Total retinal detachment and contractile movement of the disc in eyes with morning glory syndrome

Masuo Sakamoto a, Kazuki Kuniyoshi a, Shion Hayashi b, Hidetoshi Yamashita b, Shunji Kusaka a, b

a Department of Ophthalmology, Kindai University Faculty of Medicine, 377-2, Ohno-higashi, Osakasayama, Osaka, 589-8511, Japan
b Department of Ophthalmology, Yamagata University Faculty of Medicine, 2-2-2 lida-nishi, Yamagata, 990-9585, Japan

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ABSTRACT

Purpose: This study aimed to report clinical characteristics, intraoperative findings, and surgical outcomes of 2 eyes of 2 patients with retinal detachment (RD) associated with morning glory syndrome (MGS) and contractile movement of the disc.

Observations: Case 1 was a 3-year-old Japanese boy who presented with congenital microphthalmos in the left eye. The presence of total RD with MGS and contractile movement of the disc was confirmed by ophthalmoscopic examination. During vitrectomy, migration of perfluorocarbon liquid into the subretinal space was observed, suggesting a communication between the vitreous cavity and the subretinal space. Here, the retina was reattached briefly after the surgery; however, it detached again 2 months after surgery. Case 2 was a 3-month-old Japanese girl who was referred to our hospital for evaluation of RD and MGS in her left eye. Partial RD, MGS, and contractile movement of the disc were confirmed by ophthalmoscopic examination. The RD remained unchanged up to 18 months from the first visit, but atresia of the pupil was found 21 months after the first visit. Severe proliferative vitreoretinopathy and contractile movement of the disc were found after lensectomy. It was impossible to completely remove the membrane despite meticulous effort of peeling it off. At the 15-month follow-up, the retinal configuration improved but remained detached.

Conclusion and importance: Total RD with contractile movement of the disc associated with MGS is a rare condition that seems difficult to treat.

1. Introduction

In 1970, Kindler first described morning glory syndrome (MGS) or morning glory disc anomaly as a rare congenital optic nerve disc anomaly.1 It is characterized by a deep and enlarged excavation surrounding the optic disc resembling a flower of the morning glory, with a clump of glial tissue in the excavated disc, peripapillary pigmentation, and straightened vessels developing from the rim of the optic disc in a radial pattern.1,2 Although incomplete closure of the embryonic fissure of the globe3 or abnormal differentiation of the mesoblast4 has been suggested, the pathogenesis of MGS remains indefinable. The prevalence of MGS is reported to be 2.6/100,000,5 and the gender ratio is almost 1:1.3,6 Bilateral patients account for 0%–30% of total MGS,5–8 and approximately 25% of patients with MGS present with vascular and encephalic abnormalities.5,6 Except for PAX6 gene6 or syndrome MGS, the genetic link and causative agents have not been reported for this condition.10,11

The eyes with MGS usually exhibit poor vision ranging from hand movements to 0.1.5,7 Approximately 80% of MGS coexist with other ocular abnormalities such as cataract, corneal opacity, microphthalmos, and retinal detachment (RD),8 with RD constituting one-third of these patients.1,2,6,12–14 The precise mechanisms of RD in MGS is still uncertain, although exudation from vessels,1 vitreous traction,15 retinal break on the optic disc16–18 and migration of the cerebrospinal fluid from the subarachnoid space12,19 are suggested. No standard treatment exists for RD with MGS,20 although vitrectomy and laser photoagulation on peripapillary retina have been frequently attempted.

Before the term MGS was proposed1, the contractile movement of peripapillary staphyloma was reported by Wise et al.21 The contractile movement of the disc is another rare condition assumed to be caused by the fluctuation of venous pressure,22 forced lid closure,23 or light stimulation.21,22 However, the precise mechanism has not been determined.
To the best of our knowledge, this is the first report on RD and coexisting contractile movement of the disc associated with MGS and its surgical outcome. Here, we present 2 patients with these conditions who were treated by vitrectomy.

