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Pre- and postoperative OCT features and surgical outcomes of advanced retinitis pigmentosa with macular hole: case series and literature review

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Abstract

Background Macular hole (MH) is a rare complication of retinitis pigmentosa (RP) and has an adverse impact on residual visual function. At present, the underlying mechanisms are not fully understood and surgical experience is limited.

Methods We reviewed the medical records and optical coherence tomography (OCT) scans in a cohort of eight eyes of seven RP patients with MH in order to report their OCT features and vitreoretinal surgical prognosis.

Results This study includes four lamellar macular holes (LMHs) and four full-thickness macular holes (FTMHs). Pre-operative OCT revealed other macular abnormalities in all eyes, such as epiretinal membrane (ERM), cystoid macular edema (CME), lamellar hole-associated epiretinal proliferation (LHEP) and vitreoretinal traction. MH progression and subjective vision worsening were noted in one LMH eye during a seven-month follow-up. All holes closed after vitrectomy with internal limiting membrane (ILM) peeling. At final follow-up, one eye had improved vision and seven eyes remained stable compared to baseline.

Conclusions The occurrence of MH in RP is accompanied by various imaging characteristics, such as ERM, CME and LHEP, suggesting a multifactorial pathogenesis. Considering poor vision in most RP patients with potentially progressive MH, surgery appears to be effective in maintaining or improving the central vision in a period of time. Thus, vitrectomy should be performed as soon as possible and flap-assisted techniques or episcleral surgeries are needed for some special cases.

Keywords Macular hole, Retinitis pigmentosa, Optical coherence tomography, Pars plana vitrectomy

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Introduction

Retinitis pigmentosa (RP) encompasses a set of complex hereditary retinal diseases characterized by progressive degeneration of rod and cone photoreceptors. The worldwide prevalence of RP is about 1 in 4000 with well over 100 genes implicated so far [1, 2]. The clinical symptoms of RP manifest a classical order, usually night blindness is the first symptom, followed by the progressive loss of the peripheral visual field [3]. In the end stage, only a small residual central island of the visual field may remain,



which results in severely constricted vision known clinically as ‘tunnel vision’ [4]. However, macular abnormalities are relatively frequent among RP patients, leading to additional visual disturbances, such as macular atrophy, choroidal neovascularization, cystoid macular edema (CME) and vitreomacular interface disorders. Compared to others, macular hole (MH) is a rare complication of RP with a reported prevalence of 0.5–4.5% [5, 6]. Earlier studies have shown several clinical findings indicating the possible pathogenesis of RP with MH [7, 8]. More clinical features of these eyes remain to be seen, and high-resolution spectral-domain optical coherence tomography (SD-OCT) can help us understand them better.

Vitrectomy has rarely been reported for RP patients with MH, although it is widely used for MH repair in general patients. Due to the limited experience, it is important to study the outcomes and prognosis of vitreoretinal surgical treatment. Thus, our study aimed to describe the clinical features and surgical intervention outcomes of RP patients with MH.

Methods

Our retrospective review enrolled the records of seven RP patients with MH treated by vitrectomy. This study was followed the principles outlined in the Declaration of Helsinki. Approval was obtained by the Medical Ethics Committee of Qilu Hospital of Shandong University, and informed consent was obtained from all patients. Macular hole surgery was performed in eight eyes by two experienced surgeons using a standard 23- or 25-gauge three-port pars plana vitrectomy (PPV). Complete removal of the posterior hyaloid was performed in all eyes with or without the use of triamcinolone acetonide. Then about 0.2 mL ICG-diluted solution (0.25%) was gently injected into the vitreous cavity and immediately washed out with balanced solution, and the stained ILM was removed with a forceps. Atypical epiretinal proliferation, also referred to as lamellar hole-associated epiretinal proliferation (LHEP), was identified as a sticky epiretinal membrane with yellowish pigment. LHEP was also carefully removed while the parts tightly attached to the edge of the hole margin was left behind to prevent unnecessary loss of normal tissue. Two eyes experienced free ILM flap insertion into the macular hole. All surgeries ended with air-fluid exchange or silicone oil tamponade. If necessary, a concomitant cataract operation was performed.

Results

Our study included seven patients (male: 2, female:5), the mean age was 49.3 ± 13.7 years, and one patient had a bilateral presentation. All patients complained of a recent reduction in visual acuity in their affected eyes, with or

without metamorphopsia. Preoperative best-corrected visual acuity (BCVA) ranged from hand motion vision to 20/50. With dilated funduscopy, all eyes presented typical signs of RP, including peripheral retinal atrophy with bone spicule-shaped pigmentation in the mid-periphery, waxy pallor of the optic nerve head and attenuation of retinal vessels.

