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Case Report

Spontaneous hemothorax in neurofibromatosis type 1: a rare case of intercostal artery aneurysm rupture and the role of permissive hypotension

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ABSTRACT

This case report presents the effective management of a rare and life-threatening spontaneous hemothorax resulting from the rupture of an intercostal artery aneurysm in a patient with undiagnosed neurofibromatosis type 1 (NF-1). The patient exhibited acute chest pain and hemodynamic instability, requiring a multidisciplinary strategy that included permissive hypotension and interventional radiology (IR). Coil embolization successfully managed the hemorrhage, resulting in hemodynamic stabilization and positive patient outcomes. This case highlights the significance of promptly identifying spontaneous hemothorax in NF-1, the necessity of targeted resuscitation strategies, and the effectiveness of minimally invasive endovascular techniques in addressing vascular complications. This case study highlights the complexities involved in addressing an atypical presentation of an undiagnosed case of NF-1, which was further complicated by a sudden spontaneous hemothorax resulting from the abrupt rupture of an intercostal artery aneurysm. This underscores the essential importance of promptly identifying life-threatening bleeding and the necessity for a resuscitation strategy that is customized to the individual patient. Additionally, it highlights the benefits of employing a permissive hypotension protocol in stabilizing the patient's hemodynamics and preventing exacerbation of bleeding. The conclusive management utilizing coil embolization through IR effectively accomplished hemostasis. This paper aims to demonstrate that an emergency medicine physician must consider uncommon differential diagnoses when faced with a patient presenting with chest pain. It is crucial to maintain a high level of suspicion for rare, life-threatening conditions, including neurofibromatosis.

Keywords: Hemothorax, Neurofibromatosis, NF-1, Intercostal artery aneurysm, Coil embolization, Permissive hypotension, Chest pain

INTRODUCTION

Hemothorax presents a significant challenge in the emergency department, frequently necessitating a specific management strategy due to the variability in its causes and clinical manifestations. While it is a common occurrence, spontaneous hemothorax is significantly less frequent, with limited data present in case reports and case series.[1] Hemothoraces are categorized into three main types based on their cause and frequency: traumatic, iatrogenic, or non-traumatic.[2] Non-traumatic events most commonly arise as complications associated with certain cancer types when tumors invade pleural space. Conditions such as coagulopathy (including hemophilia and Glanzmann thrombasthenia), anticoagulation therapies, vascular issues (like arteriovenous malformations and aneurysms), NF-1 and neoplasms such as angiosarcoma and schwannoma, and lung cancer, are notable contributors to this phenomenon.[3]

NF-1 is recognized for its association with spontaneous hemothorax. This condition is characterized by an autosomal dominant inheritance pattern, with a prevalence of 1 in 3,000.[4] The condition exhibits a

comparable prevalence across different races and genders, typically being diagnosed in adulthood as its clinical features evolve over time. It is a multisystem disease capable of impacting any organ system, particularly connective, nervous, and vascular tissues, leading to potential manifestations such as abnormal skin pigmentation and/or skin tumors.

The pathogenesis of vasculopathy in patients with neurofibromatosis remains topic of ongoing investigation. It may occur either through direct invasion from adjacent tumors, including schwannomas, neurofibromas, or neurofibrosarcomas or as a result of vascular dysplasia characterized by thickening and a concurrent reduction in the strength of the vessel wall leading to aneurysm formation.[5,6] The rupture of fragile blood vessels or aneurysmal dilation within the thoracic cavity may result in hemothorax/potentially hemomediastinum. Patients may present with varying degrees of chest pain, ranging from mild to severe, influenced by hemodynamic changes related to the extent of bleeding and individual comorbidities and their medical history.

In research conducted by Wackers to investigate the causes of chest pain in the emergency department and the usefulness of cardiac imaging, it was revealed that pulmonary patients accounted for 2% of the total cases. Respiratory reasons were identified as the predominant underlying pathology for chest pain presentations in the emergency room. Pleural effusion, involving hemothorax, constituted 1.91% of cases.[7]

This case report emphasizes the difficulties encountered in diagnosing an unusual and rare manifestation of NF-1, which is further complicated by presence of an intercostal artery aneurysm and hemothorax. The management of patient underscored effective application of permissive hypotension to prevent worsening of bleeding, as well as the contribution of IR in attaining definitive control of the hemorrhage via coil embolization.

