

Morning glory syndrome with Moyamoya disease: A rare association with role of imaging

Janardhana Ponnatapura

Department of Radio-Diagnosis, Kempegowda Institute of Medical Sciences, Bengaluru, Karnataka, India

Correspondence: Dr. Janardhana Ponnatapura, 301/1-1, 23 Cross, 6 Block, Jayanagar, Bengaluru - 560 070, Karnataka, India.
E-mail: psjanardhan@yahoo.com

Abstract

Morning glory disc anomaly (MGDA) is a congenital optic nerve anomaly characterized by a funnel-shaped excavation of the posterior globe that incorporates the optic disc. Most cases are isolated and not associated with systemic anomalies. Systemic anomalies include midline cranial facial defects, hypertelorism, agenesis of the corpus callosum, cleft lip and palate, basal encephalocele, congenital forebrain abnormalities, and renal anomalies. We report a case of 4-year-old male child who presented with reduced visual acuity on left eye with poor fixation. The left eye demonstrated 6-diopter esotropia. Examination of fundus revealed features of MGDA. The child was further subjected to magnetic resonance imaging (MRI) and magnetic resonance angiography (MRA) of brain to rule out other associated anomalies. It demonstrated narrowing at the distal part of internal carotid artery on both sides, left more than right with prominence of lenticulostriate and leptomeningeal vessels. MRI also revealed funnel-shaped excavation of the posterior globe on the left side consistent with MGDA. Ascertaining the accurate diagnosis of MGDA guides appropriate ophthalmic management and should also prompt a search for associated intracranial abnormalities. Although the diagnosis of MGDA is typically made clinically, imaging may feed supplementary value in establishing the diagnosis and reveal the extent and character of associated ocular abnormalities, and cross-sectional imaging permits for evaluation of the globe in the setting of associated opacities of the refractive media, including persistent hyperplastic primary vitreous, which may alleviate the capacity to make this diagnosis on the basis of the funduscopy examination alone.

Key words: Magnetic resonance angiography; magnetic resonance imaging; morning glory disc anomaly; Moyamoya disease

Introduction

Morning glory disc anomaly (MGDA) is a congenital optic nerve anomaly characterized by a funnel-shaped excavation of the posterior globe that incorporates the optic disc.^[1]

In 1970, Kindler described an unusual congenital disc anomaly as “morning glory syndrome” because of its resemblance to the morning glory flower [Figure 1A].^[2]

An enlarged, funnel-shaped excavation that incorporates the optic disc characterizes it. The disc itself is enlarged, and orange or pink in color within a surrounding area of peripapillary chorioretinal pigmentary changes.^[3]

Magnetic resonance imaging (MRI) demonstrated three findings in most patients: (1) funnel-shaped morphologic pattern of the posterior optic disc with elevation of the adjacent retinal surface; (2) abnormal tissue associated

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with the distal intraorbital segment of the ipsilateral optic nerve, with effacement of the regional subarachnoid spaces; and (3) discontinuity of the uveoscleral coat.^[4]

Most cases of MGDA are isolated and not associated with systemic anomalies. Systemic anomalies include midline craniofacial defects, hypertelorism, agenesis of the corpus callosum, cleft lip and palate, basal encephalocele, congenital forebrain abnormalities, and renal anomalies.^[5]

The association of MGDA and Moyamoya vessels with midline cranial defects and central nervous system anomalies is atypical in some cases. Magnetic resonance angiography (MRA) is a noninvasive screening technique and may be used in conjunction with MRI in patients with MGDA to identify the vascular anomalies of the carotid system.^[6] Neurosurgical revascularization techniques may be considered in severe cases with strokes and hemorrhages.

Case Report

A 4-year-old male child presented with reduced visual acuity on left eye with poor fixation. His visual acuity was 20/20 in the right eye and 20/800 in the left eye with LEA symbols, which also demonstrated 6-diopter esotropia. Examination of fundus of the left optic nerve was slightly enlarged, with multiple anomalous vessels radiating circumferentially. A tuft of white material obscuring the central cup and a wide area of peripapillary pigment abnormality, with zones of hyperpigmentation in clumps, and hypopigmentation was noted [Figure 1B]. Fundoscopy examination showed enlarged left optic nerve with multiple anomalous vessels radiating circumferentially. The right optic nerve was normal. The child was diagnosed with MGDA and was subjected to other systemic examination, which was normal. MRI and MRA of brain were performed to exclude transsphenoidal encephalocele and other intracranial abnormalities.