By SD-OCT, four eyes were diagnosed with lamellar macular hole (LMH) and four with full-thickness macular hole (FTMH) (Fig. 1 and Table 1). Cases No. 1 and 7 had the presence of a foveal bump (Fig. 1a, g), and they had other similar features such as atypical epiretinal proliferation and outer retinal layer defection, both more severe in the temporal retina. Only two eyes (cases No. 2 and 3) showed posterior vitreous detachment (PVD), of which one showed incomplete PVD with apparent vitreoretinal traction (Fig. 1b) and the other one was high myopia exhibiting macular retinal detachment with complete PVD, posterior staphyloma and retinoschisis (Fig. 1c). Interestingly, they were the only two eyes who showed no atypical epiretinal proliferation. And unlike the small interretinal cystoid spaces in others, case No.2 presented a larger extent of a CME. We suspect that the formation of large cystoid spaces are related to vitreoretinal traction, and atypical epiretinal proliferation tends not to be present when mechanical retinal traction plays a dominant role in MH formation. Cases No. 4, 5 and 6 showed similar OCT images, especially atypical epiretinal proliferation at the hole margin. However, compared to case No. 4 with small FTMH and case No. 5 with LMH, case No. 6 had the largest size of the macular hole 720 μ m in diameter. To some extent, these may indicate different periods of MH development and inform us that this disease is often progressive rather than dormant.

Besides, all eyes showed the atrophy of the outer retinal layers and the macula was usually steep, especially in myopic cases (Fig. 1a, c and d). Epiretinal membrane (ERM) was noted in six eyes, although not significantly. CME was noted in five eyes. Contrary to previous reports [9], most cystoid spaces were present in the outer nuclear layer. In fact, the distribution of CME and LHEP was asymmetric in the temporal and nasal retina, and there was a corresponding relationship between the two in location (Fig. 2a, b).

In the fellow eyes of the patients with unilateral lesions, thinning and disappearing of integrity of the outer retinal layer were also observed. All eyes showed relatively good central vision (20/50 to 20/20) except one high myopic eye whose BCVA was counting fingers. One eye showed incomplete PVD (Fig. 2c). None of the eyes showed CME or LHEP. In one eye whose BCVA was 20/20, OCT demonstrated serious atrophy of the outer retinal layers and a small part of the ellipsoid zone preserved only in the

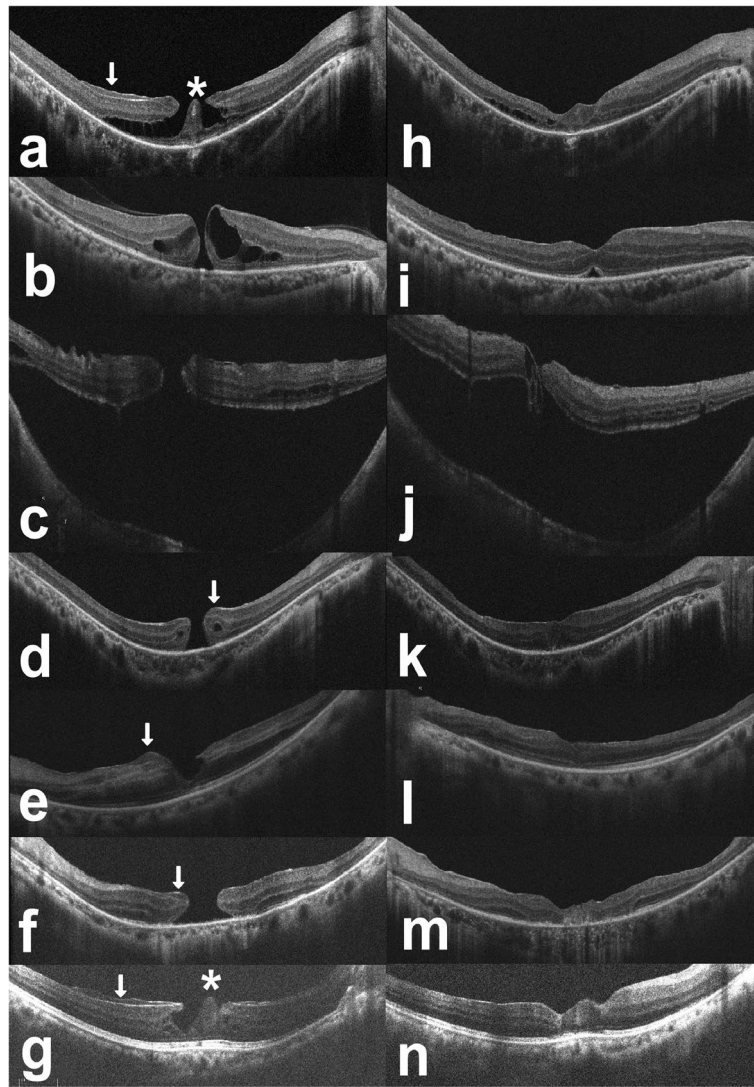


Fig. 1 Images **a-g** and **h-n** respectively corresponded to the pre- and post-operative images of seven patients in Table 1, excluding the right eye of case No. 6. **a-g** The pre-operative horizontal OCT images of seven surgical eyes. Notice the epiretinal proliferation (arrow) that isoreflective and conformed to the retinal surface. The asterisk pointed to a presence of foveal bump. **h-n** The post-operative horizontal OCT images showed the MHs were closed

