Thoracic Endometriosis Syndrome Resembling Pulmonary Embolism

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Abstract

A 32-year-old infertility patient with a previous diagnosis of stage IV endometriosis experienced shortness of breath and chest pain. She was diagnosed with a pulmonary embolism by spiral volumetric computed tomography (SVCT) and anticoagulated during hospitalization, although no history of thrombosis was ever identified. She continued to have intermittent symptoms of chest pain, back pain, and shortness of breath for the next 1.5 months. Repeat SVCT revealed a large, right-sided pleural effusion with associated consolidation but no evidence of pulmonary embolism. To obtain a definitive diagnosis, a thoracoscopic pleural biopsy was performed and showed thoracic endometriosis involving the pleura. The patient desired to retain her fertility and opted for treatment with depot medroxyprogesterone. She has been asymptomatic for 2 years with this treatment. This case illustrates the importance of recognizing thoracic endometriosis syndrome and the difficulty diagnosing this condition considering its nonspecific features.

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Thoracic endometriosis syndrome (TES) is a rare form of extrapelvic endometriosis. According to a review of all 110 reported cases, the diagnosis was made approximately 5 years after the diagnosis of pelvic endometriosis in women whose average age was 35 years.[1] Usually pneumothorax, hemothorax, hemoptysis, or pulmonary nodules predominate the clinical picture, yet the diagnosis is commonly delayed an average of 8 months after the first examination. The literature contains only one report of a woman with catamenial chest pain as her only symptom.[2] Thus, patients with nonspecific symptoms without obvious pathology are a diagnostic dilemma for the clinician.

Case Report

A 32-year-old nulligravid woman initially had pelvic pain that worsened despite treatment with nonsteroidal agents. Her uterus was 12 to 14 weeks in size, and a right ovarian cyst was seen on ultrasound examination. During laparotomy, an 8-cm posterior uterine myoma and right ovarian cyst were removed. She also had complete obliteration of the cul-de-sac and was diagnosed with stage IV endometriosis. She received a gonadotropin-releasing hormone (GnRH) agonist with add-back drug therapy (blinded from physician and patient) for 1 year as part of a randomized, placebo-controlled study evaluating this therapy in patients with endometriosis. From age 33 to 37 years her pelvic pain was controlled with nonsteroidal antiinflammatory agents.

At age 37 she experienced increasing pelvic pain, and ultrasound examination showed a $12 \times 7 \times 7$-cm unilocular cystic mass filling the cul-de-sac consistent with a peritoneal cyst. Approximately 300 ml of straw-colored fluid was drained under ultrasound guidance and was negative for cytology or infection.

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Two months later the woman developed acute symptoms of shortness of breath and chest pain. She denied other medical problems and had no history of smoking, allergies, asthma, anticoagulation problems, international travel, leg swelling, or infections. Her peak air flow on admission was 100 L/minute and although her respirations improved with albuterol, she continued to have chest pain. The chest radiograph was normal, and spiral volumetric computed tomography (SVCT) was read as high probability for pulmonary embolism. Pelvic-abdominal SVCT and venous Doppler did not reveal a thrombotic source. She was treated with intravenous heparin with the dosage adjusted according to coagulation times. Oral warfarin was added until her anticoagulation was therapeutic and she was discharged.

The patient continued to have intermittent chest pain, back pain, and shortness of breath that seemed to worsen with her menstrual cycle. Follow-up chest radiograph and SVCT scan approximately 1.5 months later did not identify evidence of pulmonary embolism but revealed a new, large, nonspecific, right-sided pleural effusion with associated consolidation (Figure 1). After consultation with a pulmonologist, the decision was made to perform needle drainage of this pleural effusion. Warfarin was stopped and thorocentesis showed an exudative effusion with a hemocrit of 2%, negative culture, and normal cytology. A presumptive diagnosis of pulmonary endometriosis was made and the woman was scheduled for thorascopic pleural biopsy for a definitive diagnosis. During the procedure bloody fluid was found in the pleural space. Histopathologic examination of biopsy tissue revealed endometrial-type stroma consistent with pleural endometriosis (Figures 2 and 3).

The patient was offered options of definitive permanent sterilization or medical treatment. She desired to retain her fertility and opted for treatment with depot medroxyprogesterone acetate. During the next 2 years she remained asymptomatic with this treatment.

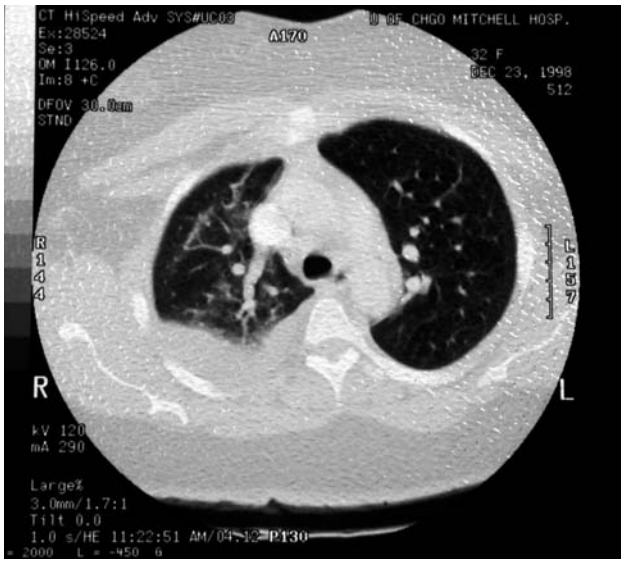

FIGURE 1. Spiral volumetric computed tomography of the lung shows a large, right-sided pleural effusion with associated consolidation.

