Successful revascularization using a saphenous vein for a ruptured brachial artery aneurysm in a patient with neurofibromatosis type I

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ABSTRACT

Vasculopathy in patients with type 1 neurofibromatosis is known. Brachial artery aneurysms in patients with type 1 neurofibromatosis are rare, but any rupture can be extremely serious. A 56-year-old woman presented to our hospital with sudden pain in her right upper arm. Computed tomography revealed a ruptured brachial artery aneurysm, and operative reconstruction using a saphenous vein graft was performed. This is one of the few case reports of such successful revascularization using saphenous vein. The pathologic findings suggest neurogenic tumor invasion, and end-to-side anastomosis was effective in avoiding hemorrhagic complications. (J Vasc Surg Cases Innov Tech 2024;10:101350.)

Keywords: Brachial artery aneurysm; Neurofibromatosis type I; Saphenous vein graft; Vasculopathy; Von Recklinghausen

Neurofibromatosis type I (NF-1) is an autosomal dominant genetic disorder first described by von Recklinghausen[1] in 1882. This disease affects ~1 in 3000 persons and is characterized by skin pigmentation called café-au-lait spots and benign neurofibromas. It can also affect the blood vessels and cause aneurysms, typically in the aorta, renal arteries, and mesenteric arteries.[2] Aneurysms in an upper extremity are rare; however, any rupture can have tragic consequences. The patient provided written informed consent for the report of her case details and imaging studies.

CASE REPORT

A 56-year-old woman presented to our hospital with sudden pain in her right upper arm. She had no history of trauma or suspicious infection. She was known to have NF-1 and dyslipidemia. The diagnosis of NF-1 was made clinically according to the revised diagnostic criteria, without genetic testing.[3] Furthermore, she had never been screened for vasculopathy related to NF-1. Her vital signs were stable. Her right upper arm was tense and significantly tender. Blood test results showed a leukocyte count of 6500/μL and hemoglobin of 12.5 g/dL. Contrast-enhanced dynamic computed tomography (CT) revealed a fusiform aneurysm, 17 mm in size, in the right brachial artery with a hematoma slightly proximal to the elbow (Fig 1). She was immediately taken to the operating room for emergency treatment.

Surgery was performed with the patient in the supine position with both upper extremities horizontal and a tourniquet around the right upper arm. We harvested ~8 cm of the great saphenous vein from her left thigh. We made a 10-cm longitudinal skin incision around the aneurysm. The hematoma around the brachial artery was removed as cleanly as possible. The aneurysm was confirmed, and the brachial artery was clamped proximally and distally, before the aneurysm was excised. The arterial wall near the aneurysm seemed too fragile for an end-to-end anastomosis at 2 cm proximal and 2 cm distal from the aneurysm; thus, we ligated both artery ends with 4-0 monofilament thread using running sutures and constructed end-to-side anastomoses using a saphenous vein graft.

We performed a histopathologic examination to determine the etiology of the aneurysm. Proliferation of small spindle cells with a myxomatous background was observed surrounding the aneurysm wall. The myxomatous areas were positive for Alcian blue staining, and the small spindle-shaped cells stained positive for SOX10 by immunohistochemistry and negative for desmin and CD34 by immunohistochemistry (Fig 2). Elastica van Gieson staining showed degeneration and tearing of elastic fibers in the aneurysm wall. Because the specimen obtained was part of the aneurysm, evaluation of the margin was not possible (Table). The patient was discharged 10 days after surgery. At 3 weeks after surgery, contrast-enhanced dynamic CT revealed no aneurysm and patency of the saphenous vein graft (Fig 3).

DISCUSSION

Vascular lesions known to be associated with NF-1 include aneurysms, arterial stenosis, and vascular malformations. In small arteries, the common histologic features include spindle cell proliferation in the intima, fibrous thickening of the intima, and smooth muscle loss.[11] These vascular abnormalities are believed to result in arterial wall weakness, leading to vasculopathy. Most of these vascular lesions are asymptomatic, and their frequency is currently unknown.[12] It is thought that the

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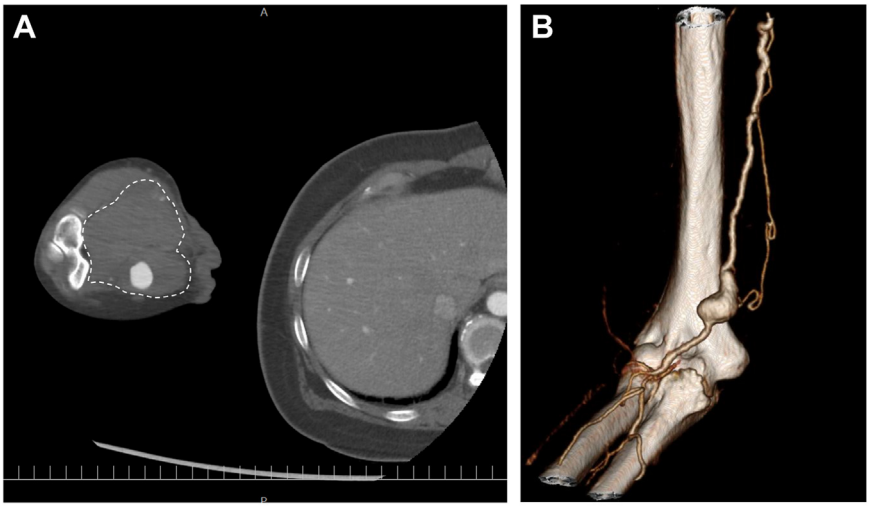

