

Extreme Duplication-type, Nonseparated Fenestration of the Basilar Artery in a Patient with Pontine Infarction: Confirmation with Virtual Arterial Endoscopy

Seong-Ryong Woo, M.D., Man-Wook Seo, M.D., Young-Hyun Kim, M.D., Hyo-Sung Kwak, M.D.*, Young-Min Han, M.D.*, Gyung-Ho Chung, M.D.*, Seul-Ki Jeong, M.D.

Departments of Neurology and Diagnostic Radiology, Chonbuk National University Medical School, Jeonju, Korea*

We present a case of a 55-year-old woman with pontine infarction and fenestration of the basilar artery that was demonstrated using virtual endoscopy. The patient had motor weakness of the right side with sudden onset. Diffusion- and T2-weighted MR images revealed high signal intensities in the left pontine regions, and an MR angiographic examination showed the double-lumen sign of the basilar artery. An extensive duplication-type, nonseparated fenestration of the basilar artery was diagnosed by virtual endoscopy and craniocaudal communications with discrete openings were observed. We report a case of a patient with basilar artery fenestration and associated brainstem infarction.

J Clin Neurol 2(1):74-77, 2006

Key Words : Fenestration, Basilar artery, Brainstem infarction, Virtual arterial endoscopy

Fenestration refers to the localized duplication of a vessel. Vascular abnormalities are divided into two main configurations: (1) duplications, which have a relatively large arterial window, and (2) arterial slits, which are assumed to be intraluminal defects.¹ Duplications can be subdivided into either partial or segmental types and either true or complete types.

The basilar artery is formed by the fusion of the plexiform, primitive, longitudinal neural arteries in the craniocaudal direction by approximately the 5th fetal week.² Duplication of the basilar artery can result from a failure of the fusion of these embryonic precursors. Both lumens of the fenestrated segments are lined by endothelial tissue and are separated by a tunica media, but they can share a common tunica adventitia.

Compared with the partial or segmental types, true or

complete duplication-type fenestrations are reported to be extremely rare.^{3,4} Only one case report of autopsy findings of duplication-type basilar artery fenestration and associated brainstem infarction has appeared in the literature.⁵ In the present article, we present a case of complete duplication-type fenestration of the basilar artery found in a patient with pontine infarction. Both proximal and distal communications of the fenestration were demonstrated with virtual arterial endoscopy, which has never been reported previously.

CASE REPORT

A 55-year-old woman suddenly developed motor weakness of the right side and dysarthria 1 day prior to

Received : December 7, 2005 / Accepted : January 31, 2006 / Address for correspondence : Seul-Ki Jeong, M.D.

Department of Neurology, Chonbuk National University Medical School, 634-18, Geumam 2-dong, Deokjin-gu, Jeonju-si, Chonbuk, 561-712, Korea
Tel: +82-63-250-1590, Fax: +82-63-251-9363, E-mail: jeongsk@chonbuk.ac.kr

admission. On admission she had mild dysarthria, especially of the lingual components, hemiparesis of the right side (motor grade III), and right facial palsy. The vascular risk profile included hypertension and type-2 diabetes mellitus. She was a nonsmoker and did not drink alcohol. Electrocardiograms and both hematological and biochemical laboratory findings were all within normal limits.

Diffusion- and T2-weighted MR images obtained the same day revealed high signal intensities in the left pontine regions, consistent with an ischemic lesion (Fig. 1-A). The source images for both contrast-enhanced MR imaging and three-dimensional (3-D) MR angiography displayed narrow segments of the basilar artery and

double-lumen signs (Fig. 1-B and C). These MR findings were consistent with a long intimal dissection or fenestration of the basilar artery. Using virtual endoscopy (Rapidia; Infinitt, Seoul, Republic of Korea), a diagnosis of an extreme fenestration was confirmed (Fig. 1-D and E). Because of the complete proximal and distal fusion shown by virtual endoscopy, we speculated that this finding was a basilar arterial fenestration of the true duplication type.

The patient was treated with heparin and later was treated with clopidogrel. Two weeks after the onset of symptoms, hemiparesis of the right side improved (motor grade V-) and she was discharged.