MRA demonstrated narrowing at the distal part of internal carotid artery on both sides, left more than right with prominence of lenticulostriate and leptomeningeal vessels [Figure 2A and B]. 3D time-of-flight image shows narrowing of supraclinoidal part of the left internal cerebral

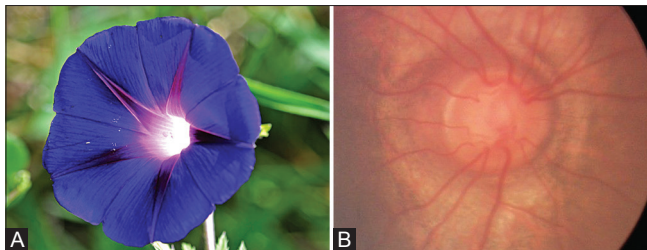


Figure 1 (A and B): (A) Morning glory flower. (B) Fundoscopic examination shows enlarged left optic nerve with multiple anomalous vessels radiating circumferentially

artery (red arrow). 3D time-of-flight image shows subtle narrowing of supraclinoidal part of the right internal cerebral artery (red arrow). The left A1 segment of anterior cerebral artery is atretic [Figure 3]. 3D time-of-flight image shows atretic A1 segment of the left anterior cerebral artery (red arrow). Prominence of bilateral middle cerebral artery is noted, left more than right [Figure 4]. 3D time-of-flight image shows prominent left middle cerebral artery (red arrow). No evidence of definite collateral vessels. No evidence of parenchymal infarct or ischemia.

MRI also revealed a funnel-shaped excavation of the posterior globe on the left side [Figure 5]. Axial constructive interference in steady state (CISS) sequence shows funnel shaped excavation of the posterior globe on the left side. No other midline defects were noted. Pituitary gland was normal.

The child was diagnosed as left MGDA with bilateral Moyamoya vessels (Suzuki type I classification). Child was started on antithrombotic and was asked for periodic check-up after 6 months.

Discussion

The association of MGDA with intracranial vascular anomalies is probably under-recognized, since carotid angiography is rarely performed in these individuals.^[6] From recent past, due to wide availability of MRI, there are case reports in the literature showing the association of MGDA with intracranial vascular anomalies.^[7-11]

The funduscopy appearance of the MGDA includes three primary features: an enlarged, funnel-shaped excavation in the optic disc; an annulus of chorioretinal pigmentary changes that surrounds the optic disc excavation; and a central glial tuft overlying the optic disc.^[12]

MGDA has been reported to be associated with other ocular anomalies and numerous intracranial abnormalities

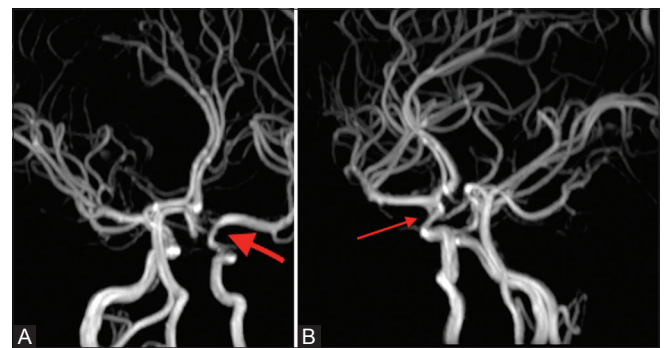


Figure 2 (A and B): (A) 3D time of flight image shows narrowing of supraclinoidal part of the left internal cerebral artery {red arrow}. (B) 3D time of flight image shows subtle narrowing of supraclinoidal part of the right internal cerebral artery {red arrow}

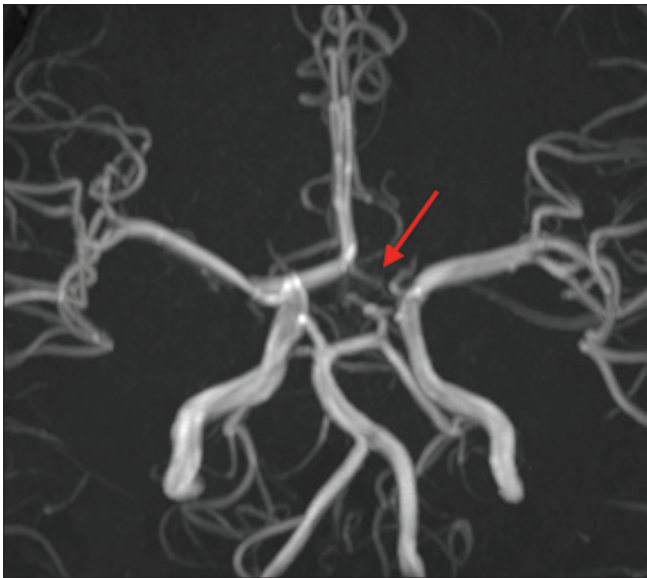


Figure 3: 3D time of flight image shows atretic A1 segment of the left anterior cerebral artery {red arrow}



