

A Repeat Case of Idiopathic Spontaneous Hemothorax

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Spontaneous hemothorax, a collection of blood in the pleural cavity in the absence of trauma, is a clinical entity usually associated with pleural malignancy. Sporadic cases of spontaneous hemothorax without identifiable underlying pathology have rarely been reported. This article discusses an unusual case of a woman who develops both a right and a left idiopathic spontaneous hemothorax within a 7-year period. A review of the medical literature is also presented.

CASE PRESENTATION

Initial Presentation

A 52-year-old woman presents to the emergency department with complaints of right chest pain and progressive shortness of breath that developed over the previous 8 hours. The patient does not smoke cigarettes and has no history of pre-existing lung disease, tumor, or hypertension. She is not taking aspirin or any other anticoagulants. Radiography and computed tomography (CT) of the chest reveal a right pleural effusion with no other evident pathology (**Figure 1**). Thoracentesis yields sanguinous fluid with an erythrocyte count of 1.8 million/mm³ and a pH of 7.4; results of cytology and culture are negative. Additional medical history includes a positive tuberculin skin test, three sputa-negative tests for acid-fast bacilli, a hepatitis screen positive for hepatitis A, and liver function tests with normal results. Surgical history includes open cholecystectomy with an excessive amount of bleeding reported in the operative note.

During the next 48 hours after presentation, the patient develops increasing shortness of breath and her hematocrit progressively falls from 34% to 22%. Results of hematologic work-up for coagulopathy are unremarkable. The patient is transfused with 3 units of packed erythrocytes and transferred to the operating room for exploratory thoracotomy.

During exploration, approximately 2 L of blood are evacuated from the pleural cavity. No pathology is identi-

fied after inspection of the lung, mediastinum, and pleura. Random pleural biopsies show reactive mesothelial cells. No further bleeding occurs postoperatively.

The patient returns for weekly follow-up for 6 weeks. Weekly chest radiography results and hematocrit levels remain normal, and the patient remains asymptomatic for 7 years.

Current Presentation

Seven years after surgery, the patient presents to her primary care physician with complaints of shortness of breath and left chest and shoulder pain. The patient is admitted to the hospital. Chest radiography reveals a left pleural effusion (**Figure 2**). Throughout hospital day one, the patient's shortness of breath progressively increases. Repeat chest radiography late on hospital day one demonstrates increasing effusion (**Figure 3**), and the patient's hematocrit falls from 31% to 25% 12 hours later. Results of hematologic work-up for coagulopathy are negative. The patient is transfused with 2 units of packed erythrocytes. A left chest tube is inserted, and 1600 mL of blood are evacuated from the left pleural cavity. The patient's symptoms resolve rapidly. No further bleeding occurs from the chest tube, and the patient's hematocrit is stable at 33%. On hospital day three, the chest tube is removed and the patient is discharged home. Results of follow-up chest radiography at 1 week, 3 weeks, and 3 months remain normal.

DISCUSSION

In 1938, Perry¹ compiled 21 cases of spontaneous hemothorax with pneumothorax. Nine of the 21 patients

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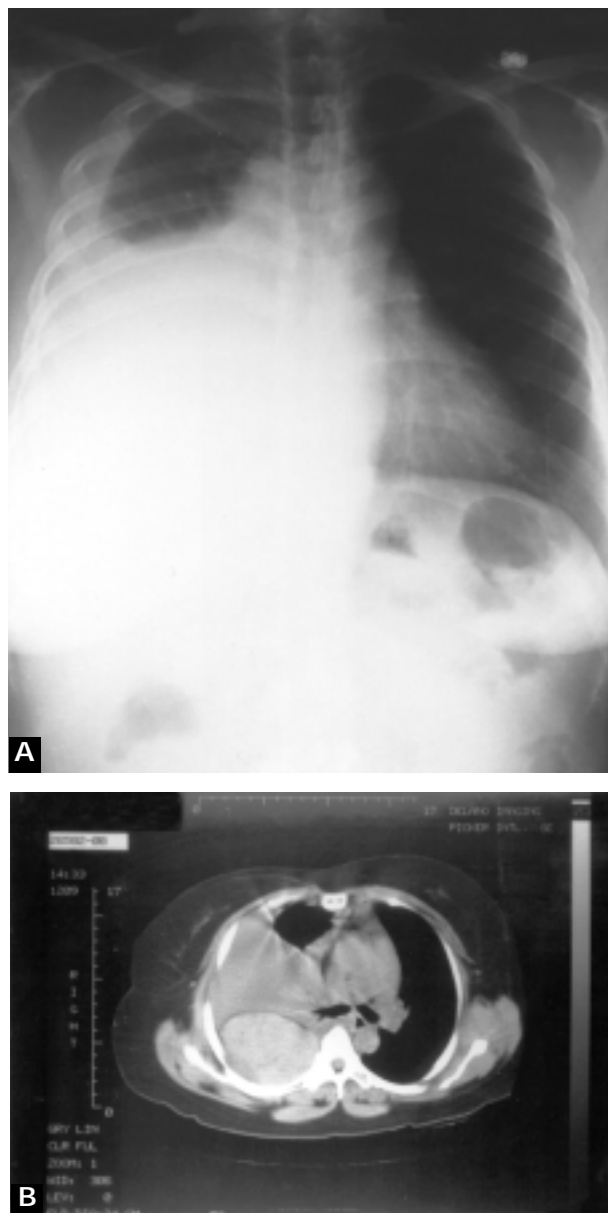


