Spontaneous Hemothorax Associated with von Recklinghausen’s Disease: Report of a Case

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A 59-year-old woman with a history of von Recklinghausen’s disease (VRD) suffered sudden chest pain. Enhanced chest computed tomography showed massive hemothorax, but no evidence of tumors or an obvious bleeding point in the thorax. After we had ensured a stable hemodynamic condition, we performed video-assisted thoracic surgery to remove the hematoma. The bleeding point was in a branch of the right subclavian artery. We performed direct surgical ligation of the bleeding vessel with a fibrin tissue-adhesive collagen fleece. Recovery was uneventful, and the patient is now doing well with no evidence of re-bleeding, 12 months postoperatively. Spontaneous hemothorax in patients with von Recklinghausen’s disease represents a critical event, and exploratory video-assisted thoracic surgery appears useful in the removal of clotted blood and reinforcement of fragile arteries for the prevention of re-bleeding. We should recognize this rare and critical condition in patients with von Recklinghausen’s disease.

Key words: von Recklinghausen’s disease, spontaneous hemothorax, video-assisted thoracic surgery

Introduction

With the exception of ruptures of the aortic aneurysm or association of pleural adhesions in the pneumothorax, spontaneous hemothorax is a rare event, though sometimes life threatening. Von Recklinghausen’s disease (VRD) is one of the causes of hemothorax. An autosomal-dominant disorder, VRD is characterized by multiple skin tumors and abnormal cutaneous pigmentation. Different associated complications have been described such as musculoskeletal anomalies and neoplasms (schwannoma, meningioma, etc.). Vascular lesions associated with VRD are rare, but can be fatal. Such lesions are characterized by stenosis, occlusion, aneurysm, pseudoaneurysm, and rupture or fistula formation in small, medium, and large arteries.

We report herein a case of critical massive intra-thoracic bleeding in a patient with VRD caused by rupture of a branch of the right subclavian artery.

Case

A 59-year-old woman with VRD developed sudden right chest pain and came to the emergency service of a local hospital at midnight. She had a history of pulmonary emphysema due to smoking, but no family history of VRD. She began to experience progressive dyspnea after presenting to the emergency services. Chest radiography showed massive pleural effusion in the right thorax, and midline shift of mediastinal structures to the left. A chest tube was immediately inserted, and
approximately 1000 ml of blood was removed (Fig. 1). She suffered progressive hypotension despite administration of intravenous fluids and dopamine support and was, therefore, urgently transferred to Nagasaki University Hospital for further examination and surgical evaluation. Physical examination showed multiple neurofibromatosis associated with café au lait spots and decreased respiratory sounds in the right thorax, with signs of hypovolemia (blood pressure 80/60 mmHg.; heart rate, 110–120 beats / min, and regular) and laboratory investigations showed anemia (hemoglobin, = 8.7 g/dl). Enhanced chest computed tomography showed massive hemothorax, but no evidence of tumors or any obvious bleeding point into the right thorax (Fig. 2). The patient was transfused with 4 packs of red blood cells and the chest tube was clamped.

We evaluated that she was in a stable condition and checked that progressive anemia was stopped after blood transfusion. The following day (12 h after admission), video-assisted thoracic surgery (VATS) was performed to remove the hematoma. The clot was carefully removed from the right thoracic cavity through a mini-thoracotomy, and continuous bleeding was found from a tortuous and dilated branch of the right subclavian artery (Fig. 3). This artery was found to be fragile, so direct surgical ligation of the bleeding vessels was performed along with the use of a fibrin tissue-adhesive collagen fleece to reinforce the vessel wall (Fig. 4). The total amount of bleeding was approximately 1600 ml.

The patient was discharged on post-operative day 7 in good condition. As of 12 months postoperatively, she is doing well with no evidence of spontaneous re-bleeding.

**Discussion**

The present report showed critical right hemothorax caused by rupture of a branch of subclavian artery in a patient with VRD.

Vascular lesions in VRD are rare, but can prove fatal.
The incidence of such lesions has been reported as 3.6%. The two main pathogenic mechanisms have been advocated for vasculopathy associated with VRD: a) direct vascular invasion from adjacent tumors such as schwannoma, neurofibroma, or neurofibrosacoma, and b) vascular dysplasia with thickened and concomitant reduced strength of the vessel wall and aneurysm formation. In the thoracic cavity, ruptures of intercostal, subclavian, and internal mammary artery aneurysms have been reported in patients with VRD. In the present case, definitive pathological and radiological findings could not be obtained, and the etiology of the spontaneous hemothorax remains unclear. However, we found a rupture of a tortuous and dilated artery. Therefore, we presume that vasculopathy might have existed. Given this possibility, we performed direct surgical ligation of the bleeding vessels and used a fibrin tissue-adhesive collagen fleece to reinforce the vessel wall. Pledged sutures might have been useful for the vascular wall.

Miura et al. reviewed 23 cases of spontaneous hemothorax associated with VRD. Patient age ranged from 17 to 63 years (mean, 42.6 years). Most ruptured arteries were intercostal arteries, followed by subclavian arteries. The source of bleeding was tumor-related rupture in 7 cases, aneurysmal rupture in 5 cases and other vascular lesions in 8 cases. Overall mortality rate was 30.4%, and 15 patients underwent emergency thoracotomy. Five patients were treated conservatively, with 2 cases treated using chest tube drainage only. The remaining 3 patients were treated by endovascular embolization for aneurysm. Diagnosis was only achieved at autopsy for 3 patients.

When spontaneous hemothorax is encountered in patients with VRD, some options are available for medical treatment and timing of the operation. In cases showing a hemodynamically unstable condition, emergent and aggressive surgical treatment is widely recommended. VATS is believed to be the best available modality for the management of clotted hemothorax. If the patient is hemodynamically stable, a more conservative approach with endovascular embolization or nonoperative management might be suitable. Endovascular treatment is a favorable, diagnostic and therapeutic option. Sakamoto et al. reported a successful endovascular stenting and coil embolization for ruptured pseudoaneurysm associated with VRD. In the present case, because the patient admitted in midnight and we ensured that she was in a stable hemodynamic condition after blood transfusion, we performed a sub-emergency operation. As a result, we were able to remove a massive hematoma sometimes causing dyspnea, and reinforced the fragile artery to prevent re-bleeding. However, even if the condition of patient appears stable, we now consider that earlier timing of operation might be favorable in order to avoid the risk of re-bleeding as fatal sudden re-bleeding has been reported 5 days after chest tube drainage. In addition, retained hemothorax can lead to infection, chronic fibrothorax, lung entrapment, and impaired pulmonary function. Therefore, we recommend earlier indication of exploratory VATS procedure or endovascular procedure for spontaneous hemothorax associated with VRD.

In conclusion, spontaneous hemothorax in patients with VRD is a critical situation that may need emergent surgical or interventional treatment. We recommended earlier indication of exploratory VATS to avoid re-bleeding and remove clotted blood even if the condition of the patient appears to be stable.

References