Case Report

Morning Glory Disc Anomaly: A Case Report

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Abstract

A rare case of congenital anomaly of the optic disc is presented to draw attention to the occurrence of this anomaly in rural India. The typical case presented with excavated, enlarged colobomatous optic nerve head filled with glial tissue surrounded by peripapillary pigmentary abnormalities. Ocular complications in affected eye may include strabismus, retinal detachment and reduced visual acuity. Contralateral eye may also be involved as a part of anterior chamber cleavage syndrome. It may have systemic associations as in Aicardi syndrome. Therefore, the comprehensive ophthalmic examination of both eyes using various ocular investigations for its early diagnosis and of associated systemic disorders and complications is essential.

Keywords: Case report, congenital anomalies, morning glory, optic disc anomaly

Introduction

Morning glory disc anomaly (MGDA) is a rare congenital malformation that results from incomplete formation of the optic nerve *in utero*.[1] This condition may have been first reported by Pedlar in 1961,[2] but the term was coined by Kindler in 1970 because of the resemblance of malformed optic nerve head with the tropical morning glory flower.[3] Usually, the condition presents unilaterally[2,4] and is reported more commonly in women.[2,5,6] However, there are rare cases of bilateral presentation.[7] The embryonic origin of this syndrome is still not known.[8] The most likely developmental interruption occurs at the 4–5 weeks stage of embryonic growth.[9] According to Pedlar,[2] the defect was secondary to faulty closure of the posterior sclera with herniation of the optic disc. Others postulated that it should be considered as primarily mesenchyme abnormality.[10] In contrast, Pollock argued that there was distal optic stalk dysgenesis, which failed to close, leaving anomalous persistence of the optic cup cavity into the optic stalk.[6]

This report reviewed the clinical features and characteristics of MGDA and its possible systemic associations, with the role of various ocular investigations in its diagnosis.

Case Report

A 27-year-old Indian boy presented for eye examination in Out Patient Department (OPD). According to his father, his right eye (RE) had been bad since he was very young. His birth history and family history were not eventful. His mental status was normal. His best corrected visual acuity for distance with cycloplegic refraction was Oculus Dexter (Right eye) (OD) −0.5 Dioptric Spherical (DS), −2.5 Dioptric Cylindrical (DC) × 110°, 6/18; Oculus Sinister (Left eye) (OS) −0.25 DS, 6/6 using Snellen's chart. His near acuity was OD J3 and OS J1+ according to Jaeger. Examination of anterior segment and intraocular pressures were within the normal range (Goldmann applanation tonometry OD = 13 mmHg; OS = 16 mmHg). There was isocoria and mild relative afferent pupillary defect (RAPD) in RE. Lang stereo test showed gross abnormality in stereopsis. His performance on Ishihara colour test using both eyes together was poor. Ocular alignment was orthophoric and extra ocular examination was full in all directions of gazes. Keratometric values were OD K1/K2 43.00/43.50 and OS K1/K2 42.00/42.50.

Ophthalmoscopic fundus examination of RE presented an enlarged and excavated funnel-shaped disc with elevated tuft of glial tissue (whitish tissue) centrally, thin radial

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retinal vessels emerging at the margins of the optic disc from under the central tissue and followed a relatively straight course to the periphery of the retina with annulus of peripapillary pigmentary changes. The *A/V* ratio was approximately 1/1.5. The foveal reflex was dull with no involvement of macula in the excavation, and peripheral retina was intact without suggestion of retinal detachment [Figure 1a and b]. Fundus of left eye (LE) was normal.

B-scan ultrasonography RE, at posterior pole, showed an anomalous excavation conoid in shape with the disc at the base. Scan of posterior pole of LE was normal.

Magnetic resonance imaging (1.5 T) of the cranium and both the orbits with gadolinium did not reveal any pathology.