CASE REPORT

A 44-year-old male with no previous medical history arrived at the emergency department (ED) experiencing acute, severe, dull, aching chest pain on the left side that radiated to the back. The discomfort intensified while lying supine and during deep inhalations, accompanied by a sensation of breathlessness, yet there was an absence of fever or cough. During the physical examination, a reduction in air entry was observed at the left lung base. Bedside ultrasound ruled out pneumothorax, while the chest X-ray indicated consolidation in the left lower zone, with a potential pleural effusion (Figure 1). Preliminary laboratory findings indicate hemoglobin at 14.1 g/L, hematocrit at 0.408 L/L, alongside negative results for troponin and D-dimer. The patient, exhibiting stable vital signs (BP 126/60 mm Hg, pulse 74 bpm, RR 18 breaths/min, and O_2 saturation 98%), was subsequently transferred to another facility as a result of bed shortages.

Upon arrival at the second facility, the patient exhibited mild tachycardia (HR 102 bpm), while his blood pressure remained stable (112/78 mm Hg) and he showed no signs of respiratory distress. A follow-up CT (Figure 2) scan of the chest was conducted, showing widespread pleural thickening and nodularity in the left hemithorax, extending along both the lateral and mediastinal pleura. This strongly indicated the presence of a primary pleural neoplasm, probably mesothelioma. Furthermore, a small pleural effusion was noted, along with volume loss in the left lung, which resulted in the secondary elevation of the left hemidiaphragm. Additionally, the absence of clear fat planes between the pleural mass and adjacent structures raises concerns regarding potential invasion. A follow-up contrast-enhanced CT scan revealed a significant left-sided pleural effusion with high density, indicative of hemothorax (Figure 3). Additionally, there was atelectasis observed in the left lower lobe and basal segments of the upper lobe, alongside multiple mass-like lesions present in the intercostal spaces and surrounding soft tissues. An aneurysm measuring 15×10 mm was identified, originating from the left intercostal artery situated between the 10^{th} and 11^{th} ribs (Figure 4). The results align closely with complicated NF-1 (NF1) associated with secondary aneurysm formation and hemothorax. The patient was transferred for urgent intervention due to a significant decrease in hemoglobin levels, dropping from 14 to 10.4 g/dL. During transfer, the patient's condition quickly worsened, presenting with intense chest pain, sweating, rapid breathing, and increased heart rate. Upon arrival at the emergency department in our facility, he presented in shock (HR 160 bpm, BP 64/45 mm Hg, RR 25 breaths/min), exhibiting significant respiratory distress and dizziness. Immediate resuscitation involved the administration of IV fluids, the transfusion of 2 units of O-negative packed red blood cells, and the initiation of norepinephrine infusion. In light of the patient's critical condition, permissive hypotension was utilized to achieve a mean arterial pressure (MAP) exceeding 65 mm Hg. This resulted in the stabilization of his blood pressure (BP 85/65 mm Hg) and an improvement in his heart rate (reduced to 144 bpm), along with a significant reduction in his respiratory distress and diaphoresis.

The individual was transported to the catheterization lab, where sedation was administered, intubation was performed, and an extra 2 units of packed RBCs were provided. Access was achieved through the right femoral artery, followed by selective catheterization of the thoracic aorta and the left 10th intercostal artery. Contrast injection demonstrated ongoing hemorrhage from the proximal region of the aneurysm. Several coils were utilized to occlude the feeding artery to the aneurysm, resulting in effective hemostasis (Figure 5). After adhering to the established protocol, an intercostal drainage (ICD) tube was placed. chest X-rays revealed Surgical sutures are along the left chest. Left increased bronchovascular markings. ICD in situ (Figure 6), and the patient was subsequently moved to the ICU for

enhanced observation. The patient maintained hemodynamic stability and, following 24 hours of observation, was moved to the ward. Patient condition improved, follow up x rays reveald improving in his general condition. the patient was released in satisfactory condition with a stable hemoglobin level of 11 g/dL.

