



Case Report

Acute Fatal Chest Pain: Spontaneous Massive Hemothorax in a Patient with Neurofibromatosis Type I

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SUMMARY

Neurofibromatosis type 1 (NF-1) is an autosomal dominant disease characterized by multiple dermatological disorders. The less frequent manifestations are vascular abnormalities. Spontaneous rupture of these pathological vessels is a rare but potentially life-threatening complication; this necessitates immediate and decisive intervention. Unstable patients should be managed with aggressive surgical intervention as soon as possible to avoid mortality. We encountered a case of massive hemothorax in a 62-year-old female with neurofibromatosis type 1 as a result of spontaneous rupture of right intercostal artery, which leads to lethal outcome despite extensive surgical intervention as well as intensive care measures.

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1. Introduction

NF-1 is a genetic disorder with a rare incidence rate of approximately 1 in 3000.¹ Arterial stenosis due to connective tissue dysplasia is a well-known vascular complication of NF-1, as well as aneurysms, which constitutes 3.6% of people diagnosed with this disease.^{2,8} Spontaneous hemothorax is one of the rare complications of NF-1. It is potentially fatal with a mortality rate as high as 36% and postoperative mortality of 33%.³ Massive and potentially fatal hemothorax caused by vascular complications is extremely uncommon. We present a rare case of massive hemothorax in a 62-year-old woman with NF-1 due to a spontaneous rupture of the right intercostal artery.

2. Case report

A 62-year-old woman with NF-1 presented to the emergency department with acute right-sided chest pain and dyspnea 30 minutes before admission. Her vital signs at triage showed blood pressure of 162/119 mmHg, the breathing rate of 22 times/min, heart rate of 52 beats/min, oxygen saturation of 59% at room air, and temperature of 37.2 °C. The patient denied any history of trauma. Physical examination revealed multiple neurofibromas over her skin and hypophonesis of her right chest. During her physical examination, the patient suddenly collapsed. The patient achieved returned of spontaneous circulation after minutes of cardiopulmonary cerebral resuscitation (CPCR).

Massive pleural effusion of the right chest was immediately detected by bedside ultrasound. Hemothorax was diagnosed with thoracentesis. An intercostal drainage tube was inserted, and

approximately 2500 ml of blood was removed. The patient received a total of 5000 ml of normal saline and six units of leukocyte-poor red blood cell. Chest X-ray showed massive right-sided pleural effusion (Figure 1). Her blood pressure dropped to 48/37 mmHg, and her hemoglobin was 5.6 g/dl, consistent with the impression of massive hemothorax causing hemorrhagic shock. Computed tomography (CT) confirmed a massive hemothorax with extravasation at the right intercostal space (Figure 2), and spontaneous intercostal artery rupture with tension hemothorax. An emergent thoracotomy was performed to determine the source of the bleeding.

Surgical exploration demonstrated 3800 ml bloody effusion over the right pleural cavity and active hemorrhage from the paravertebral region near the origin of the right intercostal artery. We identified the bleeding site and performed hemostasis with suture and hemoclip. Postoperatively, the patient was transferred to our intensive care unit with a clamped thoracostomy tube. The clamped

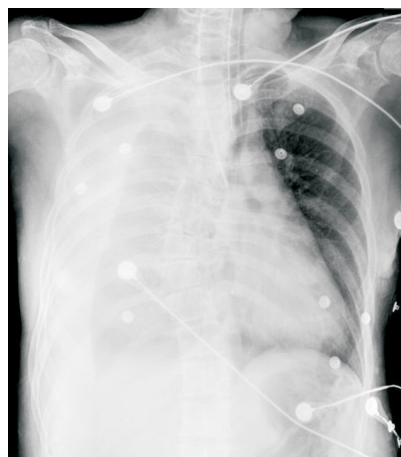


Figure 1. Chest X-ray showing massive right-sided pleural effusion.

