BRIEF COMMUNICATION

SPONTANEOUS HEMOTHORAX CAUSED BY A Ruptured INTERCOSTAL ARTERY ANEURYSM IN von RECKLINGHAUSEN’S NEUROFIBROMATOSIS

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Abstract: Aneurysms arising from an intercostal artery are very rare vascular malformations in von Recklinghausen’s neurofibromatosis, which often have a silent clinical presentation and are difficult to diagnose before rupture. We report a case of von Recklinghausen’s neurofibromatosis with massive hemothorax caused by spontaneous rupture of an intercostal artery aneurysm in a 29-year-old man. The diagnosis was eventually confirmed by percutaneous angiography and treated with endovascular embolization. During a 10-month follow-up period, the patient had a satisfactory recovery. This case illustrates that angiography and possible endovascular embolization should be the first strategy in managing hemothorax in patients with von Recklinghausen’s disease.

Key words: Aneurysm, ruptured; Embolization, therapeutic; Hemothorax; Neurofibromatosis 1; Thoracic arteries


Von Recklinghausen’s neurofibromatosis, also named neurofibromatosis type 1 (NF-1), is a well-recognized entity that is characterized by numerous neurofibromas, spots of abnormal cutaneous pigmentation, and a variety of other dysplastic abnormalities of the skin, nervous system, bones, endocrine organs and blood vessels.¹² Vascular abnormalities in patients with NF-1 have been described for large, medium-sized and small muscular arteries in the intracranial, thoracic and abdominal circulation, the most common of which are arterial stenoses or aneurysms involving both kidneys.³⁶ Due to its low incidence of 3.6% in NF-1 patients with vascular involvement, the association of arterial aneurysms has rarely been described.³⁷ These aneurysms can potentially rupture and sometimes pose a diagnostic and therapeutic dilemma. We report a case of NF-1 with rupture of the intercostal artery causing subsequent massive hemothorax. To our knowledge, this is the third reported case of treatment of this condition by endovascular embolization, and the first such report from Taiwan.

Case Report

A 29-year-old man presented at the emergency room with sudden onset shortness of breath and severe retrosternal pain radiating to his back. One week prior to admission, dull discomfort upon breathing was noted, which was not affected by position or physical activity. He had no cough at that time. Over the next few days, the pain progressed and involved his back and right shoulder. The pain was persistent and radiated to the right chest wall. The pain became more intense and suddenly exacerbated in the afternoon of the day of admission. There was no history of recent trauma or seizure. Von Recklinghausen’s neurofibromatosis had been diagnosed in this patient at the age of 8 years, with the classic symptoms of café-au-lait spots, axillary freckling and plexiform neurofibromas on the trunk and extremities, together with a familial history of neurofibromatosis.

Vital signs on arrival were systolic blood pressure 110 mm Hg, pulse rate 112 beats/minute and respiration

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rate 24 breaths/minute. Physical examination disclosed tachycardia, normal blood pressure and shortness of breath. Breath sounds were decreased over the lower one-half of the right hemithorax, with dullness to percussion and decreased tactile and vocal fremitus over the same area.

Laboratory data included the following: hemoglobin, 13.4 g/dL; white blood cell count, 11,600 cells/mm³ (differential, normal); platelet count, 391,000/mm³; normal urinalysis data; and normal values for creatinine, prothrombin time and activated partial thromboplastin time. The chest radiographs including anteroposterior and decubitus views showed a large right pleural effusion with near complete opacification of the right lower lung zone (Fig. 1).

Since the chest CT findings and thoracocentesis were consistent with right-sided hemothorax (Fig. 2), emergent thoracotomy was carried out under the diagnosis of hemothorax of unknown origin. This procedure failed to identify the cause or to stop the bleeding. Surgery was terminated since there was no evidence of pulsatile hemorrhage and definitive bleeding points could not be identified during surgery.

Emergent aortography was performed to locate the origin of bleeding. A pool of the contrast medium was found around the right seventh intercostal artery, and then selective right seventh intercostal arteriography disclosed a ruptured arterial aneurysm (Fig. 3). Subsequently, endovascular embolization was performed with a 5 mm coil (Occluding Spring Emboli, Cook, Bloomington, IN, USA), which stopped the bleeding successfully.