Figure 1. A) Chest radiography and B) computed tomography of a patient with right pleural effusion.

died; autopsies of these nine patients revealed ruptured adhesions in four patients, perforated bullae in two patients, and no pathologic findings in three patients, which Perry termed *idiopathic spontaneous hemothorax*. In addition to the three cases reported by Perry,¹ only two other confirmed cases of idiopathic spontaneous hemothorax are documented in the medical literature. Slind et al² reported a patient with spontaneous hemothorax who at thoracotomy was found to have a rent in his anterior right hemidiaphragm. Davidson³ reported a case of

spontaneous hemothorax from a ruptured intercostal artery. Within the past several years there have been anecdotal, unreported cases of patients with AIDS and Kaposi's sarcoma who have bled into the pleura or pericardium during the terminal stages of the disease.

Diagnosis

Idiopathic spontaneous hemothorax is a diagnosis of exclusion to be made after more common causes are ruled out. Conventional wisdom dictates that blood in the pleural cavity in the absence of trauma represents metastatic cancer until proven otherwise. Cytologic examination of the blood/fluid has specificity only when the examination is positive and does not rule out the presence of pleural tumor when negative. Thoracoscopic examination and biopsy of the pleural surfaces may reveal tiny tumor deposits that may not shed cells.

A generally accepted diagnostic algorithm (Figure 4) includes: the identification of blood or bloody pleural fluid by thoracentesis; evacuation of the fluid with a catheter or chest tube; submission of fluid specimens for cytology, acid-fast bacilli and fungi cultures, and pH and erythrocyte counts; CT scan of the chest looking for lung lesions and lymphadenopathy; hematologic studies for coagulopathy; and bronchoscopy with cytology and culture. If no diagnosis is achieved, the next step is thoracoscopic exploration and pleural/lung biopsy. If no positive findings are produced, idiopathic spontaneous hemothorax is the final diagnosis.

Differential Diagnosis

Hemothorax occurs most often after trauma. Hemothorax that is not associated with trauma is usually caused by metastatic tumor in the pleura, primary lung cancer, mediastinal tumor, or mesothelioma. Less often, granulomatous diseases of the lung produce pleural bleeding. Patients with coagulopathy or uncontrolled hypertension may bleed spontaneously into the pleural cavity. Less frequent causes of spontaneous hemothorax include rupture of a pleural arteriovenous malformation, bleeding from an intercostal or mammary artery, disruption of a pleural adhesion, or rupture of a bulla.

Treatment

The surgical literature contains several reports of spontaneous hemothorax that were successfully managed with thoracentesis or tube thoracostomy.^{1,4-6} One case reported by Promisloff⁴ described tube drainage alone without surgical exploration. One or more of the 12 surviving cases compiled by Perry¹ were managed with tube drainage after negative chest exploration.

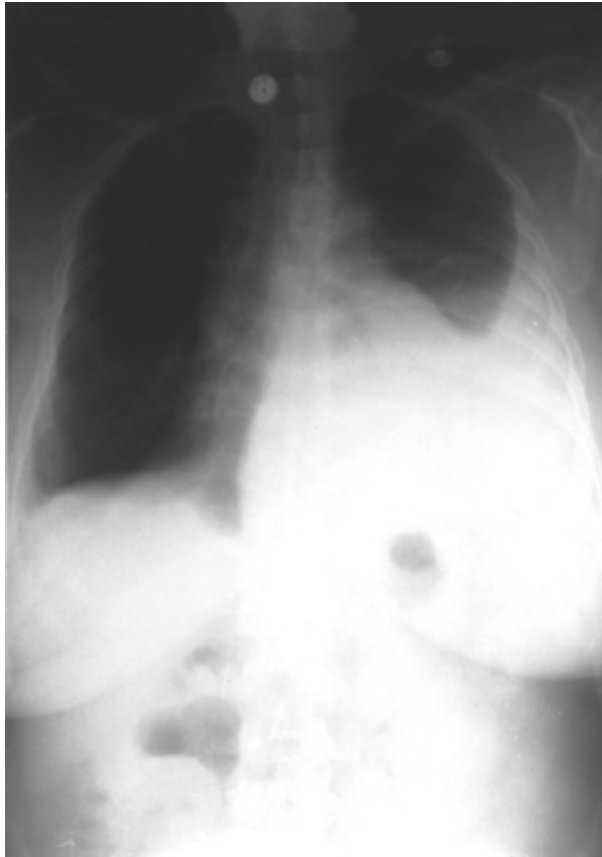


Figure 2. Chest radiography of the patient with left pleural effusion 7 years after presentation with right pleural effusion.