fovea (Fig. 2d). This may indicate that the presence of MH is a huge blow to the residual central vision of RP patients.

Three patients with LMHs (cases No. 1, 5 and 7) and two patients with FTMHs (cases No. 2 and 4) underwent vitrectomy with ILM peeling and air tamponade. Phacoemulsification and posterior chamber intraocular lens implantation were simultaneously performed in cases No. 5 and 7 for cortical cataract and posterior subcapsular cataract, respectively. These patients were advised to maintain the prone position for 3 to 5 days after surgery. Post-operative follow-up showed successful closure of all holes (Fig. 1).

Considering macular hole-related retinal detachment in case No. 3 with high myopia, he accepted vitrectomy, phacoemulsification, and ILM peeling. After covering the base of the hole with the residual part of the trimmed ILM, oil tamponade was conducted. One week later, OCT showed a failure of macular retinal reattachment, and ILM tissue inserted in the hole was observed (Fig. 1j). At 3-month postsurgery, the MH closed while the retina remained detached, despite decreased macular thickness. Due to the poor anatomical and functional results, silicone oil has not been removed so far.

In the left eye of case No. 6 with cortical cataract and a large hole, vitrectomy was performed with concomitant

Table 1 Patient demographics, clinical characteristics and clinical course

| Case no. | Gender/age | Eye | BCVA | | AL (mm) | Refraction (D) | Size of the hole | CME | ERM | LHEP | Operation |
|----------|------------|-----|--------|-----------------------|---------|----------------|------------------|-----|-----|------|---|
| | | | Pre | Post (Follow-up time) | | | | | | | |
| 1 | F/33 | OD | 20/133 | 20/133(3 months) | NA | -4.25 | NA | + | - | + | PPV,ILMP, air tamponade |
| 2 | F/46 | OD | 20/50 | 20/50(3 months) | 23.19 | NA | 119 | + | + | - | PPV,ILMP, air tamponade |
| 3 | M/53 | OS | FC | FC(12 months) | 28.26 | -11.0 | 274 | - | + | - | PHACO, PPV,ILMP, ILM transplantation,silicon oil injection |
| 4 | F/31 | OD | 20/50 | 20/40(6 months) | 26.31 | -7.25 | 365 | + | + | + | PPV,ILMP, air tamponade |
| 5 | M/59 | OS | HM | HM(3 months) | 23.29 | -1.75 | NA | - | + | + | PHACO + IOL, PPV, ILMP, air tamponade |
| 6 | F/69 | OD | 20/80 | 20/80(3 months) | 22.80 | +1.50 | 546 | + | + | + | PHACO + IOL, PPV, ILMP, air tamponade |
| | | OS | FC | 20/800(10 months) | 22.72 | +1.25 | 720 | - | + | + | PHACO, PPV,ILMP, ILM transplantation,silicon oil injection, oil removal and IOL implantation three months later |
| 7 | F/54 | OD | 20/400 | 20/400(3 months) | 22.67 | NA | NA | + | - | + | PHACO + IOL, PPV, ILMP, air tamponade |

BCVA best-corrected visual acuity, NA not applicable. AL axial length, CME cystoid macular edema. ERM epiretinal membrane. LHEP lamellar hole-associated epiretinal proliferation, F female, M male, PPV pars plana vitrectomy, ILMP inner limiting membrane peeling, PHACO + IOL phacoemulsification with intraocular lens, HM hand motion, FC finger counting

