

Access this article online

Quick Response Code:



Website:

<http://www.braincirculation.org>

DOI:

10.4103/bc.bc_10_20

Internal carotid artery origin of the anterior cerebral artery: A rare anatomic intracranial arterial variation in a child with morning glory disc anomaly and moyamoya vascular pattern; case report and review of literature

Aikaterini Solomou, Kanellos C. Spiliopoulos, Georgios Vasilagkos, Athanasios Vagionis, Petros Zampakis

Abstract:

Morning glory disc anomaly (MGDA) characterizes a congenital dysgenetic disorder of the optic disc, coexisting with arterial intracranial abnormalities, including Moyamoya vascular disease, a significantly rare disease in the European populations. We report a 2.5-year-old female child from Greece previously diagnosed with MGDA, who presented with right-hand paresis, accompanied by focal epileptic spasms, followed by an episode of brief absence seizure, as well as some arm clonic spasms. Magnetic resonance angiography scan revealed the presence of an anomalous origin of the anterior cerebral artery (ACA) from the internal carotid artery (ICA) along with vascular abnormalities, compatible with Moyamoya pattern. To the very best of our knowledge, this is the first reported case of anomalous origin of ACA from the supraclinoid ICA accompanied by severe occlusive intracranial disease (moyamoya-like pattern) in a patient with known MGDA, highlighting the embryonic character of the vascular manifestations in MGDA. It also verifies the association of Moyamoya pattern with MGDA, thus linking vascular dysgenesis as a possible cause of MGDA.

Keywords:

Cerebral arterial diseases, intracranial arterial diseases, morning glory disc anomaly, moyamoya disease, moyamoya vascular pattern

Introduction

Morning glory disc anomaly (MGDA) characterizes a congenital dysgenetic disorder of the optic disc comprising the enlargement and funnel-shaped excavation of the peripapillary posterior fundus. This disorder coexists with arterial intracranial

abnormalities, including the Moyamoya pattern of occlusive disease.^[1]

Moyamoya disease (MMD) is a stenocclusive disorder of the cerebrovascular system with chronic progression, involving the intracranial terminal portion of the internal carotid artery (ICA) and their proximal major branches, coupled

This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

For reprints contact: WKHLRPMedknow_reprints@wolterskluwer.com

How to cite this article: Solomou A, Spiliopoulos KC, Vasilagkos G, Vagionis A, Zampakis P. Internal carotid artery origin of the anterior cerebral artery: A rare anatomic intracranial arterial variation in a child with morning glory disc anomaly and moyamoya vascular pattern; case report and review of literature. *Brain Circ* 2020;6:133-8.

MRI Unit, Department of Radiology, University Hospital of Patras, Patras, Greece

Address for correspondence:

Prof. Aikaterini Solomou, MRI Unit, Department of Radiology, University Hospital of Patras, 26504, Rion- Patras, Greece. E-mail: solomou@med.upatras.gr

Submission: 17-03-2020

Revised: 15-05-2020

Accepted: 03-06-2020

Published: 26-06-2020

with the formation of an abnormal collateral vascular network.^[2] The pathogenic relevance between MMD and MGDA is a rare condition, while the incidence of MMD itself is significantly low in the European populations.^[3]

In this report, we describe the first case of the coexistence of a rare intracranial anatomic variation, in combination with a Moyamoya-like vascular pattern, in a 2.5-year-old female child with MGDA.

Case Report

A 2.5-year-old female child was admitted to our institution with right-hand paresis, accompanied by a 1-h history of focal epileptic spasms, followed by an episode of brief absence seizure (Petit Mal Seizure), as well as some arm clonic spasms.

Medical history also included prematurity (37 weeks), congenital aortic stenosis (treated with balloon valvuloplasty), congenital hypothyroidism, as well as MGDA of the left optic disc with a visual acuity of 4/10.

On presentation, clinical examination revealed right upper limb motor weakness and flaccidity. Her general clinical condition, as well as her higher mental function, was normal. The rest of the physical examination, including meningeal signs, was also normal.

