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Case Report

Morning glory optic disc anomaly associated with retinal detachment in a 9-year-old northern Nigerian male child: a case report

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ABSTRACT

Morning glory syndrome (MGS) is uncommon in Nigeria as is less commonly seen in Africans. It is one of the congenital excavated optic disc dysplasia apart from optic disc coloboma and peripapillary staphyloma. We report a case of morning glory disc anomaly (MGDA) with associated retinal detachment in a 9 year old boy, who presented with complaint of poor vision in the left eye incidentally noticed when he covered the right eye following a trivial trauma to the eye. Ocular examination revealed the best corrected visual acuity of 6/5 in the right eye and no perception of light (NPL) in the left eye. Cycloplegic refraction with tropicamide and cyclopentolate was OD: -0.50DS/-0.50DC×180 in the right eye and OS: -6.00DS/-0.50DC×180 in the left eye. He was not fixating with the left eye on cover test but demonstrated a 4-degree exotropia assessed with a prism bar. Anterior segment examination revealed a normal finding except for an afferent pupillary defect in the left eye. Funduscopy was normal in the right eye, but left eye revealed a large excavated optic disc with a central area of fibroglial tissue, multiple thin blood vessels emerging from the edge of the disc running radially in an abnormally straight course over the peripapillary retina. There were areas of peripapillary pigmentary changes and retinal detachment. MGS is an anomaly of the Optic nerve head usually presented with different ocular and non-ocular abnormalities, and it may be the cause of severe visual impairment in early childhood and subsequent development of amblyopia.

Keywords: MGDA, Retinal detachment, Northern Nigeria, 9 year old male child

INTRODUCTION

Morning glory syndrome (MGS) is a rare congenital optic disk dysplasia affecting optic nerve head.[1] It was 1st reported in 1970 by Dr. Peter Kindler. Its appearance resembles morning glory flower hence name MGS/ MGDA.[2]

It is a primary mesenchymal abnormality that present with enlarged, excavated and funnel shaped optic disc with central whitish fibrous tissue, a pigment ring slightly protrudes into the peripapillary area and straight branches of retinal vessels emerging radially from the optic disc edge.[1-2]

Studies on the disease prevalence is rare. Some cases were reported to be unilateral and sporadic, 16% of cases are bilateral.[2] A prevalence rate of 2.6/ 100,000, and the gender ratio of 1:1 have been reported by Sakamoto et al.[3] Out of the total MGS, bilateral cases account for 0%

to 30%. MGS can be associated with systemic anomalies. These may include capillary hemangiomas, hypertelorism, cleft lip, cleft palate and basal encephalocele. About 25% of patients with MGS present with vascular and encephalic abnormalities. With exception for PAX6 gene or syndromic MGS, the genetic link and causative agents have not been reported for this condition.[2-3] Study by Panyala et al reported case of MGS associated with chronic myeloid leukemia Philadelphia chromosome (CMLPC) and empty sella turcica.[4]

MGS is uncommon in Nigeria as is less commonly seen in Africans. Only one case was reported by Osaguana et al in Benin city, Nigeria.[5] Few cases were also reported in some African countries.[6-7] Majority of the cases present with inability to see, ocular deviation and leucocoria during the early period of life. Dilated fundus examination is required to establish diagnosis. Optical coherence tomography (OCT) may be needed in detecting the primary disc anomaly and also other associated retinal findings like subretinal or intraretinal fluid, retinal holes or retinal detachments.[2] B-scan echography can also help to get the details of other findings in MGS which may include optic nerve excavation, central glial tuft, microphthalmos, and retinal tissue overhanging the posterior scleral staphyloma.[2] Late diagnosis can lead to potentially serious complications such as retinal detachment.[8] MGS eyes usually present with poor vision, but can present with a different range of visual impairment from mild to severe (range 20/30-hand motion).[2] Strabismus (eso-or exodeviation) are also common, may be present in up to 70% of eyes affected by MGDA. Afferent pupillary defect and visual field deficits were also observed.[2] It is important to make prompt diagnosis and start treatment early in MGS.[8] Anisometropia should be corrected with spectacle to prevent amblyopia. Squint may require surgical correction. Retinal detachment is an associated condition that may require management surgically, although spontaneous reattachment have been reported.[8] We report a case of this uncommon congenital dysplasia affecting the optic disc with associated retinal detachment in a Northern Nigerian male child. To the best of the author's knowledge, this is first case reported in Northern Nigeria.

