Case Report

Bilateral A1 fenestrations: Report of two cases and literature review

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Abstract

Background: Bilateral fenestration of the A1 segment of anterior cerebral artery (ACA) is an uncommon anomaly. Our objective is to describe two cases with this anomaly and to review the literature.

Case Description: A 50-year-old woman presented with subarachnoid hemorrhage from a ruptured A1 aneurysm. Angiography revealed bilateral A1 segment fenestration as well as an aneurysm on the proximal end of fenestration on the right side. The second case is that of an 86-year-old woman who was found to have bilateral fenestration of A1 segment at autopsy.

Conclusion: Bilateral A1 fenestration is an uncommon anomaly that may be associated with an aneurysm. In surgical clipping of such cases, extreme caution should be exercised to inspect both arms of the fenestration since both may have multiple perforators as demonstrated in our autopsy specimen. This will be the first published pictorial demonstration of these perforators arising from the arms of fenestration.

Key Words: A1 segment, aneurysm, anterior cerebral artery, bilateral fenestration, central nervous system

INTRODUCTION

Fenestrations of the cerebral arteries most commonly occur in the anterior communicating artery, the vertebral artery, the basilar artery, and the middle cerebral artery.¹¹⁻¹³⁻¹⁵⁻¹⁶⁻¹⁸ Fenestrations of the A1 segment of the anterior cerebral artery (ACA) are uncommon and are found only in 0.14% of the specimens.¹⁴ Furthermore, bilateral A1 fenestrations have only been reported by three authors previously and there is only one case report of bilateral A1 fenestrations associated with an aneurysm involving the fenestrated segment.¹¹⁻¹⁵⁻¹⁸ We report two cases of bilateral A1 fenestrations, one of which is associated with a saccular aneurysm involving the proximal end of a fenestration.

CASE REPORTS

Case 1

A 50-year-old woman with family history of ruptured aneurysms presented with subarachnoid hemorrhage. Computed tomography (CT) angiography performed at our institution revealed a small aneurysm at the proximal end of fenestrated right A1 segment and bilateral fenestrations of the A1 segments [Figure 1a and b]. The left fenestrated A1 was found to be smaller in size than...
the right one. In addition to these findings, the origins of the posterior inferior cerebellar arteries were found to be extradural bilaterally. The patient underwent surgical clipping of the aneurysm via pterional craniotomy. Surgical exploration revealed the aneurysm just proximal to the right fenestrated A1 segment. The aneurysm was clipped successfully using a bayonet clip across the aneurysmal neck. Post-clipping angiography demonstrated complete obliteration of the aneurysm [Figure 1c and d]. Postoperative course was uneventful and the patient was discharged home neurologically intact. She was doing well for a 50-month follow-up period.

**Case 2**
An 86-year-old woman with a history of Alzheimer’s disease, depression, and hypertension passed away at a nursing home due to complications related to dementia. She did not have any known history of neurovascular diseases or complaints. Her autopsy revealed incidental bilateral fenestrations of A1 segments. The anomalous cerebral vessels (A1 segments, anterior communicating artery, and A2 segments) were isolated for further microscopic examination by cutting A1s proximally and A2s distally [Figure 2]. These vessels were then inspected, photographed, and the perforators were counted under the operating microscope (Leica, Wild M 695 surgical microscope, 5× to 40×). On each side, the fenestrated A1 segments consisted of a larger, lateral segment and a narrower medial segment. Overall, the medial segment on the right side was larger than the medial segment on the left. There were two major and four minor perforators arising from the right lateral segment, while there were two major and five minor perforators associated with the left lateral segments. For the medial segments, there were only five minor perforators on the left and four on the right.

**DISCUSSION**
Fenestration is the separation of the arterial lumen into two distinct channels, each having its own endothelial, muscular, and adventitial layers. Distally, these two channels merge into a single arterial lumen again. The fenestration of the A1 segment of the ACA is an uncommon entity which was first described by Fawcett and Blachford in 1905. They found its incidence to be 0.14% in 700 brains. Bilateral A1 fenestrations were first described by Adachi et al. in three cases in 1928. Friedlander and Ogilvy reported a case of bilateral A1 fenestration associated with an A1 aneurysm, along with 58 cases of unilateral A1 fenestrations in their literature review. In addition to the data presented in the previous literature review by Friedlander and Ogilvy, we also found that Yaşargil described various fenestrations of the A1 segment including a case with bilateral fenestration. In 1976, Perlmutter and Rhoton reported two cases of unilateral A1 fenestrations in 50 cadaver brains, and no bilateral fenestrations were found in their study. In our review so far, there are 76 reported cases of A1 fenestrations in the literature, 5 of which are bilateral including the cases presented in this study [Table 1].

Fenestrations of the cerebral vessels are associated with other vascular anomalies and saccular aneurysms. Of the...
Table 1: Seven cases of bilateral A1 fenestrations reported in the literature between 1928 and 2011

| Authors, year | Number of patients | Association with aneurysm |
|---------------|--------------------|--------------------------|
| Adachi, 1928  | 3                  | 0                        |
| Yasargil, 1984| 1                  | 0                        |
| Friedlander and Ogilvy, 1996 | 1 | 1 |
| Present study  | 2                  | 1                        |

76 reported cases, 25 were associated with an aneurysm at the fenestrated segment itself, with one interpreted as fusiform and the remaining being saccular.1-3,4,5,6,7,8,9,10,11,12,13,14,15,16,17,18,19 Saccular aneurysms involved the proximal segment of the fenestration except for one case.8 In our first case, the saccular aneurysm arose from the proximal part of the fenestration of right A1, and the left A1 was smaller than the right one.

The origin of the A1 perforators in a fenestrated A1 segment is important in both endovascular and open surgical approaches for the treatment of associated aneurysms. The A1 segment generally supplies the optic chiasm, anterior third ventricle and hypothalamus, medial third of the anterior commissure, caudate head, globus pallidus, anterior limb of internal capsule, and rostral thalamus.2,13 Perlmutter and Rhoton found that the majority of the A1 perforators originate from the lateral half of the A1 (68%).13 Thus, the origins of the major perforators should be identified if occlusion of the fenestrated arm is being considered. In the postmortem specimen, we identified the major perforator branches were originating from the larger arm of the fenestrated vessel, which is also situated laterally [Figure 2]. However, small perforating vessels originating from the small medial arm were also identified [Figure 2]. The distribution of perforators amongst the two limbs of the fenestration in the proximal ACA has not been previously demonstrated. The presence of perforators arising from both limbs suggests that perforator distribution infarction may occur if sacrifice of either limb is contemplated during treatment of fenestration-associated aneurysms.

CONCLUSION

We demonstrated two cases of bilateral fenestrations of the A1 segment of the ACA which is an uncommon variant. The fenestration of the A1 segment is known to be associated with aneurysms, and the fact that the perforators can arise from either limb is important in the planning of the management of such aneurysms.

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