The Spectrum in the Morphology of the So-Called "Morning Glory Disc Anomaly"

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ABSTRACT

Eight cases illustrating the different configurations of the morning glory disc anomaly are presented. The spectrum of the appearance is due to variability in excavation of the disc, amount of glial tissue and hyaloid system remnants in the center of the disc, and degree of peripapillary pigmented changes. There is no correlation between optic disc configuration and visual acuity. This anomaly should be suspected in infants presenting with unilateral retinal detachment.

INTRODUCTION

The so-called “morning glory anomaly” of the optic nerve head is a congenital malformation of the optic disc area that classically has been characterized by an excavated, funnel-shaped optic disc with white tissue at the center, surrounded by a pigmented raised annulus of subretinal tissue. Although this condition was recognized in the German literature by Handman in 1929, and others later, Kindler is given credit for its delineation and for likening it to the flower of the same name; we are aware of the use of the flower’s name several years before Kindler’s report (Reese AB, personal communication, 1966).

Since Kindler’s report, a large number of papers appeared and several ocular and craniofacial anomalies have been associated with the morning glory disc anomaly (MGDA) (Table 1). Although only some cases show the classical “morning glory” configuration, the term has been used broadly to describe atypical dysplastic optic discs; no attention has been given in the literature to the morphological variations in what is probably one malformation process. We present eight cases that illustrate the various configurations of the optic papilla and surrounding area in MGDA. The literature on this condition is summarized briefly, and an attempt is made to correlate final visual acuity with the various nerve head configurations.

<table>
<thead>
<tr>
<th>TABLE 1</th>
<th>Associated Anomalies With the Morning Glory Disc Anomaly</th>
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</thead>
<tbody>
<tr>
<td>Associated Ocular Anomalies</td>
<td>In Same Eye</td>
</tr>
<tr>
<td>Cataract</td>
<td>Microphthalmos</td>
</tr>
<tr>
<td>Strabismus</td>
<td>Anterior chamber cleavage</td>
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<tr>
<td>Foveal hypoplasia</td>
<td>syndrome</td>
</tr>
<tr>
<td>Retinal detachment</td>
<td>Duane syndrome</td>
</tr>
<tr>
<td>Hyaloid remnants</td>
<td>Microcornea</td>
</tr>
<tr>
<td>Ciliary body cyst</td>
<td>Optic pit</td>
</tr>
<tr>
<td>Vitreous cyst</td>
<td>Retinal vascular tortuosity</td>
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<tr>
<td>Preretinal gliosis</td>
<td>Pupillary membrane remnants</td>
</tr>
<tr>
<td>Lid hemangioma</td>
<td></td>
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<tr>
<td>Pupillary membrane remnants</td>
<td></td>
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<tr>
<td>Aniridia</td>
<td></td>
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<tr>
<td>Microphthalmos</td>
<td></td>
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<tr>
<td>Nasolacrimal duct obstruction</td>
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</table>

Associated Systemic Abnormalities

Hypertelorism
Basal encephalocoele
Absent corpus callosum
Cleft lip and palate
Renal abnormalities

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The authors wish to thank Dr. Kamel Itani from the American University of Beirut, Department of Ophthalmology, for referring case 4.
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TABLE 2
Summary of Pertinent Ocular Findings in Eight Patients With the Morning Glory Disc Anomaly

