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Intercostal Aneurysm Rupture and Massive Hemothorax associated with Neurofibromatosis Type 1

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Abstract

Neurofibromatosis type 1 involving blood vessels is rare but potentially fatal. We report a case of spontaneous rupture of a left intercostal aneurysm. The patient presented with massive left hemothorax and was successfully treated by arterial embolization. The patient had a family history of neurofibromatosis type 1.

Keywords: Neurofibromatosis; Spontaneous hemothorax; Intercostal aneurysm

Introduction

Neurofibromatosis type 1, a multi-system disease, was first described by Friedrich Daniel von Recklinghausen in 1882. It is an autosomal dominant disorder caused by abnormalities in the long arms of chromosome 17.1,2 The disease can be diagnosed by a variety of clinical criteria, including:café-au-lait spots,plexiform neurofibroma,Lisch nodules,fleckling abnormal bony lesions. The disease often involves multiple organs, mainly the central nervous system. However, vascular involvement is rare.2 Here we present a case of massive left hemothorax caused by spontaneous rupture of a left intercostal aneurysm associated with neurofibromatosis type 1.

Case presentation:

A 37-year-old woman with a past medical history of neurofibromatosis type 1 presented to the emergency department with a chief complaint of worsening dyspnea and chest pain over the prior ten days. Her body temperature of 37.2 °C, blood pressure 110/80mmHg, heart rate 90 beats/minute, respiratory rate 25 breathes/minute, and oxygen saturation was 98%. A chest examination showed decreased breathing sounds at the left lung, the trachea had shifted to the right. Her hematocrit value and hemoglobin levels were 24% and 8.5g/dl, respectively. The chest computed tomography angiography revealed mediastinum shift to the right and a large amount of left pleural effusion with a severe collapse of the left lung.Vascular structures were normal, with no evidence of aortic dissection, aneurysm, or contrast extravasation.

A diagnostic pleural puncture revealed bloody effusion, which confirmed the diagnosis of hemothorax. Video-assisted thoracoscopic exploration was conducted, 2000 ml clotted blood was evacuated using a sucker, after which we could identify an 8*12 cm posterior mediastinal mass adjacent to the descending aorta. No active bleeding was found. A chest drainage was placed in the left pleural cavity after exploration.

The patient was then transferred to the Interventional Radiology Department, where selective arteriography to the left intercostal artery was performed. It showed the presence of a 1*2 cm aneurysm in the fifth intercostal artery, which was selectively catheterized and embolized with coils, with satisfactory results.

The patient's postoperative course was uneventful. The chest tube was removed on the seventh postoperative day and the patient was discharged on the eighth postoperative day without complication. The patient was followed without recurrent hemothorax for 6 month.