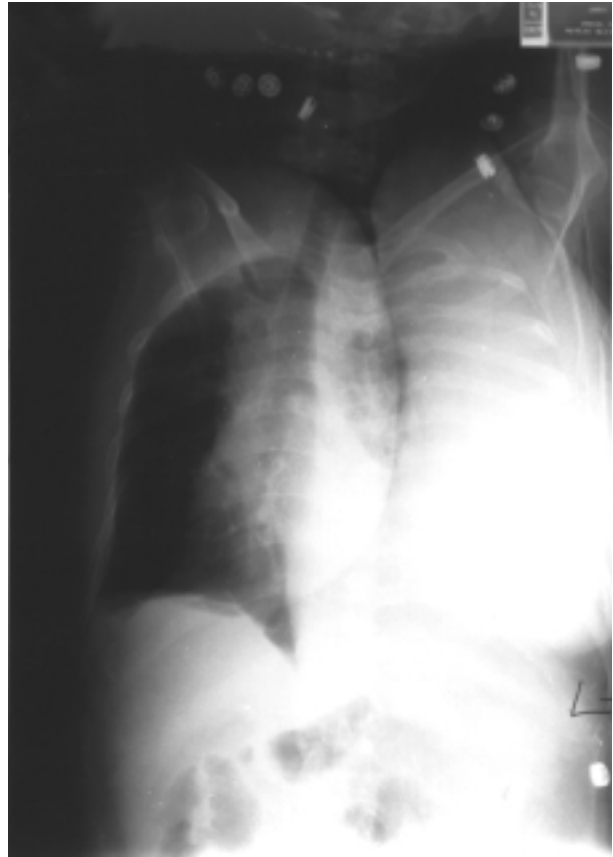


Figure 3. Repeat chest radiography of the patient performed several hours after hospital admission for left pleural effusion demonstrates increasing effusion.

Dimitri⁵ reported a case of massive idiopathic hemothorax in which thoracoscopic exploration showed no identifiable pathology or bleeding source and no further bleeding after tube drainage. In this case, multiple random pleural biopsies showed nonspecific pleural thickening and a fibrinous reaction. Yung et al⁶ reported the case of a 35-year-old man with spontaneous hemothorax who, at thoracotomy, had 2 L of blood evacuated from his pleural space with no identified source of bleeding. A chest tube was placed, and no biopsies were obtained. The patient survived.

Rationale for the Management of This Patient

The patient in this case study presented initially with a spontaneous right hemothorax. Diagnostic studies generally followed the previously noted diagnostic algorithm; however, all attempts were nondiagnostic. A few findings in the patient's medical history were notable but had no clear connection to the present intrathoracic bleeding. These findings included a positive tuber-

culin skin test but absence of sputum confirmation of active tuberculosis; a history of abnormal bleeding during cholecystectomy but currently normal clotting parameters; and a hepatitis screen positive for exposure to hepatitis A but normal results on liver function tests. Thoracoscopy was uncommon when this patient first presented, and the patient underwent an exploratory thoracotomy with no underlying pathology identified. The right pleural cavity was evacuated with a chest tube and the bleeding did not recur.

Seven years later, the patient had a similar event on the contralateral side of her chest. After blood was identified by thoracentesis, the diagnostic work-up was abbreviated and the results of the few studies that were performed were negative. The patient was thought to have sustained a second idiopathic spontaneous hemothorax. The chest was not explored surgically, and treatment was limited to evacuation of the blood by chest tube. During more than 1 year of follow-up, no bleeding has recurred.

