CASE REPORT

Rare bilateral presentation of morning glory disc anomaly

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Accepted 13 August 2016

SUMMARY

A 4-year-old girl presented with a history of poor vision and oscillation of both eyes since infancy. Ocular examination revealed the best corrected visual acuity of 2/60 in right eye and 3/60 in left eye. Horizontal pendular nystagmus was present in both eyes. Fundus examination revealed morning glory disc anomaly (MGDA) bilaterally. Radiographic imaging of the brain revealed central nervous system anomalies. The guarded visual prognosis was explained and the patient referred for low vision rehabilitation and advised yearly follow-up. MGDA is very rarely bilateral. We aim to highlight the distinguishing features of bilateral MGDA from other excavated optic nerve head anomalies which could also present bilaterally but vary in their associations, management and prognosis.

BACKGROUND

Handmann, in 1929, was the first to describe morning glory disc anomaly (MGDA) in six of his patients in the German literature. Kindler too, in 1970 published some cases with an 'unusual congenital anomaly of the optic disc'.¹ Since then, several authors have come to the conclusion that both authors described the same disease.² The anomaly was termed 'morning glory disc anomaly' because of its resemblance to the tropical morning glory flower. It is ostensibly a sporadic condition. The optic disc is usually enlarged, orange or pink in colour with a funnel-shaped excavation and whitish glial tissue covering it. The retinal blood vessels emerge from the periphery of the excavation in a radial spoke (wheel) like pattern running an abnormally straight course over the surrounding elevated peripapillary retina which has chorioretinal pigmentary changes. MGDA is more common in females (2:1) and rare in African-Americans.³ It is usually unilateral and rarely occurs as a part of multisystem disorder. A thorough PubMed search revealed that only six cases of bilateral presentation of MGDA have been reported to date. We report a case of the bilateral form of MGDA with an aim to distinguish it from other excavated bilateral optic nerve head anomalies.^{4 5}

CASE PRESENTATION

A 4-year-old girl presented with a history of poor vision and oscillation of both eyes since infancy. Family, gestational and systemic history was unremarkable. External examination revealed no soft tissue or bony abnormality. Ocular examination revealed the best corrected visual acuity of 2/60 in right eye and 3/60 in left eye. Intraocular pressure

was normal in both eyes. Pupillary reaction to light was sluggish and ill-sustained bilaterally. Horizontal pendular nystagmus was present in both eyes with a right esodeviation of 20–25° on the Hirschberg's test. The rest of the anterior segment findings were within normal limits. Fundus examination (figures 1 and 2) revealed bilaterally enlarged optic discs with central excavation and elevated pigmented borders, peripapillary chorioretinal pigmentary changes along with retinal and glial folds, cilioretinal vessels emerging from the peripheral rim of the excavation in a radial pattern and white glial tissue covering the disc and vessels in the periphery. The rest of the fundus examination was within normal limits.

INVESTIGATIONS

Ultrasonography B scans (figures 3 and 4) confirmed excavation of the optic nerve head with posterior vitreous degeneration bilaterally (videos 1 and 2). There was no evidence of retinal detachment. Anthropometry measurements revealed the height and weight to be within the normal range, but head circumference was less than third centile. X-ray films revealed a normal bony orbit and optic canals. CT scans and MRI of the brain and orbit with angiography revealed outpouching of optic nerve head, dilated right optic nerve sheath with cerebrospinal fluid (CSF), bony defect in basisphenoid with basal meningoencephalocele extending upto nasopharynx, pituitary gland herniating with meninges and CSF and mild hypoplasia of optic chiasma (figures 5–8). The clinical features and ancillary investigations established the diagnosis of bilateral MGDA.

Figure 1 Fundus photo of the right eye depicting morning glory disc anomaly.

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To cite: Dedhia CJ, Gogri PY, Rani PK. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2016-215846

Figure 2 Fundus photo of the left eye depicting morning glory disc anomaly.

Figure 3 Ultrasonography B scan of the right eye depicting morning glory disc anomaly.

Figure 4 Ultrasonography B scan of the left eye depicting morning glory disc anomaly.