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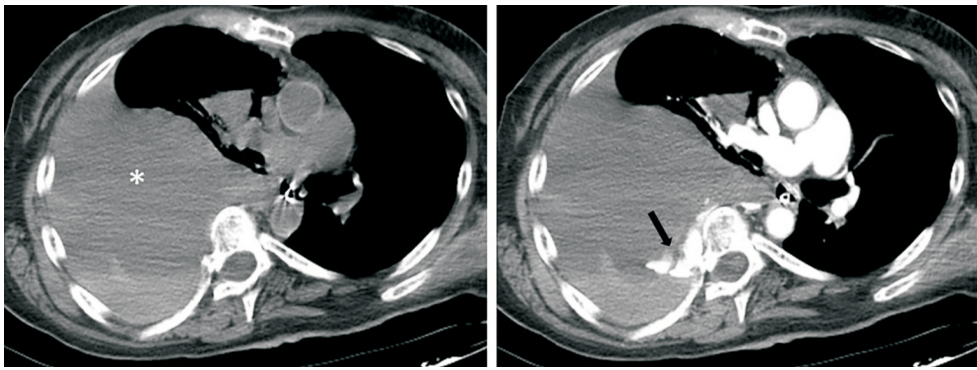


Figure 2. Left panel: CT without contrast showing massive right-sided hemothorax (asterisk). Right panel: CT with contrast showing extravasation noted at the right paravertebral region near the intercostal space (arrow).

chest tube was temporarily for massive bleeding after discussing it with the thoracic surgeon. After clamping, we closely monitored the patient's vital signs and arranged chest X-rays to reassess the patient's hemothorax condition. The patient expired on the second day of admission despite hemostasis with suture and massive transfusions for refractory severe hypovolemic shock.

3. Discussion

NF-1 is an autosomal dominant disorder linked to chromosome 17. It affects approximately 1 in 3000 people and may involve connective tissue, nerve tissue, vessels and others. The reported culprit vessels of NF-1 are usually the abdominal aorta, or the brachial, intercostal, or renal arteries.^{4,5} There are two hypotheses of the etiology of a spontaneous hemothorax in patients with NF-1. One possible cause is the arterial invasion of neurofibromas.⁶ The other one is Schwann cell proliferation, which results in the weakening of the smooth muscle and connective tissue in the wall of the artery, leading to aneurysm formation or stenosis.⁷ Both events can induce a massive spontaneous hemothorax.

The treatment for vascular complications depends on the clinical condition, location, and type of lesion, the reported treatment options include open surgery, video-assisted thoracic surgery, and endovascular intervention including coil embolization or stent placement. In NF-1 surgical repair of bleeding vessels is complicated by arterial fragility due to intimal and medial dysplasia. Miura et al. reported exploratory thoracotomy success rate was 50%.⁸ Therefore, endovascular embolization of arterial lesion is generally considered to be the treatment of choice in a stable hemodynamic situation as it is less invasive and more effective than surgical intervention. Muhammad et al. reported coiled embolization success rate was 85%.⁹ Video-assisted thoracoscopy (VATS) may be an alternative for evacuating a hematoma, especially after controlling the bleeding.

However, since the patient in our case went directly into hemorrhagic shock and cardiac arrest, an unsatisfactory stabilization of the hemodynamic situation did not allow for angiography and VATS, we considered an immediate thoracotomy as the only viable

alternative. Miura et al. consider an emergent and aggressive surgical treatment to be indicated in patients in a hemodynamically unstable condition.⁸

In conclusion, NF-1 is generally regarded as a benign disease; however, it has a potentially life-threatening vascular complication. This presents a challenge in the diagnosis and management of acute chest pain. We emphasize the importance of considering fatal spontaneous hemothorax in a patient with NF-1 who presents with acute chest pain. Early and aggressive intervention with transarterial embolization or exploratory thoracotomy with excision and ligation are critical for mortality prevention.

Conflict of interest

None.

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