

Case Report

Three-Dimensional Angiographic Demonstration of Plexiform Fenestrations of the Proximal Anterior Cerebral Artery Associated with a Ruptured Aneurysm

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A rare case of ruptured aneurysm associated with multiple A₁ fenestrations resembling plexiform network was demonstrated by 3D angiography. A 56-year-old female presented with a ruptured aneurysm in the A₂ segment of the left distal anterior cerebral artery associated with the right A₁ fenestration. The ruptured aneurysm was occluded with surgical neck clipping via interhemispheric approach without neurological deficit. Plexiform fenestrations of the right distal A₁, opposite side to the left ruptured A₂ aneurysm, were clearly visible on postoperative 3D angiography. Our case may strongly support the theory described by Paget, namely that a remnant of the plexiform anastomosis between the primitive olfactory artery and A₁ segment is the source of such fenestration.

KEY WORDS : Anterior cerebral artery fenestration · Aneurysm · Three-dimensional angiography.

INTRODUCTION

Fenestration of the anterior cerebral artery (ACA) is rare^{1-4,6,8} and is often associated with cerebral aneurysms^{3,6,8,10}. Although the pathogenesis of A₁ fenestrations has not been well described, a remnant of an embryonic plexiform anastomosis between the primitive olfactory artery and the ACA or abnormal regression are well accepted explanations for the formation of A₁ fenestrations^{8,11}. Most of the reports on A₁ fenestrations were of single fenestrations, with only a few reports of double fenestrations. We report a rare case of multiple A₁ fenestrations resembling a plexiform network accompanying a ruptured distal ACA aneurysm.

CASE REPORT

A 56-year-old female was admitted with a history of sudden headache and vomiting. Neurological examination showed no abnormal findings except for mild neck stiffness.

A brain computed tomogram was performed immediately and disclosed diffuse subarachnoid hemorrhage and focal hematoma on the interhemispheric fissure with the suspicion of the ACA aneurysm rupture (Fig. 1). Four-vessel angiography was performed and revealed a 3 × 2.5-mm saccular aneurysm on the left A₂ portion of the ACA and the right A₁ fenestration (Fig. 2). The ruptured pericallosal aneurysm was successfully clipped using an interhemispheric approach.

The patient recovered fully without neurological deficit. A follow-up angiogram performed before discharge showed that the aneurysm was completely occluded by surgical clipping, and a 3-dimensional reconstruction of the rotational angiogram clearly demonstrated multiple fenestrations in



Fig. 1. Brain computed tomography scan shows diffuse subarachnoid hemorrhage in the basal cisterns. It also shows focal hematoma in the left frontal lobe and in the interhemispheric fissure (arrow). A ruptured anterior cerebral artery aneurysm is strongly suspected.

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the distal right A₁ segment (Fig. 3). A branch of the right A₁ fenestration joined with the left A₁ and then fused with the left A₂, from which the ruptured distal ACA aneurysm arose (Fig. 3).

DISCUSSION

There have been 60 reported cases of fenestration of the anterior cerebral artery (ACA)^{3,4,6}, and the incidence of fenestration of the ACA is about 0.2-7.2% in autopsy specimens^{1,2}. The incidence of fenestration of the intracranial vessels is 0.3-0.9% on angiogram, and they are frequently located in the basilar and vertebral arteries^{7,10}. In comparison to the well-established hypothesis that basilar artery fenestrations arise from the incomplete fusion of embryologically paired longitudinal arteries⁹, the mechanism underlying A₁ segment fenestration formation is still not completely understood. Various theories concerning the pathogenesis of A₁ fenestration, including partial duplication, incomplete fusion, abnormal passage of a nonvascular structure through the precursor vasculature, and a remnant of an embryonic plexiform network between the primitive olfactory artery and the ACA, have been suggested^{8,9,11}. In his work concerning the development of the cranial artery in the human embryo, Padget⁹ clearly demonstrated that the plexiform anastomosis between the ACA and the primitive olfactory artery appears by the time the embryo reaches the 14-mm stage. Normally, there is a fenestration of the distal end of the ACA in 18- and 24-mm embryos, but no such fenestration is seen after the embryo reaches the 43-mm stage. The embryologically normal fusion process or normal regression would make such fenestrations disappear, but incomplete fusion or abnormal regression might result in a fenestration or plexiform remnant⁹. Most of the A₁ fenestrations reported thus far have been single^{3,4,6,10} fenestrations, with double^{8,12} fenestrations being reported only on rare occasions. In our case, the fenestrations were multiple and resembled a plexiform network in the distal end of the A₁, which may strongly support the theory described by Padget⁹, namely that a remnant of the plexiform anastomosis between the primitive olfactory artery and the A₁ segment is the source of such fenestrations.

Saccular aneurysms associated with fenestrations have been well-documented because they are often accompanied by cerebral hemodynamic changes^{3,5,6,8,10}. Well-known common findings, such as a contralateral A₁ hypoplasia or aplasia in aneurysms of the anterior communicating artery, are not a constant feature³ and associated cerebral aneurysms frequently occur at the proximal end of the A₁ fenestration^{3,6,8,10}. It is not clear whether the associated ruptured

left distal ACA aneurysm in our case was related to hemodynamic stress or merely coincidental because the aneurysm was located on the contralateral side of the fenestrations in the right A₁. The blood flow from a branch of the fenestrated right A₁ seeming nearly duplicated, and a certain amount of blood from the left A₁, which was not hypoplastic in comparison to the right A₁, may have affected the formation of the hypertrophic left A₂ segment. Therefore, this hemodynamic stress through the left A₂ may have played a role in the formation of a distal saccular aneurysm contralateral to the right A₁ fenestration.