<table>
<thead>
<tr>
<th>Case</th>
<th>Sex</th>
<th>Age</th>
<th>Eye</th>
<th>Vision</th>
<th>Miscellaneous Findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>M</td>
<td>22 yrs.</td>
<td>OD</td>
<td>CF</td>
<td>Vascular tortuosity in fellow eye</td>
</tr>
<tr>
<td>2</td>
<td>M</td>
<td>13 yrs.</td>
<td>OS</td>
<td>CF</td>
<td>Hyaloid remnants</td>
</tr>
<tr>
<td>3</td>
<td>F</td>
<td>16 yrs.</td>
<td>OD</td>
<td>HM</td>
<td>Exotropia</td>
</tr>
<tr>
<td>4</td>
<td>M</td>
<td>7 yrs.</td>
<td>OS</td>
<td>CF</td>
<td>Hyaloid remnants</td>
</tr>
<tr>
<td>5</td>
<td>F</td>
<td>2 mos.</td>
<td>OD</td>
<td>?</td>
<td>Vascular tortuosity in fellow eye</td>
</tr>
<tr>
<td>6</td>
<td>M</td>
<td>1 yr.</td>
<td>OD</td>
<td>?</td>
<td>Exotropia</td>
</tr>
<tr>
<td>7</td>
<td>F</td>
<td>11 mos.</td>
<td>OS</td>
<td>CF</td>
<td>Pedigree of nanophthalmos</td>
</tr>
<tr>
<td>8</td>
<td>F</td>
<td>10 mos.</td>
<td>OS</td>
<td>20/100</td>
<td>Familial aniridia</td>
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<td>Hyaloid remnants</td>
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<td></td>
<td>Optic pit and abnormal disc in fellow eye</td>
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<td></td>
<td></td>
<td></td>
<td>Microphthalmos</td>
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<td></td>
<td>Retinal detachment</td>
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<td></td>
<td></td>
<td></td>
<td></td>
<td>Esotropia</td>
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<td></td>
<td></td>
<td></td>
<td></td>
<td>Microcornea</td>
</tr>
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<td></td>
<td></td>
<td></td>
<td></td>
<td>Hyaloid remnants</td>
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<td>Exotropia</td>
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<td></td>
<td></td>
<td>Hyaloid remnants</td>
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<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Pupillary membrane in fellow eye</td>
</tr>
</tbody>
</table>

OD = right eye; OS = left eye; CF = counting fingers; HM = hand motions.

FIGURE 1: Case 1. Configuration of the optic nerve head area in MGDA. Note variability in disc excavation, gliotic tissue at center of disc, and peripapillary pigmentation in Figures 1 through 7.

FIGURE 2: Case 2. Configuration of the optic nerve head area in MGDA.

MATERIALS AND METHODS

The ocular findings, associated ocular anomalies, and other pertinent information on eight patients with MGDA are summarized in Table 2. In seven patients, the anomaly was unilateral; in case 5, the fellow disc was slightly dysplastic and an optic pit was also present. The different appearances of the discs are illustrated in Figures 1-7. No photographs of the optic nerve head are available in case 6 because a retinal detachment developed in the affected eye between the time of initial diagnosis and the first follow-up visit 1 month later, when fundus photography was planned. Three vitreoretinal surgical procedures failed to reattach the retina in this patient.

Exotropia of the affected eye was the presenting sign in cases 3, 4, and 6; the presenting sign was esotropia in case 7. Cases 1 and 2 were seen because of poor vision in the affected eye. In case 5 the MGDA was discovered when the infant was evaluated for aniridia. Case 8 was evaluated initially for nasolacrimal duct obstruction. Visual acuity was finger counting or hand motions in five cases and could not be assessed in the two infants. There were no significant errors of refraction, except for mild myopic astigmatism in most cases. In each patient, both eyes appeared to be of normal and equal size except in cases 6 and 7 where the cornea was smaller in the affected eye. Computed tomography of the orbits showed the affected eye to be smaller in size than the fellow eye in case 6. A-scan ultrasonographic studies to determine the exact antero-posterior diameter of
FIGURE 3: Case 3. Configuration of the optic nerve head area in MGDA.

FIGURE 4: Case 4. Configuration of the optic nerve head area in MGDA.

FIGURE 5: Case 5. Configuration of the optic nerve head area in MGDA. Reprinted with permission of Aeaus Press.

FIGURE 6: Case 7. Configuration of the optic nerve head area in MGDA.

FIGURE 7: Case 8. Configuration of the optic nerve head area in MGDA.

FIGURE 8: Papillary membrane remnants attached to anterior lens capsule in fellow eye of case 8.
the affected eyes have not been performed in any of the cases. The fellow eye was normal in cases 2, 4, 6, and 7. There were remnants of the pupillary membrane attached to the anterior lens capsule in the fellow eye of case 8 (Figure 8). Vascular tortuosity in both eyes was present in cases 1 and 3. Remnants of the hyaloid system were present in the involved eye in cases 2, 3, 5, 7, and 8.

Fluorescein angiography was performed in cases 3 and 4. The retinal vessels appeared to originate from the central retinal arterial circulation. The straight abnormal vascular pattern, arcade formation near the disc, and bridging retinal vessels were demonstrated (Figure 9). There was moderate staining at the nerve head in late frames (Figure 9), but there was no leakage of the fluorescein dye.

