CASE

Severe left MCA syndrome with an occluded accessory MCA

Manya Khrlobyan, DO, Edgar Olivas, MD, Lei Feng, MD, Matthew Czar Taon, MD, and Navdeep Sangha, MD

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Correspondence
Dr. Khrlobyan
mmakopyan@gmail.com

Anatomical variants of the circle of Willis have been reported in up to 60% of cases, but variations involving the middle cerebral arteries are much less common.[1] Three reported middle cerebral artery (MCA) anomalies include the accessory MCA, duplicated MCA, and fenestration of MCA. The incidence of accessory MCA (AMCA) is reported to be 0.3%–4.0%. Based on the widely accepted classification by Teal et al. in 1973, the AMCA is a variant artery arising from the anterior cerebral artery (ACA) and runs in the Sylvian fissure along with the main MCA to supply part of the MCA territory; as such, an occlusion of an AMCA can lead to severe neurologic deficits.[2] There have been very few reports of AMCA occlusion in the setting of acute ischemic stroke,[3–7] leading to clinical uncertainty for those unfamiliar with such rare variations. Here, we report a 91-year-old woman presenting with left MCA syndrome with an occluded left AMCA and severe stenosis of the main MCA trunk.

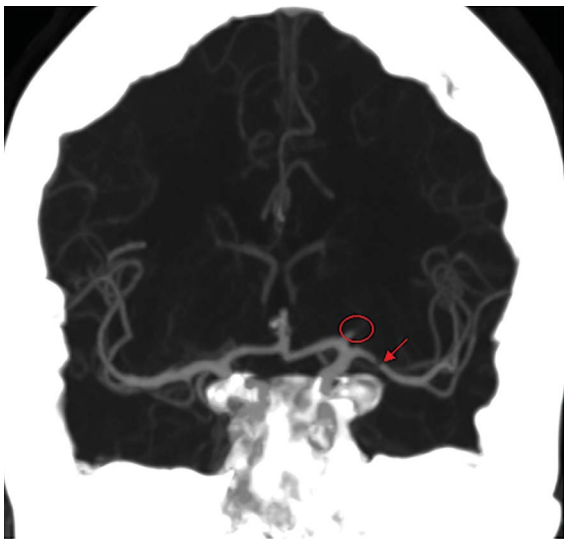
PRACTICAL IMPLICATIONS

In patients presenting with acute ischemic stroke, when the clinical presentation cannot be explained by findings seen on imaging, consider the presence of rare anatomical variations.

Case presentation

A 91-year-old right-handed woman with atrial fibrillation (not on anticoagulation), hypertension, and obesity presented to the emergency department 1 hour after acute onset of left MCA syndrome. Initial National Institutes of Health Stroke Scale (NIHSS) was 24, and her baseline modified Rankin Scale was 3. Initial CT of the head was unremarkable with an Alberta Stroke Program Early CT

Figure 1 Coronal CTA brain and neck



Left M1 focal high-grade stenosis (arrow) and proximal left A1 segment abnormality (circle).

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Department of Neurology (MK, EO, NS), Department of Neuro-Interventional Radiology (LF), and Department of Radiology (MCT), Kaiser Permanente, Los Angeles Medical Center, CA.

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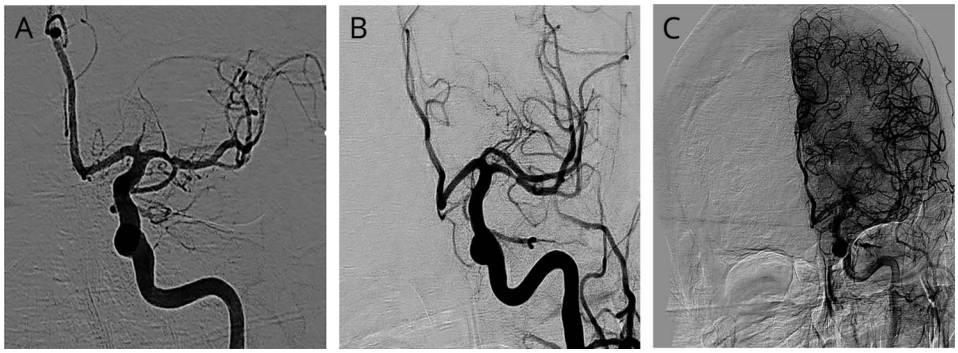
Figure 2 Angiograms



(A) Pretreatment cerebral angiogram. Left ICA cerebral angiogram demonstrating an occluded AMCA. (B) Post-treatment cerebral angiogram. Left ICA cerebral angiogram demonstrating a patent AMCA after thrombectomy. (C) Pretreatment capillary phase of cerebral angiogram. Left ICA capillary phase cerebral angiogram demonstrating decreased perfusion in the area of the occluded AMCA.