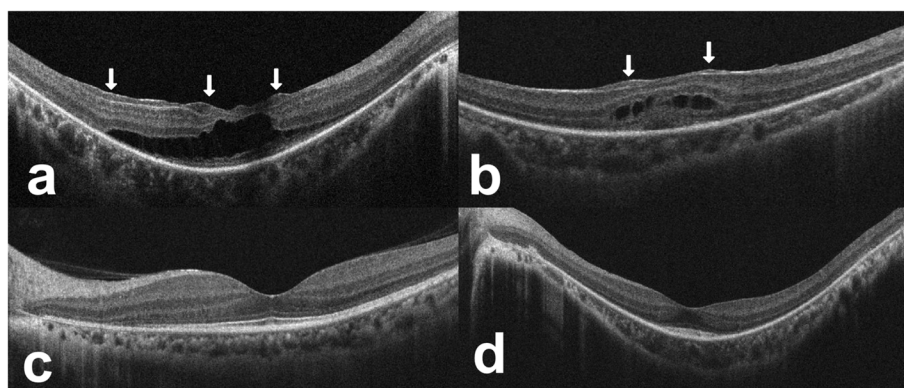


Fig. 2 **a, b** In the vertical direction, the presence of LHEP was accompanied by CME (cases No. 1 and 4). **c** OCT image of case No. 2 showed an incomplete PVD. **d** OCT image of case No. 4 showed wide defect of ellipsoid zone

removal of the cataract. Similarly, the trimmed ILM flap was used for inserting the hole and then silicone oil was injected. Ten days after surgery, OCT confirmed successful closure of the hole (Fig. 1m). Silicone oil removal as well as posterior chamber intraocular lens implantation were performed three months after vitrectomy.

For the right eye of case No.6 whose BCVA was 20/80, we initially did not intervene and kept her under regular follow-up. During the seven-month follow-up period, she reported marked subjective visual deterioration, although her BCVA remained unchanged. Of note, the hole showed a clear tendency to progress to full thickness compared to the baseline (Fig. 3). This spontaneous conversion was accompanied by the exacerbation of existing CME, ERM as well as LHEP. Progressive defection of the ellipsoidal zone was also obvious in this process. Finally, the patient underwent the same surgical protocol as cases No. 5 and 7 for LMH repair, which resulted in a good outcome without vision worsening.

At final follow-up (3–12 months postoperatively), one eye had improved vision and seven eyes remained stable compared to the baseline. Almost all patients perceived improvement in visual function. Postoperative OCT showed CME resolved significantly with reduced central retinal thickness. However, outer retinal atrophic changes were detected in all eyes except cases No. 2 and 3, especially the destruction and deletion of the ellipsoidal zone.

Discussion

Currently, the exact etiology behind MH formation in RP remains unclear and a multifactorial pathogenesis is widely proposed. Vitreoretinal interface changes are considered as a major mechanism. Principally, degenerative vitreous with the collapse of the vitreous gel and posterior vitreous detachment may lead to vitreomacular interface changes, which are likely to contribute to the

formation of CME [6]. Moreover, both the collapse of the blood-retinal barrier and dysfunction of Müller cells can result in RPE pumping failure, which may eventually provoke intraretinal fluid accumulation and CME development [10]. As a result, rupture and posterior fusion of the cysts may facilitate MH formation [11]. Besides, previous studies showed high myopia and posterior staphyloma can occur in RP eyes [12]. Myopic traction maculopathy, characterized by retinoschisis, macular hole, and foveal retinal detachment, typically contributes to the development of myopic MH [13, 14]. LHEP is an epiretinal tissue with homogenous moderate reflectivity filling the space between the epiretinal membrane and the retinal nerve fiber layer on OCT [15]. It is possible that LHEP without exerting tractional effects, originates from the middle retinal layer because it contains retinal glial cells, specifically Müller cells [16, 17]. The presence of LHEP is said to be associated with more severe macular lesions and outer retinal disruption [18]. LMH eyes with LHEP can occasionally progress to FTMH, and these patients tend to have a failure of FTMH closure [19, 20]. However, the role of LHEP in MH development is poorly understood.

Pars plana vitrectomy (PPV) with ILM peeling stands as a valuable treatment option in the general population with MH [21], but its application in RP remains controversial. To date, only small sample-sized works have been published, and we summarized eighteen previous clinical applications [6, 8, 22–37]. There were a total of 33 patients and the mean age was 46.72 ± 13.50 years (Table 2). Preoperative OCT showed that more than 60% of cases had significant vitreoretinal interface diseases, such as ERM (14%), CME (17.1%), PVD (17.1%), LHEP (14%). The median postoperative follow-up was 6 months (range 1 month to 12 years). Postoperative OCT showed central retinal thickness decreased in eyes