CONCLUSION

To our knowledge, multiple fenestrations of the A₁ that resembled a plexiform network associated with a saccular A₂ aneurysm are quite rare and this is the first to appear in the literature. When a saccular aneurysm around the anterior communicating artery is found, one should carefully review

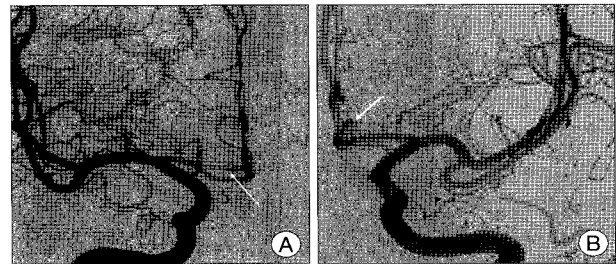


Fig. 2. Initial internal carotid artery angiogram of a 56-year-old female patient presenting with subarachnoid hemorrhage. Right internal carotid artery angiogram (A) shows a fenestration (arrow) in the right distal portion of the A₁ segment, and left internal carotid artery angiogram (B) reveals a saccular aneurysm (arrow) arising from the left distal anterior cerebral artery.

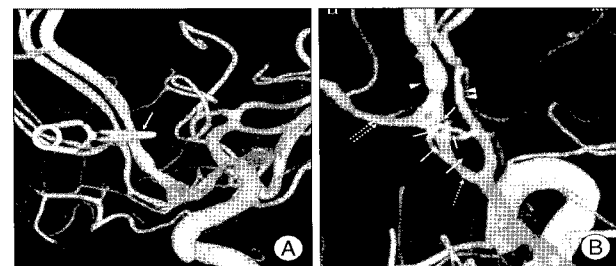


Fig. 3. The 3-dimensional reconstruction of right internal carotid artery angiogram after the surgical neck clipping. The ruptured left distal anterior cerebral artery aneurysm is completely obliterated by aneurysm clip (arrow) (A) and multiple fenestrations (arrows) resembling plexiform network of the right distal A₁ segment are clearly demonstrated (B). One of the branches of the right A₁ fenestrations (dotted arrow) joins with the left A₁ (double dotted arrows) and then fuses with the left A₂ (arrow head), seeming hypertrophic than the right A₂ (double arrow heads), from which the ruptured distal anterior cerebral artery aneurysm arises (B) (Aneurysm clip on the left A₂ is not shown here).

the cerebral angiogram using various angulation and rotation views to verify the exact location of aneurysm and associated anomalies. Three-dimensional rotational angiography is very useful in precise delineating this relationship. Hemodynamic stress could be a possible cause of aneurysm formation and subsequent rupture in relation to intracranial fenestrations.

References

1. Baptista AG : studies on the arteries of the brain. II. The anterior cerebral artery: some anatomic features and their clinical implications. *Neurology* 13 : 825-835, 1963
2. Fawcett E, Blachford JV : The circle of Willis : an examination of 700 specimens. *J Anat Physiol* 40 : 63-70, 1905
3. Friedlander RM, Ogilvy CS : Aneurysmal subarachnoid hemorrhage in a patient with bilateral A1 fenestrations associated with an azygos anterior cerebral artery. Case report and literature review. *J Neurosurg* 84 : 681-684, 1996
4. Fujimoto Y, Yamanaka K, Nakajima Y, Yoshimura K, Yoshimine T : Ruptured aneurysm arising from the proximal end of an azygos anterior cerebral artery. *Neurol Med Chir (Tokyo)* 44 : 242-244, 2004
5. Futami K, Yamashita J, Higashi S : Do cerebral aneurysms originate at the site of medial defects? Microscopic examinations of experimental aneurysms at the fenestration of the anterior cerebral artery in rats. *Surg Neurol* 50 : 141-146, 1998
6. Kachhara R, Nair S, Gupta AK : Fenestration of the proximal anterior cerebral artery (A1) with aneurysm manifesting as subarachnoid hemorrhage--case report. *Neurol Med Chir (Tokyo)* 38 : 409-412, 1998
7. Krayenbuhl HA, Yasargil MG : *Cerebral Angiography*, ed 2. New York : Thieme Medical Publishers, 1968, pp38-40
8. Minakawa T, Kawamata M, Hayano M, Kawakami K : Aneurysms associated with fenestrated anterior cerebral arteries. Report of four cases and review of literature. *Surg Neurol* 24 : 284-288, 1985
9. Padget DH : The development of the cranial arteries in the human embryo. *Contrib Embryol* 32 : 205-262, 1948
10. San-Galli F, Leman C, Kien P, Khazaal J, Phillips SD, Guérin J : Cerebral arterial fenestration associated with intracranial saccular aneurysms. *Neurosurgery* 30 : 279-283, 1992
11. Teal JS, Rumbaugh CL, Bergeron RT, Segall HD : Angiographic demonstration of fenestrations of the intradural intracranial arteries. *Radiology* 106 : 123-126, 1973
12. Von Mitterwallner F : Statistical studies on variations of the basal cerebral vessels. *Acta Anat (Basel)* 24 : 51-87, 1955