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CO- EXISTENCE OF PERSISTENT HYPERPLASTIC PRIMARY VITREOUS IN RIGHT EYE AND MORNING GLORY SYNDROME IN LEFT EYE– A RARE CLINICAL CASE REPORT

Ophthalomology

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ABSTRACT

Morning Glory Syndrome (MGS) is an uncommon congenital anomaly of the optic disc, characterized by a funnel-shaped excavation, a central tuft of glial tissue, and radially oriented retinal vessels. This condition frequently leads to significant visual deficits due to early developmental disruptions during embryogenesis. MGS is commonly associated with several ocular conditions such as retinal detachment, cataracts, and strabismus, as well as systemic disorders including basal encephalocele and Moyamoya disease [1,5,7]. A notable coexisting condition is Persistent Hyperplastic Primary Vitreous (PHPV), which shares similar developmental pathways and substantially influences the clinical presentation of MGS. This report presents a case demonstrating marked PHPV with lens colobomas in an MGS patient, emphasizing the complex developmental interrelation between these anomalies. Awareness of these associations is essential for prompt diagnosis, effective management of complications, and early detection of potentially life-threatening conditions [1–3].

KEYWORDS

Morning glory syndrome, Persistent hyperplastic vitreous, optic disc anomaly, congenital disc anomaly

INTRODUCTION

The relationship between MGS and PHPV points to a common developmental aetiology, likely rooted in abnormal embryonic fissure closure [1,2]. PHPV arises from the failure of the primary vitreous to regress, which plays a significant role in the phenotypic presentation of MGS and distinguishes it from other optic disc malformations [2]. Its frequent coexistence with lens colobomas further supports this embryologic connection [1]. Persistent vitreous can apply traction on ocular structures, exacerbating MGS abnormalities and contributing to conditions such as posterior lenticonus [3,4]. Cases involving MGS, PHPV, and anomalies like Peters' anomaly suggest a broad developmental disruption, potentially involving mutations in genes like *PAX6* [2,3]. In severe presentations, where direct fundus examination may be limited, ultrasonography becomes a critical diagnostic tool [2].

Case Report

A 4-year-old male came to Eye O.P.D with diminution of vision and involuntary eye movement in the both eye since birth. Routine eye examination showed a visual acuity of counting finger close to face in the both eyes, which was not improved by lenses. There was presence rotational and horizontal nystagmus in bilateral eyes. Pupils were normal in size, sluggishly reacting to light in both the eyes. The lens showed a posterior polar cataract in right eye and was within normal limit in left eye. A whitish mass was present centrally in the pupillary area which represented the primary vitreous a remnant of the hyaloid artery in the form of a stalk protruded into the vitreous. No other abnormalities were detected by gonioscopy in both the eyes. With rest of the anterior segment evaluation was within normal limit. Both eye intra-ocular pressure were within normal limit. On dilated fundus examination, it revealed the presence of fibrovascular stalk, or retrolental tissue, extending from the optic disc to the lens suggestive of persistent hyperplastic vitreous in right eye (fig.1)

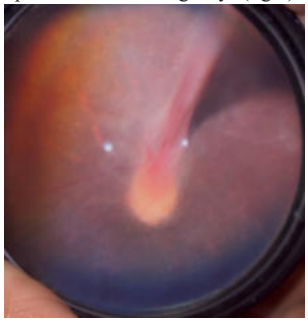
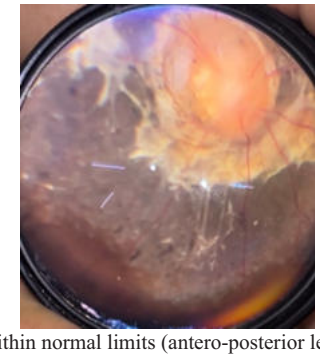


Fig. 1- Fundus photograph of Right eye showing Presence of fibrovascular stalk extending from disc to lens suggestive of P.H.P.V

The optic disc was large and funnel-shaped, with glial tuft, radial and straight-running retinal vessels coming out of it. The surrounding area of the optic disc showed pigmentary changes. This appearance is often compared to the shape and arrangement of a morning glory flower ,thus, suggestive of morning glory optic disc anomaly in the left eye. (fig.2)



