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J Case Rep Images Opthalmol 2024;7(2):13–17.
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Seeing through the glory: A rare presentation of morning glory disc anomaly

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ABSTRACT

Morning glory disc anomaly (MGDA) is a rare form of optic disc dysplasia, characterized by an excavated optic disc reminiscent of the tropical morning glory flower. Its prevalence is reported at 2.6 per 100,000 individuals. Typically observed unilaterally and manifesting in childhood, MGDA commonly presents with symptoms such as poor visual acuity, strabismus, or leukocoria.

We present a rare case of bilateral MGDA in a 40-year-old Caucasian female exhibiting preserved visual acuity despite notable retinal folds, and a possible correlation with the posterior variety of persistent hyperplastic primary vitreous (PHPV).

Keywords: Excavated disc, Morning glory disc anomaly, Morning glory syndrome, Persistent hyperplastic primary vitreous

How to cite this article

Sutedja T, Sahoo B, Hill R. Seeing through the glory: A rare presentation of morning glory disc anomaly. J Case Rep Images Opthalmol 2024;6(2):13–17.

Article ID: 100044Z17TS2024

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Received: 13 October 2024
Accepted: 20 November 2024
Published: 21 December 2024

doi: 10.5348/100044Z17TS2024CR

INTRODUCTION

Morning disc anomaly (MGDA) is a rare dysplasia of the optic disc that was discovered by Kindler in 1970 [1]. The name came from its resemblance to the tropical morning glory flower.

In this anomaly, the optical disc is often enlarged with an orangish hue and appears excavated with a central core of whitish glial tissue, in which persistent hyaloid vasculature remnants can be seen at its base. The blood vessels emerge from the rim of the central core in a radial pattern like the spokes of a wheel. The disc margins are surrounded by an annulus of peripapillary pigmentation (Figure 1) [2, 3]. In typical cases, the condition is usually unilateral and leads to very poor visual acuity [2, 4].

The prevalence of MGDA has been reported to 2.6 per 100,000. Both sexes being equally impacted. 85% of cases are unilateral, 15% are bilateral [4]. Kumar et al. (2021) reported a mean age of presentation and diagnosis of 8.8 years (range of 0.25–46 years).

In typical cases, the condition is usually unilateral and results in significantly reduced visual acuity, depending on the severity of the optic nerve anomaly [2, 4]. At initial presentation, patients often have poor vision, ranging from 6/60 to counting fingers, and may exhibit strabismus or leukocoria [5–7]. Visual field testing commonly reveals enlarged blind spots, and refractive assessments often indicate myopia and astigmatism [7]. Optimizing visual acuity as early as possible, especially during childhood, is essential to prevent amblyopia.

Morning disc anomaly can be associated with other ocular features including: serous retinal detachment, cataract, lens coloboma, foveal hypoplasia, and persistent hyperplastic primary vitreous (PHPV) [2, 3]. When associated with systemic signs and symptoms, it is known as the morning glory syndrome. Systemic associations that have been reported include cerebrovascular anomalies and facial dysmorphic features—hare lip and cleft palate [2, 3, 8].

The exact pathogenesis of MGDA remains unknown but is postulated to be related to poor and partial

development of the lamina cribrosa and an incomplete closure of the posterior scleral wall [9]. The central gliosis of the optic disc and abnormal vascular pattern may suggest an underlying primary neuroectodermal dysgenesis [10].

CASE REPORT

A 40-year-old Caucasian female was referred to the eye clinic from the optometrist with abnormal looking discs. She went to the optometrist as she noticed subtle deterioration of vision in the right eye since her last visit four years ago. Within the last 12 months, she has been diagnosed with immunoglobulin A (IgA) vasculitis (Henoch-Schoenlein purpura), with crescenteric IgA nephropathy and is currently on low dose prednisone (5 mg) and azathioprine maintenance therapy.

