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Pulmonary Endometriosis

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Abstract. Pulmonary endometriosis is manifested as either asymptomatic pulmonary nodules or as pneumothorax, hemothorax, or hemoptysis during menses. We review 84 cases of pulmonary endometriosis in the English literature and report 3 additional patients. One of our patients is the first reported to have hemopneumothorax. Catamenial pneumothorax usually involved the right chest, and occurred in young nulliparous women without pelvic endometriosis. Pleuroscopy, laparoscopy with pneumoperitoneum, and thoracotomy produced a tissue diagnosis infrequently. Hormonal suppression of ovulation and pleurodesis usually corrected this disorder. Catamenial hemothorax only affected the right chest, but occurred in older multiparous women with pelvic endometriosis. While thoracotomy or laparotomy produced a tissue diagnosis, these procedures were not curative. In contrast, our patient with this disorder was treated successfully with pleurectomy. Catamenial hemoptysis occurred in multiparous women without pelvic endometriosis. Bronchoscopy localized bleeding but never produced a tissue diagnosis. Thoracotomy produced endometrial tissue. Endometrial pulmonary nodules require a diagnosis but do not otherwise produce problems.

Key words: Pneumothorax—Pleural effusion—Hemothorax—Hemoptysis—Pulmonary nodules—Pulmonary endometriosis.

Introduction

Since 1938, when Schwarz first associated endometriosis with pulmonary lesions, others have noted four pulmonary manifestations of this disorder [1–50].

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These are: catamenial pneumothorax; catamenial hemothorax; catamenial hemoptysis; and asymptomatic pulmonary nodules. We report 3 patients who illustrate 3 of these complications of endometriosis and also reveal new aspects of this disorder not previously described. The pertinent English literature is reviewed with each case presentation. Recommendations regarding evaluation and treatment of these disorders are included.

Catamenial Pneumothorax

Catamenial pneumothorax was first described by Maurer et al. in 1958 [25]. To date 63 cases have been reported [2, 3, 10, 12, 14, 15, 18, 20, 22–29, 31, 35, 36, 38, 42, 43, 45, 46, 48–50]. These patients ranged in age from 24 to 44 years (average 33 years old). Fertility problems were uncommon, with only 3 patients reportedly nulliparous [23, 34, 41]. Fourteen (22%) of the 63 patients had pelvic endometriosis.

Recurrent spontaneous pneumothorax occurred coincident with menses but not at other times. Lillington [24] reported 1 patient with 40 probable episodes, 25 of which were documented. In individual patients, the number of probable episodes ranged from 2 to 42 (average, 14) while documented episodes ranged from 1 to 25.

Ninety-five percent occurred in the right chest, 2 were bilateral [22, 46], and only 1 proven case involved the left side alone [39]. In all of the cases, pneumothorax developed within 72 hr prior to or after onset of menses. Most of the patients complained of dyspnea and chest or shoulder pain. Chest roentgenograms during menstruation demonstrated rapid resolution of the pneumothorax, but a few were large enough to require chest tube drainage. Associated pleural effusion was never described, and Davies has noted this to be "a perplexing feature of catamenial pneumothorax" [9].

Table 1 summarizes the positive findings in 48 patients who underwent

Table 1. Catamenial pneumothorax Thoracotomy in 48 patients: positive findings[a]

Findings	Number %
Fenestrations (perforations in diaphragm)	33
Endometrial implants (diaphragm &/or pleura)	35
Defects of diaphragm and endometriosis	19
Blebs	20
Apical scarring	6
Cyst	8
Pleuritis	10
Inflammatory granuloma	2
Air leak	2
"Black lesion"	2

[a] Some patients had multiple findings

thoracotomy for catamenial pneumothorax. In only 35% of these cases were endometrial implants found and a definitive diagnosis established. Forty percent had nonspecific findings, and in 25%, no abnormal findings were discovered. Soderberg and Dahlquist [42] attributed this lack of specific pathologic findings to the lack of knowledge about this entity when the thoracotomies were performed. Shearin suggested that "more appropriate timing of the surgery . . . might provide a better opportunity to diagnose and excise areas of endometriosis that might not be apparent at other times" [39].