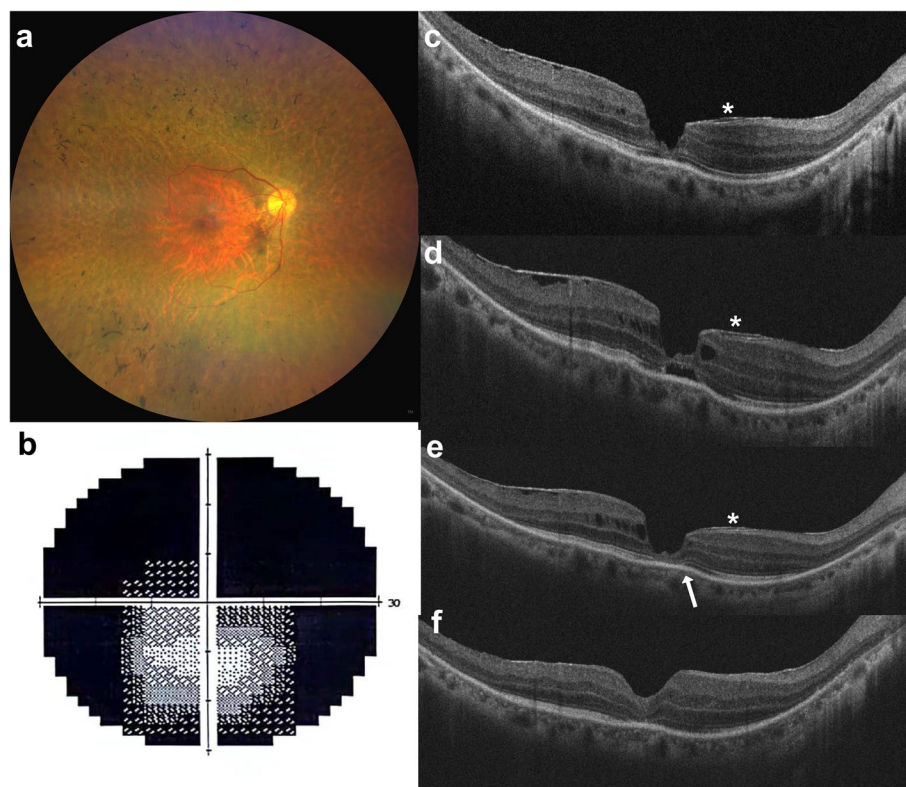


Fig. 3 **a** Colour fundus photograph at baseline revealed classical triad of waxy disc pallor, attenuated vessels and bony spicules. **b** Automated 30–2 Humphrey visual field testing at baseline (mean deviation, -25.56 decibels [dB]). **c** SD-OCT at baseline showed a LMH with inner retinal layer disruption, ERM, mild LHEP (asterisk) and CME. **d** SD-OCT obtained three months later showed a bridge-like structure at macula fovea composed of outer nuclear layer and a small part of photoreceptor inner segments with wide deletion of both inner and outer retinal layers. **e** SD-OCT obtained seven months later showed the bridge-like structure collapsed and filled in the bottom of the macular hole (arrow). **f** At 2-week post-surgery, SD-OCT confirmed a successful closure of the macular hole, with CME resolved

with CME. Among the 35 operated eyes, 22(91.4%) eyes had complete closure of the hole, 2 (5.7%) eyes failed to close the hole, and 1(2.9%) eye reopened 2 years later. Of note, five cases with retinal detachment were involved, of which four were accompanied by high myopia. All of them resulted in MH closed with vision improvement. At the last follow-up, 24(68.6%) cases had improved vision, 4(11.4%) remained unchanged, and 7(20%) worsened. In those cases with reduced vision, 3 eyes had progressive severe retinal atrophy, 2 eyes kept MH opened, and 2 had high myopia or foveal detachment.

Previous surgical treatments featured classic ILM peeling and gas or silicon oil tamponade, and flap-assisted techniques were adopted in about a quarter of cases, such as inverted ILM flap or ILM free flap transplantation, LHEP transplantation or autologous lens capsular flap transplantation. About half of the eyes became pseudophakia eventually and the average age was 47.6(28 to 67) years. Two eyes experienced more than one time of PPV and both eventually had worsening vision. In our series, cases with LMH or small FTHM were treated by

PPV with conventional ILM peeling and air tamponade. For FTHM eyes with larger hole size or macular hole-related retinal detachment, ILM free flap transplantation was performed to help bridge the hole, fill the gap, act as a scaffold and modulate Müller cell migration and proliferation [38]. Finally, all holes achieved anatomic successful closure after the primary repair. Considering that multiple vitreoretinal surgeries might not be tolerated by RP patients, selecting the best surgical option plays a crucial role in increasing the success rate of primary repair of MH. There are no clear standards for selecting which tamponade agent. The surgeon should take into account multiple factors, such as the number of surgeries, the success rate, the patient's vision needs during the recovery period and so on.

