Ruptured Aneurysm Arising from the Distal End of a Proximal A_1 Fenestration: Case Report and Review of the Literature

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A 75-year-old female patient was admitted with a history of sudden onset of headache and vomiting. Neurological examination showed no abnormal finding except mild neck stiffness. Brain computed tomography (CT) scan was performed immediately and disclosed a subarachnoid hemorrhage (Fig. 1), presumably due to a ruptured aneurysm. A three-dimensional CT angiogram revealed a fenestration at the proximal portion of the right A1 and a saccular aneurysm arising from the distal part of the A1 fenestration.

INTRODUCTION

Fenestration of the proximal anterior cerebral artery (ACA) is rare and it is usually limited to the distal half of the A1 segment. The A1 fenestration associated with cerebral aneurysms were often reported and all aneurysms related to A1 fenestration in the literature harbored at the proximal end portion of fenestration. We report a unique case of a ruptured aneurysm arising from the distal end of a fenestration in the proximal A1 segment.

CASE REPORT

A 75-year-old female patient was admitted with a history of sudden onset of headache and vomiting. Neurological examination showed no abnormal finding except mild neck stiffness. Brain computed tomography (CT) scan was...
(Fig. 2A). Cerebral angiography also showed the right A1 aneurysm associated with a fenestration. Accompanying vascular variants, including duplication of middle cerebral artery (MCA) on the right side and a right fetal-type posterior cerebral artery (PCA) in association with an unruptured internal cerebral artery aneurysm (ICA), were also noted (Fig. 2B). The ruptured right A1 aneurysm and unruptured aneurysm of the ICA were managed conservatively because the patient’s relatives refused surgical intervention owing to her long history of medical problems. The patient was discharged after a short hospital stay, and she is being well during the 37 months after the ictus.

**DISCUSSION**

A fenestration of the cerebral artery is a separation of the arterial lumen which results in the formation of distinct channels, each with its own endothelial and muscular layer\(^7\). Fenestration of the ACA other than the anterior communicating artery, with or without accompanying aneurysms, has rarely been reported in the literature\(^3,5,8-13,15,18-20\).

With very few exceptions\(^17\), the preferred sites of A1 fenestrations are usually at the distal part of the A1 segment\(^3,5,8-13,15,18-20\). Among various theories concerning the pathogenesis of distal A1 fenestrations, a remnant of embryologic plexiform anastomosis between the primitive olfactory artery and the ACA is thus far well accepted\(^14\). However, according to Teal et al.\(^16\), other fenestrations may occur as a result of partial duplication, incomplete fusion, and abnormal passage of a nonvascular structure through the precursor vasculature. The fenestration in our case was unusual because of its location at the proximal part of the A1 segment.

Owing to the cerebral hemodynamic changes in the fenestrated vessels, accompanying saccular aneurysms associated with fenestrations have been well documented and frequently occur at the proximal end of A1 fenestration\(^3,5,8-13,15,18-20\). Medial wall defects, which are more prominent at the medial and ventral walls of the proximal juncture, are known to be more prone to the development of cerebral aneurysms in the branches of intracranial artery and fenestrations\(^2,3\). Meanwhile, morphologic studies have also revealed that both proximal and distal edges of a fenestration lack the medial layer and can stimulate the formation of cerebral aneurysms at both edges in response to hemodynamic forces\(^2,4\). In our case, the aneurysm was found in an unusual site at the distal end of an A1 fenestration. We
were unable to find any report of an aneurysm arising at the distal end of A1 fenestration in the literature (Table 1). Only two cases of vertebrobasilar juncture aneurysm originating from the distal end of a fenestration have been reported\(^\text{10}\). Congenital medial wall defect of the cerebral vessel and certain hemodynamic factors in relation to the very proximal A1 fenestration are considered to be involved in the development of fenestration-related cerebral aneurysm in our patient. Hemodynamic burden around the ICA bifurcation in the present case is thought to be low owing to the multiple division/branchings in the terminal portion of the ICA; the MCA, a duplicated MCA, and two channels of A1 fenestration. As a result, the distal end of A1 fenestration has more chance to be stressed than the proximal end to constitute the proper distal A1 by flows from both channels of fenestration.

Other developmental variants including a fetal-type PCA and a duplicated MCA on the right side were also demonstrated in our patient. It remains unclear whether these vascular variations affected the hemodynamic changes in the ACA fenestration and subsequent bleeding from the aneurysm. However, the hyperplastic right ACA in our case suggests that certain hemodynamic factors can augment blood flows to the right ACA via the ipsilateral ICA.

