

Morning Glory Syndrome: MRI and Fundus Findings—A Case Report

Bo Lai, Qiuying Chen, Shuixing Zhang*

Medical Imaging Center, The First Affiliated Hospital of Jinan University, Guangzhou, China

Email: bob120355@163.com, 13678962434@163.com, *shui7515@126.com

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Abstract

Morning glory syndrome (MGS) is a rare congenital optic disc developmental anomaly characterized by a funnel-shaped optic disc excavation, peripapillary chorioretinal pigmentary changes, glial tissue overlying the optic disc, and abnormal retinal vascular distribution. We report a case of a 19-year-old female who presented with right esotropia and decreased visual acuity since birth. Ophthalmic examination revealed severely impaired vision in the right eye and absence of grade III binocular visual function. Funduscopic examination showed an enlarged right optic disc with a central funnel-shaped excavated defect, surrounding choroidal atrophy, extensive retinal pigmentary degeneration, and abnormal retinal vessels radiating from the central optic disc tissue toward the peripheral retina. Magnetic resonance imaging demonstrated microphthalmia of the right eye, a round fluid-signal lesion along the retrobulbar course of the right optic nerve communicating with a posterior defect of the right ocular wall, and adjacent optic nerve atrophy with morphological distortion. These clinical and imaging findings supported the diagnosis of right-eye MGS with sensory esotropia. The patient underwent right medial rectus recession combined with right lateral rectus resection to improve ocular alignment and cosmetic appearance. During 6 months of follow-up, visual acuity remained stable, ocular alignment improved markedly, and no retinal detachment or other ocular complications occurred. This case highlights the value of combining funduscopic examination and MRI in the diagnosis, differential diagnosis, initial anatomical assessment, and screening for associated anomalies of MGS.

Keywords

Morning Glory Syndrome, Magnetic Resonance Imaging, Fundus Examination, Imaging Diagnosis

1. Introduction

Morning glory syndrome (MGS), also referred to as morning glory disc anomaly (MGDA), is a term used to describe a distinct congenital dysplasia of optic disc. It was first systematically characterized and named by Kindler in 1970 [1]. The anomaly is pathognomically characterized by a funnel-shaped enlargement of the optic disc, central glial hyperplasia, and a surrounding elevated ring of chorioretinal pigmentary disturbances [2], closely resembling a blooming morning glory flower. The condition is predominantly unilateral and typically diagnosed during childhood following presentations of decreased visual acuity, strabismus, or nystagmus. A population-based study involving 2.1 million children and adolescents reported an incidence of approximately 2.6 per 100,000 [3]. Although mutations in the PAX6 gene have been identified in a subset of patients with MGS and other optic nerve developmental anomalies—suggesting a potential genetic predisposition [4], its precise genetic basis and pathogenesis remain incompletely elucidated. In recent years, high-resolution magnetic resonance imaging (MRI) has gained increasing clinical recognition for delineating characteristic orbital and optic nerve structural deformities, evaluating the visual pathway, and screening for concurrent intracranial anomalies. Herein, we report a case of MGS diagnosed at our institution, highlighting its characteristic fundoscopic and MRI findings to enhance clinical recognition and management of this rare entity.

2. Clinical Data

A 19-year-old female presented to our hospital on August 25, 2025, with a 19-year history of esotropia and decreased visual acuity in the right eye. According to the patient and her family, right ocular esotropia and poor vision had been noted since birth, with no obvious improvement after rest. She denied floaters, photophobia, lacrimation, conjunctival redness, ocular pain, diplopia, abnormal head posture, or other associated symptoms. She had no history of ocular trauma or infectious disease, and no known family history of hereditary disorders.

On ophthalmic examination, presenting visual acuity with spectacles was hand motion at 50 cm in the right eye and 1.2 in the left eye. Slit-lamp biomicroscopy showed no conjunctival injection, clear corneas, quiet anterior chambers, round and reactive pupils, and clear crystalline lenses in both eyes. No keratic precipitates were observed in the left eye, in which the anterior chamber was deep and the pupil diameter was 3 mm. The fundus could not be adequately visualized through the undilated pupils in either eye. Intraocular pressure measured by non-contact tonometry was 14.8 mmHg in the right eye and 12.9 mmHg in the left eye. Ocular motility testing showed limitation of abduction (−2) and elevation (−1) in the right eye. The prism corneal light reflex test estimated 60 prism diopters (Δ) of esotropia, whereas the prism alternate cover test measured 50 Δ with spectacles and 55 Δ without spectacles. Synoptophore testing demonstrated an absence of grade III binocular visual function. On 26 August 2025, undilated refraction was recorded as plano in the right eye and −2.50/0.50 $\times$ 85° in the left eye, with a cor-

rected visual acuity of 1.0 in the left eye.