Axial length within normal limits (antero-posterior length = 23.4 mm in right eye and 22.5 mm in left eye). B-scan echography revealed revealed the presence of persistent hyperplastic vitreous in right eye (fig.3) and and morning glory disc in the left eye. (fig.4)

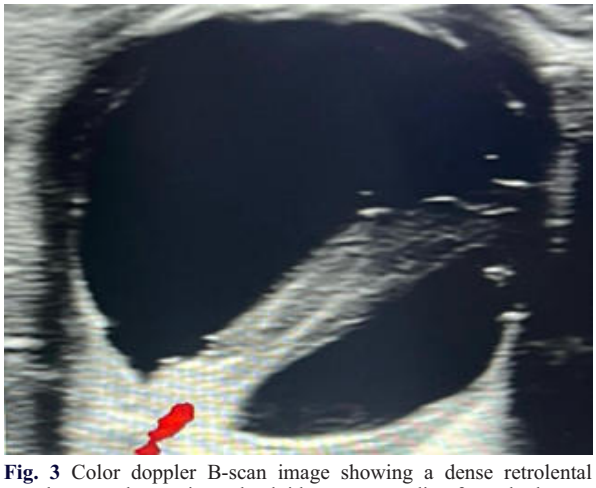


Fig. 3 Color doppler B-scan image showing a dense retrolental membrane and a persistent hyaloid artery extending from the lens to the optic disc showing blood flow within the stalk on color doppler.

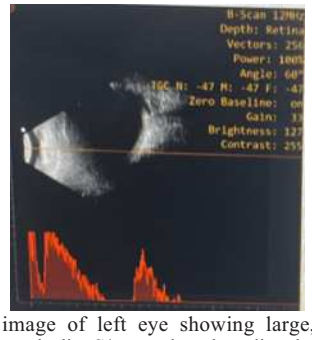


Fig.4- B-scan image of left eye showing large, funnel-shaped excavation of the optic disc S/o morning glory disc abnormality.

DISCUSSION

The co-occurrence of MGS with persistent hyperplastic primary vitreous and lens colobomas suggests a shared embryonic origin involving incomplete closure of the fetal fissure. [1,2]This closure is crucial for the normal development of ocular structures, and its disruption may lead to colobomatous anomalies affecting the retina, optic nerve, iris, choroid, and lens. [4–7]

Persistent primary vitreous significantly influences the clinical expression of MGS. The traction it generates may intensify optic disc malformations and elevate the risk of complications such as retinal detachment. [3,4] Lens colobomas reinforce the hypothesis of widespread embryonic insult, indicating the involvement of multiple ocular tissues during development. [6–9]

MGS is frequently accompanied by a spectrum of ocular manifestations:

- **Strabismus**, primarily horizontal deviations (esotropia or exotropia), is observed in approximately 70–80% of patients and often requires surgical intervention. [4–6,17]
- **Amblyopia** is another major contributor to visual impairment, usually resulting from misalignment or high refractive error. Management strategies include corrective lenses and occlusion therapy. [6–7]
- **Nystagmus**, particularly in bilateral or severely affected cases, reflects deeper involvement of the visual pathway. [8]

Additional ocular findings may include:

- **Microphthalmia**, **glaucoma**, and **leukocoria**, the latter often presenting as a white or yellowish pupillary reflex in children. [8]
- Less common associations include **corneal leucoma**, **optic nerve drusen**, **eyelid hemangiomas**, **aniridia**, and **anterior chamber cleavage syndromes**, all reflecting varying degrees of developmental interference. [9–13]

Given the complexity and range of manifestations, a multidisciplinary and integrative diagnostic approach is critical. Recognizing MGS as part of a broader spectrum of developmental anomalies rather than as an isolated optic nerve defect enables better management and patient outcomes. [10–13] This comprehensive array of defects also highlights the necessity of a holistic diagnostic and management approach for MGS, recognizing it as part of a broader developmental spectrum rather than an isolated anomaly. [14-17]

CONCLUSION

MGS represents a rare but clinically significant optic nerve malformation, typically rooted in early embryologic disruption. The frequent co-occurrence of PHPV and lens colobomas underscores the importance of understanding the syndrome's developmental basis. Given its associations with both ocular and systemic abnormalities, timely diagnosis and intervention are imperative for reducing the risk of complications and provide better visual outcomes.

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