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Title:

Ruptured subclavian artery pseudoaneurysm associated with neurofibromatosis type 1

Authors:

Shigeyuki Sakamoto, Masayuki Sumida*, Shinichiro Takeshita*, Masaaki Shibukawa, Yoshihiro Kiura, Takahito Okazaki, and Kaoru Kurisu

Institutions:

Department of Neurosurgery, Hiroshima University Graduate School of Biomedical Sciences, 1-2-3 Kasumi, Minami-ku, Hiroshima 734-8551, Japan

*Department of Neurosurgery, Hiroshima Red Cross Hospital and Atomic-bomb Survivors Hospital, 1-9-6 Sendamachi, Naka-ku, Hiroshima 730-8619, Japan

Address correspondence to:

Shigeyuki Sakamoto, M.D.

Department of Neurosurgery, Hiroshima University Graduate School of Biomedical Sciences, 1-2-3 Kasumi, Minami-ku, Hiroshima 734-8551, Japan

Phone: +81-82-257-5227, Fax: +81-82-257-5229, E-mail: sakamoto@hiroshima-u.ac.jp

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Summary

A subclavian artery aneurysm associated with neurofibromatosis type 1 (NF 1) is extremely rare. We report a case of ruptured pseudoaneurysm of the subclavian artery in a patient with NF 1 treated with endovascular surgery. A 51-year-old man with NF 1 presented with initially sudden left neck pain and continuous dysphagia. Radiologic examination showed a pseudoaneurysm of the left subclavian artery. Endovascular stenting and coil embolization was performed to prevent rebleeding, and the pseudoaneurysm was completely obliterated. Follow-up angiogram at 3 months revealed good flow through the stent without flow into the pseudoaneurysm. Our presented case is the first to describe successful endovascular treatment for a ruptured subclavian artery pseudoaneurysm associated with NF 1. Endovascular stenting and coil embolization for the ruptured subclavian artery pseudoaneurysm is very effective.

Key words: endovascular treatment; neurofibromatosis type 1; ruptured aneurysm; subclavian artery

Introduction

Neurofibromatosis type 1 (NF 1) is an autosomal dominant disorder affecting one in 3000 to 4000 individuals [7]. Clinical features of NF 1 include multiple café-au-lait spots, benign neurofibromas, optic gliomas and iris hamartomas. Vascular abnormalities associated with NF 1 are rare and are reported to occur in 0.4-6.4% of patients [8]. The most common sites for arterial lesions are the aorta, the renal or cerebral circulations. The subclavian artery is rarely affected with only a few cases of dissection, aneurysmal dilatation or rupture reported in the literature [8]. We report herein a case of ruptured pseudoaneurysm of the subclavian artery in a patient with NF 1 treated with endovascular stenting and coil embolization.

Case report

The patient was a 51-year-old man with a diagnosis of NF 1. The patient experienced sudden left neck pain followed by continuous dysphagia. He consulted with a physician 3 days after onset. His physical examination revealed the neurofibromatosis and café-au-lait spots of the skin typical of patients with NF 1. Neurological examination showed no deficits.

CT scan with contrast medium on admission showed a 3-cm aneurysm in the left subclavian artery (Fig. 1A). A diagnostic angiogram 5 days after onset revealed an irregularly shaped, almost thrombosed aneurysm, 8.0 × 5.0 mm, of the left subclavian artery compared with the aneurysm shown by enhanced CT (Fig. 1B). The left vertebral artery appeared to be absent both on the CT angiogram and diagnostic angiogram did not show the left vertebral artery. The right vertebral artery was patent on the CT angiogram and diagnostic angiogram. The aneurysm

was diagnosed as pseudoaneurysm because the aneurysm shape had changed with time, and the left vertebral artery occluded as part of a dissection which likely caused the pseudoaneurysm. Therefore, treatment with endovascular stenting and possible coil embolization was planned to prevent rebleeding.

Endovascular treatment was performed under local anesthesia 10 days after onset. After the tip of a 6-Fr Shuttle sheath (Cook, Bjaeverskov, Denmark) was placed on the left subclavian artery proximal to the subclavian artery pseudoaneurysm using the coaxial method via right femoral artery, and 4000 units of heparin were intravenously administered for anticoagulation. Preoperative angiogram revealed that the aneurysm shape had further changed (Fig. 1C). A 10 × 20 mm Wallstent RP (Boston Scientific, Natick, MA) was positioned to cover the entire aneurysmal portion, and deployed (Fig. 1D). A Prowler select LPES microcatheter (Cordis, Miami, FL) was inserted through the Shuttle sheath and its tip was positioned into the pseudoaneurysm cavity through the stent struts. A total of two detachable coils [Micrus CASHMERE (9 × 22 mm) and Micrus CASHMERE (7 × 17 mm); Micrus, San Jose, CA] were deployed into the pseudoaneurysm cavity until it was completely obliterated (Fig. 1E).

Ten days after endovascular treatment, the patient was discharged on aspirin and clopidogrel without neck pain and dysphagia. Follow-up angiogram at 3 months revealed good flow through the stent without flow into the pseudoaneurysm.

