

## Case Report

# Ruptured Aneurysm Arising from the Distal End of a Proximal A<sub>1</sub> Fenestration : Case Report and Review of the Literature

Jun Seok Koh, M.D., Ph.D.,<sup>1</sup> Eui Jong Kim, M.D., Ph.D.,<sup>2</sup> Seung Hwan Lee, M.D.,<sup>1</sup> Jae Seung Bang, M.D.<sup>1</sup>

Department of Neurosurgery,<sup>1</sup> Stroke and Neurological Disorders Centre, East-West Neo Medical Hospital, Kyung Hee University School of Medicine, Seoul, Korea

Department of Radiology,<sup>2</sup> Medical Center of Kyung Hee University, Kyung Hee University School of Medicine, Seoul, Korea

A 75-year-old female presented with subarachnoid hemorrhage. Angiography revealed a partial duplication (fenestration) in the proximal A<sub>1</sub> segment and a ruptured aneurysm at the distal end of A<sub>1</sub> 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 A<sub>1</sub> are considered to play a significant role in the development of this aneurysm. The present case is peculiar because not only the ruptured A<sub>1</sub> aneurysm was related with the anterior and middle cerebral artery duplication but also the location of A<sub>1</sub> fenestration and the origin of A<sub>1</sub> aneurysm in a fenestration are quite unusual.

**KEY WORDS :** Subarachnoid hemorrhage · Aneurysm · A<sub>1</sub> fenestration · Hemodynamic factors

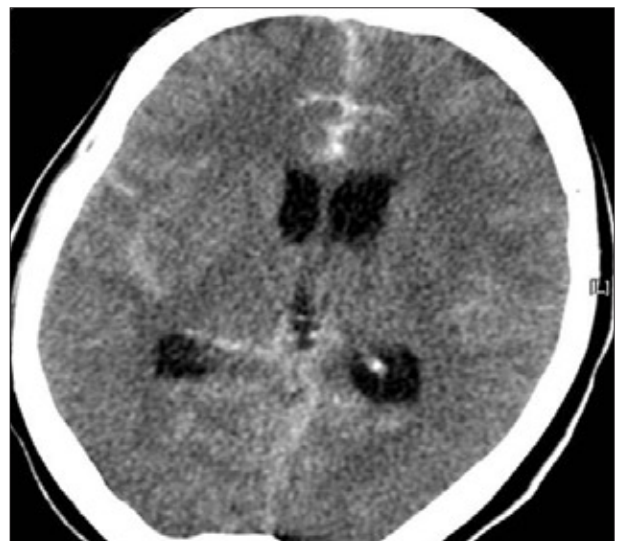
## INTRODUCTION

Fenestration of the proximal anterior cerebral artery (ACA) is rare and it is usually limited to the distal half of the A<sub>1</sub> segment<sup>3,5,8-13,15,18-20</sup>. The A<sub>1</sub> fenestration associated with cerebral aneurysms were often reported and all aneurysms related to A<sub>1</sub> fenestration in the literature harbored at the proximal end portion of fenestration<sup>3,5,8-13,15,18-20</sup>. We report a unique case of a ruptured aneurysm arising from the distal end of a fenestration in the proximal A<sub>1</sub> 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 (Fig. 1), presumably due to a ruptured aneurysm. A three-dimensional CT angiogram revealed a fenestration at the proximal portion of the right A<sub>1</sub> and a saccular aneurysm arising from the distal part of the A<sub>1</sub> fenestration



**Fig. 1.** Brain computed tomography scan shows a subarachnoid hemorrhage, especially in the interhemispheric fissure.

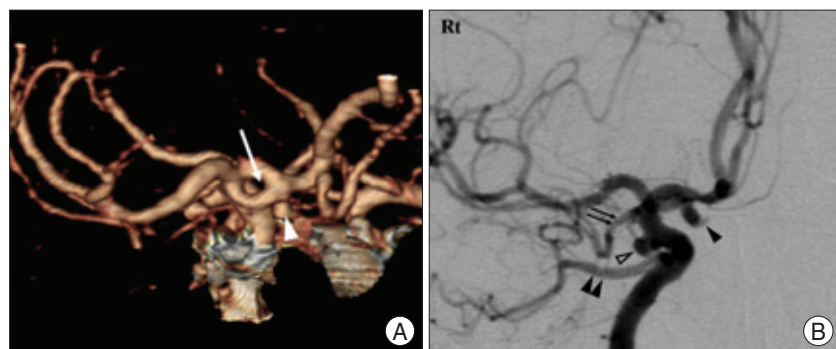
• Received : May 14, 2008 • Accepted : December 29, 2008  
• Address for reprints : Jun Seok Koh, M.D., Ph.D.  
Department of Neurosurgery, Stroke and Neurological Disorders Centre,  
East-West Neo Medical Hospital, Kyung Hee University School of  
Medicine, 149 Sangil-dong, Gangdong-gu, Seoul 134-090, Korea  
Tel : +82-2-440-6145, Fax : +82-2-440-7171  
E-mail : neurokoh@hanmail.net