Five months later, the patient returned to the hospital for a follow-up, reporting no chest pain or shortness of breath. Blood pressure measured at 125/70 mmHg, heart rate recorded at 81 beats per minute, and oxygen saturation at 99% in room air. Assessment reveals unobstructed air entry bilaterally with no additional sounds detected. The chest X-ray (Figure 7) performed reveals a blunt left costophrenic angle accompanied by haziness in the left lower zone, with no evidence of pneumothorax or hemothorax observed. Patient discharge for follow-up with outpatient clinic.

Figure 1: Inhomogeneous but a consolidation in the left lower lobe. Possible left basal effusion.

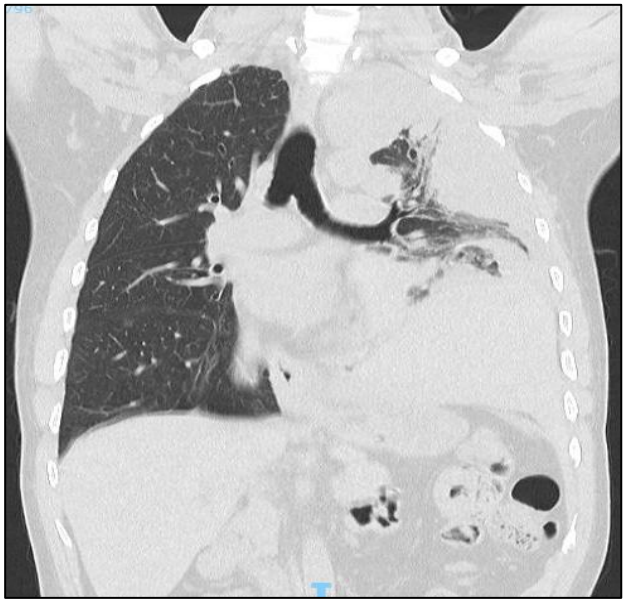

Figure 2: Plain CT of chest.

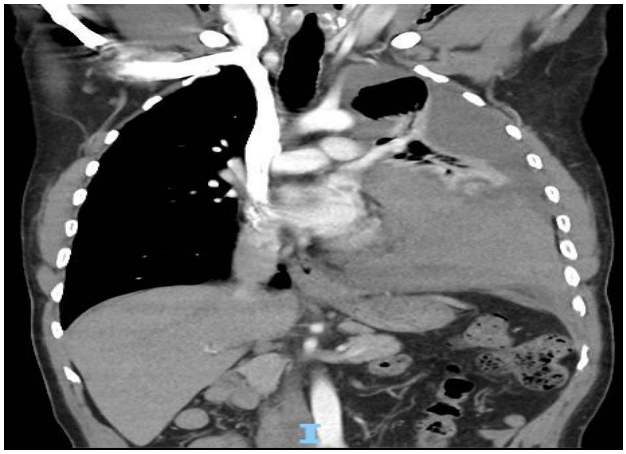

Figure 3: There is large amount of left-sided pleural effusion with high density in suggestive of haemothorax. There is atelectasis of left lower lobe and basal segments of left upper lobe.

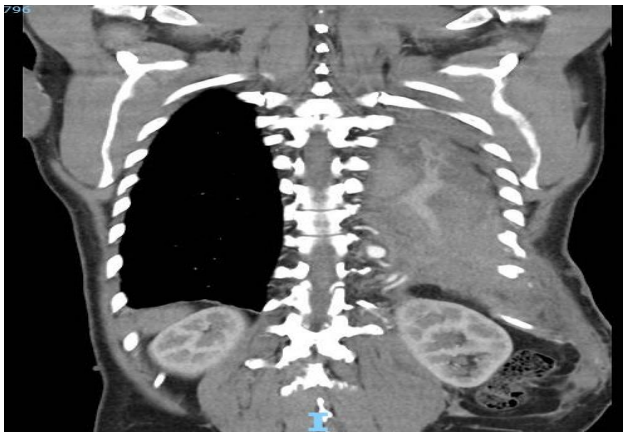

Figure 4: Aneurysm is seen arising from the left intercostal artery in between the 10th and 11th rib-measuring approximately 15×10 mm. Findings are most likely due to complicated neurofibromatosis with secondary Aneurysm formation (of left intercoastal artery) and left sided hemothorax.

