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RESEARCH NOTE

Failure of Evolutionary Pruning: a Case Report on a Young Adult with Rare Twig-like Middle Cerebral Artery and Its Implications on Worse Outcomes

Benjamin O. Sosa III* and Rhoby U. Orata

Neurosurgery Service, Department of Surgery, Victoriano Luna Medical Center,
Armed Forces of the Philippines–Health Service Command,
National Capital Region 1100 the Philippines

Twig-like middle cerebral artery (MCA) is a rare intracranial vascular pathology that was hypothesized to be an arrest on the evolutionary fusion of the plexiform arterial network in the forebrain into a single MCA artery. This anomaly predisposes affected individuals to worse outcomes such as intracranial hemorrhages due to its association with aneurysms and primitive plexiform arterial systems that are prone to rupture. In this case report, a 30-year-old male had a loss of consciousness after serving as a ceremonial guard for four straight days. The patient did not have any neurologic localizing signs but presented with occasional generalized headaches relieved by pain medications. Cranial MRA showed suspicious stenosis of the left MCA. A cranial computed tomography angiogram showed complete stenosis of the M1 segment of the left MCA. Digital subtraction angiography was also done, which confirmed the complete occlusion of the said segment with collaterals from the A1 of the anterior cerebral artery supplying the distal portions of the left MCA. These findings, hence, suggested the presence of a twig-like MCA. In this article, the prevalence, pathogenesis, arrival to the diagnosis, and clinical relevance shall be discussed.

Keywords: digital subtraction angiography, hemorrhagic events, total occlusion, twig-like MCA, vascular anomaly

INTRODUCTION

Twig-like middle cerebral artery (MCA) is a rare vascular anomaly characterized by the complete occlusion of the M1 segment of the affected MCA with extensive lenticulostriate arteries surrounding the defect and a plexiform arterial network that serves as the proximal M1 (Viso *et al.* 2021) The distal branches of the affected MCA are intact due to the presence of collaterals from the anterior or posterior cerebral arteries supplying the ipsilateral distal cerebral cortex (Viso *et al.* 2021).

*Corresponding author: benjaminolanososaiii@gmail.com

Twig-like MCA is present in 0.11–1.17% of patients who had diagnostic angiography (Viso *et al.* 2021). One of the accepted explanations of this anomaly is due to an evolutionary arrest on the development of the fetal MCA preventing the regression of the plexiform network of blood vessels into one MCA leading to occlusion (Viso *et al.* 2021).

CLINICAL PRESENTATION

A healthy 30-year-old male was on his fourth day of duty as a ceremonial guard when he felt light-headedness with associated cold sweats. He then lost consciousness and fell backward, suffering no head trauma.

During admission, the patient had no neurologic localizing signs and would only complain of occasional left-sided, non-radiating, throbbing headaches, 4–5/10 in severity that spontaneously resolved. A cranial MRI with stroke protocol showed luminal narrowing of the M1 segment of the left MCA with no further visualization of its distal segments. The patient is a military personnel and a professional athlete playing *sepak takraw* and lawn tennis.

On neurophysical examination, the patient was oriented to three spheres with intact higher cortical functioning. All cranial nerves were intact with normoflexia. The patient had no cerebellar or meningeal signs. Cranial computed tomography angiography (CTA) was done as preliminary screening in conjunction with a digital subtraction angiography (DSA) to confirm the preliminary findings. The digital subtraction images of the patient were consistent with the aforementioned vascular anomaly, as shown in Figures 1 and 2. Further, Figure 3 shows the 3D reconstruction of the cranial CTA of the patient.

No neurosurgical intervention was done on the patient because no other concomitant vascular anomalies such as an aneurysm or arteriovenous malformation were seen on the four-vessel angiogram.

