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Case report

Morning glory disc anomaly and ipsilateral sporadic optic pathway glioma

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ABSTRACT

Purpose: To present a rare case of morning glory disc anomaly in association with an ipsilateral low grade glioma.

Observations: A 5 year old male presented with a unilateral morning glory disc anomaly and an ipsilateral sporadic optic pathway glioma with chiasmal involvement. After a strict patching regimen his vision improved from 20/400 to 20/80.

Conclusions and importance: This report strengthens the recommendation for brain magnetic resonance imaging in patients with morning glory disc anomaly. Patching of the contralateral eye should be attempted since the role of amblyopia may be significant.

1. Introduction

Morning glory disc anomaly (MGDA) is a congenital anomaly of the optic disc with a characteristic appearance. Findings include funnel-shaped excavation of the a markedly enlarged optic disk opening, a central glial tuft, peripapillary chorioretinal pigmentation and spoke-like vessels radiate outward from the edge of the anomalous disc. It is typically a unilateral disorder and may result from abnormal development of the lamina cribrosa and posterior sclera.[1] Several facial and midline defects have been associated with MGDA and brain imaging is warranted in all cases. In our case brain imaging showed an ipsilateral optic pathway glioma (OPG) with chiasmal involvement. To the best of our knowledge this association has been described only once in the literature.[2]

2. Case report

A 5 year old Caucasian male was referred to our center for evaluation of possible neurofibromatosis type 1 (NF1) and for the management of unilateral right MGDA. At 2 years of age, his parents noticed that the right eye was turning out. He was examined in another center and was noted to have reduced visual acuity, exotropia and MGDA in his right eye. The initial visual acuity in the right eye was 20/400 and improved to 20/125 with patching and glasses. However, this plateaued and further evaluation was pursued. Magnetic resonance imaging (MRI) of the brain and orbits revealed an OPG involving the prechiasmatic optic nerve and right lateral aspect of the optic chiasm. At this point the patient was referred to our center. His exam revealed visual acuity of 20/125 in the right eye and 20/20 in the left eye. He had right exotropia of 20^{Δ}. There was a +1 right afferent pupillary defect. The right optic nerve appearance was compatible with MGDA (Fig. 1). The patient did not have a family history of NF1 and there were no clinical signs of NF1. Relevant MRI images are given in Fig. 2. Magnetic resonance angiography and venography was normal. Sequencing for *NF1* and *PAX6* genes was negative. After 6 months of patching (four hours per day) his vision has further improved to 20/80.

3. Discussion

We present a rare case of MGDA in association with a low grade glioma. MGDA is typically sporadic and unilateral with a characteristic appearance of the optic nerve. Rare bilateral and familial cases have been described, some of which have been reported to involve *PAX6* gene mutations.[3] Vision is usually poor with only 30% of patients achieving an acuity of 20/40 or better.[4] Since the role of amblyopia may be significant, patching of the contralateral eye should be attempted. In our case, visual acuity has improved with patching but not normalized. The pathologic anatomy of the optic disc, or the presence of OPG, or both, probably prevented further improvement.

Since a variety of facial and midline defects has been associated with MGDA, brain MRI with and without contrast is warranted in all cases. Associated pathologies include mainly hypertelorism, cleft lip and palate, basal encephalocele and agenesis of the corpus callosum.[1] However, other pathologies have been described. There is also an association between MGDA and Moyamoya disease or other cerebrovascular abnormalities.[5] Therefore, MRI together with MRA are

