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Case Report

Persistent Nonfused Segments of the Basilar Artery: Longitudinal versus Axial Nonfusion

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Summary: Embryologic development of the basilar artery occurs along two axis systems: longitudinal fusion and axial fusion. Longitudinal fusion consists of midline fusion of paired ventral arteries and reflects the simplified pattern of arterial anatomy found in the spinal cord. Axial fusion consists of fusion of the distal basilar artery, which arises from the caudal division of the internal carotid artery, to the mid-basilar agenesis to the posterior inferior cerebellar artery termination of the vertebral arteries. Persistent longitudinal nonfusion (or complete duplication) of the basilar artery is very rare, and persistent axial nonfusion is even rarer. We report one case of persistent longitudinal nonfusion of the basilar artery in a 3-year-old boy and a case of persistent axial nonfusion of the basilar artery in a 43-year-old man.

The embryologic development of the basilar artery has been described as two separate processes occurring at two levels, longitudinal and axial (1). The longitudinal system is a continuation of the pattern found in the arterial anatomy of the spinal cord, whereas axial development is representative of the cerebral anatomy. The junction of the two systems is found at the trigeminal remnant site on the basilar artery (1). The longitudinal development of the basilar artery consists of midline fusion of two paired ventral arteries in a craniocaudal manner (1, 2). The axial development of the basilar artery consists of fusion of the caudal division of the internal carotid artery, producing the distal basilar artery, to the mid-basilar agenesis to the posterior inferior cerebellar artery termination of the vertebral arteries (1, 3).

Persistent longitudinal nonfusion of the basilar artery is very rare, and persistent axial nonfusion is even rarer. We report the case of a patient with a persistent longitudinal nonfusion and a patient with a persistent axial nonfusion of the basilar artery.

Case Illustration

Patient 1: Persistent Longitudinal Nonfusion of the Basilar Artery

A 3-year-old boy presented with a generalized tonic-clonic seizure. He was a full-term infant after a normal spontaneous vaginal delivery. His medical history was notable for previous seizures and delayed development (he says only two or three words, does not walk, and does not feed himself). He was being treated with Phenobarbital. Physical examination revealed an open anterior fontanel, a slightly open posterior fontanel, a prominent forehead, wide base of the nose, and slightly posteriorly rotated ears.

A brain MR imaging and MR angiographic (MRA) examination was performed with conscious sedation. Examination demonstrated complete duplication (or longitudinal nonfusion) of the basilar artery (Figs 1A and B). Phenytoin was added to his anticonvulsant regimen. The patient remained neurologically stable.

Patient 2: Persistent Axial Nonfusion of the Basilar Artery

A 43-year-old man presented with several transient episodes of vertigo, tongue numbness, and diplopia on exertion. Each episode lasted for only a few minutes. His medical history was notable for slight hypercholesterolemia. Physical examination demonstrated normal cranial nerves; normal strength, sensation, and coordination; and normal balance. MR and MRA images of the brain obtained by his primary care physician (Fig 2A) suggested a possible basilar stenosis. Aspirin and clopidogrel were prescribed, and he was referred to our service. We performed a cerebral digital subtraction angiogram (DSA), which demonstrated persistent axial nonfusion of the basilar artery (Fig 2B–F). There are three separate nonfused segments of the basilar artery. Bilateral vertebral arteries terminate in posterior inferior cerebellar arteries. The midbasilar trunk is a separate nonfused segment that supplies bilateral anterior inferior cerebellar arteries and bilateral superior cerebellar arteries and is supplied via a persistent trigeminal remnant artery. The distal basilar territory is supplied by bilateral fetal origin posterior cerebral arteries.

He remained neurologically stable while taking aspirin and clopidogrel. His transient symptomatic episodes were thought to be a hypoperfusion syndrome related to the nonfused separate segments supplying the brain stem without the luxury of collateral flow.

Discussion

These two cases demonstrate the two processes of fusion, longitudinal and axial, of the basilar artery during embryologic development. The longitudinal system involves midline fusion of the two ventral arteries and is an extension of the simplified pattern of anatomy found in the spinal cord (1, 2). The axial system, by contrast, involves fusion of the distal basilar artery, which develops from the caudal division of

