A Case of Multisystem Endometriosis

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ABSTRACT

Catamenial pneumothorax is a rare complication secondary to pleural endometriosis. We present a case of a 37-year-old-female with a history of recurrent pneumothoraces with an associated temporal relationship to the onset of her menses. In addition to her recurrent pneumothoraces, on further evaluation, she was found to have multiple nodular masses within the omentum. A thoracoscopic biopsy was subsequently performed, which showed endometrial implants within the pleural space and within the omental cavity. The radiological features and pathogenesis of this rare disease are reviewed and discussed with reference to relevant literature.

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CASE REPORT

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A 37-year-old female with a past medical history of pelvic endometriosis presented to the emergency department in January 2006 with complaints of a three-day history of sharp right-sided chest pain and mild shortness of breath. She also complained of a 2-day history of diffuse, crampy abdominal pain, which she thought was secondary to her menstrual cycle. On physical exam there were decreased breath sounds over the right hemithorax and mild abdominal tenderness to palpation. Non-contrast chest computed tomography (CT) was obtained in the emergency department and findings showed a right sided pneumothorax (size~5-10%), as seen on the CT axial image (Fig. 1). She received supportive care and was subsequently admitted to the surgery service for monitoring. The patient did not require a chest tube. On day 2 of admission, patient showed marked improvement with no breathing difficulties and minimal chest pain and was subsequently discharged on day 3.

Three years later, in 2009, the patient was admitted to the hospital after she presented to the ED with acute chest and abdominal pain. She was again diagnosed with a pneumothorax on a chest CT, which also coincided with her menstrual cycle. CT imaging of the chest at that time showed a small right pneumothorax with a small right-sided serosanguinous pleural effusion (Fig. 1 and Fig. 2). Also seen was right-sided sub pleural nodularity (Fig. 2). Abdomen and pelvis CT revealed sub-centimeter solid omental nodules and bilateral cystic adnexal masses (Figs. 3 and 4). The subsequent pelvic transvaginal ultrasound better characterized the bilateral adnexal cystic masses measuring up to 4 cm in long axis with small amount of associated free fluid. In consideration of patient's clinical presentation and medical history, the findings were felt to be consistent with diffuse endometriosis. Subsequently the patient underwent a video-assisted thoracoscopy (VATS), which showed characteristic red and brown lesions on the pleural surface of the right hemithorax. Pathological evaluation of these lesions confirmed the presence of endometrial tissue within the pleura. The clinical course and pathology findings were consistent with multisystem endometriosis involving the pelvis, peritoneum and thorax complicated by catamenial pneumothorax secondary to pleural endometriosis.

Patient signed out against medical advice (AMA) shortly after the VATS and did not return for follow-up; thus no further information is available regarding treatment complications and/or disease progression.

DISCUSSION

Catamenial pneumothorax is lung collapse occurring in conjunction with menstruation (catamenial refers to a monthly), believed to be secondary to endometriosis within the pleural cavity. Catamenial pneumothorax remains a rare entity since Maurer and colleagues first described it in 1958 [1]. Although the number of diagnosed cases has increased significantly over the last several years due to advancements in imaging modalities and minimally invasive procedures, it is believed that this condition is still under-diagnosed with a reported incidence of 3-5%. The disease more frequently occurs in premenopausal women 30-50 years of age. [2]. Patients frequently have a history of infertility, severe endometriosis and recurrent spontaneous pneumothoraces (usually right-sided) within 72 hours of the onset of menses. The symptomatology varies significantly and most commonly presents with nonspecific findings of chest pain and difficulty breathing. Atypical symptoms such as cough and pain radiating to the neck, a secondary sign of diaphragmatic irritation [3], should yield further investigation.

In this case, our patient presented with chest pain and shortness of breath. She had a history of pelvic endometriosis and recurrent pneumothoraces, which had a temporal relation to the onset of her menses. The diagnosis of multisystem endometriosis with a history of catamenial pneumothoraces should be readily considered in a typical presentation of this "rare" disease. In our case, CT imaging readily provided the clinicians with guidance for "the next best step" in management. CT imaging clearly characterized the presence and size of the pneumothorax, and additionally revealed pleural and omental sub-centimeter nodularity, which later were biopsy proven to be endometrial implants. Additionally, CT evaluation of the abdomen and pelvis revealed bilateral non-specific adnexal cystic masses (Fig. 4),which were consistent with patient's history of endometriosis. Although not utilized in this case, MRI can be useful to further evaluate the pelvis prior to laparoscopy if the diagnosis of endometriosis is in question. *(However, it is important to note that magnetic resonance imaging (MRI) of the abdomen and pelvis is usually unnecessary and would not have provided any additional information if used in this case).

Other pathologies to consider in the differential of small pleural nodules include metastatic disease, primary pleural tumors, tuberculosis, sequelae of prior empyema, prior hemothorax, pleural plaques, and thoracic splenosis. It is difficult to definitively distinguish benign pleural diseases from malignant lesions via CT, however, there are certain characteristics that make a condition more likely to be malignant. Malignant pleural diseases tend to have nodular pleural thickening, usually >1cm, a pleural rind, and frequently have mediastinal involvement. The sub-centimeter nodularity seen on pleural and omental surfaces on CT imaging of this patient can be non-specific and the impression may include differential considerations of a rare primary pleural carcinoma or metastatic carcinoma from lung, breast, or ovarian primary malignancies. Tuberculosis should also be considered given the nodularity of the pleura, however, is highly unlikely in this patient given the lack of clinical history of exposure and lack of clinical symptoms including fever, cough, and weight loss. In our case, given the patient's young age and clinical presentation, the probability of these alternative diagnoses was significantly less likely.

