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Internal limiting membrane packing for treatment of morning glory syndrome with rhegmatogenous retinal detachment

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ABSTRACT

Purpose: This study aimed to report the ocular features, surgical methods, and surgical outcomes of a patient with morning glory syndrome(MGS) complicated with rhegmatogenous retinal detachment(RRD)

Observations: The patient was a 38-year-old Chinese woman with congenital cataract in her left eye and an artificial eye in her right eye. Ocular examination confirmed MGS complicated with RD in the left eye and revealed a retinal hole in the temporal margin of the optic disc. The retina successfully reattached after pars plana vitrectomy(PPV), silicone oil tamponade and laser photocoagulation, but the hole did not close and occurred obvious contractile movement. The retina did not detach again during the follow-up period. One and a half years later, silicone oil removal combined with internal limiting membrane and hyaloid or glial remnant plugging, autologous blood covering and C3F8 filling were performed, and the retinal hole was finally closed.

Conclusions and importance: This case is the first to report contractile movement of the retinal hole in a patient with MGS complicated with RD, and the hole was closed by internal limiting membrane tamponade combined with autologous blood coverage.

1. Introduction

Morning Glory Syndrome (MGS) is a rare congenital disorder with abnormal optic disc development, also known as Morning Glory Disc Anomaly (MGDA). As early as 1970, Kindler noticed the similarities between this congenital optic disc deformity and morning glory, and officially named it.[1] The typical features of MGS are a deep and large funnel-shaped excavation involving the optic disc, with peripapillary chorioretinal pigmentation, a clump of glial tissue mass covering the center of the optic disc, and straight radial retinal vessels at the edge of the optic cup. These characteristics make MGS look like a blooming morning glory.[2] Although binocular MGS lesions have also been documented,[3] MGS usually involves only one eye, and there is no significant difference in the right or left eye involvement rate. At present, the pathogenesis of MGS is not clear, which may be caused by abnormal development of the neuroectoderm, or it is believed that primary mesenchymal development may lead to incomplete closure of the posterior sclera wall and incomplete development of the sieve plate.[4]

The visual acuity of MGS patients varies from person to person, and the prognosis is often associated with macular involvement, retinal detachment (RD) and the development of amblyopia.[3] The most serious complication of MGS is RD, which can be seen in up to one-third of the patients. At present, the pathogenesis of RD in MGS is unknown, three mechanisms have been proposed including rhegmatogenous, tractional, and exudative. MGS-associated RD usually requires multiple operations to achieve reattachment, and the prognosis of visual acuity is often poor even after anatomic reattachment. Although pars plana vitrectomy (PPV)and laser photocoagulation have been attempted by many scholars, there is no standard treatment for MGS combined with RD.[5]

MGS is usually non-contractile (NCMGS), or in rare cases, it can be associated with constriction movements of the optic disc (CMGS). Wise et al. reported optic disc contraction movements in the case of peripapillary staphyloma for the first time.[6] This is an extremely rare phenomenon, and there are two main theories about this phenomenon: one is pressure balance, and the other is muscle contraction.

In this report, we describe a case of MGS-associated RD with an artificial eye on the contralateral eye. The detached retina successfully repaired after complicated PPV and silicone oil tamponade, however the retinal hole contracted significantly after the surgery. Then we successfully closed the hole by internal limiting membrane (ILM) flap and hyaloid or glial remnant to plug the hole, laser photocoagulation and C3F8 filling.

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2. Case report

The patient is a 38-year-old Chinese woman. Due to the ripple change of vision in the left eye accompanied by vision decline for more than 1 month, she came to our hospital for treatment. Her