Discussion

NF 1 is caused by mutations of the gene *NF 1* localized to chromosome 17q11.2 [16]. The protein product of the NF 1 gene is neurofibromin. This raises

the possibility of a link between neurofibromin deficiency and the NF 1 vasculopathy. The main hypothesis proposed by Riccardi [10] is that the vasculopathy is caused by the alteration of the normal process of vascular maintenance and repair regulated by neurofibromin.

Vascular abnormalities (arterial stenoses or aneurysms) in patients with NF 1 occur by a dynamic process of cellular proliferation, degeneration, healing, smooth muscle loss, and fibrosis [3]. Neurofibromin expression has been demonstrated in the vascular endothelial and smooth muscle cells [3]. The prevalence of vascular abnormalities associated with NF 1 is 0.4-6.4% [8]. Most patients with NF 1 vascular abnormalities are asymptomatic but have involvement of multiple vessels [8]. Oderich et al. [8] have reviewed 76 vascular abnormalities in 31 patients with NF 1. Arterial lesions were located in the aorta (n = 17 lesions), renal (n = 12), mesenteric (n = 12), carotid-vertebral (n = 10), intracerebral (n = 4), subclavian-axillary and iliofemoral arteries (3 in each). An aneurym of subclavian artery occurred in only 1 (1.3%) in 76 vascular abnormalities. Furthermore, Oderich et al. [8] have reported a review of 237 patients with NF 1 who had 320 vascular abnormalities in the English-language literature. The renal artery lesions were most common (41%) and were more often stenotic than aneurysmal. The carotid, vertebral, or cerebral artery lesions seen in 19% of patients were commonly aneurysms. Subclavian or innominate artery lesions (arterial aneurysm, dissection, rupture or stenoses) were seen in 6.3%.

Eleven cases of ruptured aneurysm of subclavian artery in a patient with NF 1, including our present case, have been reported in the literature (Table 1) [5, 6, 9, 11-15, 17]. The mean age at the time of diagnosis was 43.4 years (range, 7-61

years), with left-side preponderance, and the incidence was almost equal in men and women. The symptoms of the ruptured subclavian artery aneurysm included pain, hemothorax, expanding mass, dyspnea and dysphagia. Thoracotomy, ligation, repair, and grafting were performed as treatment of the ruptured subclavian artery aneurysm, and 4 of 11 patients underwent ligation only. Most patients improved after surgical proximal ligation only, though some suffered from upper extremity ischemia and gangrene. Therefore, ligation with grafting to reestablish blood flow to the extremity was also performed. However, surgical repair was aggressive and complex, and vessel reconstruction is limited by arterial fragility in a patient with NF 1 [4]. In contrast, a combination of endovascular stenting and coil embolization to prevent rebleeding and reestablish the blood flow peripherally has been reported in cases of extra internal carotid artery pseudoaneurysm [2]. However, there has been no reported case of ruptured subclavian artery aneurysm in a patient with NF 1 treated successfully with endovascular stenting and coil embolization.

The self-expanding stents are used to treat intra-and extra internal carotid, subclavian and other peripheral artery pseudoaneurysms. Furthermore, the combination of endovascular stenting and coil embolization has been reported in cases of extra internal carotid artery pseudoaneurysm [2]. The flexible stents may compress the pseudoaneurysm inflow tract, inducing stasis and facilitating intra-aneurysmal thrombosis; act as an endoluminal scaffold to prevent coil herniation into the parent artery, allowing tight packing of even wide-necked and irregularly shaped aneurysms; and serve as a matrix for endothelial growth [1]. Assali et al. [1] evaluated the potential for using flexible self-expanding uncovered stents with or without coiling to treat extracranial internal carotid, subclavian and

other peripheral artery posttraumatic pseudoaneurysm. They concluded that uncovered endovascular flexible self-expanding stent placement with transstent coil embolization of the pseudoaneurysm cavity is a promising technique for treating posttraumatic pseudoaneurysm vascular disease by minimally invasive methods, while preserving the patency of the vessel and side branches [1].

In the present case, we performed endovascular stenting and coil embolization to prevent rebleeding and reestablish the blood flow to the extremity. Follow-up angiogram revealed good flow through the stent without flow into the pseudoaneurysm. We used Wallstent because the smallar cell size of this stent would be more helpful in reducing the blood flow into the pseudoaneurysm and in preventing the herniation of coils into the parent artery. Although the use of covered stents may represent an alternative strategy in the endovascular treatment of the aneurysms, we did not use covered stents due to the risk of side-branch compromise. Wallstents and other self-expandable stents, as such SMART stent (Cordis, Miami, FL), have been used for stent placement with or without coiling for pseudoaneurysm [1, 2]. Other self-expandable stent or covered stent might also be one of option for management of pseudoaneurysm. We conclude that endovascular stenting and coil embolization appears to be a viable alternative to surgery.

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Figure legends

Fig. 1

(A) CT scan with contrast medium on admission showed a 3-cm aneurysm in the left subclavian artery.

(B) Diagnostic angiogram 5 days after onset revealed an irregularly shaped, almost thrombosed aneurysm of the left subclavian artery.

(C) Preoperative angiogram revealed that the aneurysm shape had changed further.

(D) Post-stenting angiogram revealed that a stent was deployed to cover the entire aneurysmal portion.

(E) Post-embolization angiogram revealed no aneurysm.

Fig1