CASE REPORT

A nine-year-old boy was referred from paediatric outpatient department of Aminu Kano teaching hospital with complaint of poor vision in the left eye incidentally noticed when he covered the right eye following a trivial trauma to the eye. The mother reported that the child has never complained of poor vision in the left eye before but the eye occasionally appears to turn inward. The child has been healthy. Antenatal and labour were uneventful. Developmental milestones were optimal. No family history of ocular diseases or congenital anomalies. Patient is the last child of 8 children from a monogamous non-consanguineous marriage. No history of similar problems, ocular diseases among the siblings or other relatives. On general examination, patient is not in obvious distress, no any facial dysmorphic features. Neurological examination was normal.

Ocular examination revealed the best corrected visual acuity of 6/5 in the right eye and NPL in the left eye. Cycloplegic refraction with Tropicamide and cyclopentolate was OD:-0.50DS/-0.50DC×180 in the right eye and OS:-6.00DS/-0.50DC×180 in the left eye. He is not fixating with the left eye on cover test but demonstrated a 4-degree exotropia with full ocular motility on both eyes, assessed with a prism bar using Krimsky method without his correction. No nystagmus noted. Anterior segment examination with slit lamp biomicroscope was normal in both eyes except for relative afferent pupillary defect in left eye. Intraocular pressures were 12 mmHg and 11 mmHg in right and left eyes respectively using keeler pulse air tonometry.

Funduscopy was normal in the right eye, but left eye revealed a large excavated optic disc with a central area of fibroglial tissue, multiple thin blood vessels emerging from the edge of the disc running radially in an abnormally straight course over the peripapillary retina. There were areas of peripapillary pigmentary changes and retinal detachment (Figure 1). Ocular B Scan revealed an axial length of 23.5 mm in the right eye and 29.7 mm in the left eye. There were echogenic vitreous strands and retinal detachment in the left eye. Right eye is sonographically normal (Figure 2). The left eye vision was NPL which is likely as a result of long-standing retinal detachment.

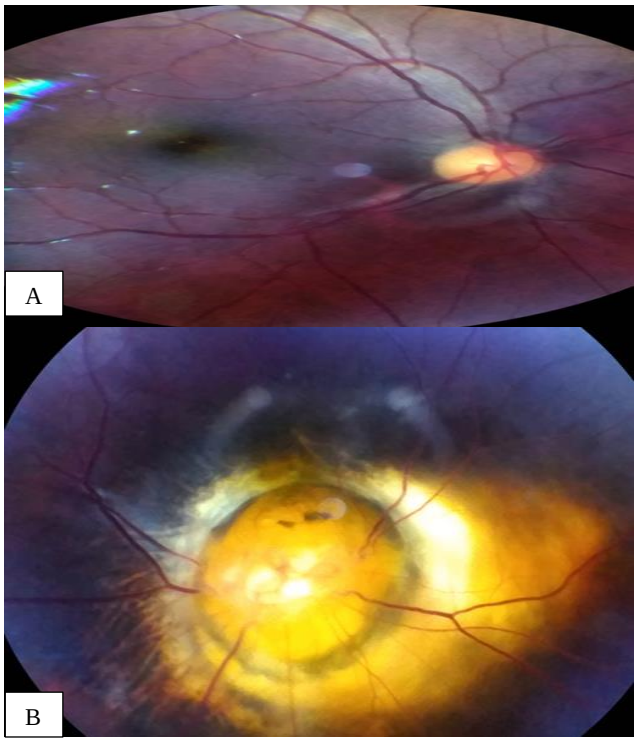

Figure 1 (A and B): Fundoscopic image of the normal eye and the eye with morning glory optic disc.