left eye had poor vision since childhood and had a history of amblyopia. The best corrected visual acuity (BCVA) was 0.5. The right eye is the artificial eye. There was no history or family history of strabismus, ocular trauma, congenital glaucoma, etc. At the first visit, BCVA was 0.12, intraocular pressure(IOP)was normal, and opacity in the central area of posterior capsule of lens. Fundoscopy examination after dilation showed the grey-white retinal eminence centered on the optic disc at 5:00–9:00. The optic disc was pale, large and excavated, with a central turf of glial tissue and peripapillary chorioretinal pigmentation. The blood vessels were straight and fanned out from the disc in a radial pattern. There was also a hole in the temporal margin (Fig. 1). Optical coherence tomography (OCT) demonstrates RD (Fig. 2). The patient was treated with 25G PPV, laser photocoagulation, and silicone oil filling under local anesthesia on September 29, 2019. During the operation, triamcinolone acetonide was used to assist the posterior vitreous detachment(PVD). The vitreous was cut as much as possible, and the subretinal fluid was drained during the gas-liquid exchange. Due to the strong adhesion between vitreous and macular area, a small hole was formed in the retina below the nose during vitrectomy, which was photocoagulated. Finally silicone oil was filled (Video 1).

Supplementary video related to this article can be found at https://doi.org/10.1016/j.ajoc.2022.101454

In the early postoperative period, BCVA improved to 0.2, most retinal was reattached in the fundus. The temporal disc hole was not closed and it contracted by showing the curved direction of the blood vessel (Fig. 3). OCT showed that the neuroepithelial layer on the temporal side of the optic disc was discontinuous, suggesting that the retinal hole was not closed. Subretinal space under the hole is present (Fig. 4). In addition, we observed the contraction and expansion of the hole under the slit lamp, which had no obvious correlation with respiration (Video 2). Retinal detachment did not occur again during follow-up, but the hole and the subretinal space under the hole still existed. BCVA remained 0.25.

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One and a half years after surgery, the patient complained of blurred vision in her left eye, BCVA was 0.15, IOP was normal, the central area of posterior capsule of lens was cloudy, vitreous cavity was filled with silicone oil, retina was attached, and the temporal margin of optic disc was the same as before. Then we performed 23G vitrectomy and silicone

Fig. 1. Fundus photography of the left eye:It can be seen that the left eye's grey-white retinal eminence centered on the optic disc at 5:00–9:00. The optic disc is large and excavated, with a central turf of glial tissue and peripapillary chorioretinal pigmentation, the peripheral blood vessels of the optic disc are radial, and a hole can be seen on the temporal edge (arrow).

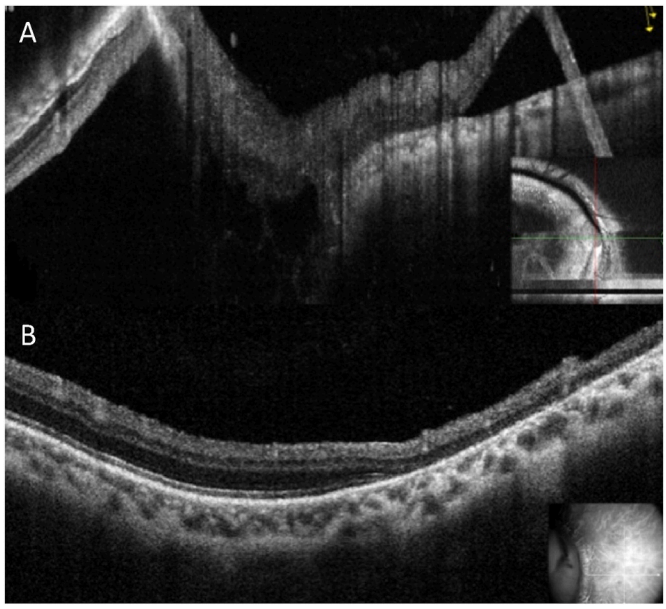

Fig. 2. A: OCT showed separation of the retinal neuroepithelial layer and retinal pigment epithelium layer on the temporal side of the optic disc of the left eye, suggesting retinal detachment light bands; B: OCT showed the retinal neuroepithelium and retinal pigment epithelium layer are not separated in the macular area of the left eye.

