Ruptured Cerebral Aneurysm of Fenestrated A1 Segment of the Anterior Cerebral Artery: Case Report and Literature Review

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Objective: Arterial fenestration is an unusual anatomic variation and is often associated with aneurysms. Aneurysm arising from the fenestration of the horizontal segment of the anterior cerebral artery (A1) is considered to be rare.

Case Presentation: We report a case of a 61-year-old man who presented with a subarachnoid hemorrhage secondary to a ruptured aneurysm of fenestrated A1 segment of the anterior cerebral artery. Cerebral angiogram revealed an aneurysm at right A1 fenestration. The aneurysm was embolized by coils, leading to complete occlusion, while preserving both channels of the fenestration. On the 11th postoperative day, the patient suffered cerebral embolic stroke due to paroxysmal atrial fibrillation and his left proximal middle cerebral artery was occluded. He is doing rehabilitation for right hemiparesis and aphasia but remains severely disabled.

Conclusion: Considering that dissection of fenestrated A1 aneurysms is sometimes difficult, and there are several branches from one or both channels of fenestration, endovascular therapy would have some advantages over the neck clipping.

Keywords ► fenestration, anterior cerebral artery, cerebral aneurysm

Introduction

A fenestration is a division of the arterial lumen, resulting in separate channel, each with its own endothelial layer and muscularis tunica; the adventitial layer may or may not be shared between channels.¹⁾ Cerebral aneurysms in the horizontal segment of the anterior cerebral artery (A1) are relatively rare (0.88%).²⁾ Fenestrations accompanying cerebral aneurysms are well-known vascular malformations. However, there have been few reports on fenestrations in the A1 segment, and the etiology of A1 fenestrations is less clear than that of basilar and vertebral artery fenestrations.¹⁾

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Most of these aneurysms have been treated with clipping. Together with a discussion of the literature, we report a case of a ruptured cerebral aneurysm in the fenestrated segment of A1 treated with coil embolization.

Case Presentation

A 61-year-old male, a hypertensive smoker, presented with a sudden onset of disturbance of consciousness during sleep and was referred to our hospital. His medical history included smoking and spinal cord infarction treated with cilostazol. He had no history of atrial fibrillation. On admission, neurological examination was unremarkable except for meningeal signs and laboratory data demonstrated no abnormal findings. CT scan performed on the day of admission (day 0) revealed Fisher group 3 interhemispheric fissure-dominant diffuse subarachnoid hemorrhage (SAH) and a diagnosis of SAH of World Federation of Neurosurgical Societies grade I and Hunt and Kosnick grade II was made (**Fig. 1**). One day after admission (day 1), cerebral angiogram revealed a fenestration in the distal right A1 segment, and a cerebral aneurysm 3.5 mm × 4.0 mm in size arising from the proximal end of the

Fig. 1 Computed tomography on admission showing subarachnoid hemorrhage in the basal cistern and inter-hemispheric fissure.

fenestration (**Figs. 2A and 2B**). Recurrent artery of Heubner was found at A1-2 junction. No cerebral aneurysms were found at any other site. The left A1 segment was not hypoplastic. On day 2, endovascular embolization of the aneurysm was performed with the patient under general anesthesia. Using Orbit Galaxy Complex Extra Soft Detachable Coils 3.5 mm × 7.5 cm (Codman & Shurtleff, Raynham, MA, USA), ED coil-10 Extra Soft type R 2.5 mm × 3 cm (Kaneka Medix, Osaka, Japan), and Target Nano 360 Detachable Coils 1.5 mm × 2 cm (Stryker Neurovascular, Fremont, CA, USA), total occlusion of the aneurysm was achieved (**Fig. 3**). The volume embolization rate was 32%. Visualization of the duplicated A1 segments forming the fenestration remained good.

Postoperative course

The patient awoke from anesthesia and did not exhibit any symptom of neurological deficit postoperatively. He progressed well but suddenly exhibited atrial fibrillation on his electrocardiogram (ECG) on postoperative day 11 (day 13). Right hemiparesis and aphasia subsequently appeared. Emergency cerebral angiography was performed, revealing occlusion of the proximal left middle cerebral artery.