Given the clinical presentation and medical history, magnetic resonance imaging (MRI) and magnetic resonance angiography (MRA) scan was performed to exclude any vascular event. The scan was performed in a 1.5 Tesla Unit (Siemens Avanto 1.5 T), under sedation using Chloral Hydrate.

Time-of-flight (TOF) MRA revealed several vascular abnormalities in intracranial circulation. More specifically, there was a severe narrowing of the left ICA, with a tortuous appearance depicted at the level of the origin of the ipsilateral ophthalmic artery [Figure 1].

The right ICA had a mild knot-like appearance and an occlusion before the carotid tip. There was also significant stenosis of the right M1 segment of the left posterior cerebral artery as well as minimal stenosis of the distal part of the left anterior cerebral artery (ACA) and the right proximal ACA [Figure 2].

Moreover, the presence of an anomalous origin of ACA from the ICA (a rare intracranial arterial finding) was also depicted. Lateral projection of the intracranial arteries better showed a vessel originating from the supraclinoid segment of the left ICA (before the origin of the ipsilateral ophthalmic artery), which runs cranially in a ventral course, anterior to the optic chiasm and medial



Figure 1: Time of flight magnetic resonance angiography shows segmental occlusion of the M1 segment of the right middle cerebral artery, severe stenosis of the terminal left internal carotid artery and the middle cerebral artery, as well as occlusion of the terminal right internal carotid artery, before the carotid tip middle cerebral artery

to the optic nerves and then characteristically turns dorsally to join the area of the anterior communicating artery [Figure 3].

Brain MRI also revealed collateral arterial network bilaterally, more visible in the left basal ganglia. Fluid-attenuated inversion recovery sequence showed linear hyperintensities along the meninges of the cortex at the left occipital lobe, ivy-like without enhancement of the paramagnetic agent on postcontrast MRIs images due to reduction of the cortical flow [Figure 4].

In addition, at the left parietal lobe, a 5-mm lesion was present without restriction on the diffusion-weighted images and high apparent diffusion coefficient, compatible with a chronic ischemic insult.

All these imaging findings could be compatible with a moyamoya pattern of vascular distribution, in combination with a rare anatomic variation of the anterior circulation.

The patient was transferred to a specialized center for further evaluation.

Discussion

To the very best of our knowledge, this is the first reported case, of anomalous origin of ACA from the supraclinoid ICA, in combination with the severe occlusive intracranial disease (moyamoya-like pattern), in a patient with known MGDA.

The term MGDA is used to characterize a rare sporadic congenital malformation of the optic nerve.^[4] The basic clinical features include a characteristic conical



Figure 2: Axial image (time of flight magnetic resonance angiography) depicts severe narrowing of the left internal carotid artery, at the level of the origin of ipsilateral ophthalmic artery



Figure 3: Lateral projection of the intracranial arteries (time-of-flight magnetic resonance angiography) shows the presence of anomalous origin of anterior cerebral artery from the internal carotid artery. The vessel runs cranially in a ventral course and then characteristically turns dorsally to join the anterior communicating artery area

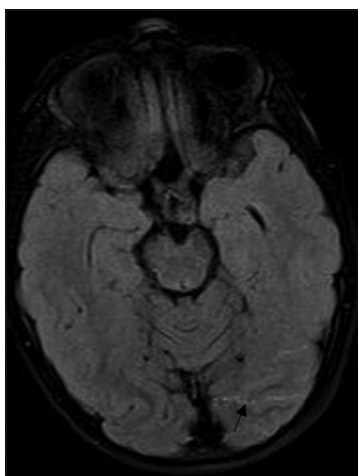


Figure 4: Axial fluid-attenuated inversion recovery image at the level of optic chiasm shows high signal within the subarachnoid space of the left occipital lobe

excavation of the optic disc, a glial tuft on the center of the optic cup, and peripapillary pigmentation. The retinal blood vessels follow a radial pattern without branching. Regarding genetics for MGDA, it has been postulated that PAX6 mutations have been associated with MGDA,^[5] but a consistent and solid genetic insult has not been recognized. Some studies have shown sporadic cases with PAX6 mutations, while others failed to demonstrate such relation.^[6] Furthermore, it has been suggested that PAX2 mutations have been associated with MGDA as well.^[7]