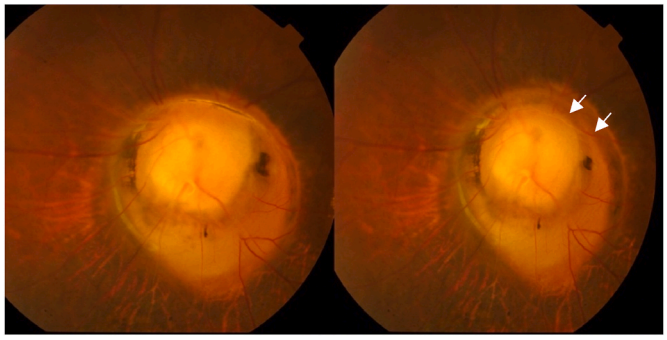

Fig. 3. Fundus photography of the left eye underwent 25G PPV with silicone oil tamponade:the retina is attached. The hole at the edge of the optic disc is not closed, and it contracted and expanded. The blood vessel pointed by the arrow contracted towards the hole.

Fig. 4. OCT of the left eye underwent 25G PPV with silicone oil tamponade: showed the neuroepithelial layer on the temporal side of the optic disc was discontinuous, suggesting that the retinal hole was not closed. Subretinal space under the hole is present.

oil extraction, combined with phacoemulsification and IOL implantation under local anesthesia on March 9, 2021. After intraoperative triamcinolone acetonide injection, traction fibroglial tissue was observed on the surface of optic disc, which was slowly removed. Then, after indocyanine green staining, we removed part of the ILM along the temporal disk margin and plugged the hole with the ILM flap and hyaloid or glial remnant. Laser photocoagulation was then applied to the hole. Use a syringe without anticoagulant to draw a small amount (about 0.3ml) of the patient's autologous blood and drop a few drops (about 0.1ml) into the hole to seal and fix the flap. Finally, C3F8 gas was filled (Video 3). In the early postoperative period, the patient's BCVA was 0.12, the intraocular pressure was normal, fundus photography and OCT showed that the retina was attached, the temporal hole of the optic disc was closed and no obvious contractile movement was observed. (Figs. 5–7). However, the patient complained that the upper visual field was blocked, and the microperimetry showed that the average retinal sensitivity (RS) was low(8.8), and the upper RS was significantly lower than the lower RS. The fixation stability was unstable (Fig. 8A). The last follow-up microperimeter showed that the overall average RS has increase(11.35), and the fixation stability was relatively stable (Fig. 8B).

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3. Discussion

To the best of our knowledge, this is the first report of contractile movements of a retinal hole in a patient with MGS-related RD and successfully closed with inner limiting membrane and hyaloid or glial remnant. Perhaps our surgical technique of the complete closure of peripapillary hole can provide a new way of thinking for other operators.

At present, the pathogenesis of RD in MGS is unknown, three mechanisms have been proposed including rhegmatogenous, tractional, and exudative. Haik et al. reported the natural course of 30 MGS patients with an average follow-up time of 10.3 years. Eleven of them had MGS-related RD.These RDs have different clinical processes, including spontaneous reattachment and eve detachment.[7] It has been confirmed that the subretinal space and the vitreous cavity are connected at the optic disc, and there is abnormal communication between the subarachnoid space and the subretinal space. Therefore, some scholars believe that the RD is exudative and is related to optic disc defects.[8] Another theory believes that the pulling of vitreous and fibroglial tissue is the main reason for MGS related RD. Lytvynchuk supported the viewpoint by using intraoperative OCT,

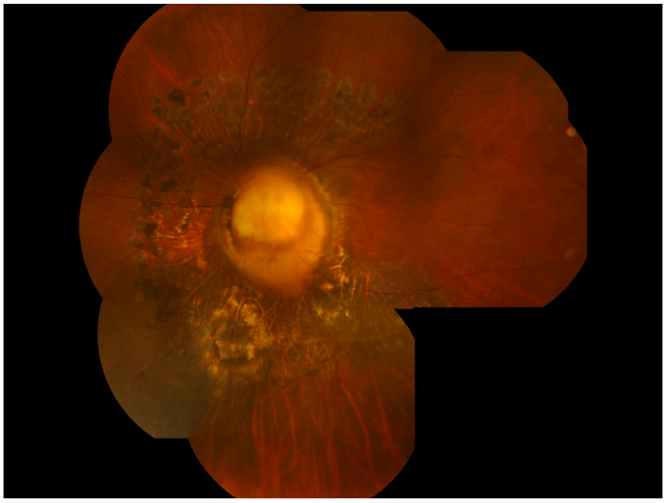

Fig. 5. Fundus photography of the left eye underwent 23G PPV, silicone oil removal, ILM packing and C3F8 gas filling:the retina was flat, the glial tissue was reduced, and the hole at the edge of the optic disc was closed.