**CONCLUSION**

To our knowledge, this is the first report of a saccular aneurysm arising from the distal end of the proximal A1 fenestration manifesting as subarachnoid hemorrhage. Congenital defect of the vessel wall and hemodynamic factors are also considered to be involved in the development of fenestration-related cerebral aneurysms.

**References**

1. Andrews BT, Brant-Zawadzki M, Wilson CB : Variant aneurysms of the fenestrated basilar artery. *Neurosurgery* 18 : 204-207, 1986
2. Black SP, Ansbacher LE : Saccular aneurysm associated with segmental duplication of the basilar artery. A morphological study. *J Neurosurg* 61 : 1005-1008, 1984
3. Crompton MR : The pathology of ruptured middle-cerebral aneurysms. With special reference to the differences between the sexes. *Lancet* 2 : 421-425, 1962
4. Finlay HM, Canham PB : The layered fabric of cerebral artery fenestrations. *Stroke* 25 : 1799-1806, 1994
5. Friedlander RM, Ogilvy CS : Aneurysmal subarachnoid hemorrhage in a patient with bilateral A1 fenestrations associated with an asygos anterior cerebral artery. *J Neurosurg* 84 : 681-684, 1996
6. Fujimura M, Sugawara T, Higuchi H, Oku T, Seki H : A ruptured aneurysm at the distal end of the basilar artery fenestration associated with multiple fenestrations of the vertebrobasilar system : case report. *Surg Neurol* 47 : 469-472, 1997
7. Hacein-Bey L, Muszynski CA, Varelas PN : Saccular aneurysm associated with posterior cerebral artery fenestration manifesting as a subarachnoid hemorrhage in a child. *AJNR Am J Neuroradiol* 23 : 1291-1294, 2002
8. Handa J, Nakasu Y, Matsuda M, Kyoshima K : Aneurysm of the proximal anterior cerebral artery. *Surg Neurol* 22 : 486-490, 1984
9. Inagawa T, Yamada T, Takada T, Taguchi H : Aneurysm of the fenestrated anterior cerebral artery. Report of two cases *Neurol Med Chir (Tokyo)* 23 : 211-215, 1983
10. Kachhara R, Nair S, Gupta AK : Aneurysms associated with fenestration of the proximal anterior cerebral artery (A1) with aneurysm manifesting as subarachnoid hemorrhage. *Neurol Med Chir (Tokyo)* 38 : 409-412, 1998
11. Korosue K, Kuwamura K, Okuda Y, Tamaki N, Matsumoto S : Saccular aneurysm arising from a fenestrated anterior cerebral artery. *Surg Neurol* 19 : 273-275, 1983
12. 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
13. Ogasawara H, Inagawa T, Yamamoto M, Kamiya K : Aneurysm in the anterior cerebral artery with persistent trigeminal artery: case report. *Surg Neurol 19* : 129-131, 1983
14. Padget DH : The development of the cranial arteries in the human embryo. *Contrib Embryol* 32 : 205-261, 1948
15. San-Galli F, Leman C, Kien P, Khazaal J, Phillips SD, Guerin J : Cerebral arterial fenestrations associated with intracranial saccular aneurysms. *Neurosurgery* 30 : 279-283, 1992
16. Teal JS, Rumbaugh CL, Bergeron RT, Segall HD : Angiographic demonstration of fenestrations of the intradural intracranial arteries. *Radiology* 106 : 125-126, 1973
17. Tran-Dinh HD, Dorsch NW, Soo YS : Ectasia and fenestration of the horizontal segment of the anterior cerebral artery. *J Neurosurg 24* : 275-283, 1973
18. Waga S, Morikawa A : Aneurysm developing on the infundibular widening of the posterior communicating artery. *Surg Neurol 11* : 125-127, 1979
19. Wakahayashi T, Tamaki N, Yamashita H, Saya H, Suyama T, Matsumoto S : Angiographic classification of aneurysms of the horizontal segment of the anterior cerebral artery. *Surg Neurol 24* : 31-34, 1985
20. Yamada T, Inagawa T, Taketa T : Ruptured aneurysm at the anterior cerebral artery fenestration. Case report. *J Neurosurg* 57 : 826-828, 1982