DISCUSSION

DSA is still considered the gold standard in visualizing vascular anatomy despite being an invasive procedure (Osborn and Digre 2023). The DSA results showed total occlusion of the proximal M1 segment of the left MCA with distal reconstitution at the mid-M1 segment from collateral branches arising from the A1 segment of the left anterior cerebral artery.

The complete stenosis of the M1 segment of the left MCA in this patient may either be acquired or congenital. Possible causes of acquired MCA stenosis are [1] atherosclerosis, [2] vascular dissection, or [3] vasculitis (Viso *et al.* 2021). These differentials would have presented with stroke-like symptoms but were not seen in the patient. Moyamoya disease was also ruled out because it affects the terminal portion of the intracranial internal

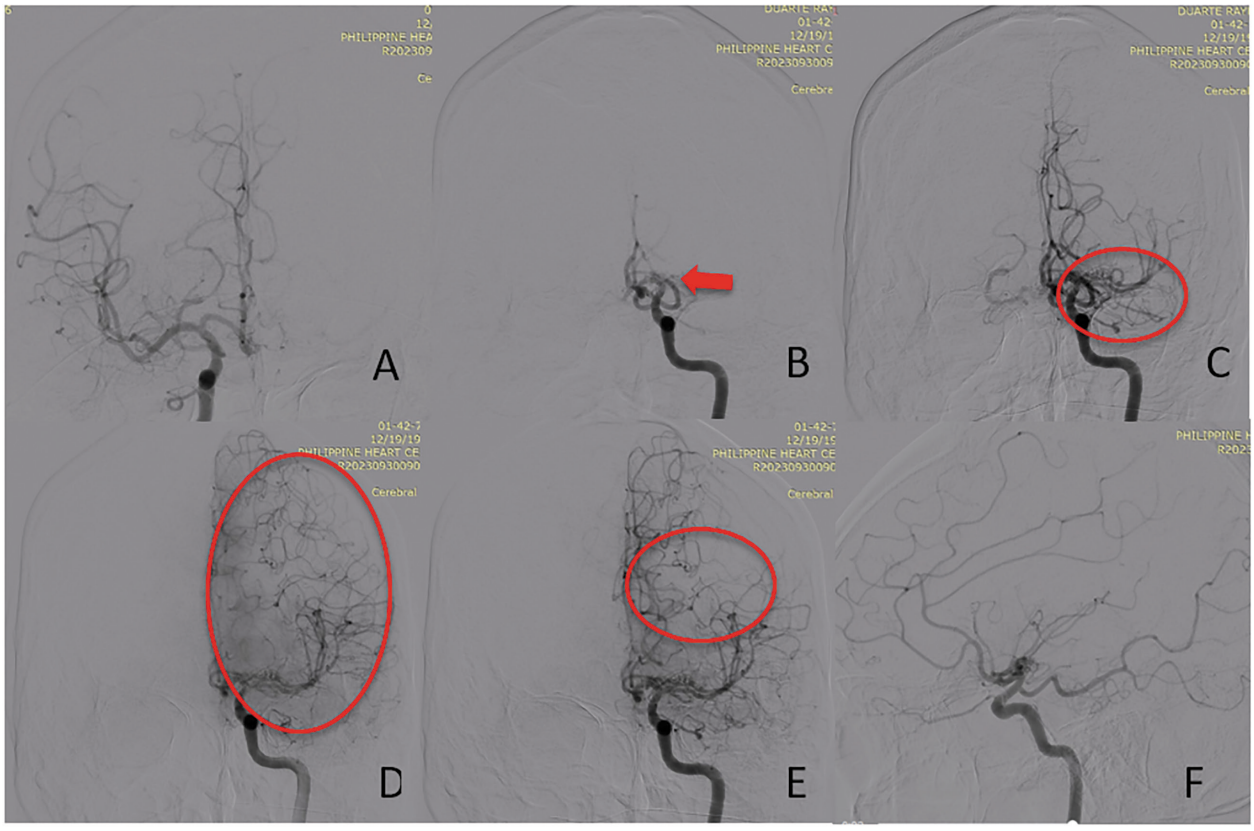

Figure 1. Common features of a twig-like MCA, as seen from the patient's DSA: [A] normal right MCA with no anomalies for point of comparison; [B] unilateral MCA occlusion; [C] lenticulostriate arteries surrounding the defect originating from the primitive plexiform arterial network that experienced evolutionary arrest; [D] normal cortical vessels beyond the occluded part of the MCA; [E] collaterals from A1 to supplying the distal left MCA; [F] sagittal view of the occluded M1 segment.