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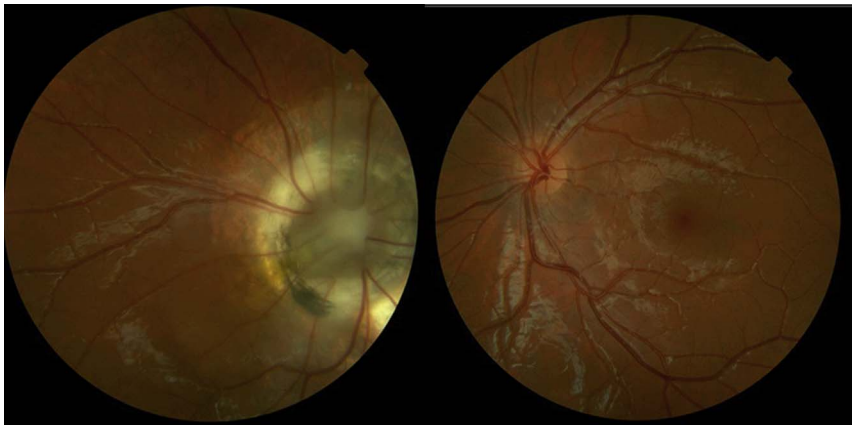

Fig. 1. Fundus photos of both eyes. The optic nerve in the right eye is markedly enlarged and has a funnel-shaped excavation. Retinal vessels emanate radially beyond a central core, and the disc itself is encircled by a partially elevated region of chorioretinal pigmentation and atrophy. This is compatible with morning glory disc anomaly. The left optic nerve and macula are normal.

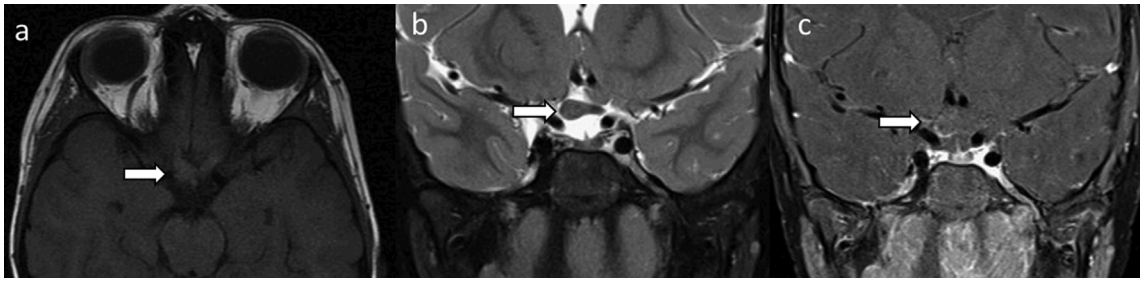

Fig. 2. Magnetic resonance images of the brain and orbits. (a) Axial T1-weighted image showing expansion of the right prechiasmatic optic nerve. (b) Coronal T2-weighted image showing enlargement of the right optic nerve and right lateral aspect of the optic chiasm with increased T2 signal (c) Coronal T1-weighted image after gadolinium injection showing minimal, thin enhancement along the margin of the lesion. White arrows indicate the right optic nerve.

recommended for MGDA patients.[5] In our case brain imaging was negative for all of the above except for an ipsilateral OPG.

Optic pathway gliomas are World Health Organization Grade I or II neoplasms found within the visual pathway that might cause vision loss in children. They are typically diagnosed under the age of 6 years and are strongly associated with NF1.[6] There is only one case in the literature describing the association between MGDA and ipsilateral OPG.[2] Interestingly, in the previous report, there was no evidence of NF1 and the glioma location was similar. Close observation with serial imaging and ophthalmology exams was recommended in both cases.

4. Conclusions

We believe that this report further strengthens the recommendation for brain MRI in patients with MGDA. Diagnosis of ipsilateral OPG with chiasmal involvement may have prognostic value as tumor progression could impact vision in the contralateral eye, which typically has good vision. Together with the knowledge that sporadic OPGs are more aggressive than NF1-related OPGs, closer follow up with serial clinical exams for contralateral optic nerve appearance and function, serial visual fields in older children and serial MRIs is needed and may result in early diagnosis of tumor progression and prompt treatment.

Patient consent

Consent was not obtained but the report does not contain any identifying information.

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Conflict of Interest

The following authors have no financial disclosures: MK, NPS, JLZ.

Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

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