Cerebral embolism due to atrial fibrillation was diagnosed, and mechanical thrombectomy was successfully performed to recanalize left middle cerebral artery. Left internal carotid angiogram indicated that the cerebral aneurysm had been embolized, and both fenestrated A1 and A2 channels were well visualized. A follow-up CT scan revealed extensive infarction in the territory of left middle cerebral artery. The patient's right hemiparesis and aphasia persisted, and he is currently undergoing rehabilitation.

Discussion

The most common sites for fenestrations of intracranial arteries to develop are, in decreasing order, the vertebral artery, basilar artery, and the middle cerebral artery.¹⁾ San-Galli et al.³⁾ have reported that fenestrations are most common in the anterior communicating artery. There have been few reports on fenestrations occurring in the anterior cerebral artery. Fawcett et al.⁴⁾ reported a frequency of 0.14% in autopsy cases. Fenestrations are often accompanied by other cerebrovascular abnormalities. Suzuki et al.²⁾ reviewed 38 cases of A1 aneurysms and found an incidence of 15.8% (6/38) for A1 fenestration. Friedlander et al.⁵⁾ detected cerebral aneurysms in 16 (27.6%) of 58 cases of A1 fenestrations. Meanwhile, Sanders et al.¹⁾ reported that according to cerebral angiogram, aneurysms that develop in fenestrations exhibit the same association as those that develop in normal bifurcation sites. In most cases, aneurysms develop proximal edge of the fenestration, and a histological examination of the fenestration tissue has shown that the loss of tunica media at the proximal edge of the fenestration makes cerebral aneurysms prone to develop at that site.⁶⁻⁸⁾ Similarly, other histological examinations of fenestrations have indicated that there is tunica media loss not only at the proximal edge of the fenestration but also at the distal edge.^{8,9)} In addition, flow phenomena at the proximal end of fenestrations, where hemodynamic stress and increased turbulence are present, may contribute to aneurysm formation.¹⁰⁾

The pathophysiologic mechanism for the development of A1 fenestration is not clearly explained. According to Padgett's illustration of a 14-mm embryo,¹¹⁾ there is a plexiform anastomosis between the anterior cerebral artery (ACA) and the primitive olfactory artery. In 18- and 24-mm embryos, there is a fenestration of the distal end of the ACA which disappears by the 43-mm stage. During the 18- to 43-mm embryos, fenestration of the distal A1 segment might occur due to the lack of fusion of the plexiform anastomosis present in the distal primitive ACA.⁵⁾ Teal et al.¹²⁾ suggested

Fig. 2 (a) Preoperative right carotid angiogram showing fenestrated A1 segment of the anterior cerebral artery associated with cerebral aneurysm in an oblique plane. (b) The 3D DSA clearly illustrating a fenestration of A1 segment with the aneurysm located in the proximal end of it.

Fig. 3 Postoperative right carotid angiogram showing complete occlusion of aneurysm and preservation of both channels of the fenestration.

that fenestrations in internal carotid system may result from true partial duplication, incomplete fusion of the network of vessels which serves as the precursor of a solitary named vessels, or incorporation of a nerve within the definitive artery.

Including our patient, there have been 19 such cases reported in the past (**Table 1**). The mean age of the patients

was 54.4 years, and the male-to-female ratio was 1:1. The right-to-left ratio was 12:7, and there were 16 cases of ruptured aneurysms. The mean age of the patients with ruptured aneurysms was 56 years, suggesting a tendency for onset in slightly younger patients than those of SAH in general. Kwon et al.¹³⁾ developed classifications based on the site of aneurysms. Type I refers to aneurysms on the proximal end of the fenestration; Type II refers to aneurysms arising from the duplicated A1 trunk, and Type III refers to multiple aneurysms within the fenestrated A1. In addition to these classifications, we suggested Type IV to refer to aneurysms at the distal end of the fenestration, with Koh et al.¹⁴⁾ reporting a case of the aneurysm at this site. Type I was the most common type, accounting for 14 of 19 cases. Type II only accounted for three cases, and an investigation of images available in the literature revealed that this type often involves cases with narrow space between channels of fenestration. Type III only accounted for one case with multiple aneurysms. Our patient was classified as Type I. Regarding treatment, many patients underwent clipping, which has been reported to offer stable treatment outcomes. However, it is reportedly difficult to perform whether there is strong adhesion between the aneurysm dome and fenestration vessel or when there is a perforating branch from the fenestration¹⁵⁾ as in the case of aneurysms of the proximal (M1) segment of the middle cerebral artery. An average 6.4 perforators arise from the A1 segment, and divide to yield average 21.9 vessels.¹⁶⁾ The possibility of

