Endovascular Trapping of Both Sides of a Cervical Fusiform Carotid Aneurysm Associated with Marfan Syndrome. Case Report

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Objective: We report a rare case of cervical internal carotid artery aneurysm associated with Marfan syndrome and the technical details of endovascular trapping.

Case Presentations: A 23-year-old male who had been diagnosed with Marfan syndrome presented with dysphagia due to an enlarged left cervical pulsating mass. Imaging studies revealed a non-thrombosed, giant cervical internal carotid fusiform aneurysm measuring approximately 3–4 cm extending to the skull base. After bypass surgery between the superficial temporal artery and the middle cerebral artery, endovascular trapping of the cervical internal carotid artery at both sides of the aneurysm was performed under flow control without positioning any coils inside the aneurysm itself. A 6F distal access catheter technique through the aneurysm and double or triple microcatheter technique were useful for tight and short-length occlusion of the artery combined with a proximal balloon guiding catheter. Dysphagia resolved after treatment due to alleviation of the compression on the recurrent nerve and disappeared completely within 3 weeks.

Conclusion: Distal access catheter and multiple microcatheters under proximal flow control can be useful for endovascular trapping of both sides of a cervical fusiform aneurysm.

Keywords ▶ Marfan syndrome, carotid artery disease, fusiform aneurysm

Introduction

Aneurysms of the cervical internal carotid artery (ICA) associated with Marfan syndrome are rare, and previous cases were usually treated by a direct surgical approach. We report a case of endovascular trapping of diseased ICA after extracranial–intracranial bypass surgery and the technical details involved in alleviation of this condition.

Case Presentation

A 23-year-old male was diagnosed with Marfan syndrome and an ascending aortic aneurysm, 4 cm in size, at the age of 16 years. Recently, the patient experienced a pulsating mass in the left cervical region, which had enlarged during the previous 6 months before he was referred to our hospital complaining of dysphagia. He did not have any apparent history of cervical or other injuries. The aneurysm consisted of three sequential bulges (45 × 36 × 34 mm, 14 × 10 × 12 mm, and 12 × 11 × 11 mm) and originated from the left ICA, extending to the height of the skull base just before the cranial opening (Figs. 1A and 1B). No thrombosis was found inside the aneurysm. Flow retardation inside the aneurysm was observed through cerebral angiograms.

Treatment strategy

Because of the progressive enlargement of the aneurysm and the risk of cerebral ischemia due to distal embolism, radical treatment was planned. Trapping of only proximal common
carotid artery (CCA) was not considered because it was reported to bring retrograde partial filling of the aneurysmal lumen. Because of the distal position of the aneurysm and the fact that it was deeply located near the skull base, it was assumed that the direct approach might be difficult; therefore, endovascular treatment was selected. A balloon occlusion test of the left ICA revealed the occlusion tolerance without any neurological deficits or any reduction in the cerebral blood flow detected on single-photon emission tomography. However, after returning to his room, he complained some somnolence might be felt during the occlusion, which was an equivocal ischemic symptom. Bypass surgery from the external carotid artery (ECA) to the middle cerebral artery was performed in order to secure cerebral blood flow because of the young age of this patient. Low flow bypass was used because high flow bypass was only applied in patients who showed ischemic symptoms during balloon occlusion in our hospital, as previously reported. To deplete the mass effect and reduce the number of coils used, not internal trapping but endovascular trapping of the cervical ICA at both sides of the aneurysm without insertion of any coils inside the aneurysm was the intended intervention.
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Treatment

Endovascular trapping was performed under general anesthesia 6 days after bypass surgery. A 9F long-sheath introducer was placed in his right femoral artery. A balloon-mounted guiding catheter (9F Optimo; Tokai Medical, Aichi, Japan) was placed and inflated in the CCA. Because the flow from the bypassed superficial temporal artery was measurable, ECA was occluded by HyperGlide balloon catheter (Covidien, Irvine, CA, USA) from 6F guiding catheter via contralateral femoral artery to prevent distal embolism through bypassed superficial temporal artery. Although the direct approach to the ICA distal to the aneurysm with microcatheters was considered possible, there was difficulty in controlling the movement of the catheters due to swaging in the large aneurysm. In order to tightly occlude the ICA distal to the aneurysm, a distal access catheter (6F Cerulean STR, 115 cm; Medikit, Tokyo, Japan) was advanced through the Optimo balloon catheter to the cervical ICA distal to the aneurysm led by the inner catheter of the 4F inner catheter (4F Cerulean STR, 125 cm; Medikit) (Fig. 2A). Under balloon occlusion of the CCA and ECA, two microcatheters of Prowler Select Plus (Codman, Dublin, Ireland) and Excelsior 1018 (Stryker...
Neurovascular, Fremont, CA, USA) were placed in parallel to achieve a double catheter technique with stable support of the 6F distal access catheter. From two microcatheters in the ICA, distal ICA obliteration was achieved with 31 coils consisting of 17 fibered coils and 14 hydrogel coils under proximal flow control (Fig. 2B). Double catheter technique was used by forming each compartment of hydrogel coils from one catheter and filling the gaps using fibered coils from another catheter. Occlusion of the distal ICA was confirmed by the stasis of contrast medium injected slowly from the distal access catheter. Subsequently, complete obliteration of the ICA proximal to the aneurysm, which was small—approximately 1 cm, was achieved with 20 coils consisting of 13 fibered coils, four hydrogel coils and three bare platinum coils in this short segment due to a triple catheter technique consisting of Prowler select plus, Excelsior 1018, and SL-10 (Stryker Neurovascular) (Figs. 3A and 3B). Triple catheter technique was used by forming each compartment of hydrogel and bare platinum coils from one pair of catheters and filling the gaps using fibered coils from another pair of catheters. Balloons in the ECA and CCA were deflated after completion of endovascular trapping. Complete occlusion was confirmed by the injection from the proximal CCA and from the contralateral ICA and vertebral artery (Fig. 4).

