Supraclinoid Internal Carotid Artery Fenestration with Associated Aneurysm: Case Report and Literature Review

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Fenestration of the supraclinoid internal carotid artery (ICA) is extremely rare and may occasionally be associated with saccular aneurysms; nevertheless, the natural history remains unclear. The authors reviewed the cases of fenestration of the supraclinoid ICA and evaluated the clinical characteristics and angioarchitecture with particular focus on the incidence for aneurysm development and rupture. Previously reported 24 cases and 1 new case from our institute were examined for the presence of aneurysms (including size and presence of bleb) leading to subsequent subarachnoid hemorrhage (SAH). Furthermore, 16 cases with sufficient information were classified into one of the three types according to the developmental condition of fenestration and fusion site: Type A (developed fenestration in which the ICA appears to duplicate [two cases]); Type B (hypoplastic fenestration fused to the ICA at the origin of the posterior communicating artery [Pcom] [six cases]); and Type C (hypoplastic fenestration fused to the Pcom itself or appeared to be a duplicated Pcom [eight cases]). In type A, the two cases had an aneurysm (100%), one of which caused SAH (50%). In type B, all six cases had an aneurysm (100%), and one of five led to SAH (20%) (one case lacked information regarding SAH). In type C, five of the eight cases had an aneurysm (62.5%), with no SAH (0%). The result suggested that the extent of development of fenestration and the location of fusion are related to the incidence of aneurysms and risk for rupture.

Keywords: fenestration, supraclinoid, internal carotid artery, aneurysm, subarachnoid hemorrhage

Introduction

Fenestration of the intracranial artery is a congenital anatomical variant defined as segmental duplication of the intracranial artery.1,2 It may occur as a result of an anomalous fusion or division of primitive embryologic vessels or plexiform network.2-4 The anterior communicating artery (Acom) is the most common site followed by the basilar and vertebral arteries.5-7

Of all cerebral blood vessels, fenestration of the supraclinoid internal carotid artery (ICA) is extremely rare and may occasionally be associated with saccular aneurysms.3,8 Owing to the progress of magnetic resonance imaging, cerebral angiography, and endovascular treatment, reports of supraclinoid ICA fenestration have been increasing slightly. To date, only 24 cases have been reported.1,3,5-8-26 Associated aneurysm may develop at the fenestration, especially on the proximal side, presumably due to vessel wall weakness,27 arterial wall defects, or unusual hemodynamic stresses.28 However, there is no further information, and the natural history of this entity and the risk for rupture remain unclear. We present a case of fenestration of the right supraclinoid ICA with a saccular aneurysm at the proximal end of the fenestrated segment that fused with the posterior communicating artery (Pcom). In addition, we reviewed previously reported cases and evaluated clinical characteristics and angioarchitecture, with a specific interest in the incidence for aneurysm development and rupture.

Case Report

A 35-year-old woman underwent a medical checkup of the brain at a local clinic. Magnetic resonance angiography incidentally revealed the aneurysm at the right ICA. Then, she was referred to our hospital. She had no symptoms or neurological deficits on admission. Digital subtraction angiography (DSA) showed the paraclinoid saccular aneurysm with a maximum diameter of 4.2 mm (Figs. 1A and B). A three-dimensional rotational angiographic reconstruction image revealed the fenestration connecting the ICA and the Pcom (Fig. 1C). The aneurysm was located at the proximal end of the fenestrated segment. We made the diagnosis of the supraclinoid ICA fenestration and the associated aneurysm. Follow-up policy was selected for this small ICA aneurysm without bleb.

Discussion

ICA fenestration is believed to be an extremely rare congenital variant. Although some cases are discovered by chance, many are pointed out by DSA study for associated aneurysms (including ruptured and unruptured lesions). The two most common findings are as follows: the fenestration is located in the supraclinoid segment (near the ophthalmic artery to the Pcom); and, second, the fenestration frequently associated with an aneurysm at the proximal end of the fenestrated segment. The ICA arises from the third aortic arch at the embryonic stage (4-5 mm), and the primitive carotid artery divides into the cranial and caudal divisions, which often has a small plexiform temporarily connecting
The failure of division or the persistence of the small plexiform may cause fenestration of the supraclinoid ICA. In some cases, including our case, the fenestration appeared to occur at the connection between the superior hypophyseal artery and the Pcom. In reports describing pathological analyses, a defect in the muscular layer of the vessel at each wall of the fenestration was identified. It is suggested that the combination of structural wall weakness and local hemodynamic stress, especially at the proximal site, is involved in aneurismal formation.

