Morning Glory Disc Anomaly in Association With Ipsilateral Optic Nerve Glioma

Morning glory disc anomaly (MGDA) is typically a unilateral congenital disorder characterized by the funnel-shaped excavation of an enlarged optic disc. Retinal vessels emanate radially beyond a central white core, and the disc itself is encircled by an elevated region of chorioretinal pigmentation. Seros and, less commonly, rhegmatogenous retinal detachment may coexist. Infants with MGDA often manifest strabismus or leukokoria. Morning glory disc anomaly is associated with other midline developmental or vascular malformations. We describe a patient with MGDA and concurrent ipsilateral optic nerve glioma.

Report of a Case. A 26-month-old previously well Asian boy had a 12-month history of exotropia. There were no associated developmental delays, dysmorphic features, or neurocutaneous stigmata. Neurological examination findings were normal. Ophthalmologic examination revealed poor fixation of the right eye with a concomitant exotropia of 40 prism diopters at distance and near. Visual acuity was 20/360 OD and 20/47 OS with Teller acuity cards and there was a trace afferent pupillary defect (early release). Indirect ophthalmoscopy demonstrated classic features of MGDA, poor foveal definition, and possible shallow serous detachment (Figure 1). The left eye was normal.

Magnetic resonance imaging studies revealed enlargement of the prechiasmatic optic nerve and right lateral aspect of the optic chiasm with minimal peripheral enhancement along the retrobulbar right optic nerve and chiasm, consistent with an optic pathway tumor (Figure 2). A retinal defect was noted in the globe near the insertion of the optic nerve, and a shallow detachment was found. The left orbit and optic nerve were normal.

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mal. Magnetic resonance angiographic and venographic findings were normal. The hypothalamic-pituitary axis was normal.

Comment. To our knowledge, we report the first case of an infant diagnosed as having MGDA and ipsilateral optic pathway glioma. Optic pathway tumors may lead to progressive vision loss and visual field defects. Our patient has no vision deficit in the contralateral eye at this time.

The etiology of MGDA is yet unknown but may result from abnormal development of the lamina cribrosa and posterior sclera. Persistent fetal vasculature in association with MGDA has been described. Vision is typically poor, with only 30% of patients achieving a visual acuity of 20/40 or better. Afferent pupillary defect is also common because the disorder is typically unilateral and retinal detachment can occur in the affected eye.

Morning glory disc anomaly has been reported in association with a variety of midline defects including hypertelorism, cleft lip and palate, agenesis of the corpus callosum, type I Chiari malformation, encephalocoele, and endocrinologic abnormalities involving the pituitary gland. Central nervous system vascular anomalies including moyamoya syndrome are seen with increased frequency. Morning glory disc anomaly is rarely associated with genetic disorders, although 2 cases have been reported in the setting of neurofibromatosis type 2, with distinctive clinical features not found in our case.

We recommend that any patient with MGDA undergo dedicated magnetic resonance imaging for evaluation of other midline defects as well as magnetic resonance angiography given the association with vascular abnormalities. Early recognition and management of amblyopia and possible retinal detachment are essential to optimize visual acuity. Our patient, whose MGDA is seen in association with optic nerve glioma, faces the additional risk of vision loss in the contralateral eye. Thus far, he has not required any tumor-directed therapy. Malignant changes in the size of the tumor and/or change in visual acuity or visual fields would prompt tumor-directed therapy to preserve vision in the contralateral eye.

This report expands on the spectrum of clinical associations with MGDA.

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Orbital Silicone Oil Granuloma Discovered During Enucleation

Silicone oil tamponade is commonly used in surgical repair of recalcitrant retinal detachments. Complications of intraocular silicone oil include corneal decompensation, cataract formation, and glau-