Funduscopy examination of the left eye showed no obvious abnormalities of the optic disc, retina, or macula (**Figure 1(A)**). Funduscopy examination of the right eye demonstrated an enlarged optic disc with a central funnel-shaped excavated defect. Peripapillary choroidal atrophy was observed, accompanied by extensive retinal pigmentary degeneration. The retinal vessels appeared to arise from the central tissue and radiated toward the peripheral retina. The arteriovenous ratio was approximately 2:3. The foveal reflex was visible, and no obvious retinal exudation or hemorrhage was identified (**Figures 1(B)-(D)**).

Magnetic resonance imaging of both globes and optic nerves was subsequently performed (**Figures 2(A)-(D)**). MRI demonstrated a smaller right globe. A round abnormal signal-intensity lesion was identified along the retrobulbar course of the right optic nerve, communicating with a posterior defect of the right ocular wall and forming a “gourd -shaped” configuration. The lesion measured approximately 0.7×0.8 cm. It showed isointensity on T1-weighted imaging and hyperintensity on T2-weighted imaging, with signal intensity similar to that of the aqueous humor. No obvious enhancement was observed after contrast administration. The adjacent right optic nerve was atrophic and morphologically distorted. The right crystalline lens, as well as the vitreous body and crystalline lens of the left eye, showed normal signal intensity. The globes were otherwise smooth and regular in contour, and the retrobulbar fat showed homogeneous signal intensity. Brain MRI showed no intracranial abnormalities.

Based on the characteristic funduscopy and MRI findings, the patient was diagnosed with morning glory syndrome of the right eye, sensory esotropia of the right eye, and ametropia of the left eye.

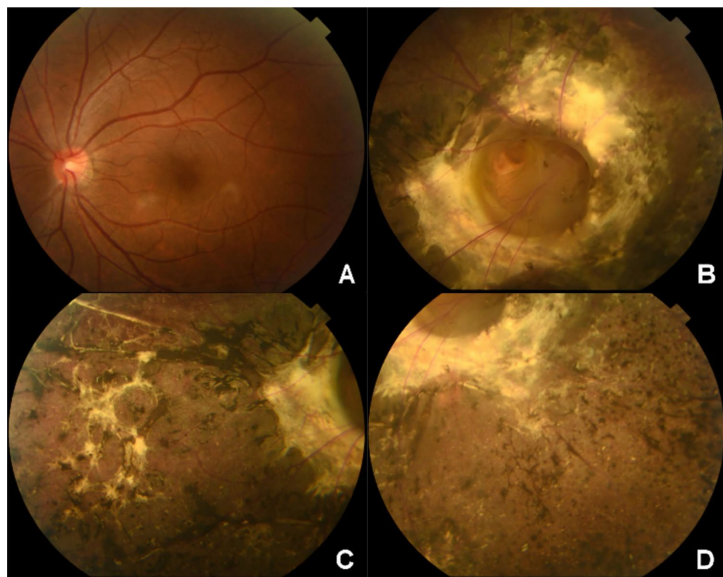


Figure 1. (A): Funduscopy appearance of the left eye. (B)-(D): Funduscopy appearance of the right eye. Enlarged optic disc, central funnel-shaped excavated defect, peripapillary choroidal atrophy and extensive retinal pigmentary degeneration.