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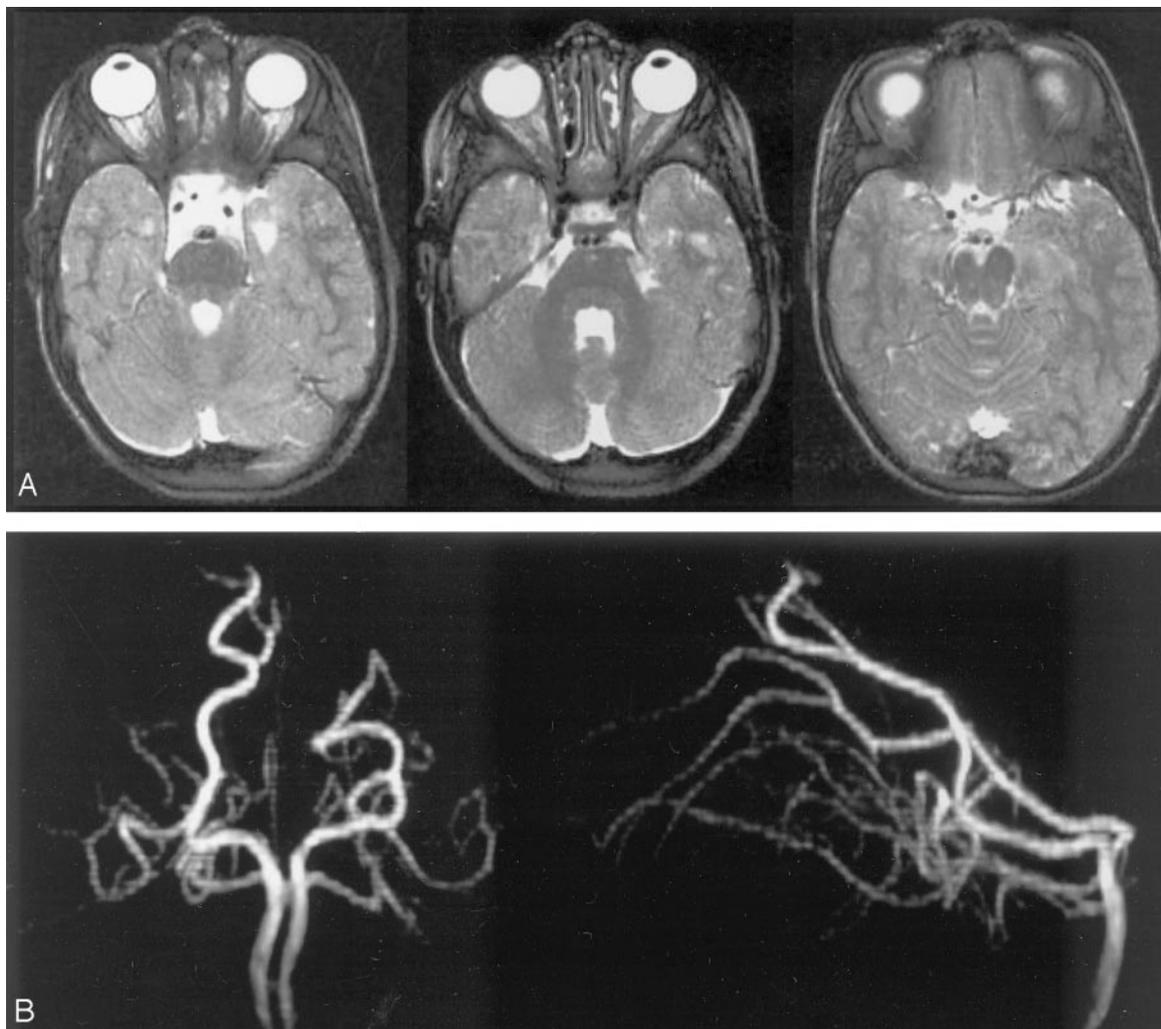


Fig 1. Patient 1 (longitudinal nonfusion of basilar artery), a 3-year-old boy who presented with generalized tonic-clonic seizure.

A, Consecutive axial images of T2-weighted brain MR imaging study demonstrate the two flow voids of the separate nonfused duplicated basilar arteries.

B, Two-dimensional time-of-flight reconstructions of MRA demonstrate the two separate nonfused duplicated basilar arteries.

the internal carotid artery, to the midbasilar agenesis, to the posterior inferior cerebellar artery termination of the vertebral arteries (1). The axial system reflects the more complex pattern of the cerebral anatomy. The "pivotal point" seems to be the site of the trigeminal remnant (1). The trigeminal artery appears embryologically at the 4-mm stage and involutes at the 7–12-mm stage (4). It arises from the basilar artery, unlike the posterior communicating artery, another of the carotid-basilar anastomosis, which arises from the posterior division of the internal carotid artery (4). A persistent trigeminal artery is estimated to occur in 0.1–0.2% of the population (4) and, in most cases, is an incidental finding. In the case of patient 2, however, the persistent trigeminal artery is vital in that it is the only supply to the midbasilar trunk segment that supplies bilateral anterior inferior cerebellar arteries, bilateral superior cerebellar arteries, and the pontine perforators. On MRA images, it could be misinterpreted as a carotid aneurysm. In some instances, the trigeminal segment may traverse the sella and can potentially be misinterpreted as a pituitary lesion.

Although partial fenestrations of the basilar artery, usually occurring at the proximal segment, which reflect the craniocaudal direction of longitudinal fusion, are thought to occur in about 5% of the population on the basis of autopsy studies (5), there have only been a few reported cases of complete duplication (or longitudinal nonfusion) of the basilar artery (6–10). Axial nonfusion is even more rare (1).

The second patient (axial nonfusion) demonstrates one of the potential pitfalls of MRA. The signal intensity dropout at the site of nonfusion on the MRA image was originally interpreted as basilar artery stenosis. Performance of cerebral DSA demonstrated this actually to be anomalous axial nonfusion of the basilar artery.