Computed tomography was performed in cases 5 and 6, and in both cases showed the cone-like widening of the optic nerve head silhouette as it leaves the globe. Case 5 had familial aniridia and has been reported elsewhere; case 4 occurred in a pedigree of recessive nanophthalmos.

Visual acuity is generally poor, ranging from 20/100 to poor light perception, although patients with good visual acuity have been reported. We have tried, through a review of published cases that included fundus photographs and visual acuity measurements, to correlate morphology of MGDA to visual acuity. We were not able to find any particular correlation between the two variables. If such a correlation were found, it would have been possible to have one predictive clue of final visual acuity in these patients. We recommend, however, correction of significant refractive errors and conservative patching of the better eye in these patients, in order to reverse any amblyopia that may be present which may be contributing to the poor visual acuity. We have been able to achieve this with case 8, where patching allowed us to reach a final visual acuity of 20/100, the patient having had poor fixation in the affected eye when first seen. Strabismus should be corrected surgically if good fixation is achieved with the affected eye, or if the deviation is cosmetically blemishing.

The three distinguishing ophthalmoscopic features of MGDA are: an enlarged and often excavated disc; a central core of white glistening tissue; and a variably elevated and pigmented ring of peripapillary tissue. Retinal vessels originate from the central retinal artery and proceed from the disc in a straight course to the retinal periphery. Arcade-shaped vascular formations are seen adjacent to the disc, and retinal vessels may divide at right angles. Usually no abnormal leakage from the disc or vessels is seen on fluorescein angiography. We have noted vascular tortuosity of retinal vessels in affected and fellow eyes in two of our patients. This observation has not been reported previously. The variable amount of fibrous tissue and hyaloid system remnants in the excavated disc, the degree of sheathing of the retinal vessels as they emerge from the disc margins, and the inconsistency of the peripapillary pigmentary changes account for the different morphologic configurations in MGDA. Additional factors such as the presence of heterotopic elements, especially myofibroblasts and adipose tissue, as demonstrated in histopathologic studies, may also alter the appearance of the dysplastic disc. Such myofibroblastic tissue may account for the contractions occasionally observed in such dysplastic discs; what previously has been reported as contractile peripapillary staphylomas may be only examples of yet another configuration of MGDA.

A number of ocular and craniofacial anomalies have been associated with MGDA and are summarized in Table 1. The most significant ocular complication of MGDA is the development of retinal detachment in up to one third or more of cases. The detachment is usually of the serous variety, and it has been suggested that an accumulation of cerebrospinal fluid under the retina through a communication between the two spaces in the malformed papilla may be responsible for the detachment. The same mechanism has been postulated to account for the serous macular and retinal detachment with optic nerve head pits. The presence of an optic pit in the fellow eye of case 5 and in a recently reported case suggests that MGDA and optic pit...
may share a common faulty embryologic developmental process.

In addition to conventional vitreoretinal surgery, some authors have recently advocated the use of optic nerve sheath fenestration to decompress the subretinal space through the abnormal communication in the nerve head and prevent recurrence of the detachment. Combined rheumatogenous/scleral retinal detachment has also been reported in MGDA and treated successfully using conventional surgical techniques. Because patients with MGDA may have retinal detachment in the involved eye, this anomaly should be suspected in infants or children with unilateral retinal detachment. Computed tomography or magnetic resonance imaging (MRI) are helpful in such cases and may demonstrate the funnel-shaped deformity of the optic nerve as it is leaving the globe. Of the craniofacial anomalies associated with MGDA, transsphenoidal encephaloceles are the most serious and should be searched for.

The embryologic defect leading to MGDA is not clear. A mesenchymal abnormality resulting in faulty closure of the posterior scleral wall and lamina cribrosa with subsequent herniation of retinal and neural tissue along with dysplasia of the nerve head is possible and is supported by some histopathologic evidence. A concomitant closure defect of the embryonic fissure at its most posterior end cannot be ruled out. Dempster et al have reconciled both views and suggest that the excavation results from a disturbance in the relative growth of mesoderm and ectoderm, but these authors also believe that the basic defect is probably mesenchymal. The associated craniofacial anomalies also support a mesenchymal defect in the pathogenesis of MGDA, because most of the cranial structures are derived from mesenchyme.

Ophthalmologists should be aware of the various configurations of the MGDA as this allows for accurate diagnosis and differentiation from inflammatory and tumorous lesions of the nerve head. Infants with unilateral retinal detachments should be evaluated for the presence of MGDA and associated cranial abnormalities.

REFERENCES

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