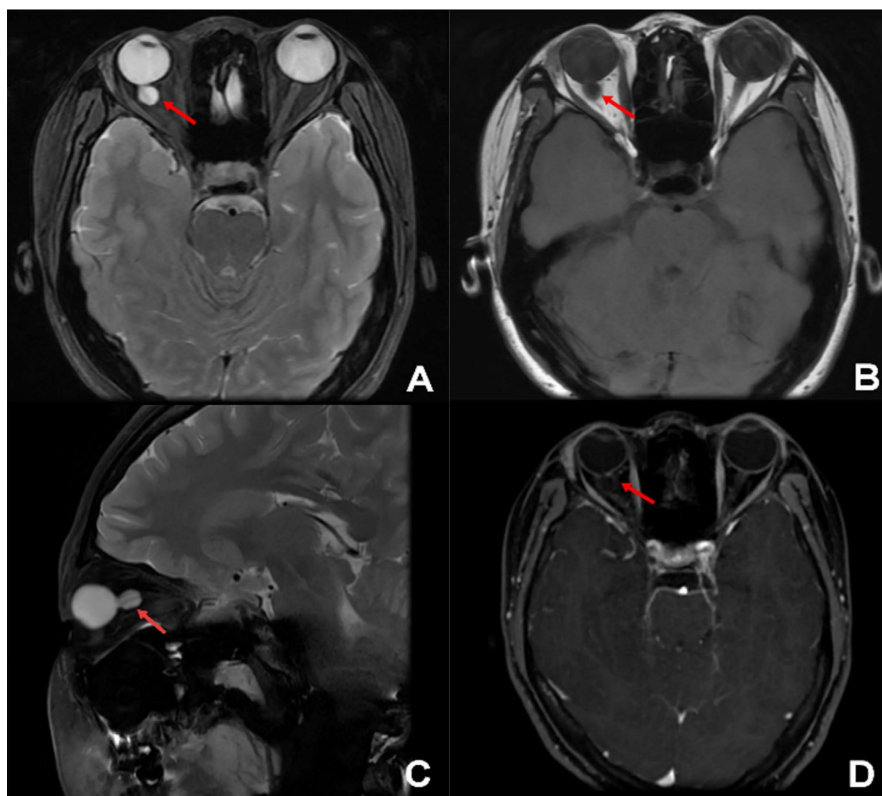


Figure 2. Magnetic resonance imaging findings of MGS. (A): Axial unenhanced fat-suppressed T2-weighted image, a round T2WI hyperintense lesion is seen along the course of the optic nerve posterior to the right globe, which is continuous with the posterior wall of the right eye, forming a gourd-shaped configuration. (B): Axial unenhanced T1-weighted image, the structure demonstrates hypointensity on T1WI sequences. (C): Sagittal fat-suppressed T2-weighted image, Sagittal view shows communication between this structure and the ocular posterior wall. (D): Axial contrast-enhanced image, no obvious enhancement is noted after contrast administration.

3. Treatment

Because the patient had long-standing severe visual impairment in the right eye, large-angle sensory esotropia, and no binocular visual function, surgery was undertaken primarily to improve ocular alignment and appearance. After preoperative evaluation showed no evident contraindications, combined right lateral rectus resection and medial rectus recession was performed under anesthesia on 27 August 2025. Following routine ocular preparation and draping, an inferotemporal conjunctival fornix incision was made. The lateral rectus was isolated, freed from the intermuscular septum and check ligament, secured with 6-0 sutures, disinserted, resected by 7 mm, and reattached at its original scleral insertion. The conjunctiva was closed with 8-0 absorbable sutures. Through a separate inferonasal fornix incision, the medial rectus was isolated and similarly freed from the surrounding fascial attachments. A 6-0 loop suture was placed 6 mm posterior to the insertion, followed by a 4-mm hang-back recession. The conjunctiva was closed with 8-0 absorbable sutures. At the end of surgery, no active bleeding was ob-

served and the globe appeared aligned. Tobramycin-dexamethasone ophthalmic ointment was applied, and the eye was patched. Postoperative recovery was uneventful.

Postoperative ocular alignment improved markedly. The prism alternate cover test and prism corneal light reflex test showed residual esotropia of 5 and 10Δ, respectively. Synoptophore examination continued to show no binocular single vision. During 6 months of follow-up, the patient underwent funduscopy examination, non-contrast brain MRI, and brain magnetic resonance angiography (MRA), no retinal detachment or other new ocular complications were documented.

4. Discussion

The characteristic funduscopy features of morning glory syndrome include an enlarged optic disc with a funnel-shaped optic disc defect, chorioretinal pigmentary abnormalities, and pale, fluffy glial proliferation overlying the optic disc [5]. In the present case, the patient had right esotropia and severe visual impairment since birth. Funduscopy examination revealed marked enlargement of the right optic disc with a central funnel-shaped excavated defect, surrounding choroidal atrophy, and extensive pigmentary abnormalities. Abnormal retinal vessels were observed radiating from the central optic disc tissue toward the peripheral retina. These findings were highly consistent with the typical manifestations of morning glory syndrome.

Previous studies have shown that patients with morning glory syndrome may exhibit several characteristic magnetic resonance imaging findings, including microphthalmia of the affected eye, enlargement of the optic disc with a funnel-shaped defect, glial-like abnormal tissue overlying the optic disc, and developmental abnormalities of the proximal optic nerve. In some patients, abnormal expansion of the optic nerve sheath and posterior scleral-choroidal structures may also be observed, and the optic chiasm and optic tracts may even be involved [5] [6]. In the present case, the right globe was smaller than the left. A round abnormal fluid-signal lesion was identified along the course of the right optic nerve within the retrobulbar intraconal space, which was continuous with a posterior defect of the right ocular wall. The adjacent right optic nerve showed atrophy and morphological distortion. These imaging findings suggest that the pathological changes of MGS may not be confined to the optic disc surface, but may also involve multiple posterior ocular and retrobulbar structures, including the sclera, choroid, and optic nerve posterior to the optic disc.