While an association with aneurysms has been shown, more detailed information regarding the natural history of this entity and the risk for rupture is scarce. To investigate the relationship between supraclinoid ICA fenestration and associated aneurysm, all relevant reports published in English found in the PubMed database were reviewed. In all, 24 cases of supraclinoid ICA fenestration were found in the published literature. In addition, one new case from the authors’ institution was added; thus, a total of 25 cases were collected and reviewed. Of these, 18 cases were reported after 2003 and, therefore, would have been evaluated with sufficiently high image quality (the other seven cases were reported before 1993). Two of 18 cases were excluded due to insufficient images or suspected dissection; the remaining 16 were classified into one of the three following categories according to the developmental condition of the fenestration and fusion site (Fig. 2): Type A (developed fenestration in which the ICA appears to duplicate); Type B (hypoplastic fenestration, which is fused to the ICA at the origin of the Pcom); and Type C (hypoplastic fenestration, which is fused to the Pcom itself or appears to be a duplicated Pcom). For these cases, the presence of aneurysms (including size and the presence of bleb) leading to subarachnoid hemorrhage (SAH) was analyzed.

All 25 cases of supraclinoid ICA fenestration are summarized in Table 1. The mean age at presentation was 42.0 years and 65% of the patients were female. In these cases, 71% had lesion-associated aneurysms. Among the available 17 cases, SAH occurred in 5; however, it appeared to include non-associated aneurysmal rupture, ICA dissection, and postoperative bleeding. A flow chart of the analysis is presented in Fig. 3.
Table 1  Clinical summary of 25 patients with supraclinoid ICA fenestration

| Reference    | Case | Age/sex | Modality       | Side | Type | AN at fenestration | AN size (mm)/bleb | SAH | Etcetera |
|--------------|------|---------|----------------|------|------|--------------------|-------------------|-----|----------|
| Yock, 1984   | 1    | 41/F    | Angiography    | R    | NA   | +                  | NA                | NA  |          |
| Yasargil, 1984 | 2    | NA      | NA             | R    | NA   | +                  | NA                | NA  |          |
| Findlay, 1987 | 3    | 28/F    | Angiography    | R    | NA   | −                  | NA                | NA  |          |
| Takano, 1991 | 4    | 51/F    | Angiography    | R    | NA   | −                  | NA                | NA  |          |
| Hattori, 1992 | 5    | 38/F    | Angiography    | L    | NA   | −                  | NA                | NA  |          |
| Banach, 1993 | 6    | 37/F    | DSA            | L    | NA   | +                  | NA                | NA  |          |
| Katsuta, 1993 | 7    | 46/F    | DSA            | R    | NA   | −                  | NA                | NA  |          |
| Tripathi, 2003 | 8    | 21/M    | DSA            | R    | C    | −                  | −                 | −   |          |
| Ng, 2006     | 9    | 34/F    | 3DRA           | R    | NA   | + (2 ANs)          | 3-4/−, 5-6/−      | −   | Insufficient images |
| Bharatha, 2007 | 10   | 73/M    | 3DCTA/DSA      | R    | C    | −                  | −                 | (+) | SAH post-tumor removal, s/o |
| Chen, 2007   | 11   | 31/M    | 3DRA           | R    | NA   | NA                 | NA                | (+) | dissection, s/o |
| Onoda, 2008  | 12   | 42/F    | 3DCTA          | L    | C    | +/proximal         | 4/−               | −   |          |
| Van Rooji, 2009 | 13   | NA      | 3DRA           | NA   | B    | +/proximal         | 2-3/−             | NA  |          |
| Baba, 2010   | 14   | 62/M    | 3DRA           | L    | C    | +/proximal         | 6/−               | −   |          |
| Plumb, 2010  | 15   | 48/F    | 3DRA           | L    | C    | +/proximal         | 8/−               | −   |          |
| Day, 2011    | 16   | 39/F    | 3DRA           | R    | B    | +/proximal         | 8.9/−             | −   |          |
| Day, 2011    | 17   | 32/F    | 3DRA           | L    | B    | +/proximal         | 3.9/−             | +   | SAH due to eclampisia, n/r/o |
| Ichikawa, 2011 | 18   | 47/F    | 3DRA           | L    | B    | +/proximal         | 7/−               | −   |          |
| Nakiri, 2012 | 19   | 47/F    | 3DRA           | R    | A    | +/proximal         | 12/−              | −   |          |
| Nakiri, 2012 | 20   | 44/M    | 3DRA           | L    | A    | + (2 ANs)          | 6/NA, 2.5/NA      | +   |          |
| Park, 2012   | 21   | 44/M    | 3DRA           | L    | B    | +/proximal         | 4.5/+             | (+) | SAH due to MCA AN |
| Uchino, 2013 | 22   | 21/M    | MRI            | L    | C    | −                  | −                 | −   |          |
| Uchino, 2013 | 23   | 40/M    | MRI            | L    | B    | +/proximal         | 2-3/−             | −   |          |
| Lee, 2018    | 24   | 65/F    | 3DRA           | R    | C    | +/proximal         | 5/−               | −   |          |
| Present case | 25   | 35/F    | 3DRA           | R    | C    | +/proximal         | 4.2/−             | −   |          |

