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MRI Evaluation of Vasculopathy and Additional Intracranial Manifestations in Morning Glory Disc Anomaly

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Abstract

Morning glory disc anomaly (MGDA) is rare, but its fundoscopic findings are well documented in the ophthalmologic literature. It is sporadic, without sexual predisposition, and usually unilateral. Furthermore, it is associated with numerous central nervous system anomalies, including vasculopathy. This case series reports four pediatric patients over a 3-year period in whom ophthalmologic evaluation identified MGDA. Magnetic resonance imaging and magnetic resonance angiography were subsequently performed to assess for associated intracranial vascular anomalies. This report is of significance because it demonstrates the spectrum of intracranial vasculopathy in this rare entity.

Keywords: Morning glory disc anomaly, intracranial vasculopathy, pediatrics

Introduction

Morning glory disc anomaly (MGDA) is a rare, sporadic entity without sexual predisposition and is associated with distinct ocular and intracranial manifestations.[1-11] Its enlarged optic disc opening is thought to represent the sequelae of defective embryonic fissure closure or a developmentally enlarged optic stalk[9] and is named after its fundoscopic resemblance to the morning glory flower. Fundoscopic evaluation demonstrates radially oriented thin vessels emanating toward the periphery of the retina.[1,9] Intracranial manifestations are ipsilateral with respect to ocular findings.[1-11] Enlargement of the ipsilateral optic nerve can be present; this may represent an optic pathway tumor.[1-3, 6,11] Intracranial magnetic resonance imaging (MRI) and magnetic resonance angiography (MRA) performed following the diagnosis of morning glory anomaly can reveal a spectrum of intracranial anomalies, which can range from normal intracranial morphology to multiple severe anomalies, including corpus dysgenesis of the corpus callosum, Chiari 1 malformations, sphenoid meningocoele, and vasculopathy with or without a Moya Moya pattern.[1-11]

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We report four cases that demonstrate the spectrum of neuroradiologic findings in MGDA, with specific attention to vasculopathy of Willis's circle.

A retrospective chart review from 2000 to 2020 yielded four patients diagnosed with morning glory anomaly. The submission was approved by the Institutional Review Board of the Mount Sinai School of Medicine in accordance with Mount Sinai's Federal Wide Assurances.

Case Reports

Patient 1: A 3-month-old female was diagnosed with morning glory anomaly OD on ophthalmologic evaluation by a neuro-ophthalmologist. MRI subsequently performed demonstrated normal ocular globe morphology without intracranial manifestations. Imaging is not included (**Figure 1a, b**).

Patient 2: A 6-year-old male presented with decreased visual acuity OS. Ophthalmologic evaluation revealed morning glory syndrome. MRI demonstrated left globe dysplasia without any additional brain anomaly. MRA demonstrated stenosis of the distal left internal carotid artery without the Moya Moya pattern (**Figure 2a-c**).

Patient 3: Eight-month-old male with morning glory OD and posterior ocular globe defect appreciated on MR. MRA revealed stenosis of the right distal internal carotid artery with a Moya Moya pattern of collateralization.

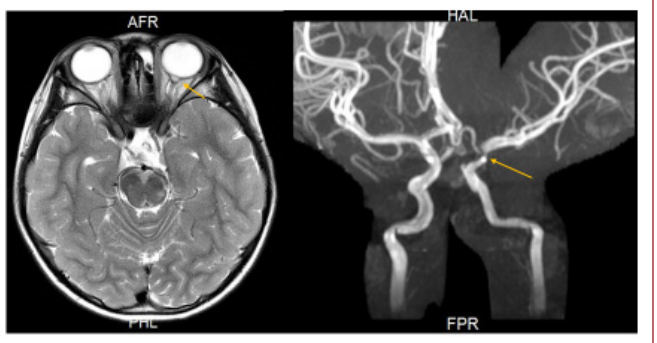

Figure 1a Figure 1b

Figure 1. a. Axial T2 weighted image reveals a defect in the left posterior globe at the optic disc insertion (arrow). **b.** Magnetic resonance angiography demonstrates stenosis of the distal left internal carotid artery (arrow).

Patient 4: An 11-month-old female diagnosed with bilateral morning glory anomaly and enlargement of the intraorbital portions of both optic nerves. Inversion recovery imaging of the nerves demonstrated abnormal signals (not shown), and post-gadolinium images demonstrated enlargement and irregular enhancement of both optic nerves compatible with optic nerve glioma. Sagittal T2-weighted imaging demonstrates sphenoid meningocoele. MRA of the cerebral vasculature demonstrates bilateral distal internal carotid artery stenosis with extension into the M1 segments of the middle cerebral arteries. MRA also demonstrates a Moya Moya pattern with collateralization and significant stenosis of the left posterior cerebral artery (not shown) (**Figure 3a-d**).

Results

This report presents four patients with a diagnosis of MGDA and associated MRI. One patient presented without intraorbital or intracranial abnormalities on MR evaluation. The second patient presented with MGDA with left ocular globe anomaly on MRI as well as stenosis of the distal left internal carotid artery without Moya Moya pattern on MRA. Two patients demonstrated ocular findings and intracranial vascular stenosis with collateralization effort compatible with the Moya Moya pattern. One of these patients was also diagnosed with a sphenoid meningocoele and neurofibromatosis type 1. Although usually unilateral, the fourth patient was diagnosed with a bilateral anomaly.