Best corrected visual acuity was 6/7.5 in the right eye, compared to 6/6 since last (and only previous) visit to the optometrist, and 6/6 in the left eye with no change since last visit. Refraction was –0.25/–0.125 Dcyl at 163 and +0.25/–0.75 Dcyl at 20 in the right and left eye respectively. Intraocular pressures were 13 mmHg in the right eye and 11 mmHg in the left eye.

Color perception and 30-2 Humphrey visual field tests were normal in both eyes.

On general examination, the patient is systemically well with no evidence of facial or eyelid abnormalities.

Ocular examination showed no manifest deviation. Anterior segment examination was unremarkable with clear lens in both eyes. The vitreous showed floating strands bilaterally. Dilated fundal exam showed normal sized, orange colored, excavated discs with a central core of white tissue in both eyes. The central core was larger and more clearly demarcated in the right eye. Retinal vessels appear to emerge from the peripheral margin of the excavation, radiating out in a radial spoke wheel pattern. Blood vessels emerging from the disc are supernumerary than usual. There is peripapillary pigmentation in both eyes; the right being worse with the pigmentation disturbance surrounding the entirety of the disc, compared to the left with involvement of only the temporal disc margin. In both eyes, there are retinal folds appearing as hypopigmented concentric lines radiating from all quadrants of the optic disc. This appears more extensive temporally as it advances toward the fovea. The macula and the peripheries are flat with no exudative pathologies, hemorrhages, tears, or detachments (Figure 2).

Optical coherence topography (OCT) discs were able to quantify the varying degree of retinal nerve fiber layer (RNFL) thickness around the neuro-retinal rim. Both eyes followed a different pattern of varying thickness. In the right eye, RNFL was thickest in the inferior quadrant, followed by superior, temporal, and, lastly, nasal quadrants (ISTN pattern). Retinal nerve fiber layer in the left eye followed a more conventional ISNT pattern. The average RNFL thickness is greater in the left eye at 146 μm, compared to the right eye at 104 μm. Optical coherence topography macula of both eyes demonstrates preservation of the foveal dip in both eyes, and an atypical folded and uneven appearance of the innermost RNFL with no epiretinal membrane seen (Figure 3).

Given that this patient's vision is satisfactory, and there is no other associated ocular features or associated neurological conditions, the patient is reassured and will continue to be followed up annually for an ophthalmic examination. Cautionary return advice for symptoms of serous retinal detachment and choroidal neovascularization were given for prompt return to clinic.

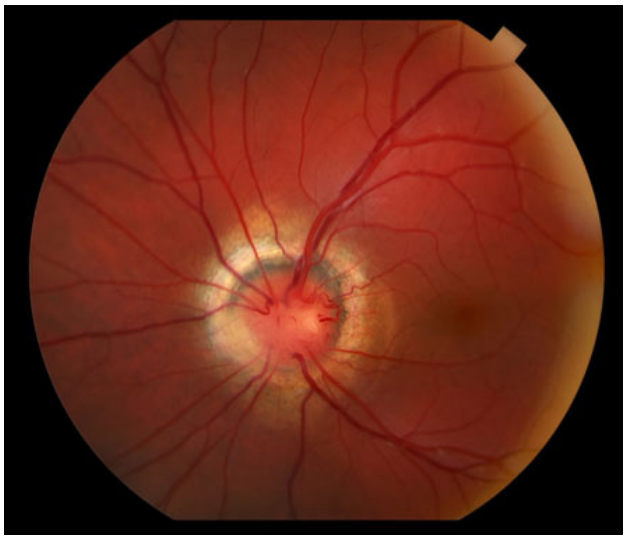

Figure 1: Typical presentation of MGDA. There is a central excavation of the optics disc filled with white glial tissue. Retinal blood vessels are seen radiating from the disc margins, accompanied by notable peripapillary pigmentation [21].