Other procedures suggested to diagnose this entity included pleuroscopy [12], and laparoscopy with pneumoperitoneum which, if followed by pneumothorax, would suggest a diaphragmatic communication [12, 41]. The diagnosis has been accepted by others based on the history and chest roentgenographic findings.

Immediate management was the same as that for pneumothorax of any cause; namely lung reexpansion, and, where necessary, chest tube drainage. Following reexpansion, patients have been treated in several ways including hormonal suppression of ovulation with testosterone, Depo-Provera, various combinations of estrogen and progesterone, and donazol (11 of 63 patients). Many patients have responded to some degree and apparently some have been cured [14, 23, 24, 46]. Total hysterectomy with bilateral oophorectomy has also been carried out [24, 36] and recurrences have been reported after hysterectomy [23, 39]. Persistent pneumothorax, a decision not to treat with hormones, and or a recurrence on hormonal therapy have been considered indications for surgical pleurodesis which has almost always been curative [8, 20, 22, 26, 39, 42, 45, 48].

In summary, invasive efforts to establish a tissue diagnosis are usually unsuccessful. The diagnosis can be presumed, however, when pneumothorax recurs during menses and there is no other obvious cause. Medical suppression of ovulation should then be initiated and surgical pleurodesis should be performed if medical treatment fails or is contraindicated.

Case Report

A 25-year-old black woman presented to the emergency room complaining of right-sided pleuritic chest pain and dyspnea that began 2 days after the onset of her menstrual period.

Two years previously, 2 days after the onset of her menstrual period, she had developed a spontaneous right pneumothorax which was treated by tube thoracostomy. Over the next 3 months, she suffered 2 further episodes of spontaneous right pneumothorax concurrent with her menses. Chemical pleurodesis with interpleural tetracycline was performed via a chest tube. Recurrence of the pneumothorax occurred and the patient underwent thoracotomy. At thoracotomy two perforations in the diaphragm, both approximately 1 mm in diameter were found. They were closed and abrasive pleuredesis was performed. There have been no further recurrences.

Our patient conforms to the general clinical description of catamenial pneumothorax. She differs from the majority of other cases in that tetracycline pleurodesis was not curative and she required thoracotomy with abrasive surgical pleurodesis.

Catamenial Hemothorax

There are nine reported cases of catamenial hemothorax [4, 6, 15, 16, 32, 33, 40, 47]. The patients were younger than those with catamenial pneumothorax, ranging in age from 23 to 42 years (average, 28). Symptoms included dyspnea and gynecologic complaints. All had extensive pelvic endometriosis and 7 had clinical ascites. All cases involved the right lung only. The roentgenographic appearance of the pleural effusions varied from slight obliteration of the right costophrenic angle to obliteration of greater than 75% of the hemithorax. Eight patients were nulliparous. Symptoms included catamenial dyspnea and chest pain associated with roentgenographic evidence of pleural effusion at the time of menses.

Thoracentesis, when reported, revealed serosanguinous fluid but this fluid was not otherwise characterized in most cases. In 1 report, Davies [9] identified endometrial cells on cytologic analysis of the pleural fluid. Pleural biopsy specimens were never diagnostic.

Thoracotomy was performed in 7 of the 9 cases; thoracic pleural endometriosis was found in 5, 1 had a "chocolate cyst" [50], and 1 had no pathologic condition verified [15]. All 9 patients also underwent laparotomy. All had extensive pelvic endometriosis, the extent of which limited the amount of resection which could be accomplished. Thus, complete total abdominal hysterectomy and bilateral salpingoophorectomy could not be performed in all patients. Only one of the 9 patients was "cured" by this procedure. Six suffered recurrences and in 2 instances the results were not reported.

Six patients underwent hormonal suppression. There were 2 apparent cures [15, 16]. One patient's condition relapsed during therapy [32], and 2 experienced relapse during discontinuation of the therapy [6, 50]. In 1, the results were not reported [33]. Prior to the availability of hormones, x-ray castration was performed in 2 patients [4, 6] and both were considered cured.

Thus patients with catamenial hemothorax rarely respond to medical management. While the diagnosis can be strongly suspected when dyspnea, chest pain, and pleural effusion recur with menses, the need for surgical therapy warrants invasive diagnostic measures to exclude other disorders. These measures include laparoscopy and pleuroscopy but frequently thoracotomy with pleurectomy has been both diagnostic and therapeutic. The high incidence of recurrence and the low incidence of cure following hysterectomy and bilateral oophorectomy raise strong reservations about this procedure.

