Case Report

Macular Hole in Morning Glory Syndrome: A Case Report and Review of Literature

Maryam Yadegari [1], MD; Alireza Zare Mehrjardi [1], MD, Ali Vafaei [*2], MD

1- Department of Ophthalmology, Imam Hossein Medical Center, Shahid Beheshti of Medical Sciences, Tehran, Iran.
2. Department of Emergency Medicine, Shahid Beheshti University of Medical Sciences, Tehran, Iran .

***Corresponding Author:** Ali Vafaei
E-mail: d_ali_vafaei@yahoo.com

Abstract

Morning glory syndrome is a rare congenital excavated optic disc anomaly that can impair vision and may be associated with ocular and non-ocular abnormalities. It is characterized by a large excavated disc with radial vessels and a central turf of glial tissue within a funnel-shaped excavation of the posterior fundus. This syndrome is more common in Caucasians than in individuals of African descent. In this case report, we describe a unique case of morning glory syndrome with a full-thickness macular hole.

Keywords: Morning Glory Syndrome; Macular; Hole; Optical Disk.

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Introduction

The term 'morning glory' was coined by Kindler in 1970 to describe a congenital excavation of the optic disc that resembles the appearance of the morning glory flower, as detailed in Kindler's 1970 study [1]. Morning glory syndrome is a rare congenital abnormality that affects the optic disc, resulting in a hollowing or excavation of the posterior part of the eye surrounding the optic disc [2]. While this anomaly typically occurs unilaterally, there have been reported instances of bilateral involvement [2-4]. It is more prevalent in females and can cause vision impairment [5]. Morning glory syndrome has also been associated with various ocular and non-ocular abnormalities [2,3]. The exact prevalence of morning glory syndrome is unknown; however, a recent study in Stockholm, Sweden reported a prevalence of 2.6 per 100,000 individuals [2]. It is more commonly observed in Caucasians and less common in African Americans [2]. Visual acuity is often severely affected, with afferent pupillary defect and visual field defects [6]. Other ocular associations include congenital cataract and persistent hyperplastic primary vitreous [7].

In this case report, we present a case of morning glory syndrome with a full-thickness macular hole (FTMH), which has not been previously reported in the affected eye. The patient provided written consent, agreeing to have her case reported.

Case Report

A 62-year-old Iranian female presented to the glaucoma clinic at Imam Hossein Eye Specialist Hospital in Tehran, Iran, with a history of poor vision and eye discomfort in the right eye since childhood. She had no significant family history of ocular diseases or congenital problems. There was no consanguinity between her parents, and none of her siblings were affected. She had no history of ocular inflammation, trauma, or previous ocular surgery. At the initial visit, her best-corrected visual acuity was counting fingers at 0.5 meters in the right eye and 7/10 in the left eye. Refraction was + 0.75 - 0.5 x 60 in the right eye and + 0.5 in the left eye. A relative afferent pupillary defect (RAPD) was observed in the right eye, while there was no nystagmus, with normal ocular motility.

Anterior segment examination revealed no abnormalities in both eyes. Fundoscopy of the right eye showed a large excavated disc with a central turf of glial tissue, peripapillary chorioretinal pigmentation, and pigmentary changes due to a full thickness macular hole (FTMH) (Figure 1). Fundoscopy of the left eye was unremarkable. Intraocular pressure was 10 mmHg in the right eye and 12 mmHg in the left eye. Macular optical coherence tomography (OCT) demonstrated a FTMH in the right eye, while the left eye showed no abnormalities. Vision in the right eye did not improve with optical correction. The patient was diagnosed as a case of morning glory syndrome.

Discussion

Morning glory syndrome is a rare congenital condition involving an excavation of the optic disc [2]. It falls within the spectrum of congenital excavated optic disc anomalies, alongside conditions like optic disc coloboma and peripapillary staphyloma [8]. Morning glory syndrome is not known to be hereditary, and its genetic basis remains unestablished [2]. The exact underlying causes of morning glory syndrome are still not fully understood. It is believed that the excavation may result from either an incomplete closure of the

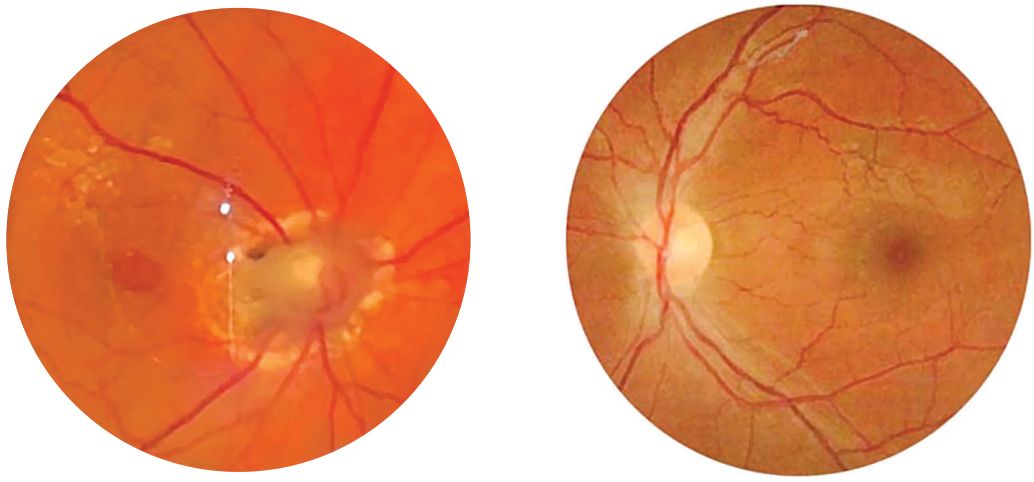

Figure 1: Fundoscopy of the right eye showed a large excavated disc with a central turf of glial tissue, peripapillary chorioretinal pigmentation, and pigmentary changes due to a full thickness macular hole (FTMH). Fundoscopy of the left eye was unremarkable