The precise etiology and pathogenesis of MGS remain unclear. Its development may be associated with mutations in the PAX6 gene [4]. Many scholars consider MGS to be a form of optic nerve coloboma or optic disc anomaly [7], and its pathogenesis may be related to incomplete closure of the superior portion of the embryonic fissure or abnormal glial development within the central optic disc [8]. However, other investigators have proposed that MGS should be regarded as a

distinct entity from optic nerve coloboma, because optic nerve coloboma is often associated with congenital multisystem anomalies, whereas MGS cannot currently be classified as part of any specific congenital malformation syndrome [1] [5]. Nevertheless, numerous associated findings have been reported, including, but not limited to, sphenoidal encephalocele [9], retinal detachment [10], congenital nasal anomalies [11], multiple sclerosis [12], moyamoya disease [13], ventricular malformations, and hydronephrosis [14]. Therefore, the relationship between MGS and other congenital developmental abnormalities warrants further investigation. These findings also suggest that brain MRI and MRA may be necessary during the diagnostic evaluation.

Although the funduscopy and imaging findings of MGS are relatively characteristic, it should still be differentiated from several other disorders, including optic nerve coloboma, optic disc pit, and peripapillary staphyloma. The typical features of optic nerve coloboma include an enlarged optic disc with a glistening white, bowl-shaped excavation. The retinal vessels are usually of normal caliber and may arise in a kinked manner from the margin of the defect. In addition, optic nerve coloboma is often bilateral and may show familial inheritance [15] [16]. In peripapillary staphyloma, the optic disc is usually normal and located at the bottom of the staphylomatous excavation. Funduscopy examination typically shows retinal vessels of normal appearance arising from the center of the optic disc [17], MGS may be associated with optic disc-centered posterior staphyloma-like changes [18]. Therefore, differentiation between these two entities requires a comprehensive assessment of optic disc morphology, retinal vascular course, and the presence or absence of central glial tissue overlying the optic disc.

In terms of treatment, the primary anatomical abnormalities of MGS cannot be reversed by surgery or medication. Clinical management mainly focuses on preserving and optimizing the patient's existing visual function, treating associated amblyopia and refractive errors, and promptly managing retinal and other ocular complications. For children within the critical period of visual development, occlusion therapy and visual training should be administered according to binocular visual function to reduce the impact of secondary amblyopia on visual outcomes [19]. Previous case reports have shown that, even in the presence of organic optic nerve abnormalities, some children may still achieve a certain degree of visual improvement after refractive correction combined with occlusion therapy, suggesting that early visual intervention has potential value [20]. For patients with concomitant strabismus, individualized treatment should be formulated according to the visual potential of the affected eye and binocular visual function. In children with favorable visual potential, strabismus correction may be considered after amblyopia treatment, whereas in patients with long-standing severe monocular visual impairment and established sensory strabismus, surgery is primarily intended to improve ocular alignment and cosmetic appearance [5]. Retinal detachment is one of the most serious complications of MGS. For progressive retinal detachment or retinal detachment involving the macula, pars plana vitrectomy combined with

retinal break management, peripapillary laser photocoagulation, and long-acting gas or silicone oil tamponade are among the most frequently reported treatment approaches [21] [22]. Complex cases often require combined procedures, such as membrane peeling, relaxing retinotomy, and silicone oil tamponade [23].

5. Conclusion

MGS is a rare congenital optic nerve developmental anomaly, and its diagnosis mainly relies on characteristic fundusoscopic findings, with MRI providing further evaluation of posterior ocular and optic nerve abnormalities. In the present case, the patient showed typical morning glory-like optic disc changes, accompanied by microphthalmia of the right eye, an abnormal posterior optic disc-related fluid-signal structure, and adjacent optic nerve atrophy. In patients with congenital monocular visual impairment and strabismus, MGS should be considered and differentiated from optic nerve coloboma, optic disc pit, and peripapillary staphyloma. Because the primary anatomical abnormalities of MGS cannot be reversed, clinical management should focus on preserving visual function, correcting strabismus when appropriate, and ensuring long-term follow-up to detect complications such as retinal detachment.

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Author Contributions

All authors participated in this work. The corresponding author, Shuixing Zhang, put forward the conception of this case report. Bo Lai collected, sorted and summarized the case materials and completed the initial manuscript writing. Qiuying Chen performed revision and refinement of the manuscript draft. All authors have read and agreed to the published version of the manuscript.

Consent for Publication

Written informed consent was obtained from the patient's legal guardian for the publication of the relevant clinical details and accompanying clinical images.

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Conflicts of Interest

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