Ruptured Aneurysm Arising from the Distal End of a Proximal A1 Fenestration: Case Report and Review of the Literature

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A 75-year-old female presented with subarachnoid hemorrhage. Angiography revealed a partial duplication (fenestration) in the proximal A1 segment and a ruptured aneurysm at the distal end of A1 fenestration. This congenital anomaly accompanying an aneurysm was associated with duplicated ipsilateral middle cerebral artery (MCA). Congenital defect of the arterial wall and hemodynamic factors at the fenestrated A1 are considered to play a significant role in the development of this aneurysm. The present case is peculiar because not only the ruptured A1 aneurysm was related with the anterior and middle cerebral artery duplication but also the location of A1 fenestration and the origin of A1 aneurysm in a fenestration are quite unusual.

KEY WORDS: Subarachnoid hemorrhage - Aneurysm - A1 fenestration - Hemodynamic factors

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 performed immediately and disclosed a subarachnoid hemorrhage, especially in the interhemispheric fissure.
(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. Fenestration of the ACA other than the anterior communicating artery, with or without accompanying aneurysms, has rarely been reported in the literature. With very few exceptions, the preferred sites of A1 fenestrations are usually at the distal part of the A1 segment. 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. However, according to Teal et al., 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. 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. 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. In our case, the aneurysm was found in an unusual site at the distal end of an A1 fenestration. We

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**Table 1. Characteristics of aneurysms arising from A1 fenestrations in the literature by year**

<table>
<thead>
<tr>
<th>Authors, year</th>
<th>Age/ Sex</th>
<th>Side &amp; site of An origin in A1 fenestration</th>
<th>Associated findings of cerebral angiogram</th>
</tr>
</thead>
<tbody>
<tr>
<td>Crompton, 1962</td>
<td>59/F</td>
<td>Lt, Distal 1/3</td>
<td>MCA &amp; VA An &amp; fenestration</td>
</tr>
<tr>
<td>Waga &amp; Morikawa, 1979</td>
<td>36/F</td>
<td>Lt, Distal half</td>
<td>ICA An</td>
</tr>
<tr>
<td>Yamada et al., 1982</td>
<td>43/M</td>
<td>Rt, Distal half</td>
<td>-</td>
</tr>
<tr>
<td>Inagawa et al., 1983</td>
<td>43/M</td>
<td>Lt, Distal 1/3</td>
<td>Dolichoectasia</td>
</tr>
<tr>
<td>Kurosue et al., 1983</td>
<td>41/M</td>
<td>Rt, Distal half</td>
<td>MCA An</td>
</tr>
<tr>
<td>Honda et al., 1984</td>
<td>50/F</td>
<td>Rt, Distal half</td>
<td>-</td>
</tr>
<tr>
<td>Minakawa et al., 1985</td>
<td>56/M</td>
<td>Lt, Distal 1/3</td>
<td>Partial duplicated VA</td>
</tr>
<tr>
<td>Wakiyoshi et al., 1985</td>
<td>41/M</td>
<td>Rt, Distal half</td>
<td>-</td>
</tr>
<tr>
<td>Ogasawara et al., 1988</td>
<td>50/F</td>
<td>Lt, Distal half</td>
<td>-</td>
</tr>
<tr>
<td>San-Gall et al., 1992</td>
<td>42/M</td>
<td>Lt, Distal half</td>
<td>Aortic origin of VA, Dolicho-basilar artery</td>
</tr>
<tr>
<td>Friedlander &amp; Ogilvy, 1996</td>
<td>33/M</td>
<td>Rt, Distal half</td>
<td>Both A1 fenestration, Azygos A1</td>
</tr>
<tr>
<td>Kachhara et al., 1998</td>
<td>75/F</td>
<td>Rt, Proximal half</td>
<td>Duplicated MCA</td>
</tr>
<tr>
<td>Koh et al., 2008</td>
<td>75/F</td>
<td>Lt, Proximal half</td>
<td>-</td>
</tr>
<tr>
<td>(current report)</td>
<td></td>
<td></td>
<td>Fetal-type PCA, ICA An</td>
</tr>
</tbody>
</table>

An: aneurysm, MCA: middle cerebral artery, VA: vertebral artery, ICA: internal cerebral artery, PCA: posterior cerebral artery, - : none reported
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. 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