embryonic fissure; making it a variant of optic nerve coloboma, or it could be a primary mesenchymal abnormality leading to scleral defects, central glial tufts, and vascular irregularities [9].

Diagnosis of morning glory syndrome is typically clinical and based on its distinctive appearance during fundoscopy [9]. It manifests as a funnel-shaped excavation at the posterior fundus, enclosing and involving the optic disc [2]. The optic disc itself is enlarged and excavated, with a central region of white glial tissue and pigmentation around the optic nerve [9]. The blood vessels exhibit supernumerary features, radiating radially from the disc's periphery, making it challenging to differentiate between arterioles and venules [2].

Visual acuity in individuals with morning glory syndrome varies widely, ranging from no light perception to normal vision, but impaired vision is common [1]. Refractive errors in these eyes are often myopic [1]. Our patient presented with poor vision, high myopic astigmatism, and visual impairment in the affected eye with morning glory syndrome. Notably, there was no history of amblyopia treatment during childhood. Poor vision in individuals with congenital structural abnormalities may result from a combination of functional amblyopia superimposed on the organic defect [2,6]. Functional amblyopia refers to poor vision due to disrupted pattern vision or abnormal binocular interaction in an outwardly normal-looking eye, while organic amblyopia stems from structural abnormalities in the eye or brain, independent of sensory input, such as structural anomalies in the visual pathway [10]. While organic amblyopia is traditionally considered irreversible, some reports suggest improvements in visual acuity with amblyopia therapy in children with morning glory syndrome [2,10]. For instance, Cavazos-Adame et al.,[11] have reported improved visual acuity from counting fingers at 0.3 meters in a child with unilateral morning glory syndrome to 20/100 after five years of amblyopia therapy.

Ocular associations linked to morning glory syndrome encompass strabismus,

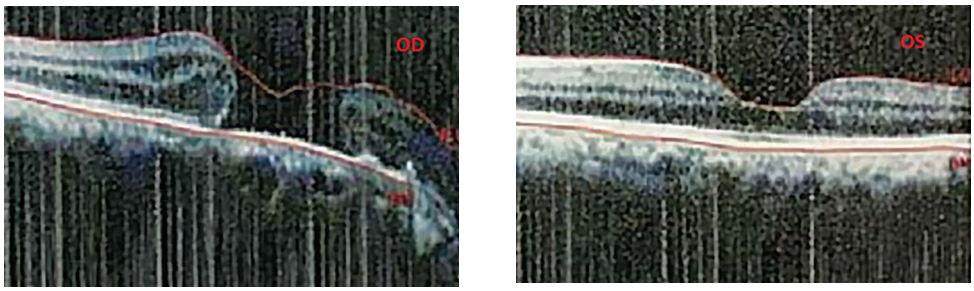

Figure 2: Macular optical coherence tomography demonstrated a full thickness macular hole in the right eye, while the left eye showed no abnormalities

nystagmus, retinal detachment, persistent hyperplastic primary vitreous (PHPV), cataract, microphthalmia, corneal leucoma, and optic nerve glioma [1,7,12,13]. Our patient had strabismus (exotropia). While strabismus associated with morning glory syndrome is typically horizontal (exotropia or esotropia), a few cases of vertical deviations have been reported [2]. Non-ocular associations include conditions such as basal encephalocele, corpus callosum agenesis, capillary hemangiomas, cerebrovascular anomalies like carotid artery stenosis and moyamoya disease, pituitary gland abnormalities, and facial abnormalities like median cleft lip and palate [1,3,14].

Optic disc coloboma is a differential diagnosis for morning glory syndrome. In morning glory syndrome, the entire disc is excavated, unlike optic disc coloboma, where the excavation is usually confined to the optic disc's inferior aspect, with a recognizable superior neuro-retinal rim [13]. Additionally, in our patient, who had high myopia, a posterior staphyloma might be considered as a differential diagnosis. However, posterior staphyloma typically involves an excavation of the posterior fundus surrounding the optic disc, while the optic disc itself remains flat without a central tuft of tissue and displays normal retinal vasculature [15,16].

Conclusion

We described a unique and rare case of morning glory syndrome with a full-thickness macular hole and the process of the successful differential diagnosis of the syndrome.

Authors ORCIDs

Maryam Yadgari:
https://orcid.org/0000-0003-0829-1861
Ali Vafaei:
https://orcid.org/0000-0002-7129-6457

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Footnotes and Financial Disclosures

Conflict of interest:

The authors declare no conflict of interest regarding the subject matter of the present study.