Common symptoms include strabismus (>90%) and reduced visual acuity (70%). Furthermore, optic vascular abnormalities, arteriovenous, and arteriole-arteriole anastomoses have been reported in the literature.^[8] The association of MGDA with ocular and neurovascular abnormalities is not uncommon. Retinal detachments constitute the most frequent ocular abnormality. Neurovascular abnormalities include midline facial malformations, basal encephalocele, agenesis of the corpus callosum, endocrine abnormalities, and MMD.^[1]

Over the past 25 years, several case reports and case series have reported the co-existence of MMD and MGDA.^[9-15] The aforementioned association has initially described by Hanson *et al.*^[16] back in 1985, while many additional cases have been reported since. Up to date, in the largest series of patients, Lenhart *et al.*^[17] reported that 45% of MGDA patients had moyamoya or other cerebrovascular abnormalities.

Therefore, further investigation of MGDA patients for intracranial vascular anomalies should be considered.^[18] Several studies have shown that MRA is a valuable tool for the diagnosis of MMD. TOF, CE-MRA in 1.5T-7T MRI have been used.^[19] The higher the magnetic field, the better results can be achieved^[20] while sensitivity rates vary from 72% to 92%; however, findings are highly specific.^[21]

This diagnostic workup was applied in our case as well. According to the 2012 guidelines for the diagnosis of MMD,^[22] all the MRA and MRI diagnostic criteria were met, concerning particularly those guidelines for diagnostic assessment in children. Hence, severe occlusion in the terminal portion of ICA bilaterally, minimal stenosis of ACA bilaterally, stenosis of the M1 segment of the right MCA, as well as the findings of the collateral vascular network bilaterally, substantiate the diagnosis of definitive MMD in our case. In fact and because of the association of our Moyamoya pattern with underlying disease (MGDA), the term Quasi MMD could be used, while it would be even more preferable to refer as Moyamoya vasculopathy or phenomenon.

It has been postulated that the syndrome could be the result of a retinal vascular dysgenesis.

The association of this syndrome with other vascular intracranial anomalies, such as MMD, verifies the presence of a possible triggering factor during embryogenesis that causes all the vascular spectrum of the disease. One possible explanation would be that the presence of embryonic vascular dysgenesis could result (due to the cerebrovascular phylogenetic plasticity) in altered blood flow dynamics during embryogenesis, which may account for intracranial coexistent anomalies.^[23]

In the same setting, other intracranial arterial distributions may occur. The presence of intracavernous intrasellar ACA is an anatomical variation of this kind. This rare variant has been previously described as a truncal variation of ACA. In this rare configuration, the vessel travels medial to ICA siphon in the pituitary fossa, emerges between the optic nerves and then it inclines posteriorly, to join the ACAs. This variety is most likely to represent a more caudal loop of the primitive dorsal ophthalmic artery and has been reported in cases of MMD.^[24] Other reported associated vascular anomalies include agenesis of the ICA, anomalous origin of the ipsilateral ophthalmic artery from the external circulation, fused pericallosal arteries, MMD and symptoms related to compression of the optic apparatus.^[23] In our case, the vessel follows the same course [Figure 3].

It is well known that in cases of severe stenosis/occlusion of the proximal carotid, there could be several arterial anatomic variations/anastomoses in the configuration of intracranial arteries, acting as a natural bypass.^[25]

MMD is a chronic, progressive type of occlusive cerebrovascular disorder. The hallmark of the disease is the progressive stenosis at the intracranial apices of the ICAs and their proximal branches, the ACAs and MCAs, as well as the formation of a compensatory collateral vascular network, characterized as moyamoya vessels. Concerning the pathophysiology, the absence of arteriosclerosis and inflammation is characteristic. The moyamoya collateral vasculature is a consequence of the reduced blood flow, and these smoke-like tortuous vascular pathways are observed near the distal portion of the carotid, in leptomeningeal vessels, on the cortical surface and the base of the skull.^[2] Undoubtedly, there is a wide variety of clinical and angiographic spectrum in MMD; thus, it has been hypothesized that a combination of genetic background, environmental as well as age-related factors could lead to different clinical and/or angiographic expression of this entity.^[26] The most consistent genetic factor of MMD in the East Asian population is polymorphism in RNF213 gene.^[27] Polymorphisms in platelet-derived growth factor

receptor and TGF β 1 genes were the first genetic places to be associated with MMD in Central Europeans by Roder *et al.* in 2010.^[28]