Using the Automated Humphrey Field analyser (Carl Zeiss Meditec, Germany) central 30-2 threshold test (Swedish Interactive Threshold Algorithm (SITA) standard, stimulus

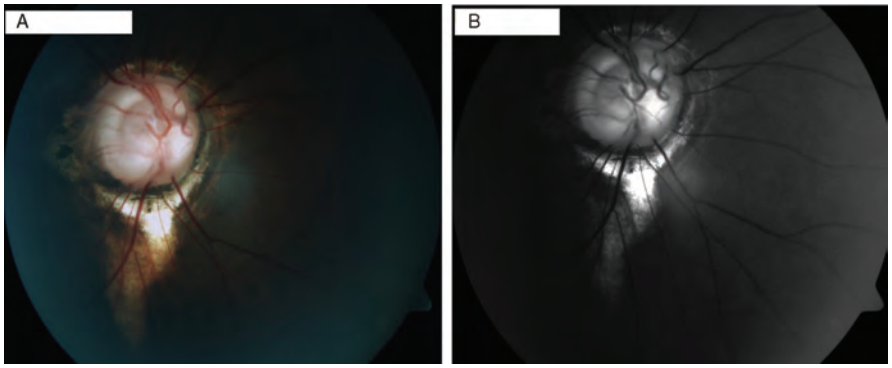

Figure 1: (a) RE coloured fundus photograph showing an enlarged and excavated funnel-shaped optic disc with an elevated central tuft of whitish glial tissue with thin radiating retinal vessels emerging at the optic disc margin from under the central tissue and following a relatively straight course to the periphery of the retina with annulus of peripapillary pigmentary changes. (b) Red free fundus photograph of right eye optic disc

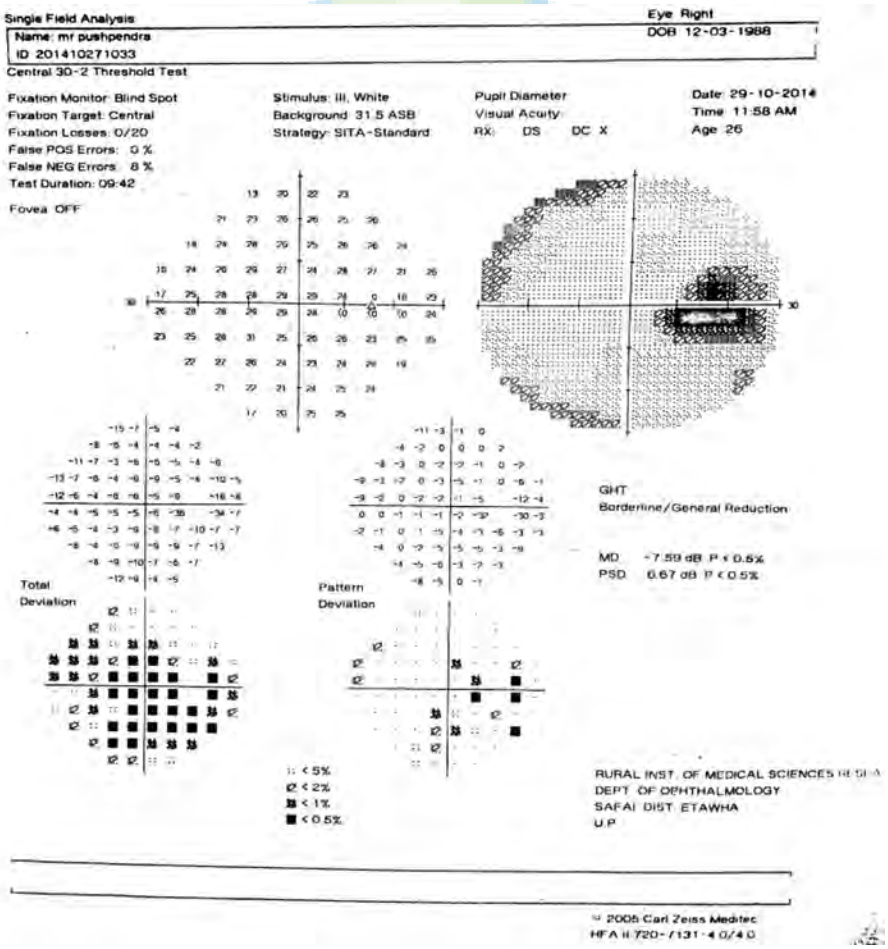

Figure 2: Automated HFA central 30-2 threshold single-field analysis of RE reveals an enlarged blind spot

III white, fixation target central and fixation monitor blind spot), single-field analysis of RE revealed an enlarged blind spot [Figure 2]. LE field analysis was within normal limits.

