EDITORIAL

Vascular variants and the evaluation of patients with acute stroke

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Neurology: Clinical Practice June 2019 vol. 9 no. 3 185-186 doi:10.1212/CPJ.0000000000000635

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Variations in the cerebral vasculature that arise during embryogenesis can predispose to stroke or lead to atypical stroke syndromes, and neurologists assessing patients with acute stroke must be aware of some of the most common variations. During fetal development, the cerebral vasculature forms through a process of sprouting, driven by target tissue growth factors induced by ischemia. The process starts when the embryo is around 1.3 mm in length with the development of 6 branchial arch arteries. Subsequent remodeling and appearance of new vessels leads to the formation of the extracranial and intracranial vasculature, a process that continues beyond birth.[1,2] The sprouting of new vessels from parent arteries (angiogenesis) and subsequent involution of anastomoses are driven by the growth of different areas of the brain, and the process is controlled through target tissue growth factors.[2] Although blood flow to the developing brain initially is through the developing carotid artery, the growth of posterior fossa structures eventually leads to the formation of an anterior and posterior circulation, connected through the circle of Willis, which is formed around week 6 or 7 of development.

Autopsy and imaging studies show that the circle of Willis is complete in only 40%–50% of people.[3,4] Processes during embryologic development can lead to anatomical variations in the site of origin of the arteries, the number of vessels, their morphology, and the persistence of fetal anastomoses.[5] The most common variation is an artery that is narrower than usual (hypoplasia) and is seen in almost 30% of people; the arteries most commonly affected are the posterior and the anterior communicating arteries, but the anterior cerebral artery (ACA) and vertebral arteries may also be affected.[3] Other common variants are conditions where 2 vessels run side by side: fenestration (when the proximal arterial lumen splits into 2 separate channels that eventually merge to reconstitute the parent artery, thought to be due to failure of midline fusion during angiogenesis) and duplication (when 2 arteries with separate origins fuse downstream to form a single vessel.) These variants are more common in the anterior circulation, particularly involving the anterior communicating artery.[2] Variants involving the middle cerebral artery (MCA) are less frequent. For example, 1%–3% of people have a duplicated MCA where both MCAs arise from the internal carotid artery (ICA). Another variant is the presence of an accessory MCA that arises from the ACA; this variant is seen in 0.3% of patients undergoing digital subtraction angiography (DSA).[2]

Two articles published in this issue of *Neurology® Clinical Practice* demonstrate how knowledge on these anatomical variants can help guide therapeutic interventions in patients with acute stroke.[6,7] In the first case, Cooke et al. describe a 37-year-old woman who had acute onset of a left MCA syndrome with aphasia and dense hemiplegia. The initial CT scan showed subtle early ischemic changes in the frontal lobe and a hyperdense MCA sign, and the perfusion study confirmed an area of penumbra involving the left MCA territory. Surprisingly, the left MCA was opacified by contrast on CT angiogram (CTA). Because of the discrepancy between the clinical and imaging findings, she underwent DSA. In this study, the MCA was clearly seen, but the lenticulostriate arteries and branches of the superior division of the MCA were missing. These findings prompted a search for an occluded accessory MCA, identified when the interventionalist found a stump at the origin of the ACA. She underwent successful

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Funding information and disclosures are provided at the end of the article. Full disclosure form information provided by the authors is available with the full text of this article at Neurology.org/cp.

recanalization of the occluded accessory MCA with a stent retriever and had a good clinical outcome.[6]

In the second case, Khrlobyan et al.[7] describe a 91-year-old woman who also had acute onset of a severe left MCA syndrome. The baseline CT scan was normal, but CTA showed a patent but narrow left MCA and a small bulge at the origin of the ACA that the authors immediately identified as an occlusion at the origin of an accessory MCA. DSA confirmed the diagnosis, and good recanalization was achieved with aspiration thrombectomy within 4 hours of symptom onset. She also had good neurologic recovery.

Neurologists who evaluate patients with acute stroke have to make quick therapeutic decisions and, aided by the clinical evaluation and multimodal imaging studies, they can now select among different treatment options (thrombolytics, endovascular procedures using a variety of stent retrievers, or aspiration.) As demonstrated by the 2 cases presented in this issue, in both of which there was a discrepancy between the clinical picture and the imaging findings that led to a search for vascular variants, a detailed knowledge of vascular syndromes and anatomical variants is necessary when interpreting imaging studies to avoid diagnostic and therapeutic errors in the acute setting. These articles focused on a relatively rare variant: an accessory MCA. Of interest, in the first case, the accessory MCA gave rise to perforating and cortical branches, whereas the normal MCA only gave rise to distal cortical ones. A review of all vascular variants is beyond the scope of this editorial, and we refer the reader to some recent excellent reviews on clinical syndromes and anatomic variants.[1,2,5,8,9] Some of the more common variants that have clinical implications are discussed below.

Variations in morphology can lead to hemodynamic compromise and stroke. Hypoplastic anterior or posterior communicating arteries lead to an incomplete circle of Willis that cannot compensate for proximal vessel occlusions. Hypoplastic ICA-external carotid artery collaterals can worsen the severity of ischemia due to anterior circulation occlusions. Variations in vessel origin can give rise to atypical stroke syndromes. When the posterior cerebral artery (PCA) arises from the ICA with minimal or no flow contribution from the posterior circulation (so called fetal PCA, a relatively common condition), patients may have posterior circulation syndromes because of ICA pathology.[5] Bilateral thalamo-mesencephalic infarcts can occur when the blood supply to both thalami arises from a single arterial trunk from 1 PCA (artery of Percheron). In patients with a hypoplastic or aplastic A1 segment of the ACA, unilateral carotid or ACA disease can lead to bilateral frontal infarcts, which can also occur when the A1 segments for each side fuse to form a single A2 segment (an azygous ACA). Variations in the number of vessels can lead to clinical-imaging discrepancies that should prompt the search for these variations. Trifurcations of the ACA and the MCA are more common, and they must be sought in patients with an apparent normal bifurcation but incomplete filling of the vascular territory on CTA or DSA. The cases presented in this issue remind us of the importance of considering accessory and duplicated vessels.[6,7]

As neurologists, we must always remember that although sophisticated imaging studies are available, our clinical evaluation and assessment should guide all our diagnostic and therapeutic decisions.

Author contributions

J.G. Merino: drafting/revising the manuscript. T. Tavarez: drafting/revising the manuscript.

Study funding

No targeted funding reported.

Disclosure

J.G. Merino is a US research editor for *The BMJ*, and *The BMJ* supports a substantial portion of his salary through a contract between *The BMJ* and his employer, the University of Maryland School of Medicine; is also the coeditor of *Blogging Stroke*, the blog of the *AHA* journal *Stroke*, which is an unpaid position; is a stroke outcome adjudicator for the NIH-funded Women's Health initiative and receives a small honorarium for each adjudication; serves in several committees and programs of the American Academy of Neurology, and the AAN reimburses his expenses for trips related to his participation in these committees and programs; and has received travel support to give talks at the Annual Meeting of the not-for-profit Mexican Academy of Neurology. T. Tavarez reports no disclosures. Full disclosure form information provided by the authors is available with the full text of this article at Neurology.org/cp.

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Neurol Clin Pract 2019;9;185-186 Published Online before print April 9, 2019
DOI 10.1212/CPJ.0000000000000635