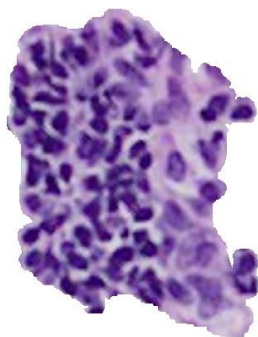


Review

Thoracic Endometriosis

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**Endometrial Cell Implant
on Pleural Mesothelium**

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Abstract: A recurrent hemorrhagic pleural effusion in a woman during her reproductive years may be the clinical presentation of thoracic endometriosis syndrome (TES). We report here a case of a recurrent bloody right pleural effusion in a young female who had a history of pelvic endometriosis. Thoracic endometriosis with pleural involvement was confirmed by pleural biopsy which showed focal involvement of functional endometrial tissue within the pleura. The patient underwent pleurectomy and talc pleurodesis without recurrence of the pleural fluid.

A hemorrhagic pleural effusion due to thoracic endometriosis may mimic other conditions including pulmonary thromboembolism, trauma, malignancy, tuberculosis, and others. Thoracic endometriosis should be in the differential diagnosis of a bloody pleural effusion in women of childbearing age, particularly in a patient with an established diagnosis of pelvic endometriosis.

Key Words: Thoracic endometriosis, pleural effusion, hemothorax

Introduction

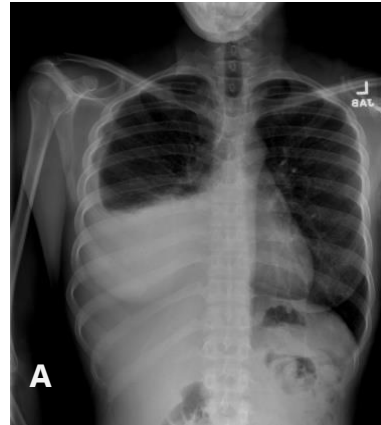
The spectrum of thoracic endometriosis (TE) is broad. It may involve lung parenchyma, tracheobronchial airways, diaphragm, pericardium, and pleura. Pleural effusion due to thoracic endometriosis is rare. It is characterized by bloody pleural fluid that occurs predominantly during a woman's reproductive years and is usually referred to as 'catamenial* hemothorax'¹. The pleural effusion in TE is predominantly right-sided. It is caused by the presence of functional endometrial tissue in pleural mesothelium. The clinical presentation of TE may include recurrent pneumothorax, hemothorax, hemopneumothorax, hemoptysis or pulmonary nodule.

The primary symptoms of TES include chest pain, dyspnea, and hemoptysis². The diagnosis of TES is usually delayed for several years because its clinical symptoms and diagnostic tests are non-specific. Treatment options include hormonal therapy and surgery.

We describe a woman with a history of pelvic endometriosis who developed recurrent right-sided pleuritic chest pain and dyspnea. She was found to have a right-sided bloody pleural effusion. Pleural biopsy revealed focal involvement of endometrial stromal tissue consistent with pleural endometriosis. The patient underwent pleurectomy and talc pleurodesis with no recurrence of the pleural effusion after six months.

Case Report

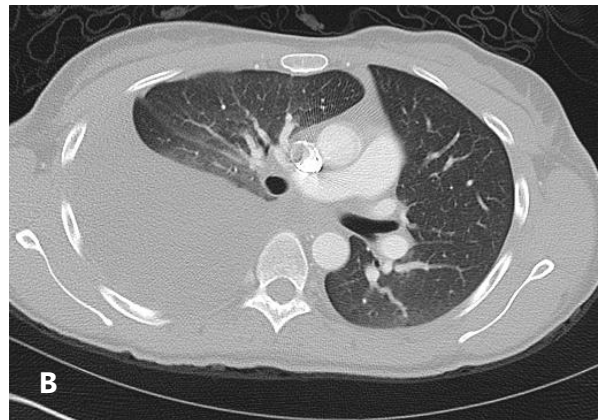
A 33-year-old non-smoking African American female presented to her primary care physician with several days of progressively worsening pain right-sided pleuritic chest and dyspnea. She had a history of pelvic endometriosis diagnosed on laparoscopic examination and was taking oral contraceptives for suppression of endometriosis. She was found to have diminished breath sounds on the right side. Chest x-ray (Image A) revealed a right pleural effusion.