Fig 1. Contrast-enhanced dynamic computed tomography (CT) views revealing a spindle aneurysm (size, 17 mm) in the right brachial artery. *Dotted line* represents the surrounding hematoma.

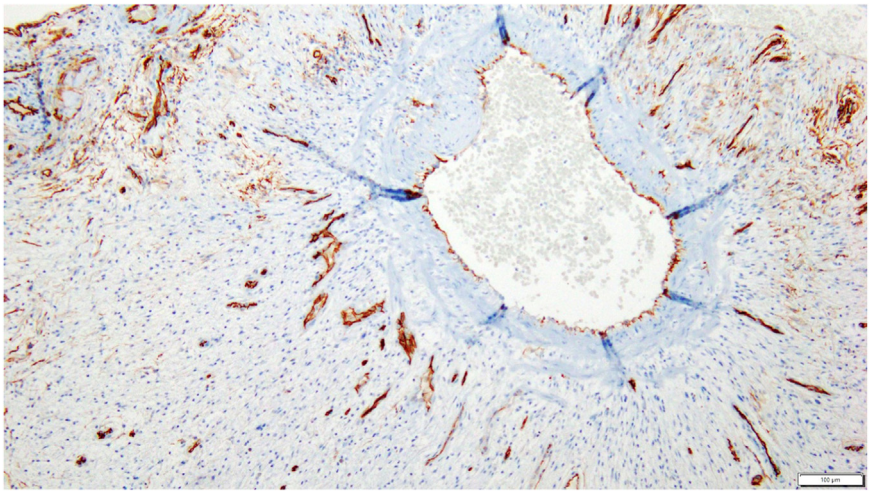

Fig 2. Proliferation of small spindle cells with a myxomatous background was observed surrounding the aneurysm wall. The small spindle-shaped cells stained positive with SOX10 immunohistochemistry.

aorta, renal artery, and mesenteric artery are the vessels most frequently forming aneurysms and that aneurysms of the arteries of the upper extremities (ie, axillary artery, brachial artery, radial artery) are less frequent.[2] Currently, no specific surveillance guidelines are available for arterial aneurysms in patients with NF-1. However, it is reasonable to consider screening for aneurysms in patients with documented vasculitis, because it is known that they often present with multiple vascular lesions.[2]

Several cases of ruptured brachial artery aneurysms in patients with NF-1 have been reported (Table).[4-10] In the previous reported cases, three of eight patients died and two survived but required upper extremity amputations. All the patients who survived without amputation had undergone vascular reconstruction with a saphenous vein graft. Degenaar et al[9] performed an end-to-end anastomotic revascularization using a saphenous vein graft, and the patient had a good outcome. Emori et al[7] also performed an end-to-end anastomotic revascularization using a great saphenous vein graft and reported intraoperative bleeding that was difficult to control due to the fragile vessels in the anastomotic site. We report another case of successful surgical revascularization using the saphenous vein. After resection of the aneurysm, each end of the artery was ligated and anastomosed at a safe distance from the aneurysm. No bleeding problems occurred intraoperatively or postoperatively, and the patient had a good clinical course. All