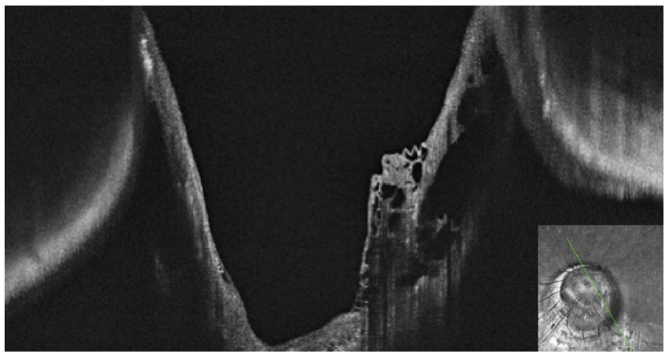

Fig. 6. OCT of the left eye underwent 23G PPV, silicone oil removal, ILM packing and C3F8 gas filling:the original temporal optic disc hole is filled with loose tissue (ILM), and the discontinuous retinal neuroepithelial layer is connected by ILM, suggesting that the retinal hole is closed.. The subretinal space of the hole still exists.

which showed that the vitreous adhered strongly to the macular area, and the fibroglial tissue around the optic disc drew the retina and further formed retinal breaks in the weakest area of the optic disc depression.[9]

However, retinal breaks are not found in all cases. For patients without obvious retinal breaks,

the cerebrospinal fluid from the subarachnoid space may accumulate in the subretinal space. In these

patients, the detached retina may spontaneously disappear or detach again.[7] However, if a retinal tear is formed, the long-existing hole may cause irreversible atrophic disease of the retina. The possibility of spontaneous regression of RD is extremely low, and surgical treatment is required. It is worth noting that, due to the decreased color contrast between the sclera and optic disc in MGS patients, fibroglial tissue may also block the hole, making it difficult to find it. Therefore, preoperative OCT, especially the three-dimensional remodeling scan, helps to better display the structure of the optic disc and determine whether there is fibrinoid tissue in the center of the optic disc and retinal breaks caused by fibroglial tissue or vitreous.

There is no standard treatment for MGS combined with rhegmatogenous retinal detachment(RRD). Many scholars have tried vitrectomy and retinal laser photocoagulation for treatment. Harris successfully attached the retina of a MGS related RRD patient using PPV, gas-liquid exchange, and postoperative laser.[10] However, considering that the most common location of retinal breaks is at the edge of the optic disc or in the retinal tissue within the optic disc, these breaks are often difficult to close, and there are no retinal pigment cells around the disc to absorb laser energy. So some people think that the peridiscal laser is ineffective.[11] Regarding that the contralateral eye of our patient is an artificial eye, the peripheral laser may affect the function of the optic papillary macular beam and thus affect the visual prognosis. We did not use the peridiscal laser during the first operation. Sen's 9 cases of MGS study showed that silicone oil filling can often get a better reduction effect, even if there is a risk of silicone oil emulsified particles migrating to the subarachnoid space.[2] Therefore, we chose silicone oil for the first operation, and in the second operation, in order to prevent the retina from detaching again after the silicone oil removal, we filled it with C3F8 gas.

Other references on MGS-RD suggest that removal of the fibroglial tissue that causes traction is critical for preventing recurrent RD.[12] We use triamcinolone acetonide to assist PVD because the posterior vitreous and macula are strongly adherent in younger patients. During the first PPV, despite our careful surgical procedures, a small hole was formed during the PVD process. We were concerned that stripping the glial tissue might cause further damage, so no special treatment was performed on the glial tissue in the optic disc area. Fundus photography