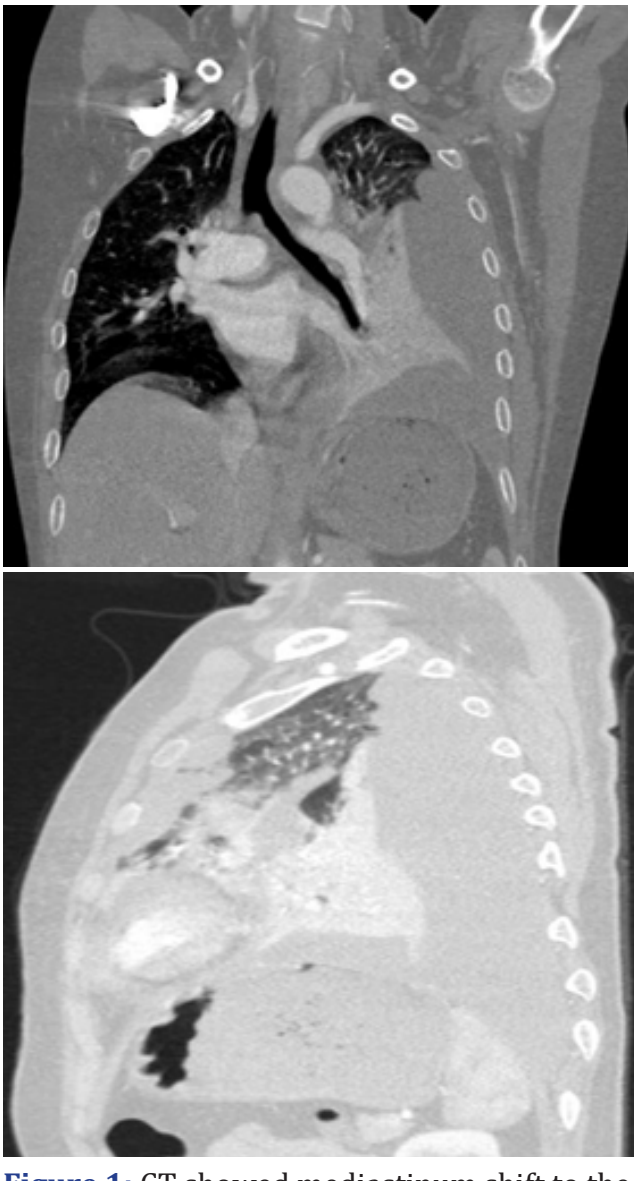

Figure 1: CT showed mediastinum shift to the right and a massive left hemothorax with a severe collapse of the left lung.

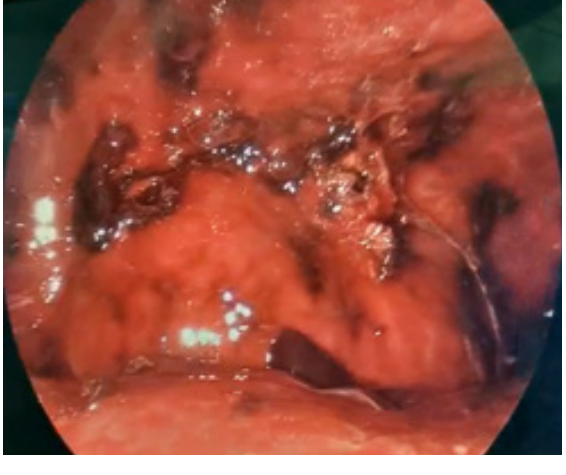

Figure 2: Thoracoscopic exploration identified an 8*12 cm pulsatile mass located under the pleura of the posterior chest wall.

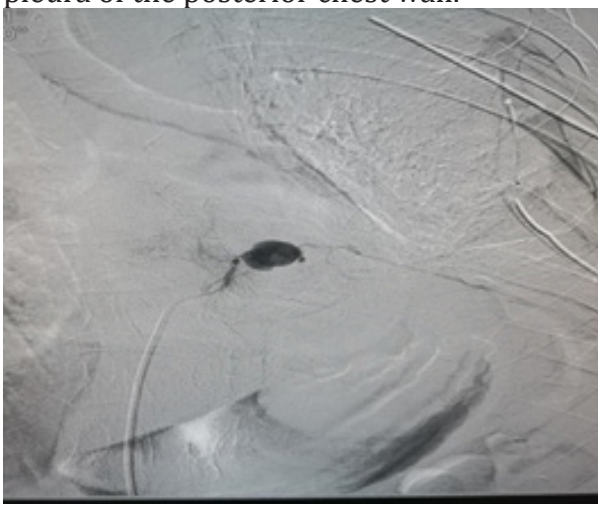

Figure 3: Selective arteriography to the left intercostal artery showed the presence of an aneurysm in the fifth intercostal artery.

Discussion:

The vascular injury caused by neurofibromatosis type 1 may involve the intercostal artery, internal thoracic artery, vertebral artery, most of which are manifested as arterial stenosis, occlusion, aneurysm, arterial rupture, and arteriovenous fistula, and the incidence rate is about 3.6%.1,2 In the present case chest computed tomography angiography didn't show any evidence of aneurysm, or active contrast extravasation. Video-assisted thoracoscopic exploration identified an 8*12 cm posterior mediastinal mass, which was obscured by the large hematoma. Intercostal aneurysm rupture was then suspected.

Because of the fragility of vascular tissue, thoracoscopy or thoracotomy to treat the disease is always difficult.3 The rapid development of endovascular treatment in recent years makes its risk far lower than that of thoracoscopy or thoracotomy.3,4 There are many successful cases of intercostal aneurysm embolization, and we believe that this technique is feasible and efficient.

In summary, spontaneous massive hemothorax from neurofibromatosis type 1 is uncommon but potentially fatal. The treatment of vascular embolization should have a broad application prospect.

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