Cirrus High Definition (Spectral Domain) Optical Coherence Tomography (HD-OCT) RE showed [Figure 3] centrally excavated enlarged disc (disc area 2.56 mm^2) with enlarged *C*:*D* ratio and loss of neuroretinal rim with reduced retinal nerve fibre layer thickness in superior, nasal and temporal quadrant. Three-dimensional reconstructed view of the optic disc cube showed central conoid excavated optic disc [Figure 4]. Normal central macular sub-field thickness implicated the sparing of macula [Figure 5]. Scan for LE was found to be within the normal range.

Fundus fluorescein angiography (FFA) was done to look for any determined retinal break or fluid in sub-retinal layer in the posterior pole. Both eye images were normal.

Discussion

The loss of vision in Morning Glory Syndrome (MGS) may be because of the presence of macular abnormalities[9] or secondary to development of anisometropic or strabismic amblyopia.[3] In our case, the cause of reduced visual acuity in RE is anisometropic amblyopia.

Usually most cases are seen as isolated ocular abnormalities.[3,9] Ocular anomalies commonly observed in the affected eye with MGS include RAPD,[5,9] visual field defects such as enlargement

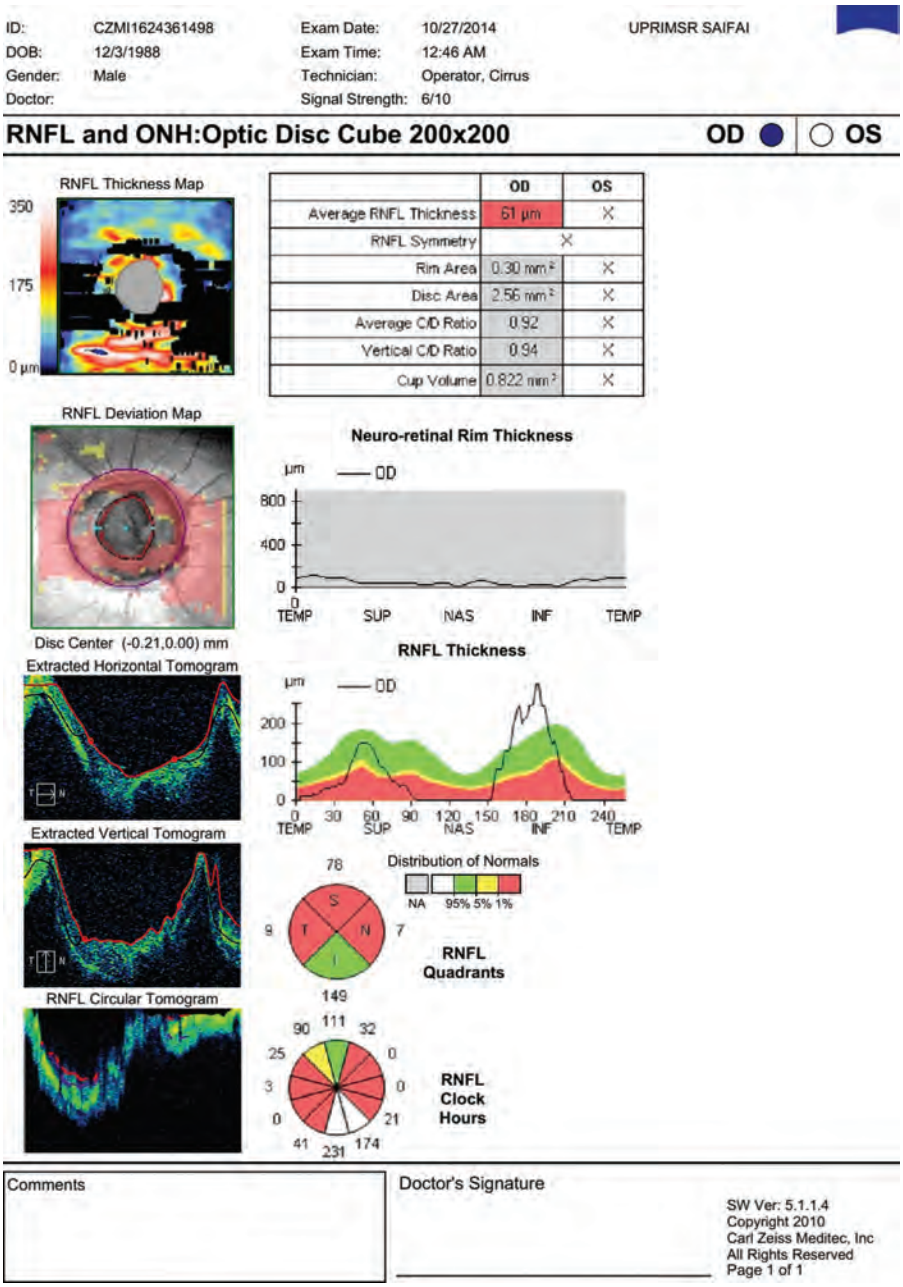

Figure 3: Cirrus HD-OCT RE shows centrally excavated disc with enlarged *C*:*D* ratio and reduced retinal nerve fibre layer thickness

