Morning Glory Syndrome: Clinical, Computerized Tomographic, and Ultrasonographic Findings

Paul Harasymowycz, MD; Lyne Chevrette, MD; Jean-Claude Décarie, MD; Nelly Hanna, OCC, MD; Maryam Aroichane, MD; Jean-Louis Jacob, MD; Jean Milot, MD; and Magda Homy, MD

ABSTRACT

Objective: To evaluate the ophthalmic, radiologic, and ultrasonographic findings in morning glory syndrome.

Patients and Methods: Retrospective review of patients’ charts.

Results: Twenty-one patients were included. Visual acuity was 20/200 or better in 50% of the eyes. Three eyes developed a retinal detachment. When computerized tomography of the orbit demonstrated peripapillary scleral staphyloma and intraocular calcifications, this correlated with poor visual acuity. Computerized tomography of the brain revealed abnormalities in three asymptomatic patients. B-scan ultrasonography demonstrated retinal tissue overhanging the peripapillary scleral staphyloma.

Conclusions: Morning glory syndrome has a spectrum of severity, with most patients retaining useful vision. Orbital and cerebral computerized tomography scan as well as B-scan ultrasonography may help with diagnosis and management.


INTRODUCTION

Morning glory syndrome was noted in the early German ophthalmic literature by Handmann,1 but it was first compared with the morning glory flower by Kindler in 1970.2 This anomaly of the optic nerve is classically described as having a funnel-shaped, excavated optic disc; a central, elevated white glial tuft; a peripapillary hypopigmented or hyperpigmented retinal ring; and radially oriented retinal vessels that extend from the outer edge of the optic disc (Fig. 1). In addition, there is variable displacement of the macular luteal pigment to the temporal edge of the disc (Fig. 2), and a spectrum of morphology may be found.3,4 Contractile membranes have also been noted.5

The morning glory disc anomaly is usually unilateral and noninherited, and it can occur as an isolated ocular finding; however, bilateral and familial cases have been reported.6,7 It is often associated, however, with ipsilateral or contra-
lateral ocular abnormalities.7-11 Up to one-third of patients have been reported to develop a non-rhegmatogenous retinal detachment, which is difficult to manage surgically.12,13 Visual acuity has also generally been noted to be poor, but a trial of contralateral occlusion therapy has been advocated.3 Systemic findings such as renal or endocrine deficiencies,14-16 brain or midline defects including cleft lip and palate, and encephaloceles may be noted.17-22 Orbital computerized tomography scans have demonstrated typical posterior scleral staphyloma as well as anterior optic nerve thickening.5,23-26

We report the clinical, orbital, and cerebral computerized tomography scan and B-scan ultrasonographic findings in a series of 22 eyes diagnosed as having morning glory syndrome at the Pediatric Ophthalmology Service of Sainte-Justine’s Hospital between 1971 and May 2002.

**Patients and Methods**

The inclusion criterion for this study was photographic evidence of the morning glory disc anomaly. Photographic charts at the pediatric clinic including such a diagnosis and those with atypical optic nerve colobomas were reviewed. The diagnosis of the morning glory syndrome was confirmed independently by three of the authors (PH, LC, MA). Optic disc morphology had to include a central glial tuft, peripapillary pigment changes, and radially oriented vessels exiting from the edge of the disc. Twenty-six charts were found, but 5 were excluded and reclassified as myelinated fibers (n = 1), myopic peripapillary staphyloma (n = 2), optic nerve toxoplasmosis (n = 1), and pseudo-papilledema (n = 1). One patient had bilateral morning glory disc anomaly.

Hospital charts were reviewed, and all of the patients were contacted for a follow-up visit. Sixteen patients were available for reexamination. A thorough pregnancy and family history and an ophthalmic examination were performed. Fourteen patients had computerized tomography scans of the brain and orbits with contrast that were reviewed. Films were noted for axial length and the presence or absence of peripapillary staphyloma, optic nerve sheath thickening or tortuosity, and calcifications. B-scan ultrasonography had been performed on 10 patients.

**Results**

Of the 21 patients who comprised this case series, 11 were male (52%) and 10 were female (48%). Ten patients had involvement of the left optic nerve, 10 had involvement of the right, and 1 had bilateral involvement.