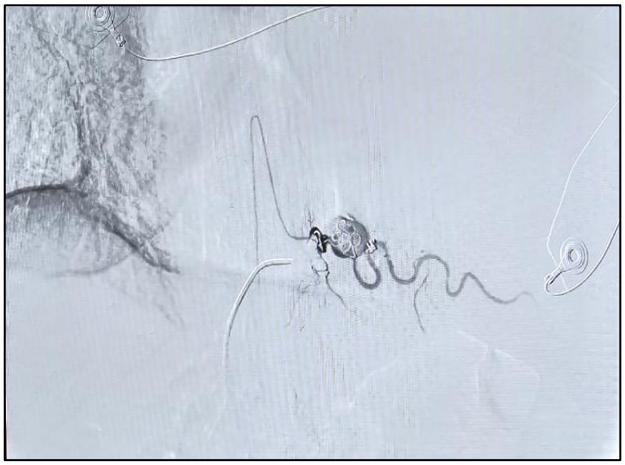

Figure 5: Several coils were utilized to occlude feeding artery to aneurysm, resulting in effective hemostasis.

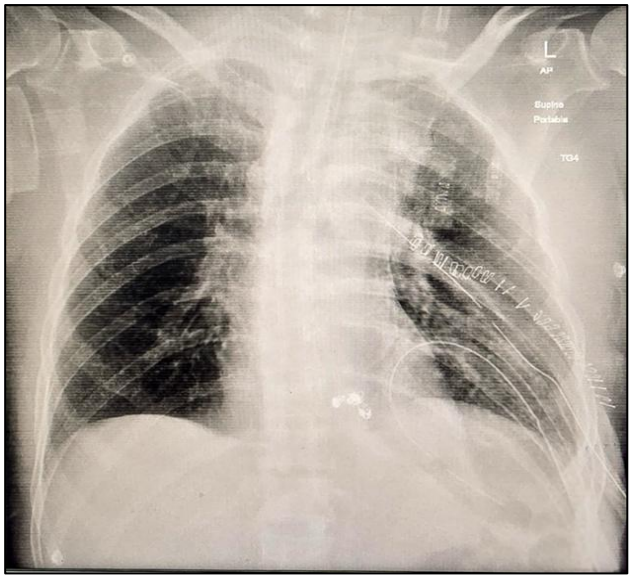

Figure 6: Post ICD surgical sutures are along the left chest. Left increased bronchovascular markings. ICD *in situ*.