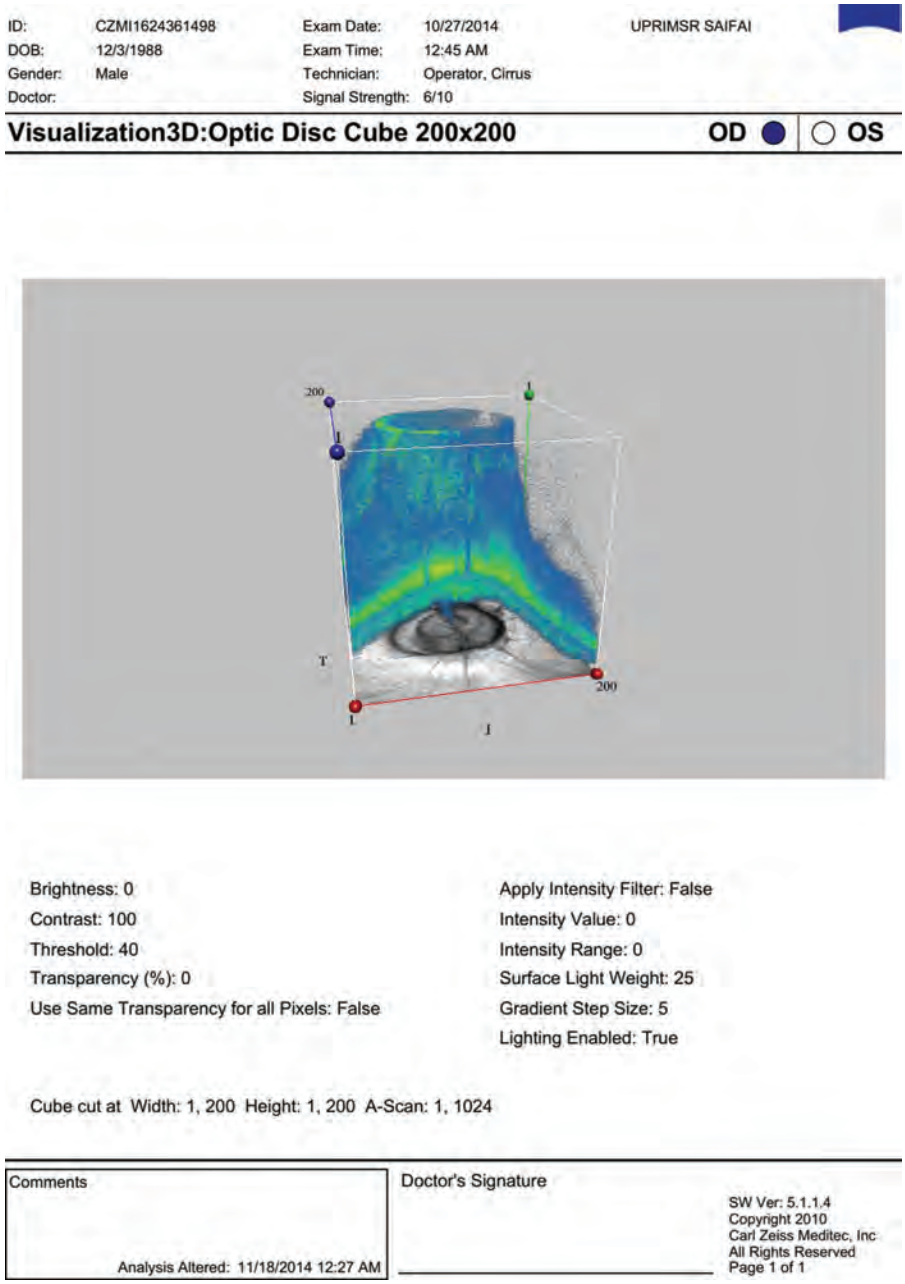

Figure 4: RE shows central conoid excavated optic disc cube in 3-dimensional reconstructed view

of blind spot and/or dense central scotoma,[11] strabismus,[3,4,12] mild–moderate myopia[3,12] and retinal detachment,[10] presence of hyaloid artery remnant,[3,9] ciliary body cyst,[9] congenital cataract,[9,12] lid haemangioma,[3,13] vitreous cyst,[9,12] posterior lenticonus, aniridia,[13] posterior sub-capsular cataract[14] and pre-retinal gliosis.[3,5]

OCT plays a significant role in early diagnosis and evaluation of possible sub-retinal fluid, thus providing information regarding the pathogenesis and associated clinical features.[15]

There are very few reports that did quantitative OCT analysis of the optic nerve head and Retinal Nerve Fibre Layer (RNFL) thickness in cases of MGDA.

OCT analysis (quantitative) of a case of isolated MGDA by Srinivasan *et al.*[16] showed an enlarged optic disc and cup with increased RNFL thickness temporally, thus violating Inferior, Superior, Nasal and Temporal (ISNT) rule and sub-normal macular thickness.

Our case report shows an enlarged optic disc and cup with thinning of RNFL and normal macular thickness.

Various systemic associations have been well documented in MGS[4-6] including congenital forebrain abnormalities such as basal encephalocele (trans-sphenoidal and spheno-ethmoidal) and endocrine alterations,[9,17] midline facial defects including hypertelorism, cleft lip or cleft palate,[4,5,17] renal hypoplasia and other renal abnormalities.[5] Rarely, it

