Case Report

Endovascular Treatment of Ruptured Intercostal Arteriovenous Fistulas Associated with Neurofibromatosis Type 1

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We report a rare case of ruptured intercostal arteriovenous fistula in a patient with neurofibromatosis type 1. The patient presented with severe back pain. Angiography revealed ruptured intercostal arteriovenous fistulas. Successful coil embolization to occlude the fistulas and the aneurysm resulted in successful recovery of the patient.

Keywords: neurofibromatosis type 1, intercostal arteriovenous fistula, endovascular treatment

INTRODUCTION

Neurofibromatosis type 1 (NF1) or von Recklinghausen’s disease is an autosomal dominant disorder caused by an abnormality of chromosome 17 and characterized by multiple skin tumors and abnormal cutaneous pigmentation,1,5 with incidence of 1 in 2500–3000 births.3) In patients with NF1, vascular abnormalities such as occlusion or stenotic disease, aneurysms, arteriovenous malformations and arteriovenous fistulas, are known to occur.4) Because of the complex nature of the lesions, treatment options are limited and carry significant risk of bleeding. We report the case of a patient with NF1 who underwent coil embolization of the ruptured intercostal arteriovenous fistulas.

CASE REPORT

A 47 year-old man with NF1 presented to our hospital complaining of sudden back pain. His past medical history was significant for right hemothorax of unknown cause 10 years prior to admission. A general examination revealed cutaneous neurofibroma and café-au-lait spots all over in his body. A contrast-enhanced thoracic computed tomography showed hematoma in the posterior mediastinum and leakage of the contrast material (Fig. 1). Rupture of the thoracic aorta was suspected, and an emergency angiogram was performed.

Digital subtraction angiogram (DSA) performed in the right transfemoral route revealed aneurysmally dilated intercostal arteries at the T9 to T11 level of the right side of the descending aorta and extravasation of the contrast media. Angiography of the T7, T8, T9, T10 intercostal arteries on the right side showed arteriovenous fistulas (Fig. 2A) and aneurysm (Fig. 2B). These ultimately drained into the azygous veins (Fig. 3A). Angiography of the right T11 intercostal artery demonstrated ruptured intercostal arteriovenous fistulas and aneurysm (Fig. 3B). Retrograde filling of the internal vertebral plexus and ascending lumbar veins were also seen.

Under local anesthesia, using Seldinger technique and the femoral approach, a 5-French long sheath (Hanaco Medical, Tokyo, Japan) and a 5-French SHK, BI catheter (Hanaco Medical, Tokyo, Japan) were placed. A microcatheter/guidewire (Cook, Bloomington, IN, U.S.) was advanced into the 8th to 11th intercostal artery. The intercostal arteriovenous fistulas and aneurysms were occluded with a total of 48 coils (1 of 30 × 4 mm, 3 of 20 × 3 mm, 1 of 40 × 5 mm; Trufill occlusion system, Johnson and Johnson, Tokyo, Japan, 11 of 5 × 2 mm, 3 of 6 × 2 mm, 6 of 4 × 2 mm, 5 of 3 × 4 mm, 11 of 3 × 5 mm, 6 of 3 × 2 mm; Cook, Bloomington, IN, U.S., 1 of 6 × 40 mm, and 1 of 5 × 50 mm platinum microcoil; Cook, Bloomington,
IN, U.S.). The post-embolization angiogram showed successful occlusion of the arteriovenous fistulas (Fig. 3C). The post-procedural course was uneventful. The follow-up CT on the 8th post-embolization day showed no change in size of the hematoma in the posterior mediastinum. Rebleeding was not seen, and the patient was discharged home in good condition.

**DISCUSSION**

The incidence of vascular lesion in NF1 has been reported to be only 3.6%. However, the frequency of blood vessel involvement in NF1 has been underestimated, primarily because lesions may be clinically silent or may be attributed to other causes once discovered. There has been an increasing awareness of vascular...
lesions in patients with NF1, including stenosis, aneurysms and arteriovenous fistulas.\textsuperscript{6} Arterial lesions seen in NF1 may include compression due to an extrinsic adjacent tumor, intramural thickening, and saccular or fusiform aneurysms due to vascular dysplasia of various types.\textsuperscript{1}

Only a few cases of NF1 associated with ruptured intercostal arteriovenous fistula have been reported. Massive hemorrhage in patients with NF1 is a rare but potentially a lethal complication, and the prognosis is poor if the patient falls into hypovolemic shock.\textsuperscript{2} Conventional treatment of intercostal arteriovenous fistulas is ligation of the feeding artery by open surgery. However, surgical treatment in patients with NF1 can be dangerous or even impossible because the natural history of this disease process is still not completely understood, and vascular fragility might exacerbate bleeding. Possible causes of bleeding in NF1 patients include bleeding by vascularized tumors of mesenchymal origin, such as ganglioneuromas or neurofibromas, and bleeding caused by rupture of weak medium in large caliber arteries.\textsuperscript{3} These arteries are either surrounded by neurofibromatous or ganglioneuromatous tissue or have weak walls caused

\textbf{Fig. 3} Late venous phase shows the azygos vein (arrow) (A). Selective angiography in venous phase of the right T11 intercostal artery shows the ruptured aneurysm (black arrow). White arrow indicates draining vein (B). Thoracic aortography after occlusion of feeders with microcoils (arrows) (C) shows no residual filling of feeding arteries (D).
by intimal proliferation, thinning of the muscle layer, and fragmentation of the elastic layer, thus making them susceptible to aneurysm formation.

Endovascular treatment of vertebral arteriovenous fistula and hemothorax in NF1 has been reported in the literature.\(^3\)\(^{14–10}\) Coil embolization is a popular, less invasive, and safe method for the control of arterial hemorrhage in cases not requiring preservation of arterial flow.\(^9,10\) In order to find the source of bleeding, the best option is to make a radiological exploration, which allows the vascular surgeons to evaluate the individual lesions meticulously, with special attention to flow patterns. In our case, bleeding was due to a rupture of the intercostal arteriovenous fistula at T11 level producing posterior mediastinal hematoma. Endovascular coil embolization was successfully carried out in the management of the ruptured intercostal arteriovenous fistulas. Although good prognosis of endovascular treatment compared with surgery has been reported in the literature, fatal rebleeding remains a life-threatening complication. A well-developed tortuous vasculature structure in NF1 patients with vascular lesions can complicate endovascular treatment.\(^10\)

Percutaneous endovascular embolization of the intercostal arteriovenous fistula offers a new, radical and effective option. It is less invasive and allows precise and selective occlusion of only the abnormal vascular structures.

In conclusion, this is a rare case of ruptured intercostal arteriovenous fistulas in NF1, which were successfully treated with endovascular technique. Endovascular approach can be safe and effective in treating a lesion that may have otherwise been fatal.

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**Reference**