Segmental Duplication of the Basilar Artery With Thrombosis

Allen D. Berry III, MD, John J. Kepes, MD, and Mark D. Wetzel, MD

Duplication or fenestration of the basilar artery, a result of an embryologic malformation, has an incidence of up to 5.26% in the general population. The most common complication of this anomaly is the formation of aneurysms. Thrombosis of a partially duplicated basilar artery developed in a 43-year-old man who complained of visual disturbances followed by seizures and coma, and who eventually died. Autopsy showed a partially organized thrombus occluding both halves of a duplicated portion of the basilar artery, old infarcts in the calcarine cortices, and a recent large infarct in the basis pontis. There was only minimal atherosclerosis of other intracranial arteries, including the vertebral arteries. Hemodynamic disturbances and turbulent blood flow at the site of fenestration may be the cause of the thrombosis that occurred in this artery. (Stroke 1988;19:256-260)

Partial duplication or fenestration is a frequent embryologic malformation of the basilar artery, having an incidence of up to 5.26% in the general population. The most common complication of this anomaly is the formation of an aneurysm at the site of the duplication, with other complications such as tortuosity and atherosclerotic dilatation occurring less frequently. We report a case of basilar artery duplication complicated by thrombosis leading to pontine infarction that in turn caused a clinical "locked-in" syndrome and eventual death. Thrombosis in a fenestrated cerebral artery, with such thrombus acting as a source of emboli, was suggested by Takahashi et al but our case appears to be the first one with pathologic confirmation.

Case Report

This 43-year-old white man was in his usual state of good health until June 1986, at which time he began to notice problems with his eyesight, including loss of peripheral vision and intermittent dysconjugate gaze, as well as headaches. Approximately 2 weeks later he developed stiffness of the neck for which he underwent chiropractic cervical spine manipulation, without relief of symptoms. Over the next week, progressing to severe headache, nausea and vomiting, and pronounced dysconjugate gaze. This was followed by generalized tonic-clonic seizures, leading to status epilepticus that lasted for several hours. The seizures were controlled after admission to a local hospital, but in the postictal state he was noted to be unresponsive with decerebrate posture. Computed tomography (CT scan) of his head was unremarkable. He was transferred to the University of Kansas Medical Center where he remained in a coma with continued decerebrate posturing. Nuclear magnetic resonance imaging (NMRI) demonstrated an increased T2-weighted image in the pons without pontine enlargement, mass effect, or other cerebral abnormalities.

Several days after admission, he regained consciousness but was found to be in a locked-in state with complete quadriplegia and paralysis of the motor cranial nerves, with the exception of the ability to curl his lip on the left side and to blink both eyes. While hospitalized, he suffered recurrent aspiration pneumonias and phlebitis of the lower extremities. He became febrile, with blood cultures positive for *Candida albicans*, and was treated with amphotericin B. He continued to deteriorate, with renal failure, and lapsed into a uremic coma, in which he remained until his death after 3 weeks of hospitalization, 6 weeks from the time of onset of symptoms.

Pathologic Findings

The brain weighed 1,530 g and demonstrated generalized edema without evidence of uncal, tonsillar, or other herniations. The arteries of the circle of Willis were found in their correct anatomic positions. The vertebral arteries were patent, and their walls showed very slight fibrous thickening in a few foci. The basilar artery showed bluish-red discoloration of its walls (Figure 1). It felt quite firm, and when viewed from the outside it appeared to be filled with a clot. Only the most rostral 3 mm of the artery had a patent lumen and thin membranous walls. The caudal half of the occluded artery appeared much wider than the rostral portion (Figure 1, arrows), but only after sectioning the artery did it become apparent that in the grossly enlarged area there was a double-barreled lumen, with both channels...
FIGURE 1. Base of the brain. The basilar artery, except for its most rostral segment, has been occluded by a thrombus. The caudal 0.8 cm is much wider than the segment rostral to it (arrows). Widened area had the reduplication of the lumen. Right vertebral artery is normal; the left was accidentally cut off at autopsy. It, too, was patent.

occluded by a red-brown firm thrombus. There was no appreciable atherosclerosis involving the wall of the artery. The thrombus extended rostrally into the portion of the artery that was single-barreled. The double-barreled portion of the artery was 0.8 cm long.

Coronal sections of the brain showed it to be essentially normal in its anterior aspects, but in the occipital lobes the cortex lining the calcarine fissures on both sides showed patchy areas of atrophy, granular softening, and slight brown and dusky discoloration. The midbrain and cerebellum appeared grossly normal, but in the upper pons the right half of the base showed marked softening and brown-red discoloration with loss of the usual anatomic landmarks, and at the level of the lower pons the same changes involved the entire width of the base, leaving only parts of the tegmentum intact (Figure 2). The medulla oblongata showed no gross abnormalities.

Histologic sections of the cerebral arteries other than the basilar artery showed no significant alterations. The intracranial portions of the vertebral arteries showed occasional foci of very slight fibrous thickening of their walls.

Sections taken from the lower two thirds of the basilar artery again showed a reduplication of the vessel, with the two branches lying very close together; both of them were occluded by a thrombus that displayed areas of organization and partial recanalization (Figure 3). At the rostral end of the reduplication, cross sections showed partial division of the lumen caused by a spur-like projection of the wall (Figure 4). The internal elastic membranes of the arterial branches were preserved in most areas but exhibited a few foci of discontinuity on Verhoeff-van Gieson elastica stain (Figure 5). Within the unorganized portions of the occluding thrombus parallel layers of fibrin (lines of Zahn) could be observed.