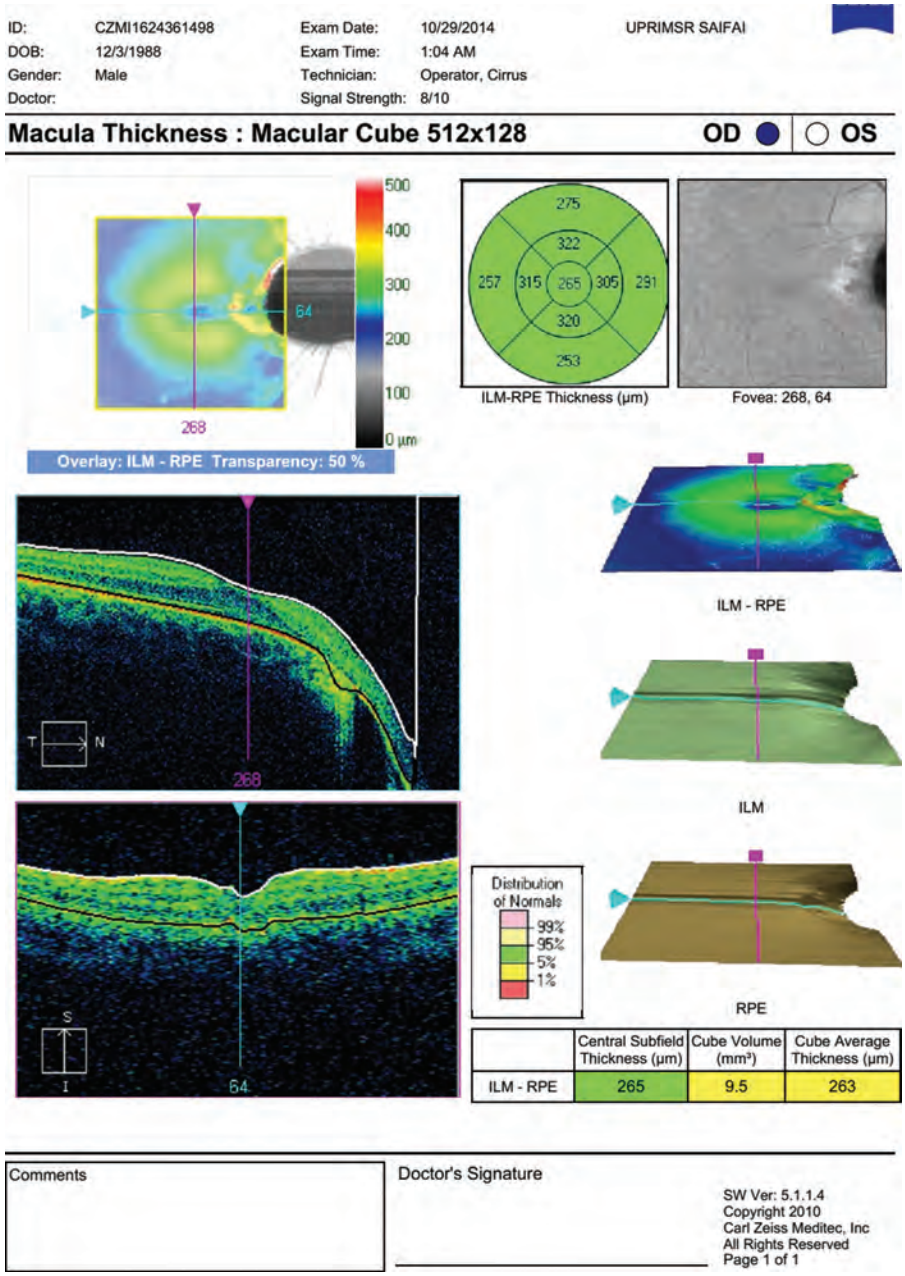

Figure 5: Cirrus HD-OCT RE macular cube (512 × 128) scan shows normal central macular sub-field thickness implicating the sparing of macula

can, however, be associated with syndromes such as Aicardi's syndrome[18] and Down's syndrome.[19]It should be differentiated from other congenital optic disc anomalies such as optic disc coloboma[12] and peripapillary staphyloma. Typical optic disc coloboma is bilateral and presents as a central crater that may resemble glaucomatous cupping.[12] In peripapillary staphyloma there is developmental weakness of the posterior sclera leading to stretching of choroid and exposure of sclera with normal appearing sunken optic disc below the surrounding retinal level.

As a result of variable presentation, various ocular investigations such as B-scan, OCT and FFA are only helpful in confirming the diagnosis and early detection of complications. In view of the presence of a tuft of whitish tissue within the colobomatous elevated optic disc and abnormal presentation of central retinal vessels, our case would be best classified as a case of isolated MGDA. It is non-progressive in nature and does not require treatment.[12] However, because of its association with other ocular anomalies, high risk for developing neuro-sensory retinal detachment and possible systemic abnormalities, early accurate diagnosis and monitoring are essential.

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Conflicts of interest

There are no conflicts of interest.

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