Endovascular treatment for a ruptured lumbar artery aneurysm in a patient with neurofibromatosis type 1

Ken Tsuchida, MD, PhD,[a] Kyosuke Kokaguchi, MD, PhD,[a] Tetsuya Hasegawa, MD, PhD,[b] Daijirou Akamatsu, MD, PhD,[c] *and* Kenji Namiki, MD, PhD,[d] *Osaki and Sendai City, Japan*

ABSTRACT

The present medical case report describes successful endovascular treatment via stent graft and coil packing for a ruptured lumbar artery aneurysm in a 55-year-old woman with neurofibromatosis type 1. Although less common, vasculopathy, such as an aneurysm, stenosis, rupture, and arteriovenous fistula, have been reported and can be a cause of death for patients with this disorder. However, only a few cases of a ruptured lumbar aneurysm have been reported. (J Vasc Surg Cases Innov Tech 2023;9:101208.)

Keywords: Embolization; Endovascular; Lumbar artery aneurysm; Neurofibromatosis type 1; Stent graft

Although the fragility of the vasculature in patients with neurofibromatosis type 1 (NF1) is well-established, the incidence of ruptured lumbar artery aneurysms (LAAs) is relatively low. In two prior reports, both afferent and efferent vessels were treated with coil embolization.[1,2] Nevertheless, in the present case, owing to the shortened afferent artery, the aneurysm was embolized with coils, and a stent graft was deployed in the aorta as a treatment modality. The potential usefulness of this approach depends on careful anatomic evaluation and appropriate device selection. The patient provided written informed consent for the report of her case details and imaging studies.

CASE REPORT

A 55-year-old female patient with a history of NF1 was referred for back pain and nausea that had continued for 1 day. The enhanced computed tomography scan showed a hematoma in the right retroperitoneal space due to the rupture of a right LAA (Fig 1). Clinical examination showed multiple café-au-lait macules and abdominal discomfort. At the age of 3 years, an intellectual disability was diagnosed, and she had a surgical history of repair of scoliosis. At 30 years of age, a neoplasm had been surgically removed from the femoral region and histopathologically confirmed as NF1. Additionally, at age 40 years, a neoplasm associated with NF1 was removed from the left gluteal region without concomitant vascular anomalies. At the current presentation, no genetic testing had been undertaken. Her hemodynamic status was stable; however, the blood test results indicated anemia (hemoglobin level, 8.7 g/dL).

With the patient under general anesthesia, emergency aortography through the left femoral artery was performed and showed an aneurysm of the first right lumbar artery. The aneurysm was successfully selected using a 4F hook-shaped catheter (model 231-42-SMAX-700-2P-SH0; Hanaco Medical).

Selective angiography revealed the presence of a very short afferent artery <1 cm from the aorta and three efferent artery branches. The aneurysm was embolized with two interlocking detachable coils (Interlock-35, two-dimensional, diameter, 15 mm $\times$ 40 cm and 12 mm $\times$ 40 cm; Boston Scientific). A 12F sheath was inserted through the right femoral artery, and a leg extension stent graft (iliac extender endoprosthesis, diameter, 16 mm $\times$ 7 cm; Excluder PLL161407 J; W.L. Gore & Associates) was deployed in the aorta distal to the renal artery. A microcatheter was left in the aneurysm at stent graft placement to allow for injection of N-butyl-2-cyanoacrylate or the placement of microcoils in the event of efferent backflow.

The final aortography findings showed that the first LAA was successfully embolized (Fig 2). No complications or blood transfusions were necessary. A contrast-enhanced computed tomography scan was performed on postoperative day 5 and showing a thrombosed lumbar aneurysm but no extravascular or other aneurysms (Fig 3). She was discharged 1 week after surgery with no complications to the facility where she was staying.

