Stent Graft Repair and Coil Embolization for Spontaneous Hemothorax

A rare case of spontaneous hemothorax of neurofibromatosis in the chest is treated with combined endovascular repair of stent graft placement and coil embolization.

BY WILMO C. OREJOLA, MD; ELIE M. ELMANN, MD; AND GREGORY T. SIMONIAN, MD

Neurofibromatosis, also called *von Recklinghausen disease*, is a relatively common genetic disorder.[1] It is rarely associated with vascular complications. Only a few cases have been reported involving life-threatening, spontaneous bleeding into the chest. Surgical management has been a challenge. However, recent advances in endovascular technology provide a less-invasive approach for treatment and prevention of recurrence.

CASE REPORT

A 40-year-old man presented in the emergency department complaining of left-sided chest pain and dyspnea. He had absent breath sounds on the left. A chest roentgenogram showed massive left effusion (Figure 1). He had a history of asthma, hypertension, and neurofibromatosis type I. A brother died from non-Hodgkin lymphoma, and a sister has neurofibromatosis.

On the day of admission, he had a syncopal episode. A contrast-enhanced computerized tomographic scan of the chest revealed a large left hemothorax, and extravasation of contrast was traced from the costocervical trunk at the origin of the superior intercostal branch (Figure 2). A nonocclusive pulmonary emboli to the right upper lobe was also noted. He was tachypneic, in sinus tachycardia, but hemodynamically stable. Initial bedside thoracentesis did not obtain any information but under ultrasound guidance, blood was obtained. He was treated with intravenous fluid, oxygen inhalation, and bronchodilators. The patient's hemoglobin was 14 g/dL.

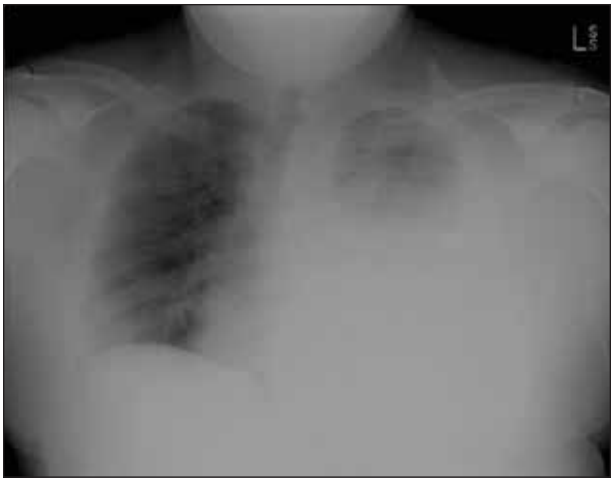

Figure 1. Chest radiograph on admission showing massive left effusion.

On the second hospital day, his hemoglobin dropped to 7 g/dL. He was brought to the operating room for left video-assisted thoracoscopic surgery. Large amounts of thrombus filled the left pleural cavity. Approximately 4,000 mL of clots and blood were evacuated. The operation was converted to a thoracotomy for better visualization. Bleeding was coming from the pleura and tissue surrounding a tumor at the thoracic inlet region where the apex of the left lung was adherent. This was covered with clots and necrotic tissues, tumor, and lymph nodes. Tissue biopsy showed neurofibroma (Figure 3). No malignant cells were identified.

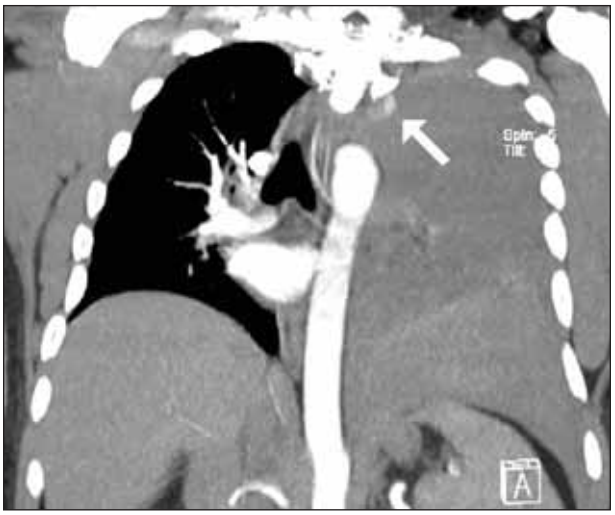

Figure 2. Coronal contrast-enhanced computed tomographic scan shows extravasation of contrast from the left costocervical trunk (see arrow).