2. Case reports

Case 1 was a 3-year-old Japanese boy who presented with congenital microophthalmos in the left eye. His parents noticed his frequent rubbing of the left eye several months before the initial visit to our hospital. The ophthalmoscopic examination revealed total RD, deeply excavated and enlarged disc, and contractile movement of the disc in his left eye. These findings led to the diagnosis of RD associated with MGS (Fig. 1A). His right eye was normal with best-corrected visual acuity of 0.6, whereas it was hand motions in the left eye. Intraocular pressure was normal in both eyes. Magnetic resonance imaging of the eye revealed that the left eyeball was smaller than the right eyeball. A deep excavation was found around the optic disc in the left eye (Fig. 1B).

Lens-sparing vitrectomy was performed in the left eye. The contractile movement of the excavated optic nerve head was observed during surgery (Fig. 2A). The movement was approximately 0.5 cycle/second, and it was not synchronized with breathing or heart pulsation nor was it influenced by intraocular pressure or light stimulation during the surgery.

Perfluoro-n-octane (PFO) was injected onto the detached retina after the creation of posterior vitreous separation. After a while, PFO seemed to be aspirated into the excavation by the contractile movement, and droplets of PFO were observed in the subretinal space (Fig. 2B, Video 1). Observing inside the excavation was impossible because of the contractile movement and deep excavation, and hence aspiration of subretinal fluid through a possible break in the excavation was impossible. Then, the subretinal fluid was drained externally through the sclera, which made the retina mostly reattached. Because the laser scars were considered to be impossible to be produced owing to the contractile movement of the disc, laser photoocoagulation around the disc was not attempted under air irrigation. Finally, 10% perfluoropropane gas was injected into the vitreous cavity. Two months after the surgery, the retina of the left eye was detached, but the parents did not wish for further treatment.

Case 2 was 3-month-old Japanese girl who was found to present with MGS in the left eye by ophthalmic examination soon after birth. Her right eye was normal. She was referred to our clinic because she developed RD in the nasal area of the optic nerve head (Fig. 3A and B). The contractile movement of MGS was observed through ophthalmoscopy. Because the macula was still attached, we decided to follow her without any treatment. After 21 months (at 2 years of age), atresia of the pupil was found in the left eye (Fig. 4A). Total RD was suspected by ultrasonography. Vitrectomy and lensectomy were performed. After lensectomy, severe proliferative vitreoretinopathy (PVR) with closed funnel was found (Fig. 4B). Despite meticulous effort to peel them off, complete removal of the membranes was difficult. Special caution was paid not to make an iatrogenic retinal break, which probably makes RD worse and spontaneous reattachment difficult. During the surgery, the contractile movement of MGS was still observed, which was not synchronized with breathing or pulsation nor was it influenced by intraocular pressure or light stimulation. After surgery, the retinal configuration improved but the retina remained detached during the 15-month follow-up (Fig. 4C).

3. Discussion

To the best of our knowledge, this is the first report on MGS with contractile movement of the disc and RD and its surgical results. Unfortunately, surgical outcomes of our patients were poor.

Haik et al. reported the natural course of patients with MGS.12 During the mean follow-up of 10.3 years, RD occurred in 11 of 32 eyes with MGS. Of these 11 eyes, 4 eyes exhibited spontaneous reattachment. However, 2 of those 4 eyes exhibited redetachment. The contractile movement of the MGS was not reported in the study. Chang et al. reported a patient with MGS with RD without apparent retinal break. Because the RD was successfully treated by optic nerve sheath opening and external drainage of subretinal fluid, they proposed the presence of a communication between the subarachnoid and subretinal spaces. Alternatively, when a small break in the optic nerve area is present in patients with RD and MGS, vitrectomy with internal drainage of subretinal fluid through the break followed by laser photocoagulation on the peripapillary retina has been reported to be effective.15–18

In patients with RD associated with MGS without apparent retinal breaks or breaks in the disc area, subretinal fluid may be derived from the subarachnoid space, and RD in these patients may spontaneously resolve and redevelop. However, if a break is present, spontaneous resolution of RD is unlikely to occur, and surgical treatment is necessary.