DISCUSSION

Neurofibromatosis is a syndrome that affects the skin and nerves and is associated with tumors of the brain,

From the Division of Vascular Surgery[a] and Division of Diagnostic Radiology,[b] Osaki Citizen Hospital, Osaki; the Division of Vascular Surgery, Department of Surgery, Tohoku University Hospital, Sendai City[c]; and the Osaki Citizen Hospital, Osaki.[d]

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Correspondence: Ken Tsuchida, MD, PhD, Division of Vascular Surgery, Osaki Citizen Hospital, 3-8-1 Furukawahonami, Osaki 989-6183, Japan (e-mail: kent@surg.med.tohoku.ac.jp).

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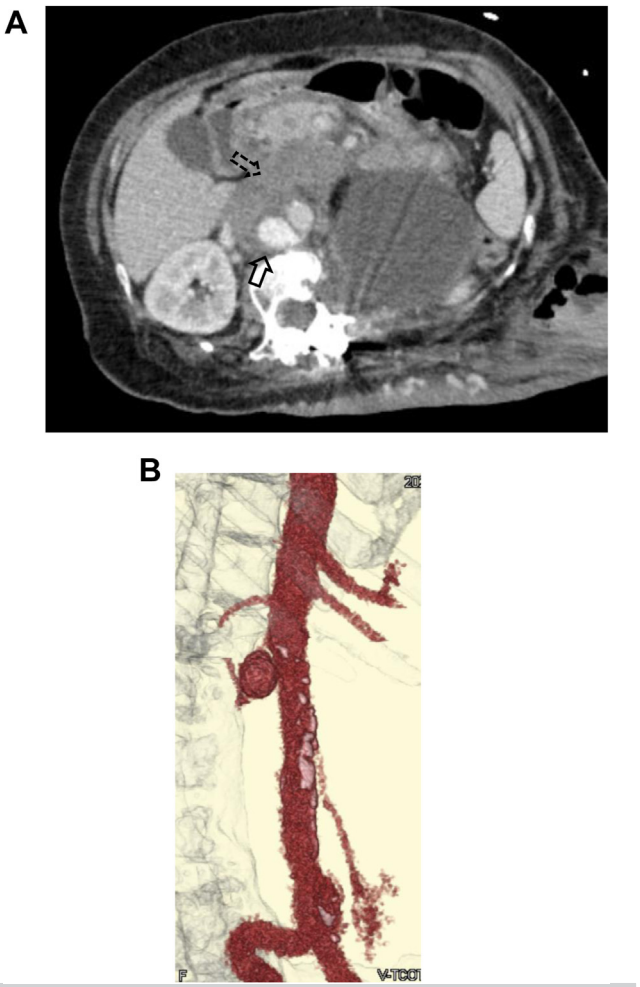

Fig 1. Preoperative computed tomography angiography. **A,** Preoperative computed tomography scan showing a hematoma in the right retroperitoneal space due to rupture of a right lumbar artery aneurysm (LAA). The *dotted arrow* denotes the hematoma; and *arrow,* a right LAA. **B,** Three-dimensional reconstructed computed tomography image.