Case Report

A 42-year-old black woman came to the emergency room complaining of dyspnea and right shoulder pain that radiated down her right chest. These symptoms began 2 days prior to the onset of her menstrual period. On 2 separate occasions during these episodes, chest roentgenograms had revealed both a right-sided pneumothorax and a small right-sided pleural effusion. With the current episode, she was noted to have a massive right pleural effusion and was admitted to the hospital. Diagnostic and therapeutic thoracentesis was performed and serosanguinous fluid was aspirated. Analysis of the fluid revealed the following: total cell count, 370,000 RBC/mm^3; hematocrit 3 vol%; white blood count, 500 cells/mm^3 with 27% eosinophils, and 18% lymphocytes; glucose 98 mg/dl (serum, 120 mg/dl); protein 4.4 g/dl (serum, 6.13 g/dl); lactate dehydrogenase, 334 mg/dl (serum 283 g/dl); amylase, 98 B.U.; Gram's stain and AFB smear revealed no organisms, and subsequent cultures produced no growth. Cytologic examination of the fluid revealed no tumor or endometrial cells. A pleural biopsy was performed and histologic examination showed chronic pleuritis with no granulomata or tumor. Cultures of the pleura produced no growth. While hospitalized, the patient became more dyspneic and the chest pain became more severe. Two days prior to her menstrual flow, a chest roentgenogram demonstrated that the pleural effusion had reaccumulated. A chest tube was placed and 4L of serosanguinous fluid was removed. The patient was treated with danazol. At the same time, two attempts at pleurodesis with tetracycline were unsuccessful, and the chest tube continued to drain 100 ml of serosanguinous fluid daily. Thoracotomy was performed and revealed 3 perforations in the diaphragm, and a 4-cm hemorrhagic cyst. The cyst was excised and the pleura was decorticated. Histologic examination revealed that the cyst was extremely hemorrhagic and that its architecture was destroyed. No endometrial tissue was found.

Following surgery, pulmonary symptoms have not recurred and the pleural fluid has not reaccumulated. However, 8 months later, the patient developed ascites and underwent exploratory laporatomy. Endometriosis involving the ovaries and uterus was found and the patient underwent bilateral oopherectomy and hysterectomy.

Our patient differs from the previous cases described in our literature review in that she is older, she is multiparous, she has had both catamenial pneumothorax and hemothorax, at thoracotomy she was found to have both a hemorrhagic cyst and fenestrations of the diaphragm, and after hormonal therapy failed she was successfully treated with surgical decortication of the pleura and removal of the diaphragmatic cyst.

Catamenial Hemoptysis

There are 7 reported cases of repeated hemoptysis associated with menses [1, 5, 11, 19, 21, 34]. These patients' ages ranged from 26 to 45 years (average, 37)

and all were multiparous. None of the patients had evidence of pelvic endometriosis. Chest roentgenograms during menses were normal or demonstrated densities presumed to be secondary to hemorrhage [41, 42]. Bleeding in the lung was localized by rigid bronchoscopy or thoracotomy. Endometrial tissue was found in all who underwent thoracotomy [40, 43, 44]. In 1, there was a cyst in the right middle lobe with endometrial tissue [44], in a second, a nodule containing endometrial tissue was found in the bronchial wall [43], and in a third a nodule in the right lung contained endometrial tissue [40].

Only 5 of the patients were treated; 3 with successful resection as above, 1 with testosterone, and 1 with x-ray castration [37]. One patient received no therapy [5], and in 1, therapy is unknown [19]. No long-term follow-up on these patients is reported.

We conclude that women who present with recurrent catamenial hemoptysis probably have pulmonary endometriosis. Nonetheless, other causes of hemoptysis might be excluded and one should attempt to visualize the bleeding site. Fiberoptic bronchoscopy is the procedure of choice and should be appropriately timed during the menstrual cycle to avoid performing the procedure after bleeding has stopped. If treatment is indicated at all, medical hormonal suppression, unless specifically contraindicated, is the initial treatment of choice. At the present time, however, there are insufficient data to predict the results of this treatment. If hemoptysis is life threatening, bronchoscopy to localize the bleeding and surgical excision of appropriate tissue should be performed.