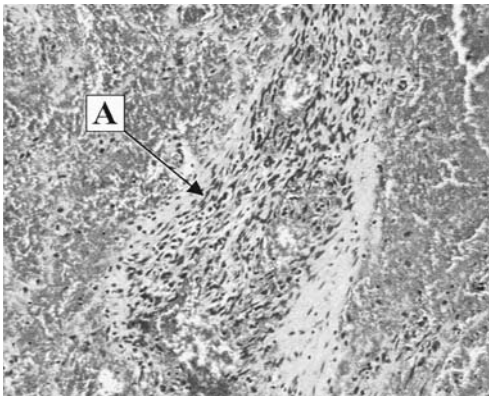

FIGURE 2. Pleural biopsy specimen. (A) Focus of endometrial-type stroma consistent with thoracic endometriosis. Surrounding parenchyma contains a significant amount of hemorrhage.

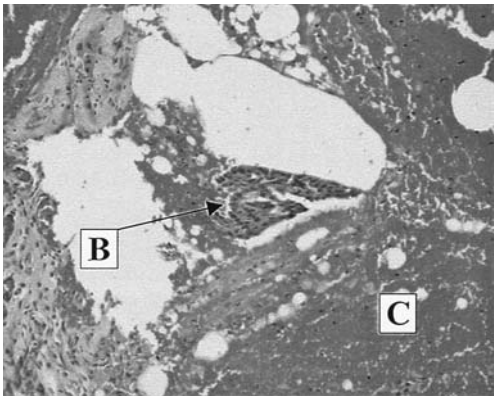

FIGURE 3. The biopsy specimen contains (B) normal lung pleural tissue with (C) frank blood.

Discussion

Despite the fact that endometriosis is a common disease, we do not completely understand its pathophysiology. Two theories are put forward as possible explanations for the development of extraperitoneal endometriosis. The metaplastic theory suggests local metaplasia of coelomic epithelium; the retrograde menstruation theory suggests that endometrial tissue escapes from the uterine cavity from the fallopian tubes to seed peritoneal tissue. Distant metastatic sites occur either from direct tissue contact through the peritoneal cavity, lymphatics, or hematologic system, or by iatrogenic implantation. Pulmonary endometriosis is thought to occur when these peritoneal implants make their way through fenestrations in the diaphragm to seed the pleural space. Of interest, fenestrations seem to be present only on the right side of the diaphragm.[3]

Over 110 cases of TES have been reported in the literature since the first one was described in 1953.[4] The largest review of the syndrome shows that the average age at diagnosis is 35 years. The syndrome is associated with pelvic endometriosis in 80% of patients[4] and is diagnosed approximately 5 years after the onset of pelvic endometriosis.[1] This and the fact that 90% of cases involved the right hemithorax seems to favor the theory of transdiaphragmatic spread from the peritoneal cavity.[4]

The syndrome can involve the pleura or the parenchyma, each with distinctive symptoms. Most of 65 cases of pulmonary endometriosis (83%) were pleural.[4] All patients in the review had pneumothorax, hemothorax, hemoptysis, or pulmonary nodules, yet diagnosis was delayed an average of 8 months after initial examination.[1]

Treatment of TES involves decreasing or eliminating estrogenic stimulation of ectopic endometrial glands. Although hysterectomy with bilateral oophorectomy may offer the highest likelihood of cure, women wanting to preserve fertility usually choose medical treatment. The GnRH agonists work by suppressing release of GnRH from the hypothalamus, but can exacerbate symptoms early in therapy.[5] Danazol has potential side effects of virilization, climacteric symptoms, and weight gain. Oral contraceptives and progesterone offer sufficient down-regulation of follicle-stimulating hormone and luteinizing hormone surges and show no difference in recurrence rates at 6 and 12 months compared with danazol.[1] Pleural implants are difficult to localize due to the multifocal nature of the disease, making surgical resection difficult. Patients with recurrent pneumothorax can be treated with surgical pleural abrasion.[6] If these therapies fail, hysterectomy with bilateral oophorectomy is the treatment of choice.

The syndrome can be difficult to diagnose, especially with nonspecific symptoms of chest pain and shortness of breath. It is a rare disease and is often not included in the differential diagnosis in women with these symptoms. Pulmonary embolism (PE) is much more common and can share the same symptoms as pulmonary endometriosis. Although our patient could have both disorders simultaneously, lack of a thrombotic source and continued symptoms even after adequate coagulation do not support this theory.

The working standard for diagnosing PE is now SVCT. Studies evaluating its effectiveness suggest that its sensitivity and specificity are 100% and 96%, respectively.[7] However, according to a critical review of the literature, sensitivity of SVCT may vary between 64% and 93%, with specificity from 89% to 100%, suggesting the need for further evaluation of this method to assess its clinical effectiveness.[8]

The clinician must include TES in the differential diagnosis when women have symptoms of PE and a history of endometriosis, catamenial pain, or hemoptysis to avoid delay in treatment.

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