Thoracic endovascular aortic repair for traction-induced aortic avulsion injury in neurofibromatosis type 1

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ABSTRACT
Rupture of intercostal artery aneurysms has been reported in patients with neurofibromatosis type 1. Many reports have demonstrated the efficacy of endovascular interventions. Herein, we present a case of successful treatment with thoracic endovascular aortic repair for traction-induced avulsion injury of the previously embolized intercostal artery. We further report the potential postoperative risk of rapid aneurysmal enlargement, possibly owing to changes in the thoracic arterial regional network. Even after successful treatment, vascular surgeons should pay attention to other aneurysmal events in the acute phase and avulsion injuries in the chronic phase. Close follow-up is essential. (J Vasc Surg Cases Innov Tech 2022;8:726-8)

Keywords: Neurofibromatosis; Intercostal artery; Thoracic endovascular aortic repair

Neurofibromatosis type 1 (NF1) is an autosomal-dominant disease. It is known for skin and musculoskeletal abnormalities, such as neurofibroma, café-au-lait spots, and scoliosis. NF1 is also associated with vascular wall fragility, leading to aneurysms or stenosis. Surgical treatment of small arterial aneurysm ruptures in NF1 is challenging. Endovascular intervention is the primary treatment choice. Here, we report a case of traction-induced aortic avulsion injury in a 42-year-old patient with a history of surgical and endovascular interventions for intercostal artery aneurysms.

CASE REPORT
A 42-year-old man with NF1 presented to our hospital with persistent acute lumbar pain, which arose after twisting the lower back 2 days prior. The patient was diaphoretic and nauseous. His blood pressure was 137/78 mm Hg, his heart rate was 82 beats/min, and his extremities were cold. He had a history of multiple treatments for intercostal artery aneurysms. To summarize, the right thoracic (Th) fourth, sixth, eighth, and tenth intercostal arteries were occluded by transcatheter arterial embolization and coiling. At the 7-month postoperative follow-up visit, the patient was in good health, and no stent-induced injury had been performed.

During the surgery, the patient had suffered massive intraoperative bleeding owing to vascular fragility and intrathoracic scarring. When the patient arrived at our hospital, a contrast-enhanced computed tomography (CECT) scan revealed a massive hematoma in the posterior mediastinum (Fig 1, A). Extravasation was seen directly from the descending aorta; however, extravasation from the distally separated intercostal artery was not observed. Contrary to the CECT findings in the past, the previously embolized right Th eighth intercostal artery was separated from the aorta (Fig 1, B/B′). Therefore, we diagnosed the patient with traction-induced aortic avulsion injury to the right Th eighth intercostal artery. A small right Th eleventh intercostal artery aneurysm was also detected (Fig 1, C). Aortic angiography revealed extravasation from the aorta (Fig 2, A). We successfully performed thoracic endovascular aortic repair (TEVAR) using a 10%-oversized graft (GORE TAG, TGMR262610J; W. L. Gore & Associates, Flagstaff, AZ) (Fig 2, B). Simultaneous intervention for right Th eleventh intercostal artery aneurysm was postponed owing to the risk of spinal ischemia. The patient was extubated in the operating room. There were no neurological complications. Considering the vulnerability of the vessel wall, the postoperative systolic blood pressure was controlled at less than 140 mm Hg.

On postoperative day 3, the patient experienced mild lumbar pain. On postoperative day 7, a follow-up CECT showed rapid enlargement of the right Th eleventh intercostal artery aneurysm, which was diagnosed as an impending rupture (Fig 3, A). We emergently occluded the aneurysm and inflow vessels using embolization and coiling (Fig 3, B). At the 7-month postoperative follow-up visit, the patient was in good health, and no stent-induced injury was detected.

DISCUSSION
NF1 is an autosomal-dominant disease that is sometimes associated with vascular lesions owing to the fragility of blood vessels. Lin et al reported a 0.8% prevalence of peripheral vascular abnormalities among 2322 patients. This report is likely an underestimation; most
patients remain asymptomatic and do not require imaging tests. The precise frequency of vascular abnormalities, therefore, remains unknown.