The age at presentation ranged from 1 month to 8 years, with a mean age of 24 months. The clinical presentation was strabismus or decreased visual acuity in all but five cases. One patient was referred to rule out papilledema and another was referred
to rule out glaucoma. Morning glory syndrome was discovered on routine eye examination in one case and during evaluation for seizure disorder in a second case. Finally, one patient presented due to a cataract in the contralateral eye.

Family history was obtained for 16 patients available for a follow-up visit. It revealed no cases of optic nerve anomalies, but 5 (31.3%) of the patients had a history of strabismus. In addition, 3 mothers reported difficulties early in the first trimester of gestation, including infection \( n = 2 \) and vaginal blood loss \( n = 1 \).

In 12 (55%) of 22 eyes, the optic nerve photographs demonstrated "macular capture": the presence of yellow xanthophyll pigment at the temporal edge. In 6 (27%) of the eyes, the disc appeared depressed in the scleral staphyloma; in 16 (73%) of the eyes, the disc appeared elevated. Three eyes had small telangiectasias on the disc surface, and three others had vessel straightening and sheathing. One patient was noted to have progressive disc elevation during a 6-year period.

Snellen visual acuity was obtained in 20 of the 22 eyes. It was measured by either Allen pictures, tumbling Es, or HOTV letters and varied between 20/20 and hand motion. Ten (50%) of these 20 eyes had a visual acuity of 20/200 or better. Six (30%) had a visual acuity of 20/40 or better, with 5 of these patients showing no macular capture on optic nerve photographs.

Strabismus was present in 19 (91%) of 21 patients, 12 (57%) of whom had esotropia and 7 (33%) of whom had exotropia. There were four cases of dissociated vertical deviation.

Cycloplegic refraction performed on the 22 eyes revealed that 13 (59%) had myopia ranging from -0.50 to -11.50 D and 7 (32%) had hypermetropia ranging from +0.50 to +8.00 D. Fourteen (64%) of the patients had anisometropia of 2.00 D or more, and 7 had less than 2.00 D or none at all. The mean visual acuity of the latter 7 patients was 6/18.

Therapy for amblyopia with full refractive correction was attempted in 15 patients with the use of occlusion \( n = 14 \) or atropine penalization \( n = 1 \). Two patients demonstrated some improvement; both had a classic morning glory disc anomaly with macular capture as well as a posterior scleral staphyloma demonstrable on computerized tomography scan. The first patient improved from 20/2000 to 20/80, and the second from hand motion to 20/200.

Several anomalies ipsilateral to the morning glory disc anomaly were found. Nine eyes (41%) were microphthalmic with pseudo-ptosis, 2 had microcornea, 1 had a posterior subcapsular cataract, 3 had anterior hyaloid remnants, 1 demonstrated posterior persistent hyperplastic primary vitreous, and 1 had several atrophic chorioretinal spots but negative TORCH titers. Non-rheumatogenous retinal detachment occurred in 3 (14%) of 22 eyes with a mean follow-up period of approximately 5 years. Combined vitrectomy and scleral buckling was
performed in one patient, with anatomic reattachment of the retina and hand motion vision postoperatively. One eye developed phthisis bulbi and was eviscerated. One patient had abnormalities in the contralateral eye that included posterior lenticonus and a subcapsular cataract.

Systemic findings included one patient with precocious adenarche with normal results on endocrinologic and neuroimaging studies and two patients with normal results on neuroimaging and a diagnosis of migraines. One patient had Joubert–Plus syndrome, with psychomotor retardation, cryptorchidism, epicanthus, hypertelorism, cerebellar vermis hypoplasia, oculomotor apraxia, bilateral morning glory disc anomaly, and posterior embryotoxon, and a visual acuity of 20/25 in both eyes. Finally, one patient had unilateral morning glory syndrome with a superior optic pit on the same disc as well as characteristics of Aicardi’s syndrome, including agenesis of the corpus callosum, convulsions, mental retardation, and bilateral chorioretinal lesions.