The two major categories of presenting symptoms are ischemia and hemorrhage. The clinical manifestations include ischemic and hemorrhagic strokes, seizures, headaches, and cognitive/or psychiatric impairment. Hyperventilation is a typical inducer of the ischemic symptoms in pediatric patients, who may also present with atypical manifestations, including syncope and visual disorders.^[2,29] Other clinical manifestations of MDD include motor weakness and hemiparesis, which can be accompanied by choreiform movements.^[30,31]

This combination of symptoms, as well as in our case, verifies the assumption that MMD should be considered in pediatric patients presenting with movement disorders or neurological deficit, as this could be the initial presentation.

In patients with moyamoya vasculopathy, the brain ischemia is crucial for the development of the characteristic clinical features, such as ischemic stroke, transient ischemic attack, and epilepsy. The ischemic phenotype of MMD pertains commonly to pediatric patients, while the rate of hemorrhage is higher in adults.^[2] A study showed that the ischemic pattern in children is more often gyral or border zone in MRI.^[32] In our case, it is unclear whether the monoparesis, involving the right hand, is due to an unidentified ischemic stroke or whether it could be associated with postictal-Todd's paresis, which refers to a focal weakness, affecting particularly one side or one limb of the body, and occurring after seizures. These seizures are usually focal motor.^[33]

The "epileptic type" is one of the three well-defined types of MMD with a presentation percentage of 7.6%, notwithstanding symptoms, lack in a defined cause apart from a relation with cortical ischemia. Children aged 10 years or younger are more frequently categorized in the epileptic type than the other age groups.^[34] A recent study, concerning the onset age of 3 years or younger and the early seizures, demonstrated that these background traits of MMD patients are associated primarily with epilepsy.^[35]

Given the fact that MMD patients carry a high risk of ischemic insults, early diagnosis, as well as comprehensive follow-up in diagnosed cases, is of paramount importance. Imaging techniques should focus on detailed angiographic and brain parenchyma status, while more advanced functional imaging techniques could be used to reveal any perfusion changes. A variety of such advanced imaging techniques is being using up to date. Diffusion and perfusion magnetic resonance,

single photon emission computed tomography, and/or advanced MRA techniques can reliably demonstrate vessels occlusion and/or parenchymal hypoperfusion or infarcts, both on symptomatic and asymptomatic patients.^[36-38] Moreover, these modern techniques can point out reperfusion candidates or evaluate surgical results.

Since MGDA can coexist with Moyamoya vasculopathy, we suggest that all such cases should undergo MRI-MRA to depict any vascular or parenchymal abnormalities that imply intracranial vascular manifestation of the disease. 1.5 or even better 3T MRA can be used in the initial workup,^[20] while digital subtraction angiography and/or perfusion techniques should be reserved in dubious MRA cases (especially when the patient is symptomatic). Follow-up MRI/MRA should be performed in cases of clinical deterioration or alteration of clinical status, while the angiographic progression of the disease could be monitored by MRAs at regular intervals.

Conclusion

Our case highlights the embryonic character of vascular manifestations of MGDA, as well as the importance of MRI/MRA for their diagnosis.

The coexistence of intracranial Moyamoya pattern with a rare intracranial arterial distribution (acting as a natural bypass), as these reliably depicted in MRI/MRA, could indicate that embryonic vascular dysgenesis may play a key role for the vascular manifestation of MGDA.

Consent for publication

Written informed consent was obtained from the patient's legal guardian (s) for publication of this case report and any accompanying images.

Declaration of patient consent

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

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Lee BJ, Traboulsi EI. Update on the morning glory disc anomaly. *Ophthalmic Genet* 2008;29:47-52.