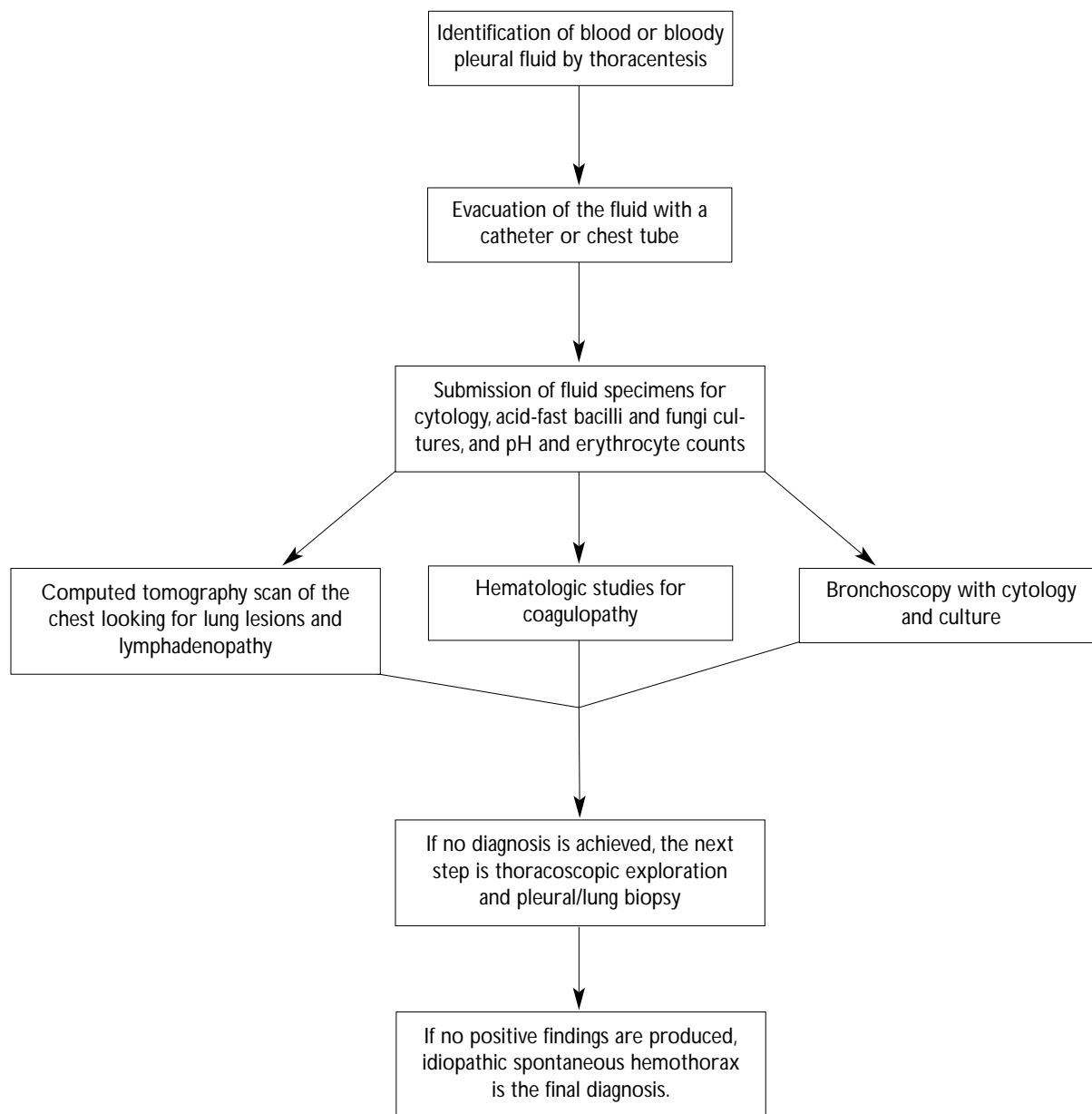


Figure 4. Diagnostic algorithm for idiopathic spontaneous hemothorax.

The cause for both episodes of bleeding may have been idiopathic, but it is reasonable to assume alternatively that some small occult event such as bleeding from a tiny ruptured arteriovenous malformation or disruption of a tiny adhesion may have been responsible for the bleeding. It seems to be reasonable, and supported by several anecdotal reports, that intrapleural bleeding is treated adequately by evacuation of the pleural blood and coaptation of the pleural surfaces.

SUMMARY

Spontaneous hemothorax in the absence of trauma or metastatic tumor is rare, and idiopathic hemothorax is even more rare. The clinical presentation is usually shortness of breath with or without chest or shoulder pain. Diagnosis of pleural effusion is made by chest radiography, and the identification of a bloody effusion is made by thoracentesis. Because of the serious and potentially lethal nature of the more common causes of hemothorax and the importance of making an

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accurate diagnosis, a diagnostic algorithm that includes the use of CT scanning and thoracoscopy may be required. If no underlying pathology is demonstrated after a thorough work-up, patients with occult causes of idiopathic hemothorax seem to be managed adequately by thoracostomy tube drainage. The patient in this case report had a second event of contralateral hemothorax sufficiently similar to the first event 7 years earlier to consider modifying the diagnostic work-up, to omit surgical exploration, and to proceed to minimally invasive treatment with a chest tube. These authors do not advocate or recommend this management for a patient with an initial spontaneous hemothorax. **HP**

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