Figure 4: 3D time of flight image shows prominent left middle cerebral artery {red arrow}



Figure 5: Axial CISS sequence shows funnel shaped excavation of the posterior globe on the left side

including midline craniofacial and skull base defects, vascular abnormalities, and cerebral malformations.^[1,12]

Establishing the correct diagnosis of MGDA guides appropriate ophthalmic management and should also prompt a search for associated intracranial abnormalities.^[3]

Table 1: Suzuki and Takaku in 1969 first described Moyamoya and its staging into six stages.^[18]

Stage I	“Narrowing of the carotid fork” Narrowed ICA bifurcation
Stage II	“Initiation of the Moyamoya” Dilated ACA, MCA, and narrowed ICA bifurcation with Moyamoya change
Stage III	“Intensification of the Moyamoya” Further increase in Moyamoya change of the ICA bifurcation and narrowed ACA and MCA
Stage IV	“Minimization of the Moyamoya” Moyamoya change reducing with occlusive changes in ICA and tenuous ACA and MCA
Stage V	“Reduction of the Moyamoya” Further decrease in Moyamoya change with occlusion of ICA, ACA, and MCA
Stage VI	“Disappearance of the Moyamoya” ICA essentially disappeared with supply of brain from ECA

ICA: Internal carotid artery, ACA: Anterior cerebral artery, MCA: Middle cerebral artery

Even though the diagnosis of MGDA is typically made clinically, imaging helps the clinician in several ways. First, when the clinical picture is puzzling, identification of specific features on imaging may help to ascertain the diagnosis and reveal the extent and character of associated ocular abnormalities like confusion often arises in distinguishing MGDA from optic nerve coloboma clinically. This distinction is important because of the implications for genetics and family counseling; MGDA is almost universally a sporadic condition whereas optic nerve coloboma is commonly familial and may occur in association with multisystem congenital malformation syndromes.^[13,14] Second, cross-sectional imaging permits for evaluation of the globe in the setting of associated opacities of the refractive media, including persistent hyperplastic primary vitreous, which may alleviate the capacity to make this diagnosis on the basis of the funduscopy examination alone.^[14] Finally, these patients may undergo imaging for unrelated clinical issues, and identification of the MGDA at imaging may be the only indicator to search for known associated intracranial abnormalities.

Moyamoya disease was first described in 1957 as a “bilateral hypoplasia of internal of internal carotid arteries.”^[15] The name of the disease comes from Japanese and means “puff of smoke.”

Changes appearing in the course of the disease include mainly the terminal parts of internal carotid arteries and/or proximal parts of middle and anterior cerebral arteries.^[16] In the affected cerebral vessels, pathological examinations do not show atherosclerotic or inflammatory lesions and the cause of stenosis is the overgrowth of the smooth muscle layer, with thrombotic changes. The disease also leads to the development of the collateral vasculature that produces a typical angiographic image, called “clouds of smoke” or “puff of cigarette smoke.”

Fluid-attenuated inversion recovery (FLAIR) sequence is very useful, as it helps to diagnose the “ivy sign,” i.e., an increase in signal intensity along the fissures and gyri of cerebral hemispheres, resulting most probably from the reduction of the cortical flow.

Angiographic criteria of the diagnosis of Moyamoya disease were established in 1998.^[17] They include stenosis or occlusion of the distal parts of intracranial internal carotid arteries and proximal parts of anterior and middle arteries, as well as the presence of collateral vasculature in the regions of the brain base, without causal disease. In case of bilateral changes, the diagnosis is considered as sure. Unilateral changes are qualified as probable. Table 1 shows Suzuki and Takaku classification of Moyamoya into six stages.^[18]

Our patient verifies the association of Moyamoya vessels with the MGDA and supports the argument that an intracranial vascular dysgenesis may underlie at least some cases of the MGDA.

MRA is a noninvasive screening technique that is widely available and should be used in conjunction with routine MRI to classify and further describe the prevalence of intracranial vascular anomalies of the carotid system in patients with anomalous optic discs of the morning glory variety.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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