Extraperitoneal Endometriosis With Catamenial Pneumothoraces: A Review of the Literature

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ABSTRACT

Objective: To present a case of recurrent catamenial pneumothorax and diaphragmatic endometriosis that was managed thoracoscopically. A review of the literature is also presented.

Methods: A 28-year-old woman presented with bloody stools, chronic constipation, and chest pain. A review of systems was positive for monthly chest pain associated with her menses. A preoperative chest x-ray revealed a right pneumothorax. Colonoscopy revealed biopsy proven endometriosis of the sigmoid colon. A pelvic computed tomography scan revealed bilateral complex, cystic and solid adnexal lesions.

Results: A right thoracoscopy was performed. A lesion on the right hemidiaphragm was excised and confirmed to be endometriosis. A wedge section of lung tissue containing a bleb was resected and also contained endometriosis. Three months later, the patient underwent laparoscopic excision of her pelvic endometriosis, including a low anterior rectal resection. Five months later, she presented again with right-sided chest pain. A thoracoscopic right total pleurectomy was performed for recurrent pneumothorax.

Conclusion: Pulmonary endometriosis may present as chest pain, shortness of breath, or hemoptysis associated with menstrual cycles. This case emphasizes the importance of a careful review of systems in patients with known endometriosis. Management now includes an endoscopic alternative and all of its known benefits.

Key Words: Catamenial pneumothorax, Extraperitoneal endometriosis, Menses.

INTRODUCTION

The first report of catamenial pneumothorax appeared in the literature in 1958. Over 100 cases have since been reported. Diagnosis is often made based on the patient’s symptoms. Extraperitoneal endometriosis has also been identified in the extremities and in the male urinary system. Several theories attempt to explain these extrapelvic sites of endometriosis. Treatment has included both medical and surgical options. With the development of advanced endoscopic techniques, pulmonary endometriosis can now be treated thoracoscopically with excellent results.

CASE REPORT

A 28-year-old woman initially presented with bloody stools, chronic constipation, and pelvic pain. During a review of systems, the patient described monthly right-sided chest pain associated with her menses. A pelvic examination revealed an anterior rectum adherent to the posterior cervix and vagina between the uterosacral ligaments. A rectal examination revealed a nodular lesion on the anterior rectum at approximately 10 cm. The patient was evaluated by a gastroenterologist and underwent colonoscopy. A submucoal-appearing lesion at 20 cm from the anal verge was biopsied and histologically reported as submucosal endometriosis overlying colonic mucosa with polypoid hyperplastic change and lymphoid aggregates. The initial workup included a preoperative chest x-ray that revealed a right pneumothorax. An abdominal-pelvic computed tomography (CT) scan revealed bilateral complex, cystic and solid adnexal lesions. A chest tube was placed resolving the pneumothorax. High-resolution chest CT did not reveal parenchymal or pleural lesions. A diagnosis of catamenial pneumothorax was made based on the patient’s history of chest pain associated with the onset of her menstrual cycles and objective findings on radiologic studies.

RESULTS

The patient underwent elective right thoracoscopy. A 1.5-cm x 1.0-cm lesion on the right hemidiaphragm was observed (Figure 1). Using endoscopic shears, the lesion was excised. The diaphragm was repaired with
interrupted 0 Ethibond sutures placed thoracoscopically and tied in succession. Next, a bleb of the right upper lobe was identified (Figure 2). A 2.0-cm x 1.5-cm x 1.0-cm wedge section of right apical lung tissue containing the bleb was resected using the endoscopic stapler. Both specimens were evaluated histologically and noted to have endometrial glands and stroma within the tissue, confirming extraperitoneal endometriosis (Figures 3 and 4). Three months later, the patient underwent laparoscopic excision of her pelvic endometriosis, including a low anterior rectal resection. Five months later, she again experienced right-sided chest pain and was diagnosed with a recurrent pneumothorax. She then underwent a thoracoscopic right total pleurectomy.