Prior empyema or prior hemothorax are benign pleural conditions that should also be considered in this patient's differential. These conditions can cause irregular or smooth pleural thickening, usually <1cm, on CT imaging. They are frequently associated with sub pleural soft tissue hypertrophy, indicative of inflammatory changes. However since the patient has not had a prior history of empyema or hemothorax, nor does she have associated inflammatory changes on CT imaging, these diagnoses are also less likely.

Pleural plaques are associated with a history of asbestos exposure, and show calcifications in 10-15% of plain radiographs, which differ from the soft tissue nodules seen with endometrial implants. Furthermore on CT, the pleural plaques are characteristically present on the superficial surface of the diaphragmatic dome and the undersurface of the lower posterolateral ribs.

Thoracic splenosis is often seen following trauma in patients with diaphragmatic defects and is frequently left sided. Patients usually are asymptomatic, however some present with chest pain or hemoptysis. The diagnosis for thoracic splenosis is usually made via nuclear imaging, specifically when findings show technetium 99m tagged RBCs accumulation within the auto transplanted spleen.

It is important to note that on imaging these diagnoses can look similar, it is the clinical history and presentation which is important to consider and can best differentiate pleural endometriosis from other pathologies including pleural plaques and thoracic splenosis [11]. Ultimately, biopsy should be performed if the clinical picture remains unclear even after taking into consideration the clinical history, presentation and imaging findings, or if a confirmative diagnosis is necessary prior to the onset of treatment. Our patient underwent a VATS with biopsy, which showed characteristic red brown lesions and subsequent pathological evaluation readily identified endometrial implants within the pleura.

It is important to mention the role of contrast enhanced CT in discussing and narrowing our list of differential diagnoses listed above. First and foremost, endometrial implants, whether in the omentum or pleural related, will show a degree of enhancement. The pattern will vary depending on the size of the implant, however enhancement is characteristic. Every differential diagnosis considered in our case can show some degree of enhancement on CT examination. The first differential diagnosis, which can be, excluded relatively easily is thoracic splenosis. Thoracic splenosis will follow the same degree of splenic enhancement on every sequence, which would not be the case in endometrial implants. Next we can discuss empyema. Although an empyema will enhance, the pattern of enhancement will be in a peripheral ring-like pattern. Additionally, we would see the "split pleura" sign as well as fluid and gas contained within the lesion. Prior hemothorax, as discussed above, would only show

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enhancement if there are soft tissue plaques and residual scarring, although calcifications are more common. The most difficult to exclude based on morphology and varying patterns of enhancement would be metastatic disease. In sum, with the exception of thoracic splenosis, the above-mentioned differentials would show a nonspecific enhancement pattern, which could make the limiting the list of differential diagnosis quite difficult. With the help of a good clinical history and basic lab tests, the diagnosis of endometriosis should be relatively clear.

Treatment for catamenial pneumothorax includes video-assisted thorascopy for resection of endometrial implants along with talc pleurodesis to help prevent recurrence. If there is diaphragmatic involvement, diaphragmatic resection should also be considered. Surgical treatment in combination with at least 6-months of medical treatment including hormone modulating agents to suppress proliferation of endometrial implants has been shown to improve prognosis. Medical treatment most commonly includes GnRH agonists, however can also include oral contraceptives. Unfortunately, as with most severe cases of endometriosis, there is a very high rate of recurrence, close to 30-40%, after treatment of catamenial pneumothorax. This can be very debilitating and serve for a poor quality of life [6]. In our case, our patient did not receive proper treatment and follow-up because she chose to sign out AMA and did not return for post VATS evaluation. It is important to emphasize the importance of follow-up to decrease morbidity and recurrence.

The pathogenesis of the recurrent pneumothoraces and of pleural endometriosis still remains a topic of debate. The number of pleural endometriosis is increasing, thus it is important to understand the pathogenesis of this mysterious disease. There are three proposed mechanisms to explain the endometrial tissue presence within the pleura:

1. Coelomic metaplasia
2. Lymphatic or hematogenous embolization from the uterus or pelvis
3. Retrograde menstruation with subsequent transperitoneal-transdiaphgragmatic migration of endometrial tissue [3]

It has proven difficult to ascertain one theory as being correct, and although not one theory can explain all of the clinical manifestations of thoracic endometriosis syndrome (TES), current data is in favor of Sampson's theory of retrograde menstruation with subsequent peritoneal implantation of endometrial tissue [4]. This theory presumes impaired clearance mechanisms at the peritoneal level that allow the endometrial tissue to survive, implant, and proliferate. The spread to thoracic structures would occur as a consequence of lymphovascular embolization [3, 5] or due to a transabdominal-transdiaphragmatic migration of endometrial tissue [3]. However, there exist inconsistencies in this theory as well. First of all, if the spread to thoracic structures were the consequence of lymphovascular embolization, we would expect a bilateral involvement and subsequent bilateral pneumothoraces. This however, is not the case as the vast majority of pneumothoraces due to TES are unilateral and right sided [5, 6, 7]. Secondly, the transabdominal-transdiaphragmatic migration theory presumes there are defects/fenestrations present in the diaphragm of patients with TES. Current data suggests that the incidence of patients with TES with concurrent diaphragmatic defects is 29-66% [7,8], and in one study 21 of 86 patients with known diaphragmatic defects never developed catamenial pneumothorax [3]. Nevertheless, with these considerations being undertaken, the unique presentation of our case provides support in favor of Sampson's theory of retrograde menstruation [9].

Omental endometriosis is a rare entity on its own, and a case described by Measday and colleagues [10] discusses the combination of an omental tag in the pelvis and a ruptured endometrial cyst as the likely pathogenesis of omental endometriosis. We believe this in part played a role in the initial endometrial tissue seeding