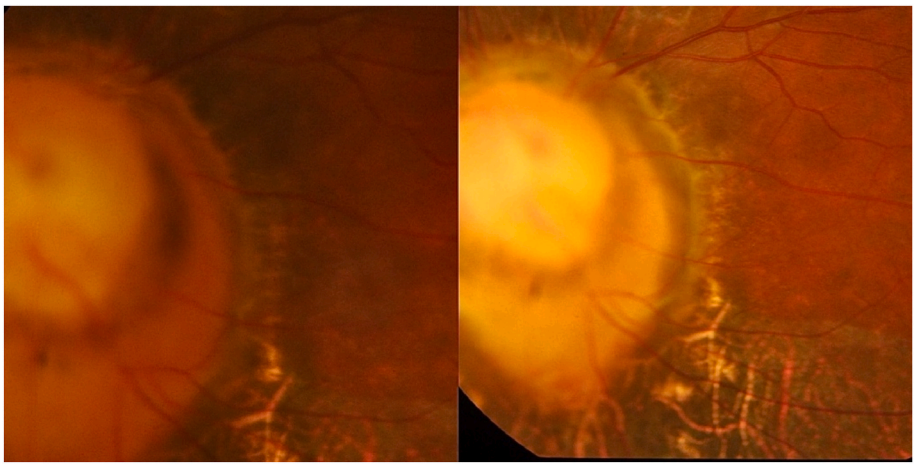

Fig. 7. Fundus photography of the left eye underwent 23G PPV, silicone oil removal, ILM packing and C3F8 gas filling:the hole at the edge of the optic disc was closed. No obvious contractile movements and vasoconstriction were observed in the fundus photography taken at two times.

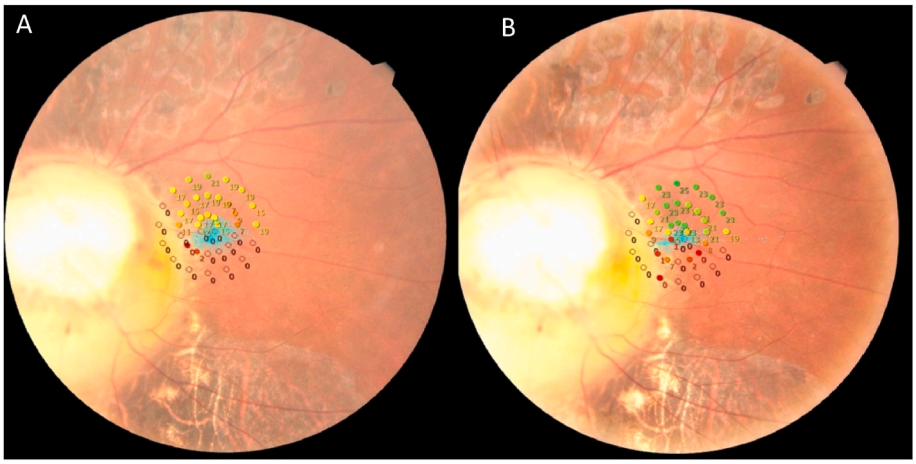

Fig. 8. (A)The early postoperative period microperimeter: the overall average retinal sensitivity (RS)was low(8.8), and the upper RS was significantly lower than the lower RS. The fixation stability was unstable(44% within 2° and 89% within 4° of the diameter circle); (B)The last follow-up microperimeter:the overall average RS increased(11.35), and the fixation stability was relatively stable(53% within 2° and 93% within 4° of the diameter circle).

after the first PPV showed that the temporal hole contracted by showing the curved direction of the blood vessel (Fig. 3), which we suspect is caused by the traction of the fibrous glial tissue around the optic disc. To prevent recurrent RD or hole enlargement, we decided to remove the glial tissue during the second PPV.