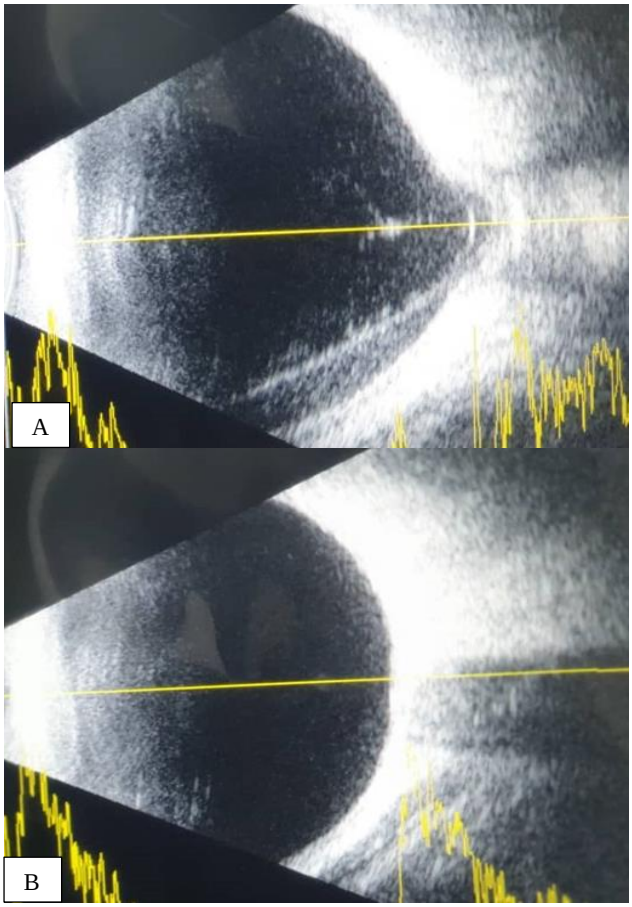

Figure 2 (A and B): The B-scan image of the normal right eye and the eye with MGS with associated retinal detachment.

DISCUSSION

To the best of our knowledge, this is the first reported case of MGS in the whole of Northern Nigeria and the second case reported in Nigeria based on the extent of our literature review.

MGS is one of the congenital excavated optic nerve head malformations like optic disc coloboma and peripapillary staphyloma.[5] MGS is non-hereditary and there is no known genetic abnormality associated with it.[9] This is also corroborated with our case report as the boy presented with no previous ocular family history of any defect. However, Altun et al reported a case of MGS in a patient with Down syndrome but apart from the trisomy of chromosome 21 that is associated with Down syndrome no other genetic defect was detected.[10] Pathogenesis of MGS is not clearly defined.[5] The excavation may be as a result of an insufficient closure of the embryonic fissure looking like a variant of optic nerve coloboma, or it can be a primary mesenchymal abnormality resulting in scleral defect, central glial tuft and vascular abnormalities.[5,11] Baksh et al reported that this optic nerve head dysplasia could be as a result of abnormal differentiation of mesodermal tissues. Specifically, imperfect closure of the posterior sclera and partial evolution of the lamina cribrosa have largely contributed in these progressive optic nerve changes, leading to anterior herniation of the disc and adjacent retina.[2] Neuroectodermal dysgenesis may be related to the final phenotype of MGS, as evidenced by the hallmark central gliosis and abnormal vascular pattern.[12] Histopathologic studies have given additional information on the pathogenesis of MDS with reports of abnormalities of posterior pole structures, which include peripapillary scleral ectasia (staphyloma), absence of lamina cribrosa, absence of choroid, reduced number of ganglion cells and reduced retinal nerve fiber layer, absence of photoreceptors, glial tuft with psammoma bodies/glial cells/fibroblasts, attenuation of vessels, and hyperplastic persistent primary vitreous.[12,13]

