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Extracranial Internal Carotid Artery Fenestration: A Case Report

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Abstract: Fenestration of the internal carotid artery (ICA) is an extremely rare congenital abnormality thought to be secondary to incomplete segmental fusion of vessels at the early stages of their development. Fenestration is usually asymptomatic and often misdiagnosed as arterial dissection. Distinction between the two can be challenging on both magnetic resonance angiography and computed tomography angiography, and digital subtraction angiography must often be utilized for precise characterization of the abnormalities. Certain imaging features, namely the length of the involved arterial segment, the regularity of luminal contour, the symmetry of arterial limbs, and the absence/presence of fusiform dilatation of either limb, produced by noninvasive angiography have the potential to help distinguish between an ICA fenestration and dissection. We report a case of an asymptomatic, true fenestration of a short segment of ICA and discuss its imaging characteristics.

Keywords: *internal carotid artery (ICA) fenestration, arterial fenestration, arterial dissection*

Case Presentation

A 71-year-old man presented with intense, intermittent, shock-like shooting pain within the right side of his face. The results of a neurologic examination and a standard laboratory workup were within normal limits. An initial magnetic resonance (MR) imaging of the brain and MR angiogram of the head and the neck without contrast were obtained to assess the possible structural causes of suspected trigeminal neuralgia.

The initial MRI of the brain was unrevealing. However, the noncontrast MR angiogram of the neck (Figures 1 and 2) showed a division of a short distal aspect of the C1 segment of the right internal carotid artery (ICA), with no gross proportional asymmetry, luminal contour irregularity, or fusiform dilatation of either of the arterial limbs. There were no areas of stenosis or luminal irregularity involving the intracranial or the extracranial vasculature. The findings were thought to be representative of a true ICA

Key Points

- Fenestration of the extracranial internal carotid artery is infrequently seen on MR and computed tomography angiography and often misdiagnosed as arterial dissection.
- The imaging features that favor a true arterial fenestration over a true dissection include shorter length of the involved arterial segment, regularity of luminal contour, proportional symmetry of arterial limbs, and absence of fusiform dilatation of either limb.
- Digital subtraction angiography is the preferred imaging modality to identify often-subtle differences between fenestration and dissection.

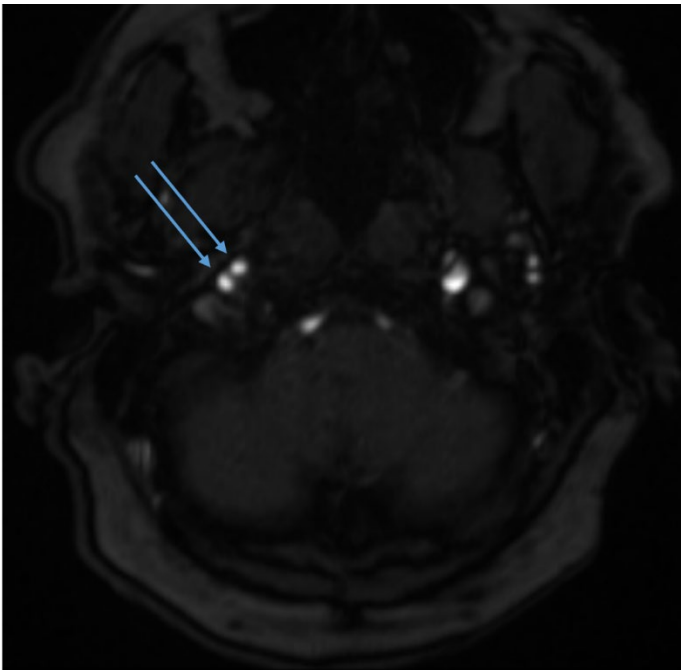
Abbreviations

ICA: internal carotid artery
MRI: magnetic resonance imaging
MCA: middle cerebral artery
MRA: magnetic resonance angiography
MIP: maximum intensity projection

fenestration, likely asymptomatic and unrelated to the patient's clinical presentation.

After being prescribed a titrated dose (with initial low dose) of carbamazepine on an outpatient basis, the patient eventually experienced significant relief of symptoms.

Figure 1. Noncontrast Magnetic Resonance Angiogram (MRA) of the Neck of a 71-Year-Old Man with Fenestration of a Distal Part of the C1 Segment of the Right Internal Carotid Artery (ICA)



Noncontrast time of flight MRA image, axial view, shows the right ICA fenestration (arrows) in cross-section with two separate, patent lumens.