Case Report

A 31-year-old black woman complained of hemoptysis and pleuritic chest pain. She had expectorated 30–50 ml of blood daily and the onset of these symptoms coincided with the onset of her menses. The patient had previously experienced 4 episodes of hemoptysis, each producing the same amount of blood and each beginning on the first day of menses. Each time she also experienced pleuritic pain in the right anterior chest and intense cramping and pelvic and lower back pain. All these symptoms stopped on the second or third day of menses. She never experienced any of these symptoms at times other than her menses. The patient had had 2 normal and uncomplicated pregnancies and all the problems described above began subsequent to the second pregnancy.

Fiberoptic bronchoscopy, performed on the third day of the patient's menses (hemoptysis had stopped by this time), revealed a normal nasopharynx; the only abnormalities seen were traces of old blood at the entrance to the right upper lobe and pale endobronchial mucosa throughout the right upper lobe. Biopsy specimens were not taken but the right upper lobe was washed. Cytologic examination revealed numerous bronchial cells, and hyperplastic cells, consistent with nonspecific inflammatory changes. All symptoms stopped by the fourth day of the patient's menses. She was advised to begin oral contraceptives but refused because she wanted to become pregnant. Subsequently,

she experienced 3 more episodes of catamenial hemoptysis. She then became pregnant and throughout this pregnancy had no further hemoptysis. She has not been seen since she gave birth.

Asymptomatic Pulmonary Nodules

There are five reports of asymptomatic endometrial pulmonary nodules [13, 17, 30, 35, 44]. All were noted as incidental x-ray findings and did not vary with the menstrual cycle. Three of the 5 lesions involved only the right lung. One patient [13], had bilateral lung nodules, and 1 patient [35] had a left lower lung nodule seen on chest tomography. This lesion had been diagnosed during an evaluation of catamenial pneumothorax involving the right lung and was never documented to be endometrial tissue. All other lesions were documented to be endometrial tissue.

One patient had clinical endometriosis. Three patients underwent thoracotomy. Granberg's patient [13] underwent needle biopsy of the lesion, and was subsequently treated with hormonal supression. The outcome is unknown Roger's patient [35] underwent diagnostic laparoscopy, and was treated successfully with hormonal supression.

Asymptomatic patients with pulmonary nodules should be evaluated in the same manner as anyone with a solitary pulmonary nodule, since no specific findings would suggest that these lesions are endometrial in origin. It is unclear whether these lesions, if identified by needle biopsy or flexible bronchoscopy and transbronchial biopsy, should be treated. All the patients cited above were treated with either hormonal suppression of ovulation or resection of the nodules. Since all these patients were asymptomatic and the nodules were benign, it is difficult to assess the therapeutic value of such treatment. Perhaps asymptomatic patients with biopsy-proven endometrial pulmonary nodules should be carefully observed, and hormonal or surgical treatment should be reserved until symptoms or findings develop.

Pathogenesis

No single hypothesis satisfactorily explains all the manifestations of pulmonary endometriosis. Barnes' suggestion that pleura, which develops from the coelomic cavity as does the peritoneum, may undergo melaplasia to form pleural endometrial tissue [2] does not explain diaphragmatic fenestrations nor the absence of endometrial tissue in many cases. Similarly, Charles' [6] postulate of "transdiaphragmatic spread secondary to pelvic endometriosis" does not account for diaphragmatic fenestrations nor for pulmonary nodules, endobronchial bleeding, the absence of pleural endometrial tissue, or recurrence following hysterectomy. Other hypotheses include bloodborne metastases of endometrial tissue [50], transdiaphragmatic spread of air or fluid through preexisting diaphragmatic perforation, check valve obstruction of the bronchus from endometrial implants and subsequent localized hyperinflation and pleural rupture

[24], idiopathic rupture of pulmonary blebs [26], and increased prostaglandin F2 with subsequent bronchoconstriction, vascular constriction, and alveolar rupture [36]. One might expect, however, that catamenial pneumothorax would be more common were the latter hypothesis correct. As with any "syndrome" this one may result from multiple origins; this opinion is supported by both the observation that few patients have more than one complication and the inability to explain satisfactorily all the findings by a single hypothesis.

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