AN: aneurysm, DSA: digital subtraction angiography, F: female, L: left, M: male, MCA: middle cerebral artery, NA: not available, R: right, SAH: subarachnoid hemorrhage, 3DRA: three-dimensional rotational angiography, 3DCTA: three-dimensional CT angiography.

excluded: one case (case 9) did not have sufficient description or images, and the other (case 11) was a case of suspected ICA dissection and dissected aneurysms from the clinical course and image findings. Of the 16 cases, two were Type A, six were Type B, and eight were Type C (Table 2); most cases had hypoplastic fenestration and those with a large fenestration were rare. In type A, two cases had an aneurysm (100%), one of which caused SAH (50%). In type B, all six cases had an aneurysm (100%), and one of five caused SAH (20%) (one case lacked information regarding SAH). In type C, five of the eight (62.5%) cases had an aneurysm, and there was no SAH (0%). The incidence for aneurysm and rupture decreased in the order type A > B > C.

In previous reviews, the rupture rate of aneurysms at the ICA fenestration has been reported to be 33–42% or more. However, these reports seem to include non-associated aneurysmal rupture, ICA dissection, and postoperative bleeding. Our research investigated the relationship between supraclinoid ICA fenestration, associated aneurysms, and SAH. Detailed examination of only more recent cases revealed that the rupture of aneurysm at the ICA fenestration occurred in < 2 of 15 cases (13.3%). Classification according to the developmental condition of the fenestration and fusion site appears to be useful for predicting the frequency of aneurysm and the risk for rupture. The incidence for aneurysm and its rupture decreases in the order type A > B > C. Moreover, the structure of type A is different from other types, and there is the possibility that developmental mechanisms and disease type are completely different. In particular, it is sometimes difficult to distinguish type A from...
blisters and dissection, but MRI vessel wall imaging and repeat angiography might be helpful. Furthermore, there is not enough opinion or consensus on treatment. Of the 13 patients with aneurysms, 9 received surgical treatments (one wrapping, four clipping, and four coiling) with uneventful postoperative course. The fenestration vessel was obstructed in three cases without any new symptoms (one clipping to type C, one clipping to type B, and one clipping to type A). Although the present study was relatively limited in scope and cases might involve other disease states (such as ICA dissection or eclampsia), the results, nevertheless, will be helpful for follow-up and determination of treatment strategies. It is necessary to consider treatment policies on an individual basis. Especially in surgical treatments, it is recommended that normal vessels should be preserved, even if fenestration has to be sacrificed.

Conclusion

We reviewed the frequency of associated aneurysm and the incidence for rupture in supraclinoid ICA fenestrations based on our original classification scheme. Although there were some limitations to this research, the following three findings are important. First, the extent of development of fenestration and the location of fusion are related to the incidence of aneurysms and rupture risk. Second, the incidence of rupture may not be as high as reported in previous studies. Third, it is recommended that normal vessels should be preserved in surgical treatments, even if fenestration has to be sacrificed. Owing to its rarity, these anatomical variations are not well understood and further accumulation of data is needed.

Conflicts of Interest Disclosure

The authors declare that they have no conflict of interest concerning the material or methods used in this study or the findings specified in this paper.

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