Almost all operated eyes followed for more than 2 years showed retinal thickness decreased and varying degrees of retinal atrophy, of which half were accompanied by progressive vision loss [24, 29, 30]. Despite the short follow-up period, this phenomenon was also obvious in our series. On the basis of retinal atrophy at the macula

Table 2 Case review of surgical treatment and postoperative outcome of RP with MH

| No | Study | Gender/age | MH stage | Associated disease | Operation | BCVA | | Follow-up time | Postoperative findings |
|----|------------------------------------|------------|-----------|--|---|---------------------|---------------------|----------------|---|
| | | | | | | Pre | Post | | |
| 1 | Lee et al., 2021 [6] | M/46 | FTMH (OD) | Retinoschisis, ERM, PPS, nuclear cataract | PHACO + IOL, PPV, ERM/ILM peeling, inverted ERM/ILM flap technique, C3F8 tamponade | 20/400 | 20/100 | 3 months | 2 MHs closed |
| | | | FTMH (OS) | ERM, PPS, nuclear cataract | PHACO + IOL, PPV, ERM/ILM peeling, anterior lens capsular flap technique, C ₃ F ₈ tamponade | 20/400 | 20/125 | 1 month | |
| 2 | Arora et al., 2021 [22] | F/60 | FTMH (OD) | PPS, foveal detachment, high myopia | PPV, ILMP, inverse ILM flap, C ₃ F ₈ tamponade, macular buckling | 2/60 | 20/40 | 2 months | MH closed, posterior pole flattened |
| 3 | Panagiotou et al., 2020 [23] | F/28 | FTMH (OD) | PVD, CME | PPV, ILMP, C ₃ F ₈ tamponade; PHACO + IOL six months after PPV for PCO | 2.5/10 | 2.5/10 | 11 months | MH closed, CRT decreased |
| 4 | Gen et al., 2020 [24] | M/36 | FTMH (OS) | NA | PPV, ILMP, SF ₆ tamponade | 20/50 | 20/25 | 3 years | MH closed, macular atrophy |
| | | | | | | | 20/40 | 9 years | |
| | | | | | | | 20/250 | 12 years | |
| 5 | Kim et al., 2019 [25] | M/55 | FTMH (OS) | Vitreomacular traction, CME, Pseudophakia | PPV, ILMP, C ₃ F ₈ tamponade | 20/67 | 20/32 | 3 months | MH closed (postoperative 6 years) |
| 6 | Enani et al., 2017 [26] | F/44 | FTMH (OD) | PCO, CME | PPV, ILMP, C ₃ F ₈ tamponade | 20/400 | 20/400 | 6 months | MH closed |
| 7 | Rush et al. 2016 [27] | F/66 | LMH (OD) | Pseudophakia, ERM | PPV, ILMP, F ₆ S tamponade | 20/60 ² | 20/30 ⁻¹ | 6 months | 2 MHs closed |
| | | | LMH (OS) | Pseudophakia, ERM | | 20/70 ⁻¹ | 20/40 ⁻¹ | 6 months | |
| 8 | Vingolo et al., 2015 [28] | M/34 | FTMH (OD) | PCO | PHACO + IOL, PPV, ILMP, | 6/20 | 10/20 | 3 years | 3 MHs closed, CRT decreased |
| | | M/49 | FTMH (OS) | Nuclear and posterior subcapsular cataract | silicon oil injection, silicon oil removal. 6 later | 8/20 | 14/20 | | |
| | | F/37 | FTMH (OD) | PCO | | 2/20 | 4/20 | | |
| 9 | Dhanashree et al., 2013 [29] | F/55 | FTMH (OD) | CME | PPV, ILMP, C ₃ F ₈ tamponade | 20/800 | 20/200 | 2 months | MH closed |
| | | M/54 | FTMH (OS) | Incomplete PVD | PPV, ILMP, C ₃ F ₈ tamponade | 20/80 | 20/120 | 2 months | MH kept unclosed |
| | | M/32 | FTMH (OD) | High myopia, incomplete PVD | PPV, ILMP, C ₃ F ₈ tamponade; PHACO + IOL one year later | 20/120 | 20/200 | 2 months | MH closed |
| | | | | | | 20/2000 | 20/2000 | 5.5 years | |
| 10 | García-Fernández et al., 2013 [30] | F/64 | FTMH (OS) | Subretinal fluid, PVD | PPV, ILMP, F ₆ S tamponade; PHACO + IOL six months later for PCO | 20/200 | 20/50 | 2 months | MH closed, severe macular atrophy |
| | | | | | | 20/200 | 20/200 | 6 months | MH closed |
| | | | | | | 20/200 | 20/200 | 2 years | MH reopened, sub-retinal fluid, intense retinal atrophy |