DISCUSSION
Seventy-five percent of pulmonary endometriosis presents as catamenial pneumothorax, 10.7% as cyclical hemothorax, and 8.3% as hemoptysis. The rest remains as asymptomatic nodules and may be discovered incidentally.10 In general, endometriosis of the pleura or diaphragm will present as cyclic pain or dyspnea associated with a pneumothorax that is usually right-sided. Parenchymal endometriosis is usually asymptomatic and may be associated with cyclic hemoptysis.11

Diagnosis of catamenial pneumothorax is most often made clinically, after the exclusion of other causes. Although not in the case presented here, computed tomography (CT) and magnetic resonance imaging (MRI) have been useful in locating the pulmonary endometriosis.12-14 MRI appears to be superior when differentiating between pleural and parenchymal disease.1

The mechanism for air entry into the thorax remains controversial. Diaphragmatic fenestrations are seen in only 19% to 33% of patients with catamenial pneumothorax.15 No obvious defect is present in the majority of patients, but microscopic passages between the peritoneal cavity...
and pleural cavity have been documented by using \(^{131}\)I-labeled albumin injected into the peritoneal cavity.\(^{16}\) It has been suggested that the genital tract may be a source of air entry.\(^{17}\) The ball-valve theory proposed by Lillington\(^{18}\) suggests that air enters the pleural space as endometriosis invades the visceral pleura and impinges upon a small bronchiole. Air can enter the space, but because the implant functions as a one-way valve, the air cannot escape. Rossi and Goplerud\(^{19}\) hypothesized that the entry of air into the pleural space results from rupture of alveoli in response to vasospasm and bronchospasm due to elevated prostaglandins during menses.

In 1927, John Sampson\(^{20}\) published his classic paper coining the term “endometriosis” and establishing retrograde flow of endometrial tissue into the peritoneal cavity as the cause of this disease. Two main problems with Sampson’s theory are (1) many women have retrograde menstrual flow and do not develop endometriosis and (2) his theory does not explain how endometriosis occurs in sites remote from the pelvis. Other theories have been developed. The coelomic metaplasia theory best explains the presence of endometriosis in the extremities. During embryologic development, the coelomic epithelium is adjacent to the mesenchymal limb buds. If the differentiating coelomic cells are incorrectly integrated into the limb buds, under certain circumstances, they may be stimulated to develop into endometrial tissue responsive to steroid hormones.\(^{21}\) This theory is also most plausible for explaining the presence of endometriosis found in the male urinary system, usually associated with high-dose estrogen treatment.\(^{6-9}\) These theories can also be applied to pulmonary endometriosis. The pleura arises from coelomic epithelium and may too be stimulated under appropriate circumstances to develop into endometrial tissue.\(^{21}\) Others have suggested that endometrial cell seeding from retrograde flow occurs after passage through diaphragmatic fenestrations, but diaphragmatic defects have been found in only 15% to 33% of patients with pleural endometriosis.\(^{22,23}\) Vascular/lymphatic transport is another classic theory. Hobbs and Bortnick\(^{24}\) investigated this theory by injecting into the ear vein of rabbits, endometrial tissue from the same animal. They found pulmonary endometriosis developed in 79% (19/24) of the animals.

Extrapelvic endometriosis and catamenial pneumothoraces have been treated with a variety of medical therapies. Successful results have been reported with danazol and gonadotropin-releasing hormone agonists (GnRH agonists).\(^{25-27}\) They are similar in their abilities to suppress disease, but both offer undesirable side effects.\(^{28}\) The most common side effects of danazol are weight gain, fluid retention, fatigue, acne, oily skin, hot flushe, growth of facial hair, emotional lability, and possible irreversible deepening of the voice. The hypoestrogenic state induced by GnRH agonists may cause hot flushes, vaginal dryness, and bone loss. The bone loss is reversible and can be decreased with postmenopausal estrogen-progestin add-back therapy.\(^{29-31}\) Other commonly used regimens for the treatment of pelvic endometriosis are continuous oral contraceptive pills (no “pill-free” week), medroxyprogesterone acetate 30 mg/d, and megestrol acetate 40 mg/d.

All of these drug therapies suppress endometriosis, but do not cure the disease, thus, the reason for failures or “recurrences.” The only definitive cure for endometriosis is surgical resection or complete destruction of the endometriotic lesion. Surgical management does not include bilateral salpingo-oophorectomy. Removal of the ovaries will not ensure relief of symptoms. Estrogen is also produced by peripheral conversion of androgens (primarily androstenedione of adrenal and ovarian origin). This conversion is catalyzed by aromatase. Aberrant aromatase expression has been identified in endometriosis.

Figure 4. Endometrial glands and stroma within the pleural bleb.
otic implants, causing local production of estradiol within the implant. Therefore, medical therapy remains a first-line treatment option, but it is not unreasonable to proceed directly to surgical management for more definitive treatment.

**CONCLUSIONS**

This case emphasizes the importance of a careful review of systems in patients with known endometriosis. Endometriosis has been identified in several sites outside of the pelvis. The most plausible explanation for its appearance in these unusual sites is the coelomic metaplasia theory. Now with the ability to perform more procedures endoscopically, surgical management becomes an even more attractive option, wherever the lesions may be.

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