(Fig. 2A). Cerebral angiography also showed the right A<sub>1</sub> 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 A<sub>1</sub> 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<sup>7</sup>. Fenestration of the ACA other than the anterior communicating artery, with or without accompanying aneurysms, has rarely been reported in the literature<sup>3,5,8-13,15,18-20</sup>. With very few exceptions<sup>17</sup>, the preferred sites of A<sub>1</sub> fenestrations are usually at the distal part of the A<sub>1</sub> segment<sup>3,5,8-13,15,18-20</sup>. Among various theories concerning the pathogenesis of distal A<sub>1</sub> fenestrations, a remnant of embryologic plexiform anastomosis between the primitive olfactory artery and the ACA is thus far well accepted<sup>14</sup>. However, according to Teal et al.<sup>16</sup>, 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 A<sub>1</sub> segment.



**Fig. 2.** Three-dimensional computed tomographic angiography (3-DCTA) and digital subtraction angiography (DSA). Craniocaudal view from subtracted 3-DCTA (A) shows the anterior cerebral artery fenestration (arrow) and an accompanying saccular aneurysm (arrowhead) arising from the distal end of a fenestration. Oblique view DSA (B) shows the right A<sub>1</sub> aneurysm (arrowhead) arising from the distal end of a fenestration. It also demonstrates that the right internal cerebral artery aneurysm (open arrow), locating proximal to a duplicated middle cerebral artery (double arrows), is associated with a fetal-type posterior cerebral artery (double arrow heads).

**Table 1.** Characteristics of aneurysms arising from A<sub>1</sub> fenestrations in the literature by year

Authors, year	Age/ Sex	Side & site of A <sub>1</sub> fenestration	An origin in A <sub>1</sub> fenestration	Associated findings of cerebral angiogram
Crompton. <sup>3</sup> , 1962	-/-	-,-	Proximal end	MCA fenestration VA An & fenestration
Waga & Morikawa. <sup>18</sup> , 1979	36/F	Lt, Distal half	Proximal end	ICA An
Yamada et al. <sup>20</sup> , 1982	43/M	Rt, Distal half	Proximal end	-
Inagawa et al. <sup>9</sup> , 1983	70/F	Lt, Distal 1/3	Proximal end	Dolichoectasia
"	43/M	Lt, Distal 1/3	Proximal end	MCA An
Korosue et al. <sup>11</sup> , 1983	41/M	Rt, Distal half	Proximal end	-
Handa et al. <sup>8</sup> , 1984	50/F	Rt, Distal half	Proximal end	-
Minakawa et al. <sup>12</sup> , 1985	56/M	Lt, Distal 1/3	Proximal end	Partial duplicated VA
Wakabayashi et al. <sup>19</sup> , 1985	41/M	Rt, Distal half	Proximal end	-
"	38/M	Rt, Distal half	Proximal end	-
Ogasawara et al. <sup>13</sup> , 1988	65/F	Lt, Distal half	Proximal end	-
San-Galli et al. <sup>15</sup> , 1992	42/M	Lt, Distal half	Proximal end	Aortic origin of VA, Dolicho-basilar artery
Friedlander & Ogilvy. <sup>5</sup> , 1996	33/M	Rt, Distal half	Proximal end	Both A <sub>1</sub> fenestration, Azygos A <sub>2</sub>
Kachhara et al. <sup>10</sup> , 1998	50/F	Rt, Distal half	Proximal end	Contralateral A <sub>1</sub> aplasia
Koh et al., 2008	75/F	Rt, Proximal half	Distal end	Duplicated MCA,
(current report)				Fetal-type PCA, ICA An

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

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 A<sub>1</sub> fenestration<sup>3,5,8-13,15,18-20</sup>. 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<sup>2,3</sup>. 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<sup>2,4</sup>. In our case, the aneurysm was found in an unusual site at the distal end of an A<sub>1</sub> fenestration. We

were unable to find any report of an aneurysm arising at the distal end of A<sub>1</sub> fenestration in the literature (Table 1). Only two cases of vertebrobasilar juncture aneurysm originating from the distal end of a fenestration have been reported<sup>1,6)</sup>. Congenital medial wall defect of the cerebral vessel and certain hemodynamic factors in relation to the very proximal A<sub>1</sub> 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 A<sub>1</sub> fenestration. As a result, the distal end of A<sub>1</sub> fenestration has more chance to be stressed than the proximal end to constitute the proper distal A<sub>1</sub> 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 A<sub>1</sub> 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.

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