In case 1, a communication between the subretinal space and the vitreous cavity was strongly suggested because of the migration of PFO injected in the vitreous cavity into the subretinal space during the surgery. Regarding the mechanism of subretinal migration of gas or silicone oil in eyes with optic disc anomalies, including MGS, Johnson et al. proposed that with the fluctuation of intracranial pressure, gas, or silicone oil may migrate from the vitreous cavity to the subretinal space through a break in the disc area. In addition, we consider that the contractile movement of the disc may have further facilitated this migration. Because achieving chorioretinal adhesion in such an eye with constant contractile movement of the disc seemed impossible, we decided not to perform laser photocoagulation on the peripapillary

![Fig. 1. Fundus photo of the left eye in case 1 showing the deeply excavated optic disc and total retinal detachment (A). Magnetic resonance image of the brain and orbits showing deep excavation in the posterior pole and microphthalmia in the left eye (B).](image-url)
At first, we did not perform surgery on both cases because of anticipated poor surgical outcome in case 1 and the attached macula at the first visit in case 2. The conditions remained unchanged for at least 18 months; however, severe PVR and atresia of the pupil were observed at 21 months after the first visit. The mechanism of sudden worsening of the conditions is unclear. Surgical treatment was not effective to reattach the retina, but the retinal configuration became better with normal intraocular pressure during the 15-month follow-up. A controversial issue regarding the treatment of RD associated with MGS is that surgery should be performed early, that is, soon after RD was found. Especially in case 2, we waited for nearly 2 years hoping for spontaneous improvement; however, severe PVR developed in the end. Earlier intervention could have been easier at least technically. However, in case 2, laser photocoagulation on the peripapillary retina was probably impossible because of the contractile movement, and surgical result could have also been poor.

During vitrectomy in case 1, we chose external drainage because we could not aspirate subretinal fluid from the disc area. We could make an intentional hole and aspirate the subretinal fluid internally, which is technically easier and faster. However, because photocoagulation on the peripapillary retina seemed impossible, accumulation of subretinal fluid from the subarachnoid space and the vitreous cavity is possible after the surgery. In that case, closing the intentional hole by laser photocoagulation would be difficult, which may make spontaneous reattachment of the retina impossible. An alternative technique is the coverage of...
inverted internal limiting membrane (ILM) flap on the disc pit, which has been reported to be effective for treating RD associated with optic disc pit. The technique may also be effective for treating RD associated with MGS; however, in our cases with the dynamic contractile movement, the ILM flap placed over the break may not stay in place.

In terms of the mechanism of contractile movement of the retina, a few hypotheses were proposed. The ectopic smooth muscle is suggested to be a cause of the contractile movement of MGS, because histological studies of eyes with MGS revealed its presence around the excavation. Wise et al. and Kral et al. reported that the contractile movement was induced by strong light stimulation not only in the affected eye but also in the healthy fellow eye. Achieving retinal reattachment in eyes with MGS, RD, and contractile movement of the disc is considered challenging.

4. Conclusion

Total RD associated with MGS and contractile movement of the disc is a rare condition that seems challenging to treat. Surgical indications for such eyes should be carefully considered.

Ethics approval and consent to participate

This research protocol was approved by the Ethics Review Boards of Kindai University Faculty of Medicine (22–132). The research protocol conformed to the tenets of the Declaration of Helsinki of the World Medical Association, and written informed consent was obtained from all guardians of the patients for the treatment and data collection after thorough oral explanation.

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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7. 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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Appendix A. Supplementary data

Supplementary data related to this article can be found at https://doi.org/10.1016/j.ajo.2020.100964.

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