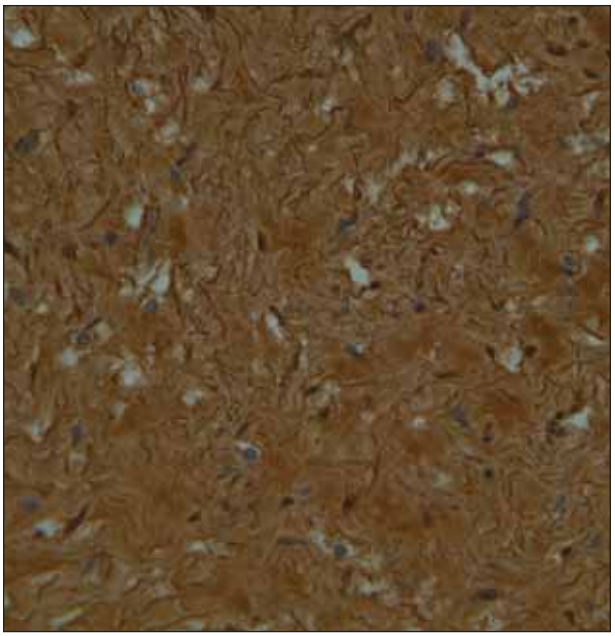

Figure 3. S-100 protein immunostain of neurofibroma taken from the apex of the left hemithorax.

Hemostasis was temporarily achieved by applying multiple 3-0 and 4-0 prolene suture ligatures at the level of the first and second intercostal spaces and the tissues surrounding the friable tumor. Packing and fibrin glue sealant were also used. Because no specific source of bleeding was identified under direct visualization, recurrence was anticipated. Given the spontaneity and life-threatening nature of hemothorax in neurofibromatosis, it was decided that a more definitive procedure should be performed in the catheterization lab the next morning.

The right femoral artery was cannulated percutaneously, and a 5-F Pinnacle sheath (Terumo Interventional Systems, Somerset, NJ) was inserted over a 0.035-inch Emerald J-wire (Cordis Corporation, Bridgewater, NJ). A 5-F vertebral Slip-Cath catheter (Cook Medical, Bloomington, IN) was then inserted over a 0.035-inch angled Glidewire (Terumo Interventional Systems) into the left subclavian artery. Selective subclavian angiography and subsequent superselective catheterization identified the left internal mammary artery, costocervical trunk, and subscapular arteries supplying the tumor.

A Wholey 0.035-inch, 260-cm wire (Covidien, Mansfield, MA) was placed into the left axillary artery, and a 6-F Terumo Pinnacle sheath was positioned at the proximal subclavian artery. A 6-F Cordis internal mammary artery catheter was placed over an angled Glidewire into the left internal mammary artery. The 5-F vertebral catheter was then also advanced over the Glidewire into the proximal left internal mammary artery, which was coil embolized with three 5-mm and two 7-mm coils (Boston Scientific Corporation, Natick, MA). Angiography confirmed thrombosis of the vessel.

The 6-F Terumo Pinnacle sheath was replaced over the Wholey wire with a 9-F 65-cm Arrow-Flex sheath (Arrow International, Reading, PA) into the left subclavian artery. A 10- X 40-mm Fluency covered stent graft (Bard Peripheral Vascular, Tempe, AZ) was then deployed into the 8-mm subclavian artery, carefully passing the origin of the vertebral artery in order to cover the origins of the costocervical and subscapular branches. There were no significant technical challenges encountered during the procedure. A completion angiogram showed the stent graft fully covering the watershed vessels of the mass including the left internal mammary artery, which was coil embolized earlier (Figure 4).