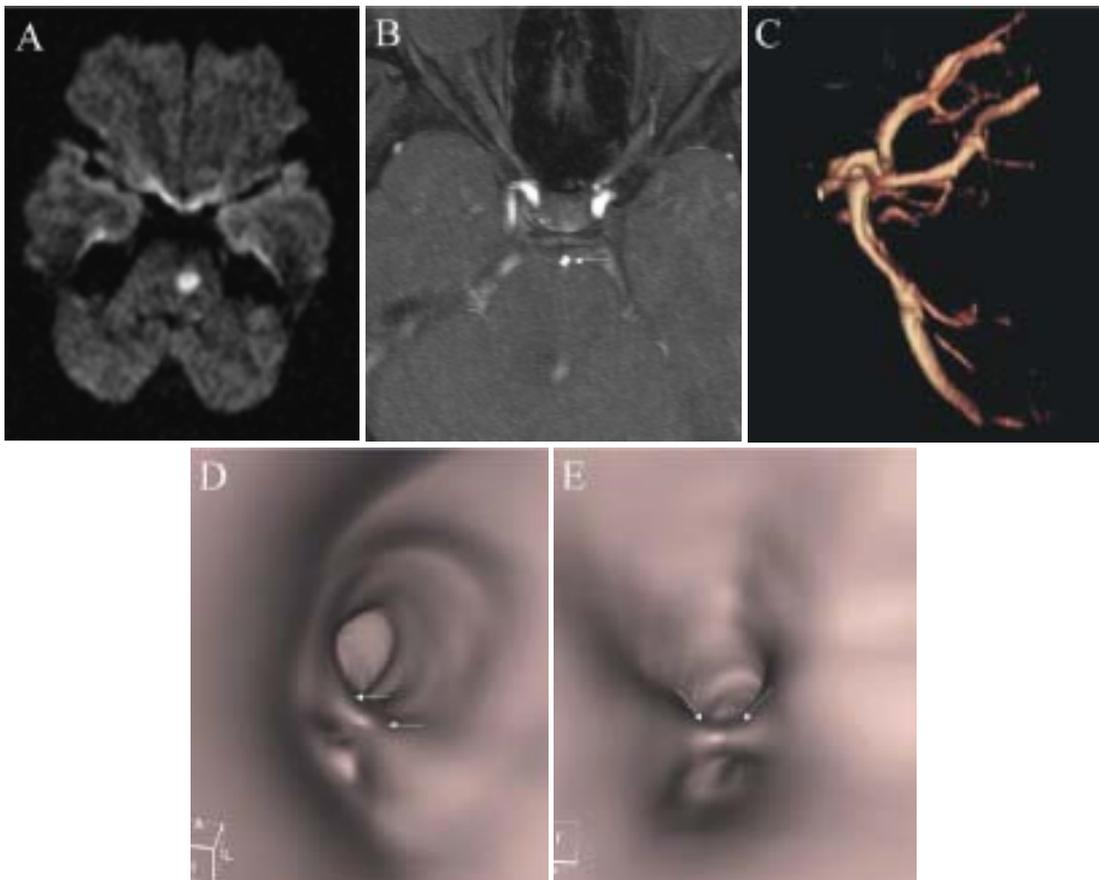


Figure 1. Imaging of a 55-year-old woman with acute pontine infarction. (A) Diffusion-weighted MR image with hyperintense signals in the left pons. (B) Source image of a contrast-enhanced MR angiogram shows the double lumens of the basilar artery (arrow). (C) Three-dimensional reconstruction image shows the double lumens and fenestration of the basilar artery. (D) Virtual endoscopy of the proximal portion of the basilar artery shows a duplicated lumen (arrows). (E) Virtual endoscopy of the distal portion of the basilar artery also shows a duplicated lumen due to complete fenestration (arrows).

DISCUSSION

The reported incidence of fenestration of the basilar artery has varied from 0.3% in angiographic examinations to 5.26% in autopsy series.^{6,7} Basilar artery fenestration is the most commonly observed form of fenestration of the cerebral arteries.⁷ The fenestration is usually short and occurs primarily in the proximal segment of the artery,⁸ with its clinical interest attributable to the possible association with aneurysms localized at the junctions of the fenestrated segments.⁹ There is known to be a defect in the media of the arterial wall that has been thought to be the predisposing factor for the prevalence of saccular aneurysms together with turbulent blood flow at the joining points.¹⁰

In the present case, the lesion was a true duplication-type fenestration, with each lumen sharing the medial wall. This finding was consistently observed from the joining portion of the two vertebral arteries to the top of the basilar artery, and each of the posterior cerebral arteries was branched from the adjoined portion of the basilar artery. The pontine infarction was located in the portion of the proximal openings.

There has been speculation about associations between either vertebrobasilar artery fenestrations and brainstem ischemia,¹⁰ or infarctions,^{5,11,12} although their relationships are controversial. Among these reports, some authors showed basilar artery fenestration and associated brainstem infarctions.^{5,11} One study showed an arterial slit-type fenestration that was confirmed by virtual arterial endoscopy,¹¹ and another study showed a true duplication-type that was confirmed by autopsy.⁵ The latter case was a true duplication-type, nonseparated form of fenestration that shared the medial wall that was confined to the caudal half portion of the basilar artery. The autopsy findings showed a partially organized thrombus that occluded both halves of the duplicated portion, suggesting possible hemodynamic disturbances and turbulent blood flow at the site of fenestration.

