A 25-Year-Old Patient With Spontaneous Hemothorax*

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A 25-year-old African-American woman with a chief complaint of progressive dyspnea and cough presented to the inpatient service after her primary care physician noted a significant right-sided pleural effusion on a chest radiograph. She had a 2- to 3-year history of intermittent, sharp, pleuritic chest pain radiating from the right subscapular region, across the anterior chest, and down the right arm that worsened with cough and accompanied this episode. Three months prior to hospital admission, dyspnea had developed with exertion and cough productive of white sputum. She sought medical attention 2 months prior to hospital admission, but two courses of outpatient antibiotic therapy did not ameliorate her condition. Her activity level deteriorated from unrestricted walking to dyspnea at rest.

She was receiving no medications, and did not drink alcohol, smoke cigarettes, or use recreational drugs. Coccidioidomycosis was previously diagnosed in a family member. The patient had noted occasional abdominal bloating, night sweats, and chills without fever, and an 8-lb unintentional weight loss over the previous 3 months. She had no known tuberculosis exposures, and a recent tuberculin skin test result was negative. She denied rash, joint pain, nausea, hemoptysis, orthopnea, hematochezia, dizziness, visual changes, dyspareunia, or dyschezia. She has a history of dysmenorrhea and was on her menses at the time of hospital admission.

In the emergency department, she was noted to have a large, right-sided pleural effusion (Fig 1). Thoracentesis revealed bloody fluid. A chest tube was inserted and drained 2.6 L of bloody pleural fluid.

On examination, the patient was a thin, African-American woman in no distress. Her chest examination revealed an appropriately placed chest tube, a soft-tissue fullness over her right scapula, which was tender to palpation, and absent breath sounds on the right side. Ascites, without organomegaly, was present on abdominal examination. The remainder of the examination was normal.

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Manuscript received December 1, 2004; revision accepted January 27, 2005.

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Figure 1. Frontal chest radiograph viewed on hospital admission revealing massive right-sided pleural effusion.
Admission chemistries, blood count, and coagulation panel were normal. Pleural fluid had a bloody appearance, with a lactate dehydrogenase level of 825 U/L and a protein level of 3.8 g/dL. Ascites fluid also had a bloody appearance, with 1.2 million RBCs/µL. Further analysis of the pleural fluid later proved to be negative for acid-fast bacilli by smear, adenosine deaminase, and polymerase chain reaction analysis. Cytology of the pleural fluid was normal. A CT scan of the chest revealed a large, right-sided pleural effusion with extension into the left chest, and a localized area of pleural thickening (Fig 2). Pelvic CT scan showed a fullness between the anterior wall of the rectum and the uterus (Fig 3), and a transvaginal ultrasound revealed a mass in the uterine cul-de-sac. The patient was taken to the operating room for thoracotomy. No subscapular mass was noted, but several 1- to 3-cm brown nodules were noted on the surface of the right lower lobe. Wedge resection of the right lower lobe and pleural biopsies were performed. Pathology showed extensive organizing pleuritis without malignancy. Smears and culture for acid-fast bacilli were negative.

What is the diagnosis?
Diagnosis: Thoracic endometriosis syndrome with catamenial hemothorax

The differential diagnosis of bloody pleural effusions is relatively narrow. Trauma, iatrogenic or otherwise, represents the most common cause of hemothorax. Other common causes of bloody pleural effusion include malignancy (primary or metastatic), tuberculosis, pulmonary embolism, and serositis from collagen vascular diseases such as rheumatoid arthritis and systemic lupus erythematosus. Less common causes include pancreatitis and pseudocyst, leaking thoracic aneurysm, Meig syndrome from pelvic malignancy, and thoracic endometriosis syndrome with catamenial hemothorax.

Clinical Features

Since its identification as a clinical entity in 1953, the thoracic endometriosis syndrome has remained a fleeting diagnosis. Diagnosis is often delayed for months or even years, a testament to the high index of suspicion the clinician must hold for its wide range of clinical presentations. Symptoms, signs, and pathologic states that recur in the chest with menses remain a hallmark of the disease. Such phenomena, termed catamenial by Dr. Lillington in 1972 (from the Greek: κατάμενα for concerning, and μήν for month) may take on a number of clinical manifestations, including pneumothorax, hemothorax, hemoptysis, pulmonary nodules, chest pain, dyspnea, and others.

Catamenial hemothorax represents the second most common manifestation of thoracic endometriosis syndrome, occurring in 14% of known cases, and affects the right side > 80% of the time. Concomitant pelvic endometriosis was found in 100% of cases. The majority of these patients were young, nulliparous, black women. Intraoperative findings are reported to include chocolate-brown and bloody fluid, dense adhesions with a dense nodular peel, chocolate- and blue-colored nodules, and orange-red plaque-like lesions. Importantly, only 71% of reported cases taken to surgery resulted in diagnostic pathology. One study reported that pathologic identification of one case of thoracic endometriosis syndrome was only possible after a second set of samples from the original block was inspected. Thoracic endometrial tissue also undergoes cyclic expansion and recession concomitant with the menstrual cycle, and this has been postulated as a reason for the discrepancy between pathologic data and clinical diagnosis.

Treatment

The goal of medical therapy for the thoracic endometriosis syndrome is to minimize estrogen secretion. A number of hormonal agents, which mimic anovulation, pregnancy, or menopause, are utilized to achieve this objective. Danazol, a derivative of 17α-ethinylestradiol, minimizes the midcycle surge of leutinizing hormone, thus mimicking the chronic anovulatory state. Progestational agents have been given to induce atrophy of endometrial tissue. Gonadotropin-releasing hormone agonists minimize follicle-stimulating hormone and leutinizing hormone secretion, inducing hypogonadotropic gonadism. Oral contraceptive pills, which cause amenorrhea, have also been used.

In general, medical therapy alone is insufficient for the treatment of pleural manifestations of thoracic endometriosis. Recurrence often necessitates pleural inspection, with resection of endometrial implants. Repair of diaphragmatic fenestrations has also been suggested. Hysterectomy and oophorectomy have also been performed as a surrogate for hormonal therapy, with varied results.

In our case, further questioning determined our patient had worsening chest pain and tenderness during menses. Furthermore, her chest tube output precipitously dropped from 200 to 300 mL of bloody fluid per day to close to zero coincident with the cessation of menses. In addition, thoracotomy was performed several days after her menses, which makes it less likely that we would have been able to identify active endometrial tissue from our biopsy samples. We presume the implants found at thoracotomy and the organizing pleuritis identified by pathology represents the residua of endometrial tissue that has reseeded since the end of menses. Based on these clinical features, we ascribed a clinical diagnosis of catamenial hemothorax with thoracic and peritoneal endometriosis to our patient. She was initially placed on danazol with plans to ultimately treat her with oral contraceptive pills. At 1- and 2-month follow-ups, her pleural disease had abated and her ascites had markedly improved.

REFERENCES


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