Discussion

MGDA is a rare congenital syndrome without sexual predisposition. It is usually unilateral although it can be bilateral, as in the fourth submitted case.[10] Fundoscopic findings of MGDA are well documented in the ophthalmology literature.[1-11] MGDAs may present with enlargement of the optic nerve, and neoplasms can be suspected, such as optic pathway gliomas in neurofibromatosis type 1, as demonstrated in the fourth patient.[1-4,6,11]

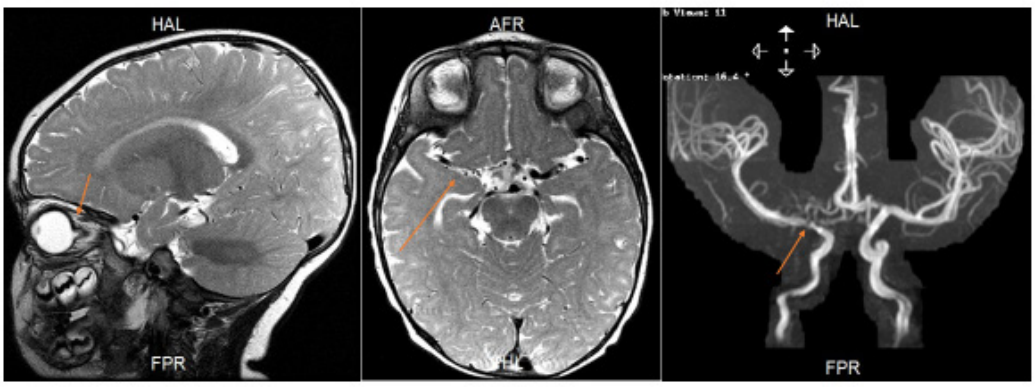

Figure 2a Figure 2b Figure 2c

Figure 2. a. Sagittal T2 weighted image demonstrated a defect in the right posterior globe (arrow). **b-c.** Axial T2 weighted and MR Angiographic images demonstrated collateral vasculature at the level of the distal right internal carotid artery stenosis (arrows).

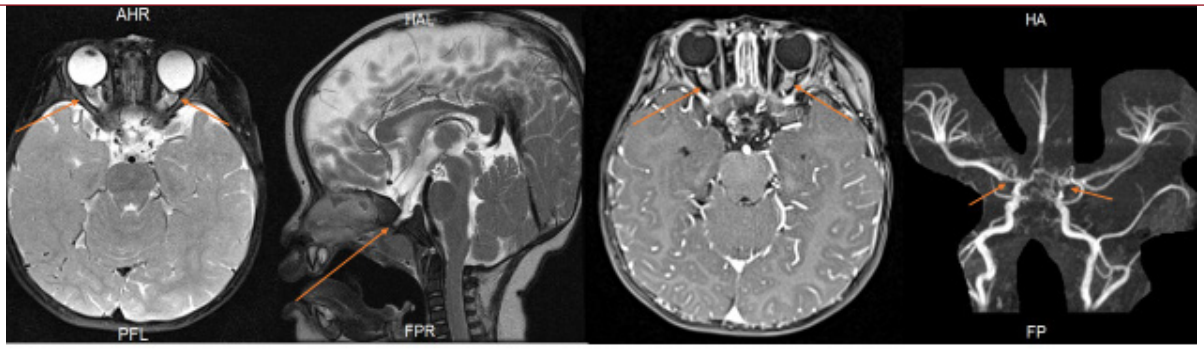

Figure 3. a. Axial T2 fat saturated image demonstrates bilateral ocular defects at the optic disc insertion level (arrow). **b.** Sagittal T2 weighted image demonstrates a sphenoid meningocoele (arrow). **c.** Axial post gadolinium images reveal enlargement and patchy enhancement of both intraorbital optic neves (arrow). **d.** Magnetic resonance angiography reveals bilateral distal Internal Carotid Artery stenosis with Moya Moya pattern (arrow).

Morning glory anomaly is associated with midline defects, such as dysgenesis of the corpus callosum, Chiari 1 malformations, sphenoid meningocoele, and intracranial vasculopathy, such as distal integral carotid artery stenosis and Moya Moya collateralization effort.[5,6,8] Pituitary dysfunction is also established, presumably related to basal cephalocoele development.[7,9]

There are multiple associated intracranial anomalies in morning glory anomaly.[1-11] As a result, when morning glory anomaly is detected, neuroimaging is of value in the patient's clinical assessment.

In summary, this case series demonstrates the constellation of intracranial manifestations of morning glory anomaly with specific emphasis on multiple patterns of intracranial vasculopathy. Therefore, neuroimaging is an important component of evaluation for this condition.

Informed Consent: Retrospectively study.

Author Contributions: Heller GD: Surgical and Medical Practices, Concept, Design, Data Collection or Processing, Analysis or Interpretation, Literature Search, Writing.; Kumar MS: Surgical and Medical Practices, Data Collection or Processing, Analysis or Interpretation.

Conflict of Interest: The authors have no conflicts of interest to declare.

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