Table 2 (continued)

| No | Study | Gender/age | MH stage | Associated disease | Operation | BCVA | | Follow-up time | Postoperative findings |
|----|------------------------------|------------|---------------|--|---|------------------------|-------------|----------------|--|
| | | | | | | Pre | Post | | |
| 11 | Hagiwara et al., 2011 [31] | M/55 | FTMH | Cortical cataract | PHACO + IOL, PPV, ILMP, SF ₆ tamponade; repeated PPV | 20/200 | 20/333 | NA | MH closed, RPE atrophy |
| | | M/36 | FTMH | Cortical cataract | PHACO + IOL, PPV, ILMP, SF ₆ tamponade | 20/50 | 20/29 | NA | MH closed |
| 12 | Jin et al., 2008 [32] | M/19 | FTMH (OS) | PVD | PPV, ILMP, C ₃ F ₈ tamponade | 20/100 | 20/67 | 1 month | 4 MHs closed, CRT decreased |
| | | M/56 | MH (OD) | High myopia, retinal detachment, proliferative vitreoretinopathy | | 20/1000 | 20/667 | NA | |
| | | F/69 | FTMH (OD) | PVD | | 20/286 | 20/50 | 6 years | |
| | | F/36 | FTMH (OS) | CME | | 20/400 | 20/67 | NA | |
| 13 | Nakamura et al., 2005 [33] | F/49 | FTMH (OD) | ERM | PPV, ILMP | 20/200 | 20/667 | 14 months | MH closed, RPE atrophy |
| 14 | Artemiya et al., 2002 [34] | M/24 | FTMH | LHEP | PPV, ILMP, gas tamponade | 20/100 | 20/29 | NA | MH closed |
| 15 | SAPERSTEIN et al., 2001 [35] | F/63 | FTMH (OS) | Mild nuclear sclerotic cataract, subretinal fluid | PPV, ILMP, C ₃ F ₈ tamponade | 20/200 | 20/30 | 21 months | MH closed, subretinal fluid resolved |
| 16 | Hirokazu et al., 2001 [36] | M/41 | FTMH (OS) | Pseudophakia, high myopia, retinal detachment, PPS, vitreoretinal adhesion | PPV, ILMP, F ₆ S tamponade | 12/200 | 60/200 | 6 months | Retina attached, MH closed |
| 17 | Liu et al., 2020 [8] | M/34 | FTMH (OS) | LHEP, macular detachment, high myopia | PPV, ILMP, LHEP transplantation, C ₃ F ₈ tamponade | HM | FC | 20 months | Retina attached, MH closed |
| | | F/46 | FTMH/LMH (OD) | HELP, foveal detachment | PHACO + IOL, PPV, ILMP, LHEP transplantation, C ₃ F ₈ tamponade | 20/200 | 20/250 | 6 months | Fovea attached, MH closed |
| | | F/45 | LMH (OS) | LHEP, retinoschisis, PPS | PPV, ILMP, LHEP transplantation, C ₃ F ₈ tamponade | HM | FC | 6 months | Retina attached, MH closed |
| | | F/55 | LMH (OS) | Pseudophakia, PCO, LHEP, CME | PPV, ILMP, LHEP transplantation, C ₃ F ₈ tamponade, posterior capsulotomy | HM | FC | 6 months | MH closed, CRT decreased |
| | | M/67 | LMH (OS) | LHEP | PHACO + IOL, PPV, ILMP, LHEP transplantation, C ₃ F ₈ tamponade | 20/200 | 20/40 | 2 months | MH closed |
| 18 | Yan et al., 2018 [37] | NA | 4FTMH | NA | PPV, ILMP, SF ₆ or C ₃ F ₈ or silicone oil tamponade | 2 improved, 1 worsened | 1 unchanged | ≥ 9 months | 3 MHs closed, 1 kept unclosed after 3 times of PPV |

Abbreviation: F female, M male, BCVA best-corrected visual acuity, PPV pars plana vitrectomy, FTMH full-thickness macular hole, LMH lamellar macular hole, PHACO + IOL phacoemulsification with intraocular lens, ERM epiretinal membrane, ILMP inner limiting membrane peeling, PPS posterior pole staphyloma, CRT central retinal thickness, CME cystoid macular edema, PVD posterior vitreous detachment, NA not applicable, LHEP lamellar hole-associated epiretinal proliferation, PCO posterior capsular opacification, HM hand motion, FC finger counting

caused by long-term damage to the photoreceptor or retinal pigment epithelial loss, further retinal thinning after vitrectomy appears to be the main reason for vision deterioration and hole reopening [30]. These atrophic changes may also be secondary to mechanical insult to the retina, phototoxicity by endoillumination, or injury to the retinal pigment epithelium by indocyanine green [31, 39]. Given the natural progression of RP, it is difficult to ascertain the role of surgical trauma in postoperative retinal atrophy. However, from the current perspective, surgery is not without merit, particularly for patients with good vision and early stage of MH. It may be more meaningful to extend patient follow-up and evaluate the stabilization time of patients' postoperative vision, rather than just the improvement of postoperative vision.