2. Scott RM, Smith ER. Moyamoya disease and moyamoya syndrome. *N Engl J Med* 2009;360:1226-37.
3. Hever P, Alamri A, Tolia C. Moyamoya angiopathy – Is there a Western phenotype? *Br J Neurosurg* 2015;29:765-71.
4. Kindler P. Morning glory syndrome: Unusual congenital optic disk anomaly. *Am J Ophthalmol* 1970;69:376-84.
5. Azuma N, Yamaguchi Y, Handa H, Tadokoro K, Asaka A, Kawase E, *et al.* Mutations of the PAX6 gene detected in patients with a variety of optic-nerve malformations. *Am J Hum Genet* 2003;72:1565-70.
6. Guerra-Junior G, Spinola-Castro AM, Siviero-Miachon AA, Nogueira RG, Lemos-Marini SH, D'Souza-Li LF, *et al.* Absence of mutations in Pa×6 gene in three cases of morning glory syndrome associated with isolated growth hormone deficiency. *Arq Bras Endocrinol Metabol* 2008;52:1221-7.
7. Markitantova Y, Simirskii V. Inherited eye diseases with retinal manifestations through the eyes of homeobox genes. *Int J Mol Sci* 2020;21:1602. <https://doi.org/10.3390/ijms21051602>.
8. Brodsky MC, Wilson RS. Retinal arteriovenous communications in the morning glory disc anomaly. *Arch Ophthalmol* 1995;113:410-1.
9. Ponnatapura J. Morning glory syndrome with Moyamoya disease: A rare association with role of imaging. *Indian J Radiol Imaging* 2018;28:165-8.
10. Bakri SJ, Siker D, Masaryk T, Luciano MG, Traboulsi EI. Ocular malformations, moyamoya disease, and midline cranial defects: A distinct syndrome. *Am J Ophthalmol* 1999;127:356-7.
11. Quah BL, Hamilton J, Blaser S, Heon E, Tehrani NN. Morning glory disc anomaly, midline cranial defects and abnormal carotid circulation: An association worth looking for. *Pediatr Radiol* 2005;35:525-8.
12. Fernández-Fernández S, Vázquez-López M, Carrasco-Marina LL, Vela-Valdecabres C, Cortés-Valdés E, Arregui-Sierra A. An association between moyamoya disease and morning glory anomaly. *Rev Neurol* 2003;37:541-4.
13. Krishnan C, Roy A, Traboulsi E. Morning glory disk anomaly, choroidal coloboma, and congenital constrictive malformations of the internal carotid arteries (moyamoya disease). *Ophthalmic Genet* 2000;21:21-4.
14. Sathyan S, Chackochan M. Morning glory disc anomaly and facial hemangiomas in a girl with moyamoya syndrome. *Indian J Ophthalmol* 2018;66:1644-6.
15. Massaro M, Thorarensen O, Liu GT, Maguire AM, Zimmerman RA, Brodsky MC. Morning glory disc anomaly and moyamoya vessels. *Arch Ophthalmol* 1998;116:253-4.
16. Hanson MR, Price RL, Rothner AD, Tomsak RL. Developmental anomalies of the optic disc and carotid circulation. A new association. *J Clin Neuroophthalmol* 1985;5:3-8.
17. Lenhart PD, Lambert SR, Newman NJ, Biousse V, Atkinson DS Jr., Traboulsi EI, *et al.* Intracranial vascular anomalies in patients with morning glory disk anomaly. *Am J Ophthalmol* 2006;142:644-50.
18. Auber AE, O'Hara M. Morning Glory syndrome. *MR imaging. Clin Imaging* 1999;23:152-8.
19. Oh BH, Moon HC, Baek HM, Lee YJ, Kim SW, Jeon YJ, *et al.* Comparison of 7T and 3T MRI in patients with moyamoya disease. *Magn Reson Imaging* 2017;37:134-8.
20. Jin Q, Noguchi T, Irie H, Kawashima M, Nishihara M, Takase Y, Gong H, *et al.* Assessment of Moyamoya disease with 3.0-T magnetic resonance angiography and magnetic resonance imaging versus conventional angiography. *Neurol Med Chir (Tokyo)* 2011;51:195-200.
21. Yamada I, Suzuki S, Matsushima Y. Moyamoya disease: Comparison of assessment with MR angiography and MR imaging versus conventional angiography. *Radiology* 1995;196:211-8.
22. Guidelines for diagnosis and treatment of moyamoya disease (spontaneous occlusion of the circle of Willis). *Neurol Med Chir (Tokyo)* 2012;52:245-66.
23. Burbank NS, Morris PP. Unique anomalous origin of the left