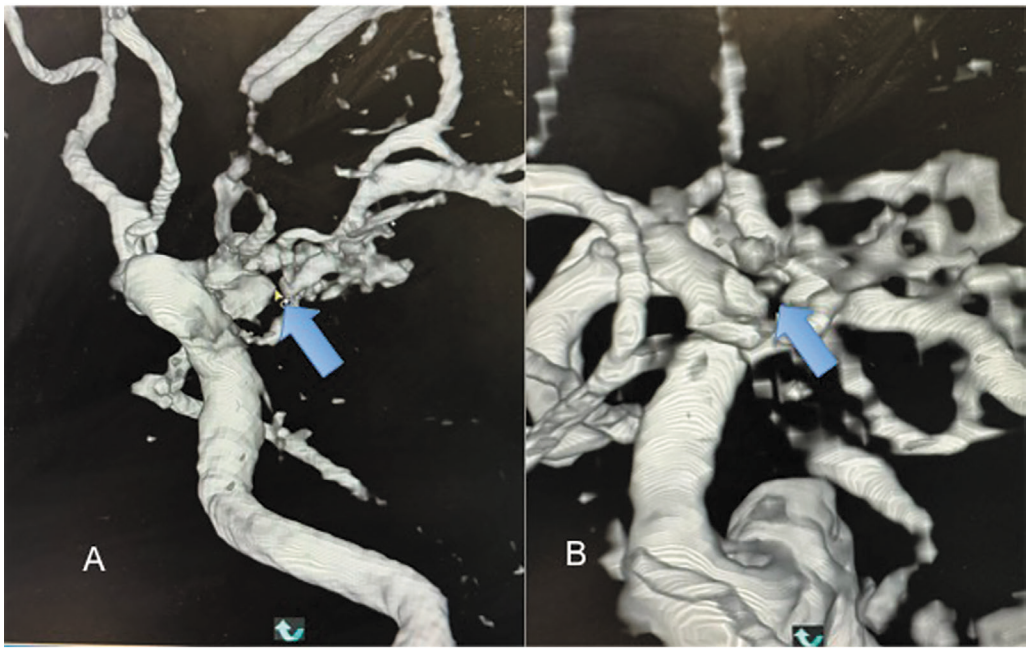

Figure 2. 3D reconstruction of the DSA done on the patient showing the occluded M1 segment. The blue arrow points to the occluded M1 segment of the left MCA.