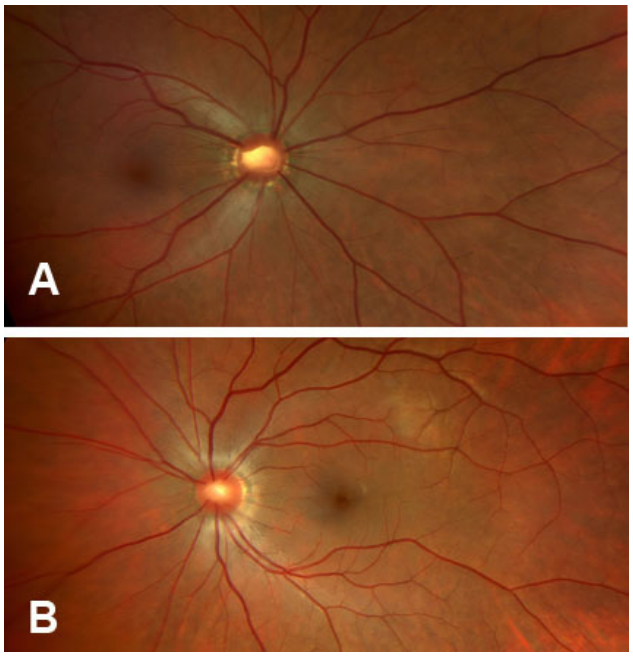

Figure 2: Fundal photos of the right (A) and left (B) eyes. Note the well-demarcated borders of excavation and the hyaloid vasculature underneath the white glial tissue of the right eye.

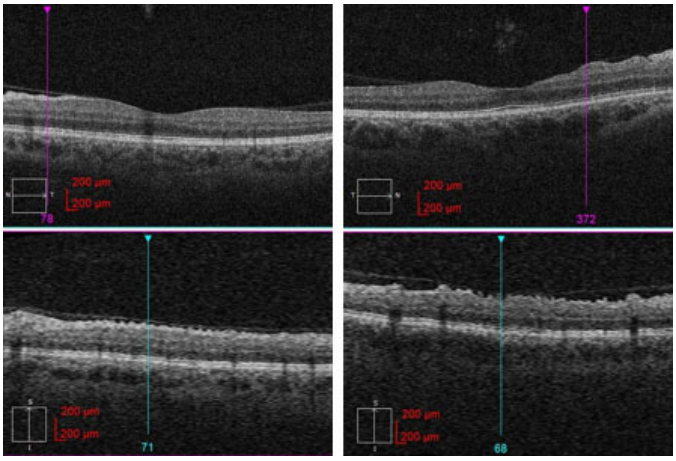

Figure 3: OCT demonstrating retinal folds in the macula. Despite the prominence of these folds, the patient's vision is well preserved.

DISCUSSION

Morning glory disc anomaly is predominantly a unilateral disease, with only 15% of cases reported as bilateral. It often presents in childhood with poor vision, strabismus, or leukocoria [2, 4]. Here, we present an atypical case of bilateral MGDA in an adult patient with no prior ocular history. The typical pattern of visual loss elicited in MGDA ranges from 6/60 to counting fingers [5]. Apart from the subtle decline in vision in the right eye over four years, from a best corrected visual acuity (BCVA) of 6/6 to 6/7.5, the patient remains asymptomatic despite the prominent retinal folds and surface wrinkling observed.

The retinal folds seen on examination of the posterior pole and macula scans are features of posterior PHPV—also known as falciform retinal septum or ablation falciformis congenita—that are known to be associated with MGDA [9]. Persistent hyperplastic primary vitreous occurs as a failure of the primary vitreous and the hyaloid vasculature to regress. Fei et al. (2013) indicate a high incidence of MGDA associated with PHPV, suggesting that the failure of the primary vitreous to regress is more likely in the presence of an optic disc defect.