- anterior cerebral artery. *AJNR Am J Neuroradiol* 2005;26:2533-5.
24. Lasjaunias P, Berenstein A, terBrugge K. *Surgical Neuroangiography: Clinical Vascular Anatomy and Variations*. 2nd ed., Vol. 1. Berlin: Springer-Verlag; 2001.
 25. Zampakis P, Panagiotopoulos V, Petsas T, Kalogeropoulou C. Common and uncommon intracranial arterial anatomic variations in multi-detector computed tomography angiography (MDCTA). What radiologists should be aware of. *Insights Imaging* 2015;6:33-42.
 26. Bang OY, Chung JW, Kim DH, Won HH, Yeon JY, Ki CS, *et al.* Moyamoya disease and spectrums of RNF213 vasculopathy. *Transl Stroke Res* 2019. <https://doi.org/10.1007/s12975-019-00743-6>.
 27. Kamada F, Aoki Y, Narisawa A, Abe Y, Komatsuzaki S, Kikuchi A, *et al.* A genome-wide association study identifies RNF213 as the first Moyamoya disease gene. *J Hum Genet* 2011;56:34-40.
 28. Roder C, Peters V, Kasuya H, Nishizawa T, Takehara Y, Berg D, *et al.* Polymorphisms in TGFB1 and PDGFRB are associated with Moyamoya disease in European patients. *Acta Neurochir (Wien)* 2010;152:2153-60.
 29. Kuroda S, Houkin K. Moyamoya disease: Current concepts and future perspectives. *Lancet Neurol* 2008;7:1056-66.
 30. Maheshwari S, Anthony A, Kushwaha S, Singh S, Desai R, Madan D. Moyamoya disease presenting as alternating hemiparesis with relapsing remitting hemichorea: An unusual manifestation. *J Pediatr Neurosci* 2018;13:514-6.
 31. Kraemer M, Trakolis L, Platzen J, Schwitalla JC, Bersano A, Albrecht P, *et al.* Movement symptoms in European Moyamoya angiopathy – First systematic questionnaire study. *Clin Neurol Neurosurg* 2017;152:52-6.
 32. Cho HJ, Jung YH, Kim YD, Nam HS, Kim DS, Heo JH. The different infarct patterns between adulthood-onset and childhood-onset moyamoya disease. *J Neurol Neurosurg Psychiatry* 2011;82:38-40.
 33. Anaththanam S, Hassan A. Mimics and chameleons in stroke. *Clin Med (Lond)* 2017;17:156-60.
 34. Burke GM, Burke AM, Sherma AK, Hurley MC, Batjer HH, Bendok BR. Moyamoya disease: A summary. *Neurosurg Focus* 2009;26:E11.
 35. Mikami T, Ochi S, Houkin K, Akiyama Y, Wanibuchi M, Mikuni N. Predictive factors for epilepsy in moyamoya disease. *J Stroke Cerebrovasc Dis* 2015;24:17-23.
 36. Choudhri A, Zaza A, Auschwitz T, Klimo P, Mossa-Basha M. Noninvasive vascular imaging of moyamoya: Diagnosis, follow up, and surgical planning. *J Pediatr Neurosurg* 2014;3:13-20.
 37. Li J, Jin M, Sun X, Li J, Liu Y, Xi Y, *et al.* Imaging of moyamoya disease and Moyamoya syndrome: Current status. *J Comput Assist Tomogr* 2019;43:257-63.
 38. Gburek-Augustat J, Sorge I, Merckenschlager A. The “ivy-sign” in moyamoya disease-from MRI pattern to diagnosis. *Neuropediatrics* 2020. doi: 10.1055/s-0040-1708546.