Discussion

Vascular fenestration is thought to be secondary to failed or incomplete segmental fusion of embryonic vessels.¹ From this perspective, arterial fenestration is a division of an arterial segment into at least two patent limbs that have separate intimal and muscular layers and sometimes a shared adventitial layer, depending on the level of vascular fusion during the embryonic development, and reunite to a single lumen distally.^{1,2} While some embryogenic mechanisms have been proposed to explain fenestrations of the middle cerebral arteries (MCA), the distal vertebral, or the basilar arteries, there is no clear understanding of the pathogenesis of cervical ICA

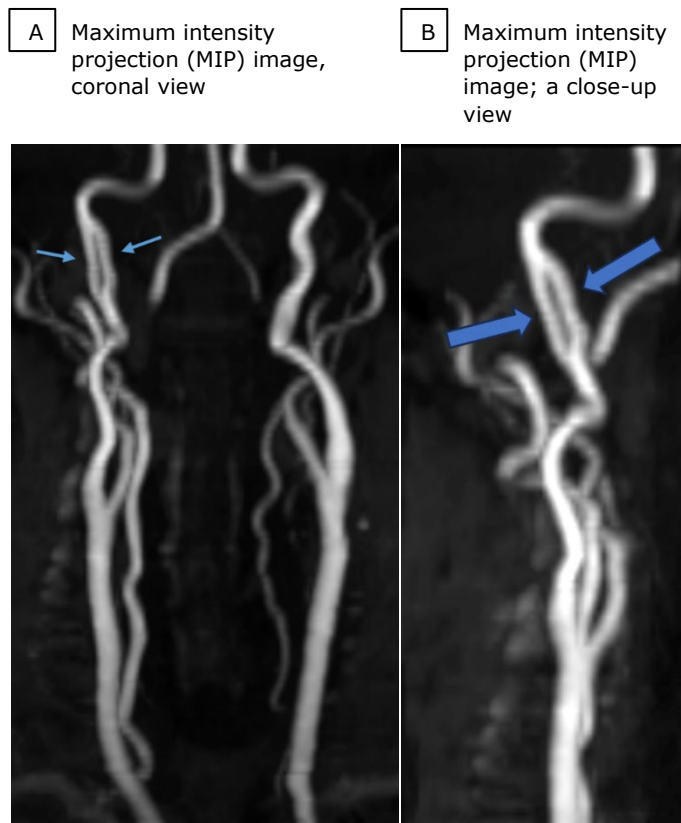
fenestrations.³ Gailloud et al³ argued, although controversially,⁴ that apparent fenestrations of the extracranial ICA are in fact sequelae of arterial dissections, actual arterial dissections, or so-called pseudofenestration with a patent false lumen.

Within the intracranial and the extracranial vasculature, fenestration has been well documented in the anterior communicating arteries, the vertebral and the basilar arteries, as well as the anterior cerebral arteries and the MCAs in both angiographic and postmortem studies.¹ However, true fenestration of the ICA is much less frequently seen and is thought to be an extremely rare congenital variant, with less than 30 cases of intracranial ICA fenestrations reported, according to Liang et al.⁵ There have been even fewer documented cases of extracranial cervical ICA fenestration in the literature.⁴

Without a definite history of trauma or known underlying arterial pathology (eg, fibromuscular dysplasia), it can often be difficult to distinguish between fenestration and dissection based on imaging alone.³ However, there are a few imaging findings, contour irregularity involving the arterial lumen, a proportional asymmetry of the limbs of the arterial segment, and a fusiform dilatation of one or both of the limbs, that can suggest the presence of an arterial dissection.³ Notably, an association has been described between fenestrations and aneurysm development at the proximal part of the fenestration, hypothesized to be secondary to flow-related weakness or defects in the tunica media.^{1,5,6} The length of the abnormality can also be a helpful differentiation tool, as fenestrations typically extend over a shorter distance up to a few centimeters, while dissections usually extend over a longer arterial segment.³ Lastly, the location of the abnormality can be another differentiation tool, as ICA dissections are commonly located in the immediate proximity of the skull base.³

In most cases, digital subtraction angiography is the preferred imaging modality to identify subtle contour irregularities, intimal injury, as well as vascular limb asymmetry and dilatations involving the intracranial and the extracranial vasculature.^{3,5} However, given the wide applicability and noninvasive nature of CT and MR imaging, fenestrations can be identified on these imaging modalities first. Because of a lack of

Figure 2. Noncontrast Magnetic Resonance Angiogram (MRA) of the Neck, of a 71-year-old Man with Fenestration of a Distal Part of the C1 Segment of the Right Internal Carotid Artery (ICA)



(A) MIP image, coronal view, of the noncontrast MRA of the neck shows fenestration (A, arrows) of the C1 segment of the right ICA, with the two separate arterial limbs having smooth luminal contours, symmetric proportions, and no associated aneurysmal dilatation. The remaining extracranial neck vessels are within normal limits.

(B) A close-up view of the right ICA shows the fenestration (B, arrows) with the two arterial limbs without contour irregularity or associated aneurysmal dilatation.

understanding of the pathogenesis of fenestrations, the difficulty in differentiating between fenestration and dissection on cross-sectional imaging, as well as the necessity to determine a risk of cerebrovascular events associated with fenestrations and to make an appropriate selection of patients who might benefit from invasive angiography, the differential diagnosis of fenestration should always include

consideration of dissection.^{1,3-5} However, when imaging shows a short ICA segment duplication with a regular luminal contour, a proportional symmetry of arterial limbs, and the absence of fusiform dilatation of either limb, the diagnosis of true ICA fenestration may be warranted.

Author Contributions

Conceptualization, G.H.; Acquisition, analysis, and interpretation of data, Z.W. and A.Z.; Writing – original draft preparation, Z.W.; Review and editing, G.H. and Z.W.; Supervision, G.H. All authors agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. All authors had full access to all the data in the study and take responsibility for the integrity of the data and the accuracy of the data analysis.

Disclosures

None to report.

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