Another case of extreme fenestration of the basilar artery was reported in a retrospective MR angiographic study.⁴ In that study, 10 basilar artery fenestrations were observed from 600 brain reviews (1.7%) and one case

showed a total duplication. The shape of the fenestration was different in the proximal and distal parts of the basilar artery; in the proximal two-thirds of the artery, a true duplication-type fenestration that shared the medial wall was noted, but in the distal one-third of the artery, the artery was fully separated, with each basilar artery terminating in a posterior cerebral artery. However, it was not mentioned whether this finding was combined with a brainstem infarction.

Because of the 'double-lumen sign' in the present case, a basilar artery fenestration might be misinterpreted because it is known as a pathognomonic sign of arterial dissection.¹³ Even though neuroimaging has improved in both resolution and 3-D capabilities, it remains difficult to diagnose fenestrations of intracranial arteries. This is consistent with the large discrepancy in the reported incidence of fenestration of the basilar artery between angiographic examination and autopsies.^{6,7}

Virtual endoscopy is a new diagnostic method that uses computer processing of 3-D image datasets, such as CT or MR imaging scans, to provide simulated visualization of specific organs.¹⁴ The ability to obtain an internal view of a vessel facilitates qualitative analyses and could improve the pretherapeutic visualization of vascular variants, stenosis, and obstruction of cerebral vessels.¹⁵ Virtual endoscopy in patients with fenestration, as in the present case, has both improved the anatomical identification of vessels and prevented misdiagnoses of fenestration as either dissection or thrombus.

In summary, we have reported a case of a patient with a duplication-type, nonseparated fenestration of the basilar artery and associated brainstem infarction. Even though a causative linkage between arterial fenestration and cerebral ischemia was not established, a reliable diagnosis of the vascular condition in the affected lesion was important clinically. From this point of view, virtual arterial endoscopy may enhance diagnostic capabilities, especially in certain cases.

REFERENCES

1. Lasjaunias P, Braun JP, Hasso AN, Moret J, Manelfe C. True and false fenestration of the vertebral artery. *J Neuroradiol* 1980;7:157-166.

2. Padgett DH. The development of the cranial arteries in the human embryo. *Contrib Embryol* 1948;32:205-261.
3. Goldstein JH, Woodcock R, Do HM, Phillips CD, Dion JE. Complete duplication or extreme fenestration of the basilar artery. *AJNR Am J Neuroradiol* 1999;20:149-150.
4. Uchino A, Kato A, Takase Y, Kudo S. Basilar artery fenestrations detected by MR angiography. *Radiat Med* 2001;19:71-74.
5. Berry AD 3rd, Kepes JJ, Wetzel MD. Segmental duplication of the basilar artery with thrombosis. *Stroke* 1988;19:256-260.
6. Sanders WP, Sorek PA, Mehta BA. Fenestration of intracranial arteries with special attention to associated aneurysms and other anomalies. *AJNR Am J Neuroradiol* 1993;14:675-680.
7. Wollschlaeger G, Wollschlaeger PB, Lucas FV, Lopez VF. Experience and result with postmortem cerebral angiography performed as routine procedure of the autopsy. *Am J Roentgenol Radium Ther Nucl Med* 1967; 101:68-87.
8. Takahashi M, Tamakawa Y, Kishikawa T, Kowada M. Fenestration of the basilar artery. Report of three cases and review of the literature. *Radiology* 1973;109:79-82.
9. Yoon SM, Chun YI, Kwon Y, Kwun BD. Vertebrobasilar junction aneurysms associated with fenestration: experience of five cases treated with Guglielmi detachable coils. *Surg Neurol* 2004;61:248-254.
10. Small SL, Fukui MB, Reinmuth OM. What is a 'normal anatomic variant'? Transient left lateral medullary ischemia in a patient with a fenestrated left vertebral artery. *Neurology* 1994;44:1358-1359.
11. Scherer A, Siebler M, Aulich A. Virtual arterial endoscopy as a diagnostic aid in a patient with basilar artery fenestration and thromboembolic pontine infarct. *AJNR Am J Neuroradiol* 2002;23:1237-1239.
12. Schievink WI, Hunter SF, Marsh WR, Aksamit AJ, Jack CR. Traumatic occlusion of one limb of an intracranial arterial fenestration: an uncommon cause of stroke. *Neurology* 1996;47:277-278.
13. Drapkin AJ. The double lumen: a pathognomonic angiographic sign of arterial dissection? *Neuroradiology* 2000; 42:203-205.
14. Rubin GD, Beaulieu CF, Argiro V, Ringl H, Norbash AM, Feller JF, et al. Perspective volume rendering of CT and MR images: applications for endoscopic imaging. *Radiology* 1996;199:321-330.
15. Kim DY, Park JW. Visualization of the internal carotid artery using MRA images. *Magn Reson Imaging* 2005;23: 27-33.