Spontaneous Hemothorax in Patients with von Recklinghausen’s Disease

Spontaneous massive intrathoracic bleeding is rare except for the rupture of aortic aneurysm or pleural adhesions in association with pneumothorax. We encountered two cases of critical massive hemothorax in patients with von Recklinghausen’s disease (type I neurofibromatosis). Case 1; a 59-year-old female suddenly experienced severe back pain followed by syncope and shock. The hemothorax was caused by a bleeding of diffuse type neurofibroma of the parietal pleura and she underwent thoracotomy and surgical ligation of the bleeding vessels. Case 2; a 46-year-old male suddenly suffered back pain and fainted while driving. An intercostal aneurysmal rupture caused a spontaneous hemothorax and he underwent chest tube drainage followed by endovascular coil embolization. We reviewed 23 cases reported in the literature, including our two cases. Spontaneous hemothorax in patients with von Recklinghausen’s disease is a life-threatening syndrome and may require emergency surgical or endovascular embolization. (Jpn J Thorac Cardiovasc Surg 2005; 53: 649–652)

Key words: von Recklinghausen’s disease, type I neurofibromatosis, spontaneous hemothorax

Takashi Miura, MD, Yozo Kawano, MD, Masao Chajo, MD, Michiyio Miyawaki, MD, Hiromu Mori, MD,* and Katsunobu Kawahara, MD.

Spontaneous massive intrathoracic bleeding is rare except for the rupture of aortic aneurysm or pleural adhesions in association with pneumothorax. We report two cases of critical massive intrathoracic bleeding caused by von Recklinghausen’s disease (VRD) (type I neurofibromatosis).

Cases

Case 1. A 59-year-old female with VRD suddenly suffered severe back pain followed by syncope. She was immediately referred to a local hospital while in a state of shock. She had an arterial pressure of 60/30 mmHg with severe anemia and was found to have a left hemothorax. Chest roentgenography showed complete opacification of the left hemithorax and a mediastinal shift to the right (Fig. 1A). Computed tomography showed a massive hemothorax, a partial thickening of the pleura and a depression of the anterior chest wall with costal defects (Fig. 1B). A dissecting aneurysmal rupture was considered likely, and she was immediately transferred to our institution for surgical treatment. On arrival at our hospital, she had received orotracheal intubation and oxygenation therapy. Her systolic blood pressure had fallen to 40 mmHg and she was unconscious. Transesophageal echocardiogram revealed no evidence of a dissecting aneurysmal rupture, and we started blood transfusion and performed an emergency thoracotomy. Massive amounts of blood flooded out from the pleural space and her blood pressure resumed to normal range. Approximately 3,000 ml of fluid and clotted blood were removed. The source of bleeding was an erosion of the fifth intercostal vessels involved by a flat and friable soft tumor, extending to the parietal pleura and atrophic intercostal muscle of the anterior chest wall. We performed ligation of the eroded intercostal vessels and partial excision of the tumor, and hemostasis was successfully achieved. Total blood loss was 7,680 ml. The associated chest wall defect was repaired with Marlex mesh sheet. The patient required re-thoracotomy for removal of the pleural hematoma on the 1st postoperative day. Her postoperative course was uneventful and she was discharged 50 days after surgery.
She has been asymptomatic for the past 10 years. Histological findings of the resected tumor showed short spindle shaped tumor cells proliferating and involving subpleural fat tissue and intercostal muscles in H.E. stain. Focally, the arterial wall was involved with tumor cells and Wegner-Meissner bodies were also recognized. Tumor cells were also stained with neuron specific enolase (NSE) and S-100 protein and the diagnosis was diffuse neurofibroma.

**Case 2.** A 46-year-old male suddenly suffered back pain and fainted while driving a car. He was transferred to an emergency hospital by ambulance. On physical examination, there was no definite injury or focal neurological deficit, but he manifested dermal tumors and café-au-lait-spots. He was not in a state of shock and on admission laboratory data revealed no anemia. Computed tomography showed a right hemothorax and a low density mass at the thoracic paravertebral region. Under the suspected diagnosis of hemothorax caused by spinal tumor, he was referred to our institution for further examination and treatment. Chest tube drainage was immediately instituted after admission and approximately 1,200 ml of bloody fluid was removed, and there was no further significant bloody discharge. Enhanced computed tomography revealed only a meningocele and a flat soft-density mass at the thoracic paravertebral region. As the cause of bleeding was suspected to be due to a vascular lesion associated with VRD, a digital angiogram was performed on the 16th hospital day. The findings revealed a 6 mm aneurysm of the right sided 7th intercostal artery, but no definite extravasation (Fig. 2A). His condition was complicated by empyema, and so he received chest tube irrigation and intravenous antibiotic therapy. After his recovery from empyema,