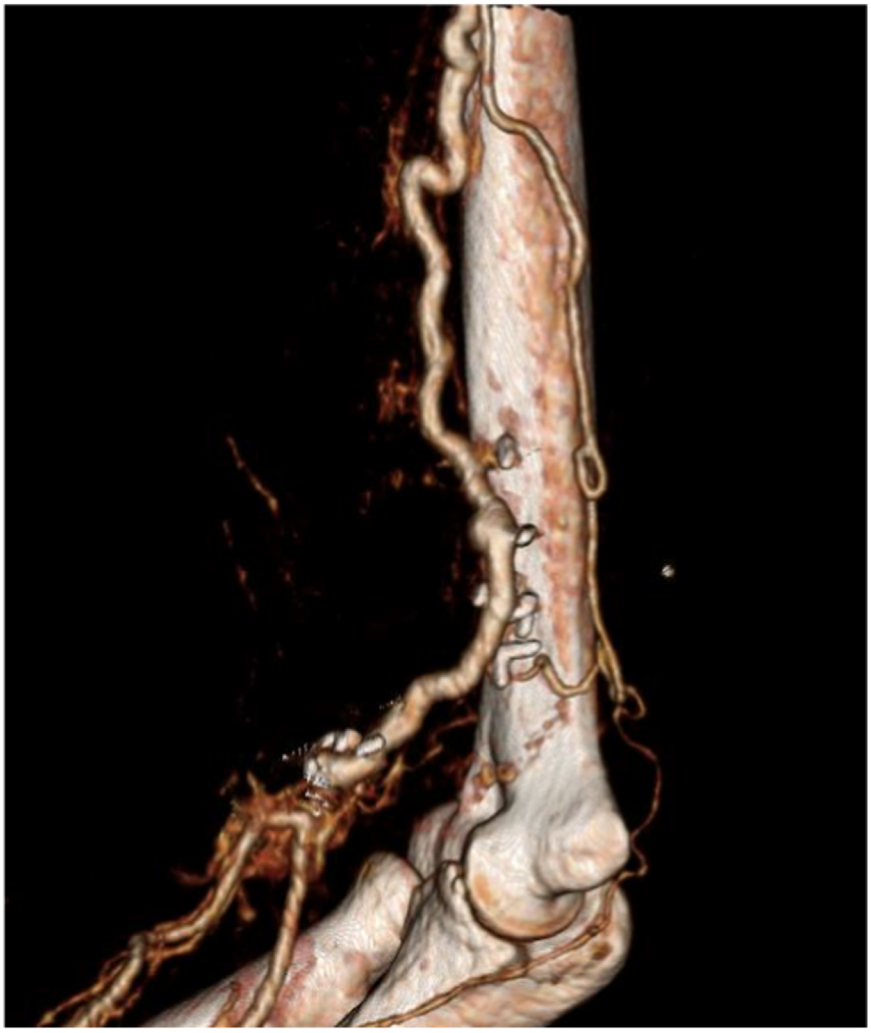

Fig 3. Three weeks after surgery, contrast-enhanced dynamic computed tomography (CT) revealed no aneurysm and patency of the saphenous vein graft.

previous successful cases had used an end-to-end anastomosis. Because the vessel wall near the aneurysm is likely to be fragile, even if apparently normal by visual inspection, it is appropriate to securely ligate the vessel near the aneurysm and perform an end-to-side anastomosis to avoid hemorrhagic complications.

We used the great saphenous vein graft, as in the previous literature. NF-1 is considered to affect almost all vessels, including veins. Concern exists that using venous grafts in young patients could result in a risk of aneurysmal degeneration.[13] However, the saphenous vein is commonly selected as a graft, and we could not find a report of graft rupture or reoperation due to a graft aneurysm.

In patients with NF-1, the vessel wall is thought to become fragile, and an aneurysm can form due to direct invasion of neurofibroma tumor.[11,14] S-100 immunohistochemistry is considered less specific for nerve cells than SOX10 immunohistochemistry because the former reacts with other mesenchymal and epithelial cells, and SOX10 immunohistochemistry has higher sensitivity and specificity for melanocytes and Schwann cells.[15] In the present case, SOX10-positive spindle cell proliferation was seen in the artery wall, suggesting tumor invasion of neuronal origin.

However, postoperative CT showed no residual or recurrent aneurysms. However, it is not clear whether the neurofibroma invading the artery had been completely resected pathologically. Although no studies have reported the recurrence of aneurysms after revascularization, it might be wise to test for the absence of tumor cells in the vessel margins with a rapid intraoperative pathology test to reduce the risk of recurrence.

DISCLOSURES

None.

Table. Reported cases of brachial artery aneurysm[a]

Pt. No.	Investigator	Age, years; sex	Side	Surgery	Outcome
1	Saitoh et al,[4] 1998	28; F	Left	Resection after ligation	Died 9 days after surgery
2	Tidwell et al,[5] 1998	30; F	Right	Failed reconstruction; amputation	Alive, but required amputation
3	Jeong et al,[6] 2008	35; F	Right	Resection after ligation	Died 4 days after surgery
4	Emori et al,[7] 2010	53; F	Left	Reconstruction with vein graft (end-to-end anastomosis)	Alive; limb well conserved
5	Balanescu et al,[8] 2019	34; F	Right	Compression bandaging/ reconstruction with vein graft (end-to-end anastomosis)	Alive; limb well conserved
6	Degenaar et al,[9] 2019	48; F	Left	Reconstruction with vein graft (end-to-end anastomosis)	Alive; limb well conserved
7	Lee et al, 2021 (patient 1)	33; M	Left	Embolization; amputation	Alive; but required amputation
8	Lee et al,[10] 2021 (patient 2)	32; M	Right	Embolization; amputation; additional embolization	Died 7 days after second embolization
9	Present case	56; F	Right	Reconstruction with vein graft (end-to-side anastomosis)	Alive; limb well conserved

F, Female; *M*, male; *Pt. No.*, patient number.
[a]All patients alive without amputation had undergone reconstruction with a vein graft.

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