**Outcome and follow-up**

Preservation of complete occlusion was demonstrated with carotid echography on the second day and cerebral angiography on the seventh day. Multiple, small postoperative embolic cerebral infarctions were asymptomatic as observed through MRI. Dysphagia due to compression on the recurrent nerve by the thrombus inside the aneurysm followed, but naturally resolved completely within 3 weeks.

The patient recovered fully without any sequelae. Follow-up cerebral angiography was performed at 7 months, and complete occlusion and disappearance of the cervical aneurysm were confirmed. Axial magnetic resonance (MR) imaging showed marked reduction of cervical fusiform carotid aneurysm (Fig. 5).

**Discussion**

Marfan syndrome is an autosomal dominant connective tissue disorder commonly due to mutation of the fibrillin-1 (FBN-1) gene that causes disruption of elastic fibers in large- and medium-size arteries. Consequently, the syndrome predisposes the individual to aneurysm formation and arterial dissection. The patients with Ehlers-Danlos syndrome, which is a disease of the collagen, have abnormal fragile vessel walls of the whole
body and are susceptible to hemostatic difficulties. However, most patients with Marfan syndrome have only cardiovascular complications and neurovascular complications are rare. Dissecting aneurysms of the extracranial cervical arteries in Marfan syndrome are most frequently caused by an extension of aortic dissection. In this case, neither apparent aneurysm in the proximal CCA nor any dissecting aorta was found.

Cervical internal carotid aneurysm is found in association with a number of infections including syphilis and Salmonella enteritidis, trauma, Ehlers–Danlos syndrome, and Marfan syndrome. Patients with this kind of aneurysm often show evidence of ischemic cerebrovascular disease in addition to dysphagia, pharyngeal discomfort, and dysgeusia due to space-occupying lesions that compress the lower cranial nerves. The benefit of early treatment for these aneurysms was confirmed in a long-term observational study, which showed a low incidence of spontaneous remission.

Some investigators have reported excision of the aneurysm and end-to-end anastomosis and some investigators have performed endovascular trapping with or without bypass. This type of aneurysm commonly extends to the skull base. As the cranial opening of the ICA (carotid canal) is deep-seated, cranial nerve palsy such as peripheral facial paralysis may occur. An endovascular approach is advantageous for trapping of the cervical ICA distal to the aneurysm. Although placing coils in the affected arteries may cause unfavorable effects such as severe inflammation, it was reported that covered stent graft or coils placed in the axillary and internal mammary arteries of a Marfan patient did not cause such effects for 9 months.

Because this patient did not show apparent occlusion intolerance of the ICA, only trapping ICA without bypass surgery could be performed safely. Because a nearly normal life expectancy is expected in a patient with Marfan syndrome and late de novo aneurysm formation following therapeutic ICA occlusion was reported, this patient might suffer another aneurysmal formation during his rest of long life. We decided to perform bypass surgery to contribute to the anterior circulation distal to the occluded ICA and lessen extended load from the ICA occlusion.

Trapping the arteries at both sides of a giant aneurysm without endosaccular coil insertion is advantageous in preventing the mass effect and also better with respect to cosmetic retention. Previous reports of endovascular treatment include only proximal occlusion and consequent protrusion of the coils placed in the distal segment into the aneurysm, which means that it is often difficult to advance the microcatheters to the distal portion and control them for tight packing. In our case, various backup methods, including the triple coaxial system and double and/or triple microcatheter techniques, were useful. Triple microcatheter technique was superior in trapping the shorter segment because another microcatheter was always available for the filling of the compartment formed by the coils from double microcatheters. It is also necessary to pay attention to the postoperative changes according to the thrombotic potential of the aneurysm such as airway obstruction and recurrent nerve palsy, even though this was temporary in our patient. Long-term outcome and postoperative course of this kind of aneurysm are unknown and further follow-up is required.

## Conclusion

In conclusion, aneurysms limited at the cervical ICA are rare in Marfan syndrome. Endovascular trapping of both sides of the aneurysm is useful. It is necessary to pay attention to the postoperative changes according to the thrombotic potential of the aneurysm. Proximal flow control by a combination of balloons and a multi-coaxial system can attain minimally invasive endovascular trapping.

## Disclosure Statement

All authors do not have any potential conflicts of interest or disclosures.

## References