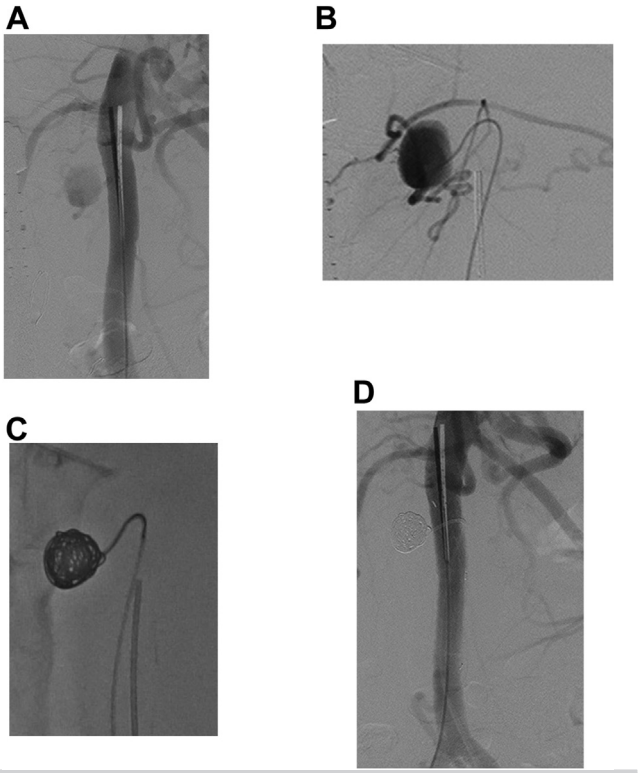

Fig 2. Angiography and endovascular treatment. **A,** Aortography showing a right lumbar artery aneurysm (LAA). **B,** Selective angiography of a right LAA showing that the afferent part of the lumbar artery is very short, with multiple efferent branches of the aneurysm. **C,** The aneurysm was packed with a coil using an Interlock-35. **D,** The aneurysm is no longer visible in the final contrast-enhanced scan. No dissection or other collateral damage was observed.

spinal cord, organs, skin, and bones. NF1 represents 96% of all neurofibromatosis cases, NF2 3%, and schwannomatosis 1%. NF1 is the most frequent subtype and is characterized by café-au-lait spots, benign neurofibromas, and iris hamartomas.[3] In addition to bilateral vestibular schwannomas, NF1 is characterized by aneurysmal and occlusive vascular abnormalities. Vasculopathy in NF1 includes arteriovenous malformations, aneurysms, and stenosis, with 0.4% to 6.4% of patients affected.[4] The most common sites of aneurysm formation in patients with neurofibromatosis include the carotid and vertebral arteries, aorta, mesenteric arteries, renal arteries, and cerebrovascular system. Vascular disorders are the second leading cause of death in those with NF1, after cancer.[3] Studies have reported treatment of peripheral artery aneurysms such as those in the renal artery, internal carotid artery, intercostal artery, and subclavian artery.[5] Nevertheless, the incidence of LAAs in those with NF1 is very rare, and only two cases treated via endovascular embolization with coils have been reported.[1,2]

In the present report, we describe the first case of effective endovascular therapy for an LAA in a patient with NF1 using a stent graft and coil packing. In our patient, given the location of the aneurysm and presence of a hematoma, an open surgical procedure was deemed excessively invasive, and an endovascular technique was selected. Individuals with a diagnosis of NF1 are considered to have elevated susceptibility when subjected to general anesthesia. This results from complications arising from tracheal intubation and laryngoscopy, in addition to the patients' cardiovascular conditions and pulmonary fibrosis.[6] Although regional anesthesia is preferable, general anesthesia was chosen for our patient because of her intellectual disability and inability to stay immobile during the procedure.

In the two previously reported studies, the afferent and efferent arterial branches were occluded using coil

