotemporal retina still remained(Figure 2A,
The OCT at one month after surgery proved that the retinoschisis was in inner nuclear layer (Figure 2D). OCT scans showed some schisis-like changes at inner and outer nuclear layers during the follow-up (Figure 2E, 2F). Retinoschisis in right eye remained stable during follow-up.

**Conclusion**

Foveoschisis in nanothalmols or in PM has been reported. While previous reports present a foveoschisis always combined with RPE abnormalities, or papillomacular folds caused by microphthalmos\(^{[1,4]}\). The case presented here is different from previous reports. There were no signs of RPE abnormalities, foveoschisis, and macular folds. The schisis appears in peripheral retina and presents as a bullous type. Similar retinal findings have been described in association with nanophthalmos by Dhrami-Gavazi E et al. in 2009\(^{[7]}\). However, in his three cases, there was no schisis involved the posterior region of retina, the schisis was just localized in peripheral. Paplliomacular folds are common in posterior nanophthalmos because of scleral growth independent of neuroretinal growth\(^{[1-3]}\). But in this case, macular folds in left eye were compressed by the bulla, which might be interpreted as a non-congenital sign. The macular was normal in the fellow eye.

As all reports mentioned, uveal effusion syndrome (UEF) and glaucoma were very common in nanophthalmos. But there was no
symptom of UEF in this case, such as ciliary body detachment, leopard-spot in FFA, and retinal detachment. The mechanism of retinoschisis is unclear. The nanophthalmos have thick sclera, uvea, and retina. We presumed that these thicken tissue may contribute to the irregular outflow of the eye, which causes the cystic degeneration of the retina.

Although there have been some reports about the MFRP gene mutation correlated with the nanophthalmos\cite{6,9}, in this case, there was no family history. His mother had used some medicine unknown to treat arthritis when pregnant, which might be a major doubting point.

The management was very challenging. According to our long term observation of X-linked retinoschisis, 82% (9 out of 11 eyes) the schisis or retinal detachment progressed and visual acuity decreased during the mean 34.7 month’s follow up.\cite{8} In vitrectomy group (n=17 eyes), foveoschisis resolved in all eyes and 94% (16 out of 17 eyes) retinal reattached and the visual acuity regained. Vitrectomy may be an effective and essential treatment for patients with progressive X-linked retinoschisis to prevent a deterioration of vision. Though the mechanism is different between X-linked retinoschisis and acquired retinoschisis, the schisis in our case extended to posterior retina and visual acuity decreased, so the surgery was performed. The outcome after surgery proved that vitrectomy is effective for preventing the schisis progression.
and the visual acuity decrease.

In conclusion, in this rare case, if schisis is limited in peripheral retina, it may stay stable. While it progressed to the posterior pole, vitrectomy may prevent the progression of acquired retinoschisis.

**Content**

Written informed consent was obtained from the patient for publication of this Case report and any accompanying images. A copy of the written consent is available for review by the Editor of this journal.

**List of abbreviations**

PM  posterior microphthalmos
OD  right eye
OS  left eye
OU  both eyes
BCVA  best correct visual acuity
FFA  fluorescence angiography
UEF  uveal effusion syndrome
OCT  Optical coherence tomography

**Competing interests**

All authors declare that they have no financial or non-financial competing interests.
Authors’ contributions
The work presented here was carried out in collaboration between all authors. YSS was the major contributors in writing the manuscript. GY and HYS were the operator. LXL was the academic advisor. All authors read and approved the final manuscript.

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**Figure legends**

**Figure1 Baseline statue of the patient.**

1A: ASP OS: smooth thinner schisis retina nearly contact to the len
1B: FP OD: Retinoschisis limited in inferotemporal peripheral retina of right eye.
1C: FP OS: Retinoschisis presented a bullous, smooth, thinner surface in left eye.

1D: UBM OS: showed normal anterior camber.

1F: OCT OD: normal fundus with a thick neurosensory retina and a thick choroid (533 μm)

1G: OCT OS: macular folds caused by the elevated schisis compression.

ASP: anterior segment photography; FP: fundus photography; UBM: ultrasound biomicroscopy; OCT: Optical coherence tomography

**Figure 2** The fundus photography and OCT follow up post-operation.

2A: one-month after the bullous collapsed after surgery, the schisis cavity still remained in inferotemporal retina.

2B: FP OS: one-year follow up. the bulla collapsed, laser spot in schisis region.

2C: OCT OS: one-month follow up: macular region was flat.

2D: OCT OS: one-month follow up: schisis cavity was in inner nuclear layer in inferotemporal retina.

2E: OCT OS: three-month follow up: macular region was flat, schisis-like changes was at inner nuclear layer of retina.

2F: OCT OS one-year follow up: macular region was flat, schisis-like
changes was at inner and outer nuclear layer of retina.