Chest x-ray showing pleural effusion

She was treated with oral antibiotics for presumed pneumonia with a parapneumonic effusion and referred to a pulmonologist for evaluation. She was admitted to hospital.

CT angiogram of the chest (Image B) demonstrated a large right pleural effusion. No pulmonary embolism or mass lesion was seen.

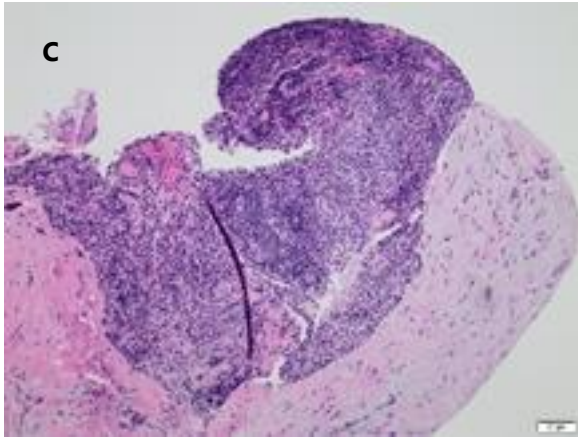


Ultrasound-guided thoracentesis produced 1500 cc of bloody pleural fluid. The pleural fluid RBC count was 611,000 and WBC count 159, total protein 5.1 g/dl, LDH 304 U/L, cholesterol 77 mg/dl, consistent with exudative pleural fluid. Cytological examination was negative for malignancy. Microbiology testing, including AFB

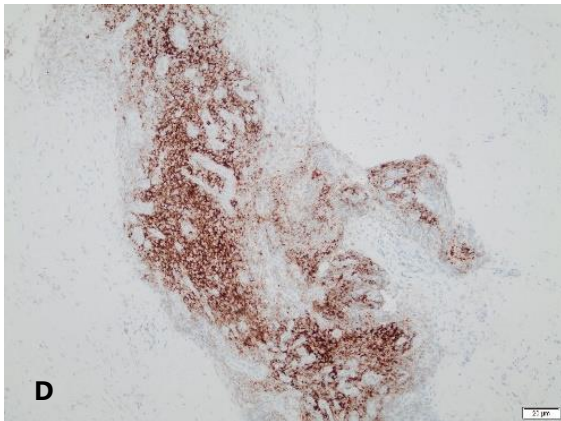
* Catamenial: Associated with menstruation. Deriv. Greek; Catamenia καταμήνια: kata (κατα): by, in accordance with, and mén (μήν): month. The English word menses (pl.) is derived from the same root.

culture, was negative. She was found to have no hematologic or collagen vascular diseases.

In the subsequent few weeks, the patient had repeated thoracenteses for recurrent right pleural effusion with test results similar to the first one. She then underwent diagnostic thoracoscopy which showed a normal appearing parietal pleura with no obvious lesion or abnormality. A random pleural biopsy revealed focal endometrial stromal proliferation. (Image C)



An immunohistologic study (Image D) was positive for the presence of endometrial tissue consistent with pleural endometriosis.



Discussion

Thoracic endometriosis (TE) is a rare disease occurring predominantly in women of re-

productive age. Most patients with TES have associated evidence of pelvic endometriosis³. TES is characterized by functional endometrial tissue within the pulmonary parenchyma including airways, lung tissue, and pleura⁴. These ectopic sites may slough and hemorrhage in conjunction with the hormonal changes of the menses.

Pericardial and diaphragmatic involvement has been reported as well⁵. Presenting symptoms include chest pain, dyspnea and hemoptysis. The most common pulmonary complication of TES is pneumothorax, followed by hemothorax, hemoptysis and pulmonary nodule. Catamenial pneumothorax and hemothorax due to TES are almost always confined to the right side, whereas lung parenchymal endometriosis can be unilateral or bilateral³. Radiographic findings are non-specific.