Sieving and Fishman reported a significant myopic shift of the mean spherical refractive error of RP patients from the mean of a normal population [40]. A study of 365 RP patients in Koreans reported that the prevalence of high myopia was up to 16.1% (40 of 249 eyes) [41]. Moreover, Komori and coworkers reported a high incidence of steep macular curvature in non-highly myopic RP eyes, and the steepness was affected by the degree of photoreceptor degeneration [42]. Although the role of high myopia in RP remains to be elucidated, retinal degeneration and visual acuity deterioration are more apparent in patients with high myopia [43]. The occurrence of myopic traction maculopathy further complicates the RP patient's condition. Because of the poor retinal adhesion to the underlying surface and the posterior staphyloma that may produce an inverse traction to impede retinal adhesion, the surgical repair is challenging [44, 45]. Retinal traction cannot be completely eliminated by means of vitreoretinal surgery alone since retinal stretching caused by posterior staphyloma and other components are still present [46]. Just like our high myopic patient with an axial length of 28.2 mm, the retina failed to attach in the short term, despite silicon oil injection. In contrast, Arora et al. combined macular buckling and vitrectomy to treat a myopic macular hole foveal detachment associated with posterior pole staphyloma in a RP eye, which resulted in MH closed with posterior pole flattening [22]. Therefore, to flatten the concavity of the posterior part of the eye and release the inverse traction caused by the posterior staphyloma, episcleral surgeries such as posterior scleral reinforcement and macular buckle are necessary [47].

Spontaneous closure of MH in three eyes was once described [8, 48]. In other cases, MH tends to progress to a larger lesion in the degree and depth of retinal disruption. Based on previous studies, surgical treatment is usually considered beneficial and a successful closure of the hole is achieved in the majority,

notwithstanding the long-standing retinal dysfunction as well as the potential risks of vitrectomy. Thus, surgery is worth attempting in order to remain or improve the central vision in some cases, although the ultimate prognosis of these patients may still be poor. We believe that the criteria for assessing the feasibility of surgery should not be limited to improved vision post-operatively, and that stopping the vision loss caused by MH is equally relevant. For the patients themselves, if early surgery allows them to maintain relatively good visual acuity for a certain period of time, this is a good enough indication that they can benefit from the surgery. Several times surgeries are unadvisable, which would lead to undesirable consequences, such as visual acuity worsening and severe retinal atrophy. Careful consideration before vitrectomy is important, and screening RP with OCT is highly recommended to follow-up the patients, evaluate the natural history of the disease as well as identify those patients who could benefit from PPV.

In summary, the occurrence of MH in RP is accompanied by various macular abnormalities, suggesting a multifactorial pathogenesis. In our cases, microincision vitrectomy surgery for RP with MH appears to be effective and safe for helping stabilize the remnant vision for a period of time. Therefore, screening for MH in RP patients is vital, and surgical management should be selected as soon as possible if the hole progresses. However, not all surgeons are suitable to try this treatment as it requires extensive experience and subtle manipulation, sometimes referrals are advisable. Surgeons must be very cautious to decide whether or not to indicate surgery for such cases, due to the potential risks that might outweigh the potential benefits for these already poor prognosis patients.

Abbreviations

| | |
|--------|---|
| MH | Macular hole |
| RP | Retinitis pigmentosa |
| OCT | Optical coherence tomography |
| LMH | Lamellar macular hole |
| FTMH | Full-thickness macular hole |
| PPV | Pars plana vitrectomy |
| ILM | Internal limiting membrane |
| LHEP | Lamellar hole-associated epiretinal proliferation |
| SD-OCT | Spectral-domain optical coherence tomography |
| PVD | Posterior vitreous detachment |
| CME | Cystoid macular edema |
| ERM | Epiretinal membrane |
| BCVA | Best corrected visual acuity |

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Authors' contributions

SYL and SYY concept and design, made the interpretation of the information, and drafted the initial manuscript. WSH and XMH carried out the collection of the reported studies. ZYT, ZJ and CY reviewed and revised the manuscript. All authors read and approved the final manuscript.

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Availability of data and materials

The datasets used and/or analysed during the current study are available from the corresponding author on reasonable request.

Data availability

No datasets were generated or analysed during the current study.

Declarations**Ethics approval and consent to participate**

This article has been reviewed and approved by the Medical Ethics Committee of Qilu Hospital of Shandong University. It adheres to the tenets of the declaration of Helsinki. Written informed consent has been provided by all patient and confidentiality of the patients identity is maintained as much as possible.

Consent for publication

Written informed consent was obtained from the patients in this case series for publication of this article and any accompanying images. A copy of the written consent is available for review by the Editor of this journal.

Competing interests

The authors declare no competing interests.

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