Conclusion

Fusion of the basilar artery during embryologic development occurs longitudinally and axially. Whereas longitudinal nonfusion of the basilar artery (or complete duplication) is very rare, axial nonfusion is even more so. The MRA findings in the patient with axial nonfu-

FIG 2. Patient 2 (axial nonfusion of basilar artery), a 43-year-old man who presented with transient episodes of vertigo, tongue numbness, and diplopia.

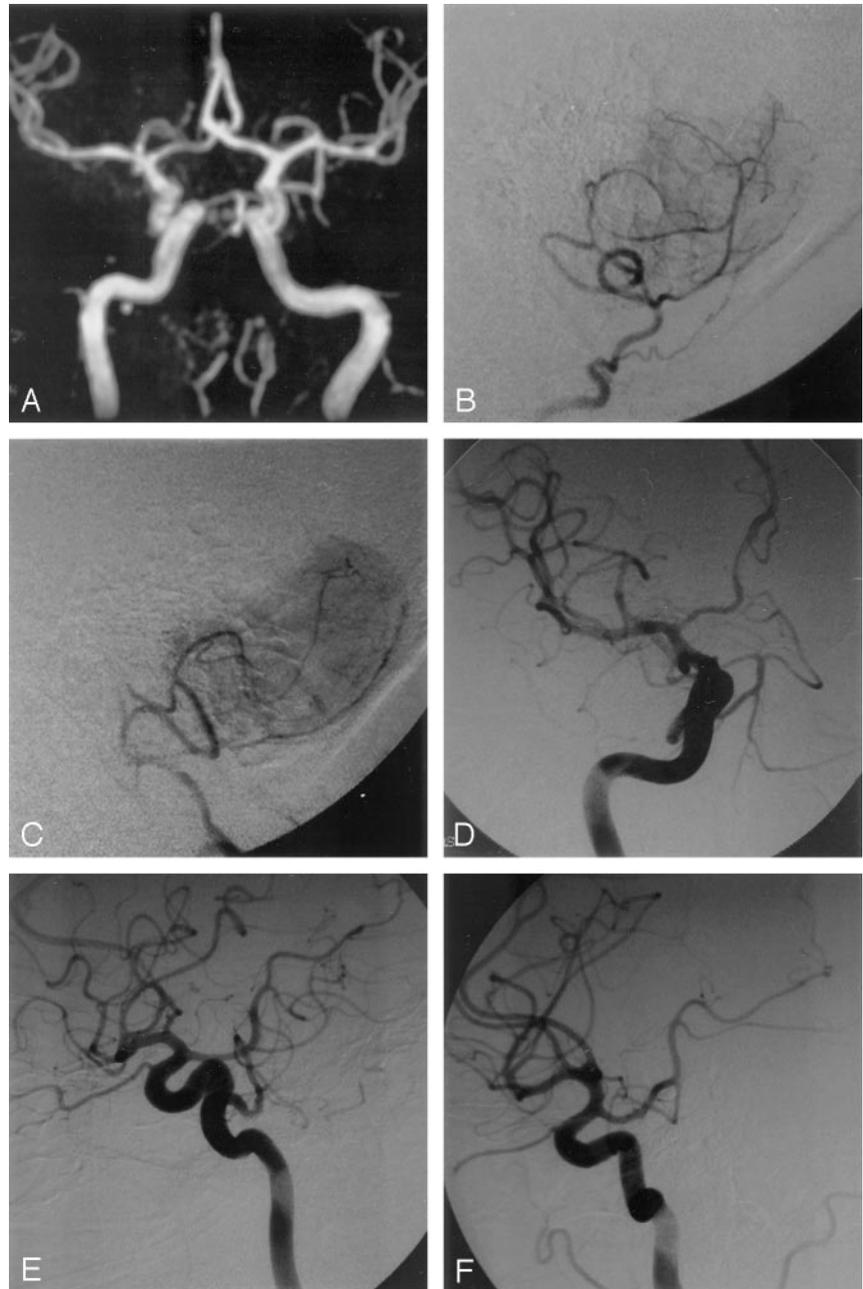
A, Two-dimensional time-of-flight reconstructions of MRA demonstrate signal intensity dropout at the basilar artery trunk. This was originally interpreted as basilar artery stenosis.

B, Lateral projection left vertebral artery injection DSA demonstrates the left vertebral artery terminates in the left posterior inferior cerebellar artery.

C, Lateral projection right vertebral artery injection DSA demonstrates that the right vertebral artery terminates in the right posterior inferior cerebellar artery.

D, Anteroposterior projection, and E, lateral projection right internal carotid artery injection DSA, demonstrate the separate nonfused midbasilar trunk segment supplying the bilateral anterior inferior cerebellar arteries and bilateral superior cerebellar arteries and supplied via a persistent trigeminal remnant artery. There is a fetal origin right posterior cerebral artery, which supplies a separate distal basilar segment.

F, Lateral projection left common carotid artery injection DSA demonstrates the fetal origin left posterior cerebral artery.



sion was originally interpreted as basilar stenosis demonstrating a potential pitfall in MRA compared with conventional cerebral angiography.

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