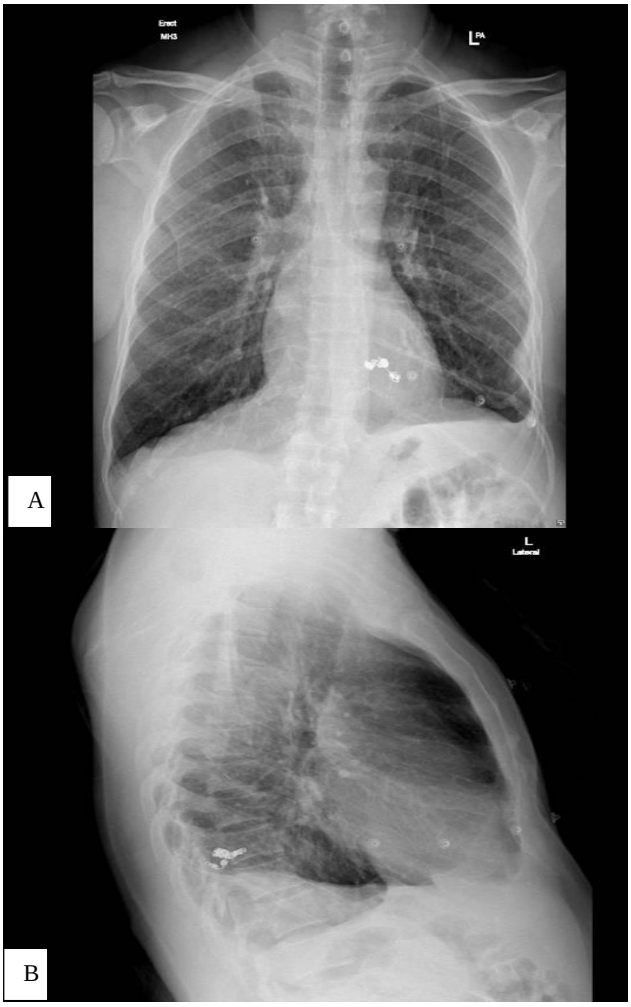

Figure 7 (A and B): Chest X-ray reveals a blunt left costophrenic angle accompanied by haziness in the left lower zone.

DISCUSSION

The challenges in the emergency department encompass not only clinical judgment and the early diagnosis of hemothorax as a primary cause of chest pain but also the identification of the underlying cause of hemothorax and the selection of the most effective management plan for patients, particularly those presenting with hemodynamic instability. The primary complication associated with hemorrhagic shock is the "lethal triad," which consists of acidosis, hypothermia, and coagulopathy. The lethal triad arises when tissues experience hypoxia and depend on anaerobic metabolism, resulting in the production of lactic acid. Acidosis and hypothermia both impede the clotting cascade, thereby reducing the body's capacity to coagulate and halt hemorrhage.[8]

In a patient presenting with hemorrhagic shock, two primary options are available for resuscitation. The initial objective is to attain complete resuscitation, ensuring a target systolic blood pressure exceeding 90 mmHg, alongside the implementation of a massive transfusion protocol and the use of vasopressors. Numerous patients exhibited side effects associated with fluid overload, including edema, hemodilution, and an inflammatory response, as well as metabolic and cellular alterations that may impact the pulmonary, cardiac, and coagulopathy systems.[9] The second option involves permissible hypotension or hypotensive resuscitation, wherein the mean arterial blood pressure is intentionally maintained below physiological levels. Permissive hypotension functions solely as a temporary strategy prior to the implementation of hemorrhage control or surgical intervention. Permissive hypotension may offer greater cost-effectiveness and enhanced treatment efficiency, leading to improved outcomes. This method is frequently utilized to manage acute hemorrhagic volume depletion resulting from significant trauma.

The impact of permissive hypotension on mortality reduction is extensively documented in numerous studies. The implementation of permissive hypotension results in a reduction of postoperative recovery time compared to the immediate normalization of an individual's blood pressure.[9] Permissive hypotension may be employed in the management of nontraumatic conditions such as leaking abdominal aortic aneurysm, dissecting aneurysm, pulmonary contusion, and bleeding duodenal ulcer. The procedure may also be utilized in patients undergoing neurovascular surgery.

Numerous sources concur that permissive hypotension can be attained in patients with a MAP of approximately 50 mm Hg or a systolic blood pressure (SBP) ranging from 80 to 90 mm Hg. Fluid infusions ranging from 100 to 200 mL may be administered to maintain the patient within acceptable pressure limits during evaluation.[9]

The management of patients with leaking aneurysms remains a subject of debate. Treatment options depend on

the hemodynamic stability of the patient. Endovascular embolization is warranted in cases of hemodynamic stability. Thoracotomy with surgical ligation is indicated as an alternative in cases of active bleeding accompanied by hemodynamic compromise. The prognosis of NF-1 in patients with haemothorax indicates a disease mortality rate of 36% and a postoperative mortality rate of 33%. Recent cases indicate that coil embolization yields optimal results.[10]

CONCLUSION

This case demonstrates the benefits of a multidisciplinary approach, incorporating emergency medicine, radiology, IR, cardiothoracic surgery, and intensive care, in achieving optimal outcomes for patients with rare and complex conditions.

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