Fig. 1. Chest radiography shows a large right pleural effusion with near complete opacification of the right lower lung field.

Fig. 2. Computed tomography scan of the chest reveals a large amount of right-sided high-attenuated fluid collection associated with a well-circumscribed high-density mass, consistent with hemothorax and hematoma formation. The mediastinal structures were pushed to the left.

Fig. 3. Selective angiography of the right seventh intercostal artery reveals the aneurysm, and extravasation of the contrast medium displaying the origin of hemorrhage.
Two weeks later, the patient had an uneventful recovery and he remained well in the ensuing 10 months.

**Discussion**

Spontaneous hemothorax is an infrequent clinical entity that is usually caused by coagulopathy, hyper-vascular tumors, remote traumatic injury or bleeding from a primary non-traumatic vascular source.\(^6\) Von Recklinghausen’s neurofibromatosis associated with a ruptured intrathoracic artery aneurysm is an extremely rare condition. Spontaneous rupture and hemorrhage of these aneurysms is usually unexpected and may sometimes represent a more serious complication. There are 2 major distinct pathologic mechanisms explaining this condition: 1) vessel wall replaced by neurofibromatosis tissue such as schwannoma, neurofibrom, or neurofibrosarcoma; or 2) aneurysmal formation caused by vascular dysplasia of small vessels with disarrangement of the smooth muscle and fragmentation of the elastic layer.\(^2,4,5,9\)

Spontaneous hemothorax caused by a ruptured intrathoracic artery aneurysm may manifest with a variety of symptoms. For example, rupture into the pulmonary parenchyma may cause hemoptysis,\(^10\) or rupture into the mediastinum may mimic a dissecting aortic aneurysm accompanied by radiating back pain and subsequent shock.\(^11,12\) Our patient had dyspnea and chest pain. CT scan could only reveal massive spontaneous hemothorax and led to a suspicion of intrapulmonary bronchial artery or extrapulmonary intercostal artery aneurysm rupture due to a history of congenital disease. Definitive diagnosis should be confirmed by selective angiography to facilitate therapeutic embolization with steel coils.

Our literature review found 24 reported cases of NF-1 with spontaneous hemothorax,\(^2,4,5,7,13-17\) including 6 vascular schwannomas and 18 ruptured intrathoracic artery aneurysms. Of these aneurysms, the subclavian or intercostal arteries were the most common to rupture. The general guidance for surgical intervention is chest tube output of more than 200 mL/h for 4 hours.\(^16\) However, surgery may sometimes fail. In our patient, surgery was terminated because there was no evidence of pulsatile hemorrhage and definite bleeding points could not be identified. Moreover, the patient began to develop signs of metabolic acidosis. However, in the previously reported cases identified in our review, surgical intervention was still the option in 14 of 18 cases, with 9 cases demonstrating an uneventful recovery. The remaining 6 patients were treated by a non-operative management including hemostasis and endovascular embolization with a satisfactory outcome.

Angiography with endovascular coil embolization should be the preferred method in patients with NF-1, not only because it precisely demonstrates a wide range of vascular malformations but also because it obviates the need for surgery.\(^5,7,13\) Other imaging studies, including Doppler echocardiography, CT angiography or even magnetic resonance angiography were much less sensitive to detect the presence of arterial aneurysms. These methods can be used for a non-invasive screening, if clinically necessary.\(^3,13\) To control bleeding in this emergent condition with a possible aneurysm rupture, as in our patient, we advocate percutaneous angiography prior to surgical intervention.

In cases of NF-1 with massive spontaneous hemothorax due to a ruptured intercostal artery aneurysm, early recognition of the potentially fatal but curable complication of von Recklinghausen’s disease is essential. Appropriate treatment is paramount for the patient’s survival. The utilization of the endovascular techniques appears to be the first choice in managing spontaneous hemothorax, especially when the patient has a history of von Recklinghausen’s disease.

**References**