Score (ASPECTS) of 10. She received IV tissue plasminogen activator within 20 minutes of arrival to the emergency department with very minimal improvement of her symptoms. CT angiography (CTA) of the brain and neck demonstrated focal high-grade stenosis of the left M1 segment and a small bulge from the proximal left A1 segment indicating aneurysm or occluded AMCA (figure 1, A and B). An emergent cerebral angiogram was performed, confirming the occlusion of the M1 segment of left AMCA, which supplied the frontal lobe with good collaterals from the adjacent MCA and ACA branches (figure 2, A and B). The AMCA came off at a rather sharp angle from the proximal A1 segment along with the underlying stenosis in the mid-AMCA segment. Given the anatomy, small caliber of the AMCA, and the underlying stenosis, a decision was made to attempt aspiration thrombectomy rather than the commonly used stent retriever. The patient underwent aspiration thrombectomy of the left AMCA with thrombolysis in cerebral infarction 2b recanalization (figure 2C). Time from symptom onset to recanalization was 4 hours. At the time of discharge, NIHSS was a 2 and modified Rankin Scale was noted to be 3.

Discussion

The patient's clinical presentation was consistent with a left MCA syndrome, and the severity of her symptoms pointed toward a large vessel occlusion. On initial review of CTA, a left M1 segment stenosis with patent distal vessels was the most evident finding, but the patient's severe clinical symptoms were puzzling and would be best explained by the occlusion of M1 or dominant M2. Cerebral angiogram then confirmed an AMCA branching from the A1 segment. In addition, the AMCA was seen to have lenticulostriate arteries, whereas the main stenotic MCA trunk did not. Furthermore, the territory supplied by the main MCA trunk was dependent on the AMCA for collateral supply due to its severe proximal stenosis. These findings on cerebral angiogram explained why the patient presented with full left MCA syndrome.

Embryologically, there have been several proposed explanations for the origins of the AMCA. Uchiyama[2] considered that MCA forms as a fusion of lenticulostriates, and when the fusion is incomplete, AMCA, duplicated MCA, or double MCA trunk can develop. In a double MCA trunk, the larger trunk is defined as the main MCA trunk and the distal trunk, which is typically smaller, arises from the ACA and defined as the AMCA. In our case, it is difficult to define the main trunk according to Uchiyama's classification as both trunks were equal in size. This configuration could be congenital, as it incorporated all the frontal branches, or compensatory due to the underlying stenosis of the main MCA trunk.

There have been a number of reports involving aneurysms of the AMCA but only a few case reports describing the AMCA in the setting of acute stroke.[3–7] This case represents one of the first acute ischemic strokes secondary to an occluded AMCA undergoing successful thrombectomy and demonstrates that an occluded AMCA can lead to severe neurologic deficits and thrombectomy can yield a good clinical outcome.

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Appendix Authors

Name	Location	Role	Contribution
Manya Khrlobyan, DO, M.S	Kaiser Permanente, Los Angeles Medical Center	Corresponding Author	Major role in drafting and editing case report.
Edgar Olivas, MD	Kaiser Permanente, Los Angeles Medical Center	Co-author	Major role in drafting and editing the case report. Senior resident during patient presentation.

Appendix *(continued)*

Name	Location	Role	Contribution
Matthew C. Taon, MD	Kaiser Permanente, Los Angeles Medical Center	Co-author	Acquired, formatted, and organized images for the case report.
Lei Feng, MD	Kaiser Permanente, Los Angeles Medical Center	Co-author	Major role in editing the case report. Neurointerventional radiologist during patient presentation.
Navdeep Sangha, MD	Kaiser Permanente, Los Angeles Medical Center	Final author	Major role in drafting and editing the case report.

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