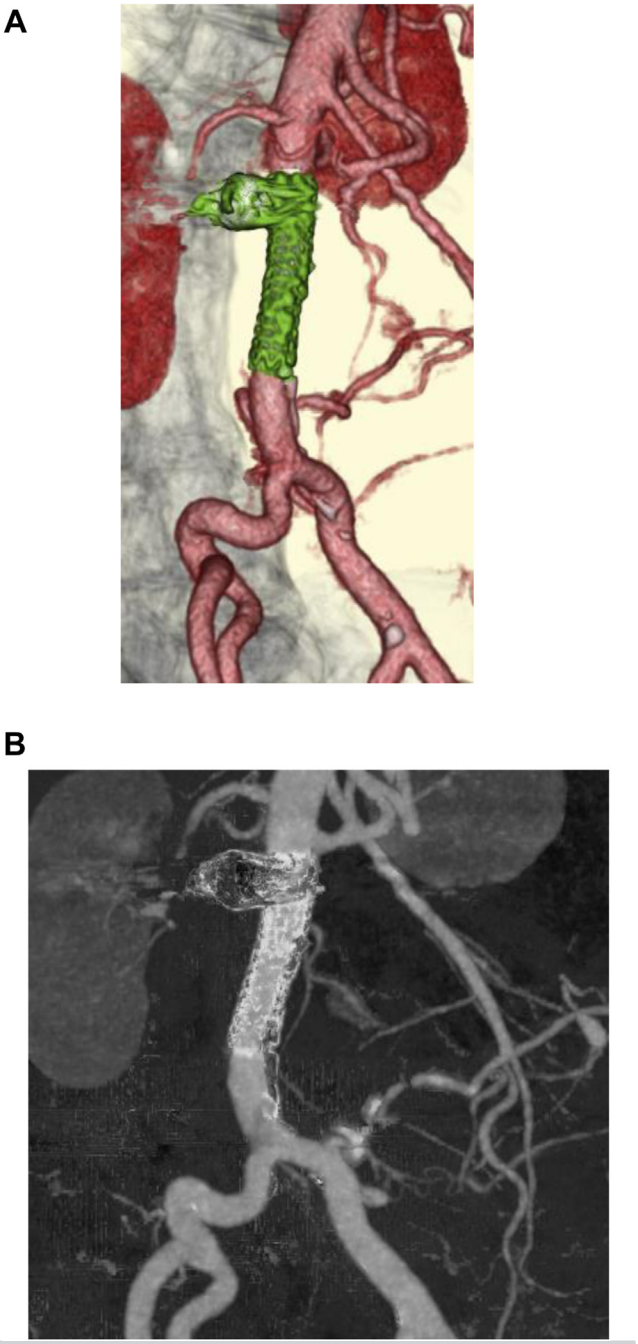

Fig 3. Postoperative computed tomography angiography indicating a thrombosed lumbar artery aneurysm (LAA) with no extravasation and no additional aneurysms. **A,** Three-dimensional reconstructed computed tomography image. **B,** Maximum intensity projection.

embolization.[1,2] However, in the present case, because of the presence of numerous efferent arterial branches and the likelihood of coil migration into the aorta owing to the limited afferent artery, it was decided to embolize the aneurysm with a coil, and stent graft interpolation was performed within the aorta to interrupt the afferent vessel.

In 2014, the Japan Medical Safety Research Organization issued a warning regarding the need to carefully evaluate the risk of vascular injury during surgery or other invasive procedures near the great arteries in patients with NF1 owing to their vascular fragility.[7] In the current and expanding endovascular era, endovascular aortic repair (EVAR) has emerged as a treatment option for abdominal aortic aneurysms linked to NF1.[8,9] Because of the less direct intervention with EVAR compared with open surgery, EVAR can prevent vascular fragility-related complications. However, studies have reported vascular fragility-related complications in patients who underwent EVAR, including ulceration of the intima,[8] distal end perforation of the stent graft,[10] and formation of a secondary aneurysm.[4,9,11] Therefore, the size of the stent graft should be carefully chosen.

In typical EVAR, a stent graft that is 10% to 20% larger than the diameter is used. However, the aim in the present case was to obstruct the afferent branch of the lumbar artery; thus, a stent graft of appropriate size should be chosen. In the present case, the aortic diameter was 16 to 14 mm, and an iliac Extender stent of 16 mm $\times$ 7 cm seemed optimal. No balloon dilation was performed because of vascular fragility.

Furthermore, LAAs associated with NF1 are very rare, and no treatment strategy has been established. In terms of the invasiveness and vascular vulnerability, endovascular treatment might be preferable to open surgery. Coil embolization of both the afferent and the efferent vessels is a minimally invasive treatment but can be anatomically or technically difficult. Although a detailed anatomic assessment and appropriate device selection are required, coil packing of the aneurysm and interpolation of a stent graft to occlude the afferent artery could be useful treatment options.

In cases for which an endovascular approach is being considered for a patient with NF1, it is imperative to take great care to prevent any injury to the delicate vascular endothelium. Furthermore, physicians must perform a thorough examination of the entire body, because multiple vascular lesions could be present. Vigilant monitoring during the follow-up period is also obligatory for patients with NF-1.

CONCLUSIONS

Endovascular treatment of an LAA in a patient with NF1 using a stent graft and coil packing was performed for our patient. Stent graft placement and coil packing can be useful and therapeutically successful if the indications are considered and the device is selected appropriately.

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