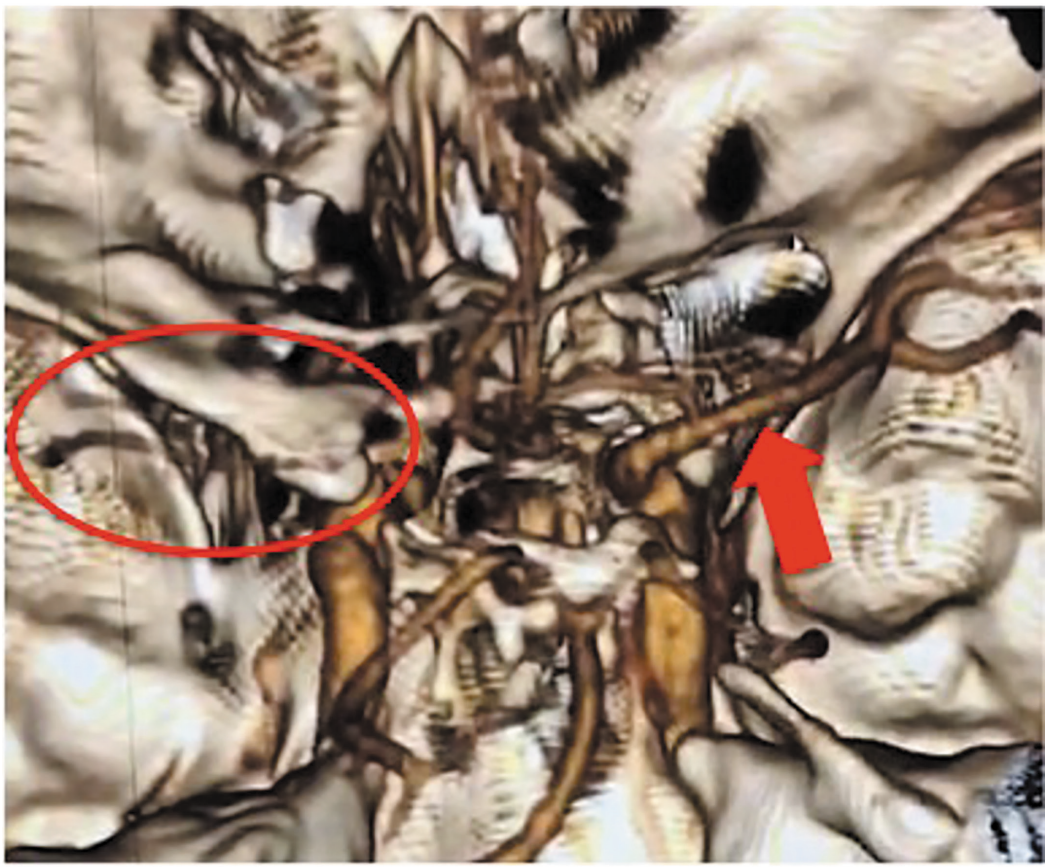

Figure 3. 3D reconstruction of the cranial CTA showing the occlusion at M1 due to the non-visualization of the left MCA. The red circle on the left shows the non-visualization of the left MCA because the contrast was not able to pass due to the M1 occlusion. In contrast, the red arrow on the right shows a clear visualization of the right MCA, which had no anomaly.

carotid artery and requires the presence of an abnormal vascular network in the vicinity of the stenosis bilaterally (Kuroda *et al*. 2022). Since the patient was asymptomatic with collaterals from A1, the patient's condition could be congenital.

Despite the benign course of the patient, twig-like MCA is usually associated with an increased risk for hemorrhagic events because the inherent morphologic anomalies in this vessel are linked to increased odds of aneurysm formation (Viso *et al* 2021; Gailloud *et al* 2002). The fetal arterial twigs are remnants of the primordial MCA, which have thinner lumens and a less developed muscularis layer, hence making it prone to adventitial tear and eventual rupture (Viso *et al.* 2021).

According to Klopfenstein *et al.* (2005), asymptomatic patients with MCA stenosis should not be considered for surgical intervention because the risks outweigh the benefits. Surgical treatment may cause iatrogenic vascular injury to the affected MCA that can cause stroke-like symptoms. For symptomatic patients, stenting and surgical bypass are the available options. Intracranial stenting is warranted if the stenosis of the proximal portion of the MCA is > 60% and with a history of transient ischemic attack (Kim *et al.* 2004).

CONCLUSION

In general, the prevalence of twig-like MCA in the population has not been fully elucidated because the majority of the reported cases are incidental findings or have concomitant intracranial hemorrhagic events. The association between hemorrhagic events and the possible presence of twig-like MCA is of clinical significance because of the possible association of this vascular anomaly to other vascular malformations such as aneurysms or persistent fetal MCA.

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