DIFFERENTIAL DIAGNOSIS

MGDA needs to be distinguished from other congenital disc lesions, especially optic nerve coloboma and peripapillary staphyloma. Unlike MGDA, optic nerve coloboma is often bilateral, familial, commonly associated with iris/ciliary/retinal colobomas and multisystem genetic disorders and without any racial or gender predilection. Optic nerve coloboma is rarely associated with basal encephalocele, unlike MGDA.^{3 6} On fundus examination, the disc lies centrally (symmetrically) within the excavation in MGDA, whereas excavation lies asymmetrically (usually inferiorly) within the disc in cases of optic nerve coloboma. Also, the absence of central glial hyperplasia, peripapillary depigmentation and anomalous retinal vasculature distinguishes optic nerve coloboma from MGDA.^{3 6} Peripapillary staphyloma is characterised by a comparatively deeper cup-shaped excavation, a relatively normal, well-defined optic disc along with the absence of glial and vascular anomalies unlike MGDA which has less deep, funnel-shaped excavation in a grossly anomalous, poorly-defined optic disc with a central glial bouquet and anomalous vascular pattern.^{6 7}

Video 1 Ultrasonography B scan of the right eye depicting morning glory disc anomaly.

Video 2 Ultrasonography B scan of the left eye depicting morning glory disc anomaly.

Figure 5 Sagittal T2W MRI of the brain through right optic nerve showing outpouching of optic nerve head. T2W, T2-weighted MRI of the brain.

TREATMENT

The guarded visual prognosis was explained. The patient was referred for low vision rehabilitation and paediatrician for pituitary deficiency.

OUTCOME AND FOLLOW-UP

The patient was advised yearly follow-up.

DISCUSSION

Children with bilateral optic disc abnormalities usually present with nystagmus and poor vision. The developmental interruption

Figure 6 Axial and sagittal T2W MRI of the brain showing dilated right optic nerve sheath with cerebrospinal fluid (CSF). T2W, T2-weighted MRI of the brain.

Figure 7 Axial CT scan images of brain (inferior to superior) showing herniation of meninges and CSF into the nasopharynx. CSF, cerebrospinal fluid.

Figure 8 Sagittal and coronal T1W MRI of the brain showing a bony defect in basisphenoid with basal meningoencephalocele (yellow arrows) extending up to nasopharynx. Pituitary gland (red arrow) is seen herniating with meninges and CSF. Mild hypoplasia of optic chiasma is also seen (blue arrow). CSF, cerebrospinal fluid; T1W, T1-weighted MRI of the brain.

most likely occurs at 4–5 weeks of embryonic growth.⁸ The defect results secondary to faulty closure of the posterior sclera and embryonic fissure combined with herniation of the optic disc. Thus there is a failure of normal neuroectodermal development along with primary mesenchymal abnormality.⁸ Common ocular associations of MGDA are non-rhegmatogenous retinal detachment (most common), strabismus, congenital cataract, persistent hyaloid remnants, lid haemangiomas, preretinal gliosis, lens colobomas, chronic simple glaucoma, Duane's retraction syndrome, microphthalmia, anterior chamber cleavage syndrome and hypertelorism.⁹ Systemic associations include midline craniofacial defects, frontonasal dysplasia, depressed nasal bridge together with an osseous defect in the anterior skull base with herniation of pituitary-hypothalamic structures inferiorly into the defect, defects of the sella turcica, duplication of the pituitary stalk and caudal displacement of the floor of the third ventricle,¹⁰ basal encephalocele and agenesis of the corpus callosum, midline cleft lip and cleft palate,¹¹ hypoplasia

of optic chiasma, pituitary deficiency (endocrine dysfunction), hypoplastic cerebellar vermis and malformed occipital lobe, parieto-occipital porencephaly, mild hydrocephaly of lateral ventricle, brain atrophy and enlarged thalami,¹² Moyamoya vascular pattern,¹³ PHACES association,¹⁴ CHARGE syndrome,¹⁵ and neurofibromatosis type 2.¹⁶

There is no definitive treatment for this anomaly. Patients should have a complete general physical examination and growth evaluation for early diagnosis and treatment of pituitary deficiency. The condition can be managed by correcting refractive errors, giving occlusion therapy in selected cases for amblyopia management, monitoring for retinal detachment, use of low vision aids in bilateral cases, referral to appropriate specialty, optimising the conditions at home and at school in an attempt to ensure that impaired vision does not impede overall development and education. Counselling of parents is essential to explain to them the anomaly with its prognosis.

Learning points

- ▶ Morning glory disc anomaly (MGDA) is usually unilateral with very rare bilateral cases reported.
- ▶ Differentiating MGDA from other excavated congenital optic disc anomalies like optic nerve coloboma and posterior staphyloma is important as they could also present bilaterally but vary in their associations, management and prognosis.
- ▶ A thorough evaluation should be carried out in all cases of MGDA to detect various systemic associations.

Acknowledgements The authors acknowledge Dr Foram Gala, Neuroradiologist, Lifescan Imaging Centre, Mumbai for helping us with the interpretation of CT-MRI scan images and Professor Jill Keefe, University of Melbourne, Australia for helping with the proof-reading of the manuscript.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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