

Idiopathic Spontaneous Rupture of an Intercostal Artery

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ABSTRACT

Spontaneous rupture of an intercostal artery is exceptionally rare without an inciting event such as trauma or nontraumatic arterial wall weakening. This report details the diagnosis and treatment of a 47-year-old man who presented with a spontaneous hemothorax from an intercostal artery. There are very few reports in the literature documenting spontaneous intercostal artery rupture without associated illness or injury.

INTRODUCTION

Spontaneous rupture of an intercostal artery is a rare but life-threatening surgical emergency requiring prompt diagnosis and intervention for optimum outcome. Spontaneous intercostal artery hemorrhage (ICAH) may be complicated by hemothorax, hematoma formation, and/or retroperitoneal bleeding, which contributes to significant morbidity and mortality in these patients. The majority of intercostal artery bleeds result from trauma; however, nontraumatic predispositions (ie, Ehlers danlos, Marfans disease, Neurofibromatosis Type I) that result in arterial wall weakening and aneurysmal formation also contribute.¹⁻³ What is exceptionally rare is an idiopathic spontaneous intercostal artery bleed with nontraumatic or traumatic inciting factors. We present a case of a 47-year-old white man with a spontaneous hemothorax. To our knowledge, this is the third report in the literature documenting spontaneous intercostal artery rupture without associated illness or injury.⁴

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CASE PRESENTATION

A 47-year-old man with no significant medical history arrived at an outside hospital with shortness of breath and severe right-sided chest pain several hours following sexual intercourse. He denied any history of trauma. A CT scan completed at the hospital revealed a right hemothorax and prompted transfer to our level I trauma center. The patient was hypotensive and tachycardic on arrival even with appropriate resuscitation during transfer and had an oxygen saturation fluctuating between 70% to 90%. Physical examination revealed decreased breath sounds on the right side and mild tenderness over his right flank.

While in the emergency department, the patient became acutely unresponsive and required immediate intubation. A right chest tube was placed and drained 750 cc of frank blood. He was transferred to the surgical intensive care unit (SICU) but continued to have frank bleeding from his chest tube, resulting in further hemodynamic instability. This prompted an emergency surgical exploration of his chest in the operating room. A right posterior-lateral thoracotomy was performed and a large hemothorax evacuated from the right thoracic cavity. At this point no active intrapleural bleeding was noted but a large extrapleural hematoma had formed, separating the pleura from the chest wall. Here was noted frank, active extrapleural bleeding from an intercostal artery close to the 11th rib just medial to the spine. A second incision was made over the 11th rib to access this bleeding vessel and subsequent control of bleeding was achieved with ligation.

The chest tube was replaced and both incisions were closed. The patient was taken back to the SICU. Four hours postoperatively, significant output was noted from the chest tube and the patient was brought back to the OR for re-exploration. The incision over the 11th rib was re-opened and active bleeding was noted, this time from an anterior intercostal artery remote from the site of the original bleed. The vessel was controlled with suture ligation and the incision closed. He did well overnight with no

further acute bleeding and was extubated on postoperative day (POD) 2. His chest tube was removed on POD 3; he was tolerating a normal diet and his pain was controlled on oral narcotics. He was discharged home on POD 4 and has had no further instances of intercostal bleeding on follow-up examination. He was referred as an outpatient to the cardiology clinic for evaluation for vascular dysfunction. Though he did not undergo genetic testing, he was evaluated by a cardiology specialist and deemed to have no evidence of any vasculitis or arterial dissection.

DISCUSSION

This report discusses the third reported case of a spontaneous intercostal bleed documented in the literature. Our patient suffered this spontaneous intercostal artery bleed with no known predisposing factors, significant medical history, or traumatic insult.