In the second operation, after removal of the glial tissue, we stripped part of the inner limiting membrane along the temporal disc rim and plugged it over the temporal retinal hole. It has been reported that ILM combined with long-standing gas tamponade can effectively treat RD associated with optic disc pits.[13] Compared with simple ILM dissection, ILM packing preserves part of the ILM flap. The ILM flap is rich in glial cell debris, which can serve as a scaffold for cell proliferation, induce the proliferation of glial cells in the retina and on the surface of the ILM, and promote hole closure.[14] In addition, the ILM flap can also act as a barrier to prevent the migration of vitreous and cerebrospinal fluid to the subretinal. However, considering that the ILM flap is usually thin and the hole is relatively large, it is difficult to ensure that the ILM flap remains in place if residual subretinal fluid spurts onto the hole. Moreover, the dynamic contraction motion of the hole weakens the sealing ability of the simple ILM flap. Due to the strong adhesion of glial tissue, we tried to plug the hole with the ILM flap and hyaloid or glial remnant. Mohammed suggested that the ILM flap should be fixed with heavy fluid before the gas-liquid exchange, but the process of removing the heavy fluid would make the operation more difficult, and the residual heavy fluid may have toxic effects on the retina.[15] Therefore, we also used autologous blood to fix the ILM flap according to our experience in macular hole surgery. Nada et al. have reported successful treatment of optic disc pits-related macular detachment using autologous blood.[16] Autologous blood containing various growth factors and collagen can promote the coupling adhesion between RPE and neuroepithelial layer and promote the proliferation and migration of cells. The hole had been closed in the early postoperative period and loose ILM was seen in the hole (Fig. 6). However, studies have shown that the recovery of retinal microstructure after ILM tamponade is slower than simple ILM dissection. Iwasaki et al. believe that packing may lead to excessive cell proliferation and affect postoperative recovery.[17] This may also be the reason why the patient's BCVA after the second operation was worse than before the operation, and the patient complained of occlusion of the upper visual field. But what is exciting is that from the results of the recent follow-up, the patient's fixation stability and RS have been significantly improved. We believe that the structure and function of the retina will slowly recover over time.

Different from the general RRD, the retinal hole in this case can contract and previous reports of MGS cases are only about the contraction of the optic disc.[18,19] This is an extremely rare phenomenon, and there are two main theories about this phenomenon: one is pressure

balance, and the other is muscle contraction. The pressure balance mechanism points out that the contraction movement of the hole is related to the respiratory cycle, intraocular pressure changes, and eye movements. When the instantaneous pressure gradient changes in the subarachnoid space and the subretinal space, due to the abnormal communication between the two spaces, the fluid flows back and forth along the optic nerve, causing contraction and expansion.[14] However, in this case, there was no synchronized contraction of the hole during the forced eye closure or Valsalva maneuver.

Another hypothesis is that smooth muscle is the origin of contraction. In normal eyes, there are non-vasoconstrictive smooth muscle cells in the choroid and sclera, and abnormal smooth muscle will appear in the case of optic disc defects. Lee et al. have demonstrated the contraction movement around the edge of the optic nerve in the computer analysis in the horizontal direction, which supports this muscle contraction mechanism. They believe that more obvious contraction movement can be observed when the fibroglial tissue is removed.[20] Our patient is similar with this phenomenon, so we are more inclined to the second muscle contraction mechanism.

4. Conclusion

Usually MGS-related RD patients require multiple operations to achieve retinal reattachment. In this case, PPV combined with ILM packing, colloidal tissue removal, laser photocoagulation, and silicone oil filling successfully flatten the retina and seal the hole. It is worth noting that although the ILM packing can increase the closure rate of the hole, there may be a delay in the recovery of the retinal structure. Due to the poor contrast of the posterior pole and the presence of fibrous tissue on the surface of the optic disc, retinal breaks are difficult to find before surgery in such patients. At the same time, it also causes certain difficulties in removing the ILM during surgery, which should be paid attention to by the surgeon. In addition, we speculate that the contractile movement of the hole in MGS patients is related to the contraction of ectopic smooth muscle, but has nothing to do with the respiratory cycle and intraocular pressure changes.

Patient consent

Consent to publish this case report has been obtained from the patient(s) in writing.

Ethics approval and consent to participate

The study followed the tenets of the Declaration of Helsinki, informed consent was obtained from the patient after explanation of the nature and possible consequences of the study. The study was approved by the Ethics Review Boards of Wuxi Second People's Hospital (Jiangsu, China).

Consent to publish

All clinical data were presented after obtaining signed informed consent for their publication from the guardians of the patients.

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Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

Declaration of competing interest

The authors have no proprietary interest in any aspect of the report.

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