Morning glory disc anomaly and PHPV may share a genetic basis involving key regulators of retinal and optic nerve development. The PAX6 gene is essential for ocular morphogenesis and is expressed in the developing central nervous system [10, 11]. Mutations in PAX6 have been associated with optic nerve malformations, including optic disc coloboma and hypoplasia, morning glory syndrome, and PHPV [10, 11]. Additionally, the ATOH7 gene codes for an important transcription factor, Atoh7, that is crucial for regulating early retinal ganglion cell (RGC) differentiation. Mutations in ATOH7 can impair RGC formation, resulting in persistent hyaloid vasculature and abnormal blood vessel and neural development [12, 13].

Morning glory disc anomaly can be a syndrome when associated with systemic illnesses. Systemic associations that have been reported include cerebrovascular anomalies and facial dysmorphic features [2, 3, 8]. Our patient does not have any features of these neurological or facial anomalies. Rather, she was recently diagnosed with IgA vasculitis within the last 12 months, raising the possibility of an underlying pathogenic autoimmune mechanism that could influence the symptomatic onset of this atypical case of MGDA. Immunoglobulin A vasculitis is associated with ocular conditions including episcleritis, scleritis, anterior uveitis, and retinal vasculopathies [14, 15]. The latter ocular manifestation can cause ischemic neuropathy, in which a pale and edematous optic disc is often seen on fundoscopy [14]. This is the first reported case of a patient with IgA vasculitis and MGDA, with no known reported association between the two conditions.

While MGDA is usually diagnosed by funduscopic examination alone, the case presented was atypical and diagnosis was proven to be challenging. The patient had good visual acuity in both affected eyes, size of the disc appears normal, and there were no significant peripapillary atrophy that has been widely reported in typical cases of MGDA [2, 3].

Differential diagnoses of congenital excavated optic disc anomalies, including optic disc coloboma and peripapillary staphyloma, were considered based on the fundus appearance. Optic disc coloboma typically presents with an inferior excavation extending toward the choroid, lacks a central glial core, and shows normal disc vasculature [16]. It is often bilateral and associated with systemic conditions, such as CHARGE (Coloboma ocular, Heart defects, Atresia or stenosis of the choanae, Retardation of growth and/or development, Genitourinary anomalies, and Ear abnormalities) syndrome, which includes eye, heart, and ear abnormalities [17]. Peripapillary staphyloma also features an excavation at the optic disc base, lacks a central glial core, and preserves normal retinal vasculature [18, 19]. The presence of central glial tissue and a distinct vascular pattern arising from the disc margins helped narrow the diagnosis, aligning it more closely with the characteristics of morning glory disc anomaly.

Additionally, OCT imaging was essential in the diagnostic process. In MGDA, traction from PHPV can result in retinal folds, whereas optic disc coloboma and peripapillary staphyloma typically display smooth, intact retinal layers [19, 20].

As of current, there is no definitive treatment for MGDA. Treatment strategies include correction of refractive errors, regular monitoring for retinal detachments and neovascularization, and referral to appropriate specialties (if systemic conditions are involved). In our case, the anomaly appears to be an incidental finding and requires no urgent treatment. It is possible that MGDA is a spectrum, in which anomalies range from asymptomatic to serious visual defect with guarded prognosis.

CONCLUSION

Morning glory disc anomaly is a rare optic disc dysplasia characterized by an excavated disc with a central glial core and radial vascular pattern. While it typically presents unilaterally and leads to significant visual impairment, this case demonstrates an atypical presentation with bilateral involvement and no visual symptoms. Although, MGDA may be linked to genetic defects and systemic conditions, no correlation with autoimmune disorders such as IgA vasculitis has been previously reported. Early detection, through detailed fundal examination and OCT imaging, and regular monitoring, is vital for managing potential complications and preserving vision in affected individuals.

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Author Contributions

Theodore Sutedja – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Baswati Sahoo – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Robert Hill – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all

aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Guarantor of Submission
The corresponding author is the guarantor of submission.

Source of Support
None.

Consent Statement
Written informed consent was obtained from the patient for publication of this article.

Conflict of Interest
Authors declare no conflict of interest.

Data Availability
All relevant data are within the paper and its Supporting Information files.