Table 1 Summary of reported cases of A1 fenestration aneurysms

Author	Age (years)	Gender	Laterality	Ruptured/Unruptured	Location	Treatment	Outcome
Yamada et al. ⁷⁾	1982	M	rt	Ruptured	Proximal	Clipping	Death
Korosue et al. ¹⁷⁾	1983	M	rt	Ruptured	Proximal	Clipping	Death
Handa et al. ¹⁸⁾	1984	F	rt	Ruptured	Proximal	Clipping	GR
Wakabayashi et al. ¹⁹⁾	1985	M	rt	Ruptured	Proximal	Clipping	Disabled
Minakawa et al. ²⁰⁾	1985	M	lt	Ruptured	Proximal	Clipping	?
Ogasawara et al. ²¹⁾	1988	F	lt	Ruptured	Proximal	Clipping	VS
San-Galli et al. ³⁾	1992	M	lt	Unruptured	Proximal	Clipping	GR
Friedlander et al. ⁵⁾	1996	M	rt	Ruptured	Proximal	Clipping	GR
Kachhara et al. ²²⁾	1998	F	rt	Ruptured	Proximal	Clipping	GR
Taylor et al. ²³⁾	2000	M	lt	Ruptured	Trunk	Clipping	GR
Wanibuchi et al. ¹⁵⁾	2001	F	lt	Unruptured	Proximal	Clipping	GR
Ihara et al. ²⁴⁾	2003	F	rt	Ruptured	Trunk	Clipping	GR
Evans et al. ²⁵⁾	2006	F	lt	Ruptured	Trunk (fusiform)	Embolization	Disabled
Koh et al. ¹⁴⁾	2009	F	rt	Ruptured	Distal	Conservative	GR
Mantazis et al. ²⁶⁾	2011	M	rt	Ruptured	Proximal	Embolization	GR
	2011	?	lt	Unruptured	Proximal	Embolization	GR
	2011	F	rt	Ruptured	Proximal/	Embolization	GR
Mitsuhashi et al. ²⁷⁾	2011	F	rt	Ruptured	posterior		
Kwon et al. ¹³⁾	2013	F	rt	Ruptured	Proximal	Clipping	GR
Present case	2016	M	rt	Ruptured	Proximal	Embolization	Disabled

M: male; F: female

branches from channels of fenestration can be easily assumed. There have also been recent reports on coil embolization, which has resulted in favorable outcomes. Considering that in the case treated by neck clipping of the aneurysm, dissection of the fenestration aneurysms is sometimes challenging and the perforating branch is sometimes sacrificed, the benefits of coil embolization can be understood. It could be particularly useful for Type I cases in which the dome of aneurysm is adherent to surrounding fenestration channels. The embolization performed on our patient was not technically difficult, and the aneurysm was successfully embolized. The patient progressed well postoperatively. On postoperative day 11, paroxysmal atrial fibrillation appeared on his ECG, after which cerebral embolism occurred. However, as the embolism was of the opposing side middle cerebral artery, we do not believe that it was related to the treatment we implemented. The patient had no history of atrial fibrillation and did not undergo any anticoagulation therapy pre- or postoperatively.

Many cases of cerebral aneurysms in fenestration sites involve ruptured aneurysms, and were small size of aneurysms which are prone to rupture.¹⁵⁾ Aggressive treatment should be considered if the aneurysm is detected when unruptured. Furthermore, the fenestration site may not necessarily show up on performing cerebral angiography. If an aneurysm is noted in the A1 segment, a treatment policy should be considered, keeping the possibility of a fenestration in mind.

Conclusion

We treated a 61-year-old man who developed a ruptured aneurysm in the A1 fenestration. A1 fenestration is rare, and there have been few reports on cerebral aneurysms at this site. Although coil embolization was successful and the patient progressed well, he also suffered from cerebral embolism caused by paroxysmal atrial fibrillation leading to right hemiparesis and aphasia. As there have also been reports indicating that cerebral aneurysms that occur in fenestrations are likely to rupture, aggressive treatment should be considered if an unruptured aneurysm is detected.

Disclosure Statement

None of the authors or co-authors have any conflict of interest to declare.

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