Spontaneous hemothorax in NF1 is a rare but life-threatening complication caused by massive bleeding into the thoracic cavity. This event can be caused by a rupture of an intercostal artery aneurysm, for example. Surgical hemostasis during thoracotomy is challenging because of the friable vessel walls. Separating a bleeding vessel from the surrounding tissue is also difficult.² Fedoruk et al³ reported an operative mortality of 33% in their review. Our patient had a history of left thoracotomy, which was difficult to manage, leading to massive bleeding. Friable vessels made even ligation challenging. In addition, the left thoracic redo approach was impractical because of scarring; therefore, an endovascular intervention was a reasonable choice.

Multiple treatments for recurrent intercostal artery aneurysms have been reported in the past, as in this case.⁴ In addition, this case highlights the possibility of an avulsion injury, even after the treatment of intercostal artery aneurysms. Our patient's onset of lumbar pain

Fig 1. (A) A contrast-enhanced computed tomography (CECT) taken on arrival revealed a massive hematoma in the posterior peritoneum. Extravasation occurred directly in the aorta. The distance between the aorta and the previously embolized right thoracic eighth intercostal artery (B) differed from that in the prior image (B'). A left thoracic eleventh intercostal artery aneurysm (red arrow) was also detected (C).

Fig 2. (A) Aortic angiography shows an extravasation (red arrow) from the aorta. (B) The aortic stent graft was successfully deployed without endoleak.
implied that a rapid lumbar twisting motion led to pull-away traction power between the relatively unfixed aorta and the previously occluded, fixed, and friable intercostal arteries. In this situation, aortic stent implantation is a reliable and minimally invasive procedure because the artery distal to the bleeding site had already been occluded.

Preoperative scrutinization of the bleeding site is crucial; misdiagnosing an avulsion injury is a risk. Detecting intercostal artery bleeding is challenging. Multiple treatments of the intercostal arteries and a history of surgery for scoliosis deteriorate the quality of the CECT images. If bleeding had occurred from the nontreated intercostal artery, TEVAR could not have managed the bleeding from the distal end of the ruptured intercostal artery. We confirmed this using late-phase CECT images and angiography of the proximal aortic arch before TEVAR. No regional collateral arteries showed extravasation.

Another risk factor is aortic injury. Nakai et al reported rapid aneurysmal development after endovascular aortic repair in NF1. Aortic wall fragility leads to dissection, and true or pseudoaneurysm formation and touch-up ballooning should be avoided. The size of the stent graft was challenging to determine. The primary purpose is to stop bleeding; however, an oversized graft could lead to aortic injury. To our knowledge, there is no consensus on the graft size for NF1. We chose a 10% oversized graft and there was no endoleak or early phase aortic injury.

The present case also shows a possible sequela of endovascular aortic repair in NF1. After TEVAR, the right Th eleventh intercostal arterial aneurysm rapidly enlarged, causing new lumbar pain, indicating impending rupture. Moro et al reported a left colic artery aneurysm rupture 22 days after abdominal aortic stent placement in NF1. The mechanism of rapid aneurysmal change seems to be the same. In this case, the aortic stent graft occluded several other intercostal arteries, causing changes in the regional blood flow distribution. This change might have increased the blood flow and arterial pressure of the aneurysm and led to the quick degeneration. The timing of follow-up CT scan should be carefully planned; follow-up CT scans 1 week and 1 month after the intervention may be recommended. Newly recognized pain should not be overlooked. Moro et al reported a left colic artery aneurysmal rupture 4 days after the aneurysm was detected. If a new aneurysm is detected, prompt embolization or surgery should be performed.

CONCLUSIONS
This study offers two critical findings. First, traction-induced aortic avulsion injury can occur even after the occlusion of intercostal arterial aneurysms, and TEVAR can be a potential intervention. Second, rapid enlargement of an additional intercostal artery aneurysm may occur after aortic stent implantation. Close follow-up is essential.

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