MGS is usually sporadic as also seen in our study. Contrary to some studies including our study, pathogenic variants in PAX6 gene have been revealed in some families.[2,5,9,11,13] Furthermore, cavitary optic disc anomalies (CODA)-a disease spectrum that include MGS, megalopapilla, optic disc coloboma, and optic disc pit-are known to be inherited in an autosomal dominant fashion through mutations in the MMP19 gene.[14]

MGS can be diagnosed clinically by its characteristic appearance on fundoscopy as described by Osaguona and Momoh, Baksh et al and this was also witnessed in our reported case (Figure 1), in that there is a funnel shaped excavation of the posterior fundus which encloses and includes the disc.[2,5] The disc is large and excavated with a central area of white glial tissue and peripapillary chorio-retinal pigmentation. The blood vessels are supernumerary, fan out in a radial manner from the periphery of the disc and it is hard to distinguish between arterioles and venules.[5] In some instances, the adjacent macula can be involved in the larger area of excavation, a phenomenon known as “macular capture.” Other fundus findings may include subretinal fluid and retinal detachment, as in our reported case where we have peripapillary retinal detachment, as shown in Figure 2 above.[2] Traction may likely contribute to the pathogenesis of retinal detachment in Morning Glory Disc possibly by creating retinal breaks with subsequent accumulation of fluid beneath the retina leading to retinal detachment. The sources of the fluid are speculated to be from vitreous cavity and or cerebrospinal fluid.[15] The degree of visual acuity in eyes with MGS is variable from no light perception to normal vision, but in most cases, it is severely impaired which may lead to low vision cases and eventually present as visual handicap.[5,11] The refractive error in eyes with MGS is commonly myopic.[5,7] Our patient had poor vision with a high myopic astigmatism, Strabimus (exotropia) and impaired vision (no light perception) in the eye with MGS. There was no history of treatment of amblyopia in childhood in our patient and other orthoptics work-up tests was not done on him due to absence of stereopsis, which is as a result of anomalies of binocular vision, e.g. suppression, abnormal retinal correspondence associated with his case

(MGS). He also had none of the ocular abnormalities associated with MGS such as nystagmus, persistent hyperplastic primary vitreous (PHPV), cataract, microphthalmia, corneal leucoma and optic nerve glioma as reported by other studies.[5] We did not find any non - ocular associations of MGS with our patient, such as basal encephalocele, corpus callosum agenesis, capillary hemangiomas, cerebrovascular anomalies such as carotid artery stenosis and Moya Moya disease, pituitary gland abnormalities and facial abnormalities such as median cleft lip and palate as reported in many studies.[5,9]

Optic disc coloboma, optic nerve pit, advanced glaucomatous optic neuropathy and peripapillary staphyloma are the differential diagnosis of MGS. In MGS, the whole disc is excavated unlike in optic disc coloboma in which the excavation is contained within the optic disc usually in the inferior aspect of the disc and the superior neuro-retinal rim is recognizable.[5] Another differential diagnosis in our patient who also had high myopia is posterior staphyloma. In posterior staphyloma there is excavation of the posterior fundus surrounding the optic disc; the optic disc is usually flat, without a central tuft and the retinal vasculature is normal.[5] Posterior staphyloma is unlikely in our patient because the posterior fundus as well as the disc was excavated and there was a central tuft of tissue on the disc with abnormal retinal vasculature.[5]

CONCLUSION

MGS is an anomaly of the Optic nerve head usually presented with different ocular and non-ocular abnormalities such as reduced vision, strabismus, high refractive errors, retinal detachment, and various binocularity dysfunction. MGS can exist in our African population, so it should be considered always when evaluating any child with unilateral and or bilateral visual impairment as it may be the cause of severe visual impairment in early childhood and subsequent development of amblyopia.

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