The patient was discharged without complications. One- and 2-year follow-ups have shown no recurrence of hemothorax based on clinical examination and chest roentgenogram.

COMMENTS

Neurofibromatosis type I is a dominant autosomal disease characterized by spots of increased skin pigmentation (often referred to as *café au lait*), peripheral nerve tumors, and a variety of dysplastic abnormalities of the skin, nervous system, bones, endocrine organs, and blood vessels. Vascular involvements have been described as stenotic or aneurysmal alterations in large

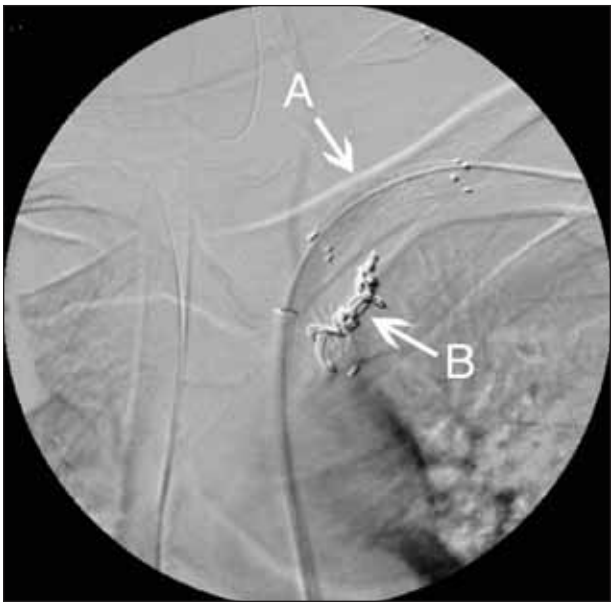

Figure 4. Angiogram shows the stent graft in the left subclavian artery (arrow A) and coil embolization of the internal mammary artery (arrow B).

vessels (commonly the renal arteries and the aorta and its branches) and/or dysplastic features in smaller vessels.[1] This has occurred only in 3.6% of neurofibromatosis patients studied.[2]

Although the incidence is low, this vascular complication has been reported to cause life-threatening spontaneous hemothorax when associated with ruptured aneurysmal disease of the intercostal,[3,4] thyrocervical,[5] and internal mammary arteries.[6] Including this case report, no other sites of spontaneous rupture of a neurofibroma in the chest have been reported.

Surgical repair of dysplastic vessel and friable neurofibromatosis tissues has been a challenge, which includes primarily conservative direct suturing and packing to achieve hemostasis.[7] However, when a specific source of bleeding is identified, endovascular coil embolization has been used effectively, as reported in ruptured thyrocervical trunk[5] and intercostal artery aneurysms.[4]

Although stent grafts have been used for treating pseudoaneurysm of the subclavian and brachiocephalic arteries from trauma,[8] stent graft repair for bleeding neurofibromas has not been reported. In our case, coil embolization to the internal mammary artery had to be performed separately because it also fed the tumor through the intercostal branch, and the stent graft cover had to spare the origin of the vertebral artery superiorly. We report this first case of combined endovascular repair and coil embolization in massive spontaneous hemothorax associated with neurofibromatosis.

Coil embolization of the left internal mammary artery combined with stent graft placement in the subclavian artery appears to be an effective endovascular means to avoid recurrence in this clinical situation. The life-threatening nature and spontaneity of hemothorax associated with neurofibromatosis could not be overemphasized. Furthermore, bleeding neurofibromas in the chest have been reported only in the thoracic inlet region. This may warrant further studies so that clinicians will be more prepared in dealing with this rare but life-threatening vascular complication. ■

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