Sections of the pons in the grossly described areas

FIGURE 2. Recent infarct of pons with involvement of the entire base and paramedian areas of the tegmentum.
of discoloration and softening showed a typical subacute infarct in a state of partial resolution, mostly populated by lipid-laden macrophages (gitter cells) and showing only occasional astrocytes. In contrast, the infarcts of the calcarine cortices appeared to be older because a much larger contingent of fibrous astrocytes surrounded the rather narrow linear zones of infarction.

In addition, although gross examination of the brain did not suggest it, there were many microscopic areas of abscesses and tiny granulomas with macrophages and multinucleated giant cells in gray and white matter alike, all containing pseudohyphae of Candida organisms, reflecting the patient's terminal Candida sepsis. Some microabscesses abutted on the periphery of infarcted areas in the pons, but the infarcts themselves appeared to be of the bland, nonseptic variety. Careful scrutiny of the wall of the basilar artery and the thrombus itself failed to show the presence of fungi.

The general autopsy findings included bilateral candidal pyelonephritis, but in other viscera fungi were not identified histologically. There was no evidence of endocardial thrombi, cardiac valvular vegetations, or any other likely source for embolism to the basilar artery. Postmortem blood cultures were positive for Candida albicans.

Figure 3. Section through the basilar artery 4 mm rostral to its lower end: two separate lumens are seen, both filled with thrombus that shows partial organization and recanalization. Median walls of the two arteries are fused. Reticulin stain, ×22.

Figure 4. At the rostral end of the reduplicated segment of basilar artery, the two halves communicate and are separated only by a sharp spur extending about half way into lumen. Focus of organization is seen on the right. Gomori's trichrome stain, ×20.
Discussion

Reduplication or fenestration of the basilar artery is a peculiar congenital anomaly of that structure. As well documented by Padget, the basilar artery develops from paired primitive longitudinal neural arteries that are formed in the 4–5-mm embryo during its first stage of development. These vessels course longitudinally along the ventral portion of the hindbrain and form focal connections with each other across the midline. During the second stage of development, at 5 weeks’ gestation, there is gradual fusion of the channels to form the basilar artery. Fenestration of the basilar artery is believed to be the result of incomplete fusion of the paired arteries.

As can be expected from these embryologic data, the spectrum of fenestration of the basilar artery is wide, ranging from a small window between the arterial segments to partial or even complete duplication. Fenestration occurs most commonly at the proximal (caudal) end of the artery but may involve the vessel anywhere along its course. Such fenestration or segmental reduplication of the basilar artery is not rare. Stopford, the first to describe this anomaly in 1916, encountered one example in 150 necropsies, whereas Wollschlaeger et al recorded an incidence of 5.26% from 291 postmortem examinations.

Most cases of basilar artery occlusion are due to atherosclerosis with superimposed thrombosis; many other causes exist, including embolism, arteritis, infections, clivus fracture, dissecting aneurysms, fibromuscular dysplasia, or "idiopathic thrombosis." In previous reports and reviews on basilar artery occlusion, there have been no cases described having basilar artery fenestration as a cause.

In our case, the possible role of chiropractic manipulation in providing an additional complicating factor must be commented on. While such a role cannot be ruled out with absolute certainty, it is likely that the thrombotic process that had begun prior to the patient’s seeking help for its symptoms (visual disturbances) from a chiropractor would have run its course, leading to complete occlusion without added physical trauma to the vertebrobasilar system. The thrombus at autopsy showed areas of different ages and stages of organization. It can be assumed that, when the thrombotic occlusion was still incomplete, the basilar circulation already became incapable of providing adequate blood supply to areas at the “end of the line,” such as the calcarine cortices of both sides, resulting in the older infarcts found in these areas at autopsy, and clinically leading to disturbances in cortical vision. (Alternately, small emboli detached from the basilar artery thrombus during its formation and swept to the calcarine area may also have been responsible for these symptoms.) Early ischemic changes involving the pontine gaze center may have been responsible for the symptoms and signs of dysconjugate gaze. Eventually, complete thrombotic occlusion of both barrels of the fenestrated basilar artery led to massive infarction of the basis pontis, which in turn resulted in quadriplegia, the locked-in syndrome, and ultimately the patient’s death.

Hemodynamic alterations and turbulence may make the fenestrated artery a more likely site for thrombosis than a normal artery. Black and Ansbacher noted endothelium-lined partial intraluminal septa within the fenestrated artery that can appear as spurs (as in Figure 4). Such spurs could act as points of turbulence and initial sites for potential thrombosis.

The possibility of thrombosis within a fenestrated artery was first postulated by Takahashi et al, who, using angiography, observed bilateral occlusion of the posterior cerebral arteries associated with fenestration of the extracranial portion of the left vertebral artery. Embolization from a suspected thrombus in the fenestrated vertebral artery was entertained in their case even...
though the authors considered their findings to be coincidental events. Our case documents thrombosis as one of the potential complications associated with fenestrated or double-barreled cerebral arteries.

Acknowledgments
The authors are grateful for the technical help of Ms. Janine Flaschhoen for preparing the microscopic slides and for the secretarial assistance of Ms. Geri Kruger.

References

KEY WORDS • basilar artery • fenestration • thrombosis
Segmental duplication of the basilar artery with thrombosis.
A D Berry, 3rd, J J Kepes and M D Wetzel

Stroke. 1988;19:256-260
doi: 10.1161/01.STR.19.2.256

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://stroke.ahajournals.org/content/19/2/256

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published in Stroke can be obtained via RightsLink, a service of the Copyright Clearance Center, not the Editorial Office. Once the online version of the published article for which permission is being requested is located, click Request Permissions in the middle column of the Web page under Services. Further information about this process is available in the Permissions and Rights Question and Answer document.

Reprints: Information about reprints can be found online at:
http://www.lww.com/reprints

Subscriptions: Information about subscribing to Stroke is online at:
http://stroke.ahajournals.org//subscriptions/