Spontaneous intercostal bleeding is a rare occurrence. When diagnosed, usually it is associated with underlying etiologies—namely trauma, anticoagulation or bleeding disorders, lung infections, and/or predisposing medical conditions such as Neurofibromatosis Type I (Von Recklinghausen's disease, NF Type I), Systemic Lupus Erythematosus (SLE), and/or uncontrolled hypertension.¹⁻⁵ Most data on intercostal bleeds are from case reports, the most common cause being trauma and most common nontraumatic cause Neurofibromatosis Type 1.¹⁻³

To our knowledge, there have been 6 case reports demonstrating spontaneous intercostal bleeds. Only 2 of these were associated with no predisposing medical conditions or probable etiologies, making our case the third.¹⁻⁷

Presenting symptoms noted in the literature are highly variable and may include abdominal pain, dyspnea, thoraco-abdominal masses, flank pain, and/or shoulder and back pain.¹⁻⁷ Deciphering the probable location of the culprit intercostal is aided by clinical exam. In the literature and in our case, the most common location of intercostal spontaneous rupture is the 10th or 11th intercostal vessel and the majority of these patients were not on anticoagulants.^{1,5,6}

Probable underlying etiologies in these cases included violent coughing, uncontrolled hypertension, and SLE or NF1.¹⁻⁷ In this case, the patient presented several hours following sexual intercourse, but denied any trauma or violent coughing. Other than our case, the case reported by Matthew et al⁷ described a male patient presenting with dyspnea and left loin pain with a subsequent finding of abdominal wall hematoma and hemothorax due to 10th intercostal spontaneous, idiopathic bleed. Similar to our case, blood was discovered in the thoracic cavity with hematoma formation, and the resultant area of bleeding in relationship to the rib was also similar in pattern to our case.⁷

Moon et al⁵ described a case of a 45-year-old male patient who

ceased taking his hypertensive medication 5 months prior to presentation and suffered a spontaneous intercostal bleed.⁵ Although their patient had problematic coughing spells, that also may have been an etiological factor of the intercostal rupture such as in the case report by Lu et al,² they documented that the patient's intercostal artery rupture occurred prior to the onset of coughing, pointing to increases in blood pressure as an etiological factor.

On the nontraumatic side, Neurofibromatosis Type 1 (von Recklinghausen's disease) is the most common cause of intercostal rupture. This is an autosomal dominant condition that affects connective tissue and vasculature causing arterial stenosis due to intimal dysplasia of large vessels as well as development of numerous neurofibromas on the skin.¹⁻³ Other medical conditions also have been associated with the development of spontaneous hemothorax. Multiple hereditary exostoses, in which the patient experiences growth of multiple osteochondromas in childhood, has been reported to cause bleeding due to the protrusion of a lesion into the pleural space.⁸ Isolated hyperostosis of a single rib also has been shown to have caused spontaneous hemothorax following erosion of the rib through an intercostal artery.⁹ One report described hemothorax developing as the result of spontaneous rupture of a hydatid cyst, a concern in areas where sheep and cattle are raised.⁹

On physical exam of our patient, we found nothing suggestive of Neurofibromatosis, and following his recovery we referred him as an outpatient for evaluation for vasculitis. Although our patient did not undergo genetic testing, his past medical and family history were evaluated and his physical exam did not reveal any evidence of a vasculopathy. However, it is possible that he carries a missed diagnosis of a condition that may predispose him to spontaneous intercostal arterial rupture and resulting hemothorax. It is therefore incumbent upon the surgeon in such cases to consider this possibility and to be prepared for any potential problems during the operative procedure that may result from such disorders. Additionally, these findings merit further investigation once the patient is stabilized in order, if possible, to determine a cause for his hemothorax.

Overall, it is imperative that any clinician encountering new onset hematoma/retroperitoneal bleed with unknown and obscure etiology at least consider intercostal bleed as a differential, particularly in patients with predisposing conditions such as trauma or NF Type I. In addition, as evidenced by our report and others, when intercostal artery hemorrhage is suspected prompt intervention can be lifesaving.

CONCLUSION

Spontaneous rupture of intercostal arteries in the setting of no underlying etiology such as trauma, NF1, and/or anticoagulation is extremely rare. Surgeons and other clinicians observing

new onset hemothorax/hematoma formation, shortness of breath, and atypical symptomology that does not correlate with clinical history should consider intercostal hemorrhage and/or hematoma formation especially in the context of underlying disease mechanisms that may weaken the arterial wall. Close follow-up of patients who are diagnosed with spontaneous arterial bleeds must be conducted to rule out underlying etiology.

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