Spontaneous rupture of an intercostal artery in a patient with neurofibromatosis type 1

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Abstract

A 48-year-old man with neurofibromatosis type 1 (NF1) presented with a right pleural effusion. A 3D computed tomography (CT) angiogram showed an aneurysm of the right 11th intercostal artery. He had no history of chest trauma so we diagnosed a spontaneous rupture of the aneurysm causing a massive effusion. We opened his pleural cavity and found lacerated pleura and active bleeding in the posterior 11th intercostal space. After controlling the active bleeding, we treated a persistent oozing from the region of the 10th–12th vertebrae with pressure hemostasis by absorbable oxidized cellulose packing. The next day, the patient gradually developed a paraplegia affecting both lower limbs. Magnetic resonance imaging (MRI) showed spinal cord compression at the level of the 9th and 10th vertebrae. We evacuated the cellulose and coagulum. The patient’s paraplegia improved and within six months he was walking without a crutch.

Keywords: Neurofibromatosis type 1; Spontaneous rupture of the peripheral artery; Hemothorax; Absorbable oxidized cellulose; Paraplegia

1. Introduction

Neurofibromatosis type 1 (NF1) disease may primarily involve any tissue in the body, including connective tissue, nerve tissue, the vasculature and others. Vascular signs and symptoms associated with NF1 include renovascular hypertension [1] and neurologic abnormalities [2, 3]. Spontaneous rupture of a major artery is, however, extremely uncommon. We report the case of a 48-year-old man with NF1 who suffered a spontaneous rupture of his right 11th intercostal artery.

2. Case report

A 48-year-old man with NF1 presented with a right pleural effusion. Several days before admission, he had experienced a worsening dull pain in his right upper back. Computed tomography (CT) of his chest revealed a right pleural effusion. His hemodynamic status was stable, with a hemoglobin of 13.9 g/dl and white blood cell count of 12,500/µl. His chest pain subsided 3 hours after admission. A 3D CT angiogram showed an aneurysm or pseudoaneurysm of the right 11th intercostal artery (Fig. 1). The next day, the pleural effusion had increased in volume and his hemoglobin had dropped to 12.3 g/dl. As he had no history of chest trauma, we diagnosed a spontaneous rupture of the intercostal aneurysm causing a massive effusion. A radiologist refused coil embolization due to the fragile nature of the vascular tissue in NF1. We scheduled an urgent right thoracotomy to control the hemothorax.

We opened his pleural cavity by a standard right thoracotomy via the 10th intercostal space. After removal of a massive blood coagulum and effusion, we did not find an aneurysm or pseudoaneurysm, but we found lacerated pleura and active bleeding in the posterior 11th intercostal space. Soft tissue edema around the ribs prevented identification of the aneurysm despite careful inspection near the spine. We controlled active bleeding from the 11th intercostal artery, possibly including the distal end of the aneurysm, but the region of the 10th–12th vertebrae continued to ooze. Without resection of ribs or vertebrae, we used pressure hemostasis by absorbable oxidized cellulose packing to control the bleeding from this area. We could not find the proximal end of the aneurysm.

The next day, the patient gradually developed paraplegia affecting both lower limbs. Magnetic resonance imaging (MRI) revealed that the cellulose packing had swollen in the spinal canal and, in combination with a blood coagulum, was compressing the spinal cord at the level between the 9th and 10th vertebrae (Fig. 2). We successfully evacuated the cellulose and coagulum. A postoperative MRI showed that we had relieved the spinal cord compression and a chest CT showed resolution of the aneurysm. We transferred the patient to another hospital for rehabilitation and his paraplegia improved and he could walk without a crutch within six months.

3. Discussion

NF1 is an autosomal dominant disorder linked to chromosome 17. It affects approximately one in 3000 people and may primarily involve any tissue of the body, including...
connective tissue, nerve tissue, the vasculature and others [4]. In addition, compression of the gastro-intestinal or urinary tracts by visceral neurofibromas may generate serious complications. The propensity for malignant transformation in any organ also leads to increased morbidity and mortality in sufferers of the disease. Arterial stenosis due to intimal or medial dysplasia is a well-known vascular complication of NF1 and some cases of arteriovenous malformations and aneurysms have also been reported [4–7]. Arterial lesions may be located in the aorta or in renal, mesenteric, carotid-vertebral or intracranial arteries.

Signs and symptoms vary according to the size and location of the vascular lesions and include ischemic symptoms or mass effects from the involved artery. Renovascular hypertension due to renal artery stenosis [1] and neurologic abnormalities due to intracranial aneurysms [2, 3] are both well recognized syndromes. However, spontaneous rupture of a major artery is extremely uncommon. Cases of ruptured intercostal, popliteal, subclavian, intracranial and retroperitoneal arteries have been reported [8–10].

CT and/or MRI scans are required for anatomical diagnosis. The treatments for vascular complications depend on the location and type of lesion. Endovascular coil embolization is used for an aneurysm or pseudoaneurysm. Percutaneous transluminal angioplasty or bypass grafting are used for stenotic lesions [1, 4, 6]. Open surgery includes clipping aneurysms, excision of aneurysms with or without grafting, and excision or ligation of arteriovenous malformations [5, 8, 9]. However, open surgery is often difficult due to the fragile nature of the vascular tissue in NF1 [10]. Late vascular problems include the occurrence of additional vascular lesions and close follow-up is thus required in patients with NF1.

Our patient developed chest pain and a hemothorax. Our radiologist judged that coil embolization would be technically difficult due to the fragile nature of the vasculature in NF1. The patient’s hemodynamic condition was stable and we observed his progress overnight expecting spontaneous hemostasis. His hemodynamic status was unchanged the next morning, but the hemoglobin level had dropped. Thus, we scheduled an urgent operation. We could not detect the precise oozing point or the arterial wall during his right thoracotomy so we were unable to obtain pathological findings of an aneurysm or pseudoaneurysm. The preoperatively recognized aneurysm might be a pseudoaneurysm of the intercostals artery.

We used pressure hemostasis by absorbable oxidized cellulose packing to control the bleeding around the vertebral. Although we controlled his bleeding, the patient gradually developed paraplegia the day after surgery. We successfully retrieved the swollen cellulose packing. Using cellulose packing for pressure hemostasis around the spine carries the risk of causing spinal cord compression. In such cases, an additional left thoracotomy might be useful to
control bleeding because the right intercostal arteries can be accessed along the descending aorta situated on the left side of the spine.

4. Conclusions

We have reported a rare case of spontaneous rupture of the 11th intercostal artery in a patient with NF1. We used pressure hemostasis by absorbable oxidized cellulose packing to control bleeding around his vertebrae because we could not identify the precise point of arterial rupture. The patient gradually developed paraplegia the day after surgery. MRI revealed the cellulose packing had swollen in the spinal canal. We successfully evacuated the cellulose and coagulum, and the patient’s paraplegia resolved within six months of the operation.

References