There are three major theories regarding the pathogenesis of TES. The first and most accepted theory is retrograde menstruation with endometrial tissue implanting in the pelvis and migrating up the abdomen and through a diaphragmatic fenestration followed by ectopic implantation in the thorax. The second is vascular or lymphatic microembolization. The third, coelomic metaplasia⁶, posits that pelvic and pleural mesothelial stem cells, tissues of the same mesenchymal origin, are pathologically induced to differentiate into endometrial cells.

Patients with thoracic endometriosis typically have a history of infertility and pelvic surgery and present with catamenial hemoptysis, pneumothorax, or a pulmonary nodule. Our case had the elements of a hemothorax, a history of infertility, and pelvic surgery. Bronchoscopy with BAL and pleural fluid analysis are rarely helpful for the definitive diagnosis of thoracic endometriosis. Thoracotomy or VATS (Video Assisted Thoracoscopy) is usually required to establish a tissue diagnosis⁷.

First line treatment for TE is medical therapy, which includes hormonal therapy (danazol or GnRH analogs, e.g., leuprolide) to suppress the endometrial tissue by creating a hypo-estrogenic state. The recurrence rate with hormonal therapy is up to 50%. VATS with chemical pleurodesis and or decortication can be useful for treatment and prevention of further recurrence. It is superior to hormonal therapy. A combination of medical hormonal and surgical therapy is the most effective treatment⁸. In our case, the patient preferred a surgical approach. She underwent pleurectomy and talc pleurodesis without recurrence of pleural effusion in long-term follow-up.

Learning Points

It is imperative to keep thoracic endometriosis in the differential diagnosis in a young fe-

male with a history of infertility and pelvic surgery who presents with catamenial chest pain, dyspnea, hemoptysis, recurrent pleural effusion, pulmonary nodule, or pneumothorax. The diagnosis of TES is frequently delayed for a few years after the initial symptoms because it is an extremely rare disease with non-specific routine diagnostic testing. A tissue diagnosis to confirm TES is needed in most cases.

Treatment options include using hormonal therapy to suppress endometrial tissue growth and to prevent pelvic seeding, or surgery with chemical pleurodesis which is superior to hormonal therapy in preventing recurrence. A combination of medical and surgical treatment is needed most in advanced cases.

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References

1. Channabasavaiah AD, Vempilly Joseph J. Thoracic Endometriosis, *Medicine*, 2010; Volume 89, number 3:183-188. Lippincott Williams & Wilkins
2. Bhattacharjee S, DebJ, Saha R, Chakrabarti S, Mukherji J, Tapadar S.R. Pleural Endometriosis: An exceptional cause of hemorrhagic pleural effusion, *J. of Obstetrics and Gynecology of India*, 2014, 64(S1):S100-S104.
3. Joseph J, Sahn Steven A, Thoracic Endometriosis: New observations from an Analysis of 110 cases. *American Journal of Medicine*, 1996, Volume 100; 164-170
4. Ziedalski TM, Sankaranarayanan V, Chitkara R, Alto P. Thoracic endometriosis: A case report and literature review. *Journal of Thoracic and Cardiovascular surgery*, 2004 Volume 127, number (5); 1513-1514.
5. Senseig DM, Serlin O, Hawthorne HR. Pericardial endometriosis. An experimental study in Dogs. *JAMA*, 1996;198:645-7.
6. Sevinc S, Unsal S, Ozturk T, Uysal A, Samancilar O, Ors Kaya S, Ermete S. Thoracic Endometriosis syndrome with bloody pleural effusion in a 28 year old women. *J Pak Med Assoc*, 2013, Vol 63, No 1; 114-16
7. Inoue T, Kurokawa Y, Kaiwa Y, Abo M, Takayama T, Ansi M, Satomi S. Video-Assisted thoracoscopic Surgery for Catamenial Hemoptysis. *Chest* 2001; 120:655-658.
8. Machairiotis et al. Extrapelvic endometriosis: A rare entity or an under diagnosed condition? *Diagnostic Pathology* 2013, 8:194.

