Complete Duplication or Extreme Fenestration of the Basilar Artery

Jonas H. Goldstein, Richard Woodcock, Huy M. Do, C. Douglas Phillips, and Jacques E. Dion

Summary: We describe a 42-year-old man with complete duplication or extreme fenestration of the basilar artery. We review the developmental anatomy and embryology and discuss the possible clinical implications and associated findings of this anomaly.

The term fenestration refers to a localized duplication of a vessel. While fenestration of the basilar artery has been reported to be as high as 6% in postmortem studies, its angiographic prevalence has been described as ranging from 0.04% to 0.6% (1). Basilar artery fenestration most commonly occurs in the lower half of the vessel (2), extending for a length of less than 5 mm (3, 4). Complete duplication of the basilar artery, with each vertebral artery continuing separately to form a posterior cerebral artery, has been described previously (5, 6).

We present a case of complete duplication or extreme fenestration of the basilar artery demonstrated by catheter digital subtraction angiography (DSA).

Case Report

A 42-year-old obese man with rhizomelia and mild mental retardation was admitted to the emergency department with abrupt onset of a left temporal headache accompanied by nausea and vomiting. A noncontrast head CT scan was obtained and showed no evidence of subarachnoid hemorrhage. Lumbar puncture revealed equivocal findings for subarachnoid hemorrhage. Diagnostic cerebral angiography was requested to exclude an aneurysm or vascular malformation.

Cerebral angiography showed the left vertebral artery as arising from the aortic arch and giving origin to the thyrocervical trunk in its cervical course. Likewise, the thyrocervical trunk on the right arose from the vertebral artery. The left vertebral artery injection did not show reflux into the contralateral vertebral artery. Although the ipsilateral posterior cerebral, superior, anterior inferior, and posterior inferior cerebellar arteries were present and normal, their contralateral counterparts were not identified (Fig 1A), with the exception of transient opacification of the right superior cerebellar artery via a small communicating branch. The right vertebral artery injection revealed similar findings (Fig 1B), with the addition of two small bridging vessels to the contralateral vertebral artery, one at the proximal and one at the distal segment. Additional vascular anomalies included the left pericallosal artery arising from the right anterior cerebral artery, and the right ophthalmic artery arising from the internal carotid artery cavernous segment. The posterior communicating arteries were not identified and the remainder of the examination failed to reveal a cause of subarachnoid hemorrhage. Subsequently, an MR imaging/MR angiographic examination was performed, with the MR angiogram confirming findings on the conventional angiogram (Fig 1C and D). The patient was clinically stable for 48 hours and was discharged.

Discussion

The embryology of the basilar artery has been well described by Padget (7). Fusion of the embryonic longitudinal neural arteries into a single basilar artery occurs in a craniocaudal direction (8) by approximately the fifth fetal week (7). Fenestrations or “windows” within the basilar artery occur as a result of failure of fusion of the neural arteries and of regression of the bridging arteries that connect the longitudinal arteries. The term fenestration has been used interchangeably with partial duplication (9) or segmental duplication (10–12). Although partial or segmental duplications (fenestrations) have a reported rate of occurrence of up to 5% to 6% (11), true or complete duplication is extremely rare (4–6).

This case illustrates complete duplication or extreme fenestration of the basilar artery, with each vertebral artery terminating in a posterior cerebral artery. Given the patient’s rhizomelia and mental retardation, we were curious as to whether this vascular anomaly was part of a chromosomal syndrome. A normal karyotype result had been obtained several years earlier, however. This nonfusion of the embryologic longitudinal neural arches was thought to be an incidental finding. It was interesting to note both a small superior and inferior connecting artery (Fig 1B); we hypothesized that the superior connecting artery was composed of the P1 segments of the posterior cerebral arteries and the terminal basilar artery, which together may represent a posterior cerebral vessel analogous to the anterior communicating artery. These two small bridging arteries were not thought to be subjected to the same type of hemodynamic alterations as true fenestrations, and therefore did not indicate an increased risk of aneurysmal formation sufficient to justify further surveillance.
Fig 1. Left and right vertebral artery angiograms (A and B) and MR angiograms (C and D) in a 42-year-old man with rhizomelia and mild mental retardation.

A, Anteroposterior projection of left vertebral angiogram shows filling of basilar artery to the left of midline (straight arrow). There is opacification of a common trunk for the AICA and PICA (curved arrow), with no filling of the right AICA/PICA trunk. A small branch is seen from the distal basilar artery (arrowhead).

B, Anteroposterior projection of right vertebral angiogram shows filling of basilar artery to the right of midline (straight solid arrow). There is opacification of a common trunk for the AICA and PICA on the right (curved arrow), with no filling of the left AICA. A branch from the distal basilar artery (arrowhead) opacifies left PCA (open arrow).

C, Angled anteroposterior projection of MR angiogram shows two vertebrobasilar systems (arrows), each filling an ipsilateral PCA. The distal basilar cross-filling branch faintly opacifies (arrowhead).

D, Anteroposterior projection of MR angiogram shows proximal portion of the two vertebrobasilar systems (arrows).

References