Computerized tomography of the orbit with contrast was obtained in 15 eyes. Posterior peripapillary staphylomas were demonstrated in 12 cases (80%) and ranged in severity: 8 were large and funnel shaped and 4 were milder in appearance. Thickening, or ectasia, of the optic nerve sheath complex was present in 8 eyes (53%) (Fig. 3). Optic nerve tortuosity was found in 6 eyes (40%); microphthalmos in 6 eyes (40%); and ocular, scleral (Fig. 4), or optic nerve calcifications in 4 eyes (27%). There was a correlation between the presence of a large funnel-shaped posterior scleral staphyloma (classic), calcifications, or both and poor visual acuity in all cases (Table).

Computerized tomography of the brain with contrast was performed in 16 patients. In the two patients mentioned previously, one with Joubert–Plus syndrome and the other with Aicardi-like syndrome, imaging showed partial cerebellar vermis hypoplasia in the former and agenesis of the corpus callosum in the latter. Of 14 neurologically asymptomatic patients, 3 (21%) had demonstrable findings. One patient had a large temporal lobe arachnoid cyst measuring 12 × 33 mm ipsilateral to the eye with the morning glory disc anomaly as well as temporal lobe hypoplasia. Another scan revealed idiopathic frontal lobe leukomalacia. Finally, a third patient had an arachnoid cyst of the medullary velum as well as an unspecified vascular malformation adjacent to the pineal gland.

B-scan ultrasonography had been performed in 10 patients. Excavation of the optic nerve was present in all (100%) of these cases, calcifications were noted in 8 (80%), a central glial tuft in 6 (60%), and microphthalmos in 5 (50%). Interestingly, on review of the B-scan ultrasonography, retinal tissue overhanging the posterior scleral staphyloma was noted. This novel sign, termed the “overhang sign,” was demonstrated in seven cases (Fig. 5). In one case with retinal detachment and no fundus visualization at the time of B-scan ultrasonography, the overhang sign was also clearly visible.
DISCUSSION

This is the largest case series of morning glory syndrome published to date. This study confirms that the morning glory disc anomaly is rare, is non-hereditary, and affects males and females equally. The mean age at presentation was 24 months, with strabismus, poor vision, or both being the usual presenting signs. This series also indicates the spectrum of severity for morning glory syndrome, with most of the patients in this study having useful (> 20/200) vision. Strabismus and anisometropia are common, and patients may respond to occlusion therapy. Ipsilateral and contralateral ocular abnormalities are also common.

Although some have purported that morning glory syndrome arises from a defect in the embryonic fissure, it is clear from histopathologic studies that retinal pigment epithelium is present in the scleral defect and that a mesodermal hypothesis is more likely. In this and other studies, many of the ipsilateral and contralateral ocular abnormalities, such as posterior embryotoxon, included structures derived from neural crest cells. Abnormalities of the optic nerve sheath described here, as well as midline facial defects with encephaloceles, also arise from neural crest cells, thus raising the possibility that morning glory syndrome may be a neurocris-topathy.

As opposed to other studies, which quote a retinal detachment rate of 30%, retinal detachment was seen in only 14% of the eyes in this study with a mean follow-up period of 5 years. With a longer follow-up period, this percentage may increase. In one case, successful surgical reattachment was achieved. Thus, surgery may be warranted in these cases.

Computerized tomography of the orbits may demonstrate typical scleral staphyloma and intracocular calcifications, both of which may be associated with a poor visual prognosis. Morning glory syndrome with retinal detachment and calcifications can be confused with retinoblastoma and should be considered in the differential diagnosis. Also, the thickened and tortuous optic nerve seen in morning glory syndrome may be confused radiologically with an optic nerve glioma.

Computerized tomography of the brain should be performed, as neurologic abnormalities are often found. In comparison with other studies, however, no cases of basal encephalocele were found in this study. However, this may reflect a selection bias because cases had been identified from a photographic database. In addition, no cases of moyamoya disease were detected on computerized tomography of the brain in this series, but magnetic resonance angiography was not performed.

B-scan ultrasound may show intracocular calcification and, more importantly, retinal tissue overhanging the staphyloma (overhang sign), a novel sign of morning glory syndrome. This sign, which was present in the patient with poor visualization due to retinal detachment, may prove to be helpful in similar cases and thus may narrow the differential diagnosis.

Morning glory syndrome has a spectrum of severity, with most patients retaining useful vision. Orbital and cerebral computerized tomography scan as well as B-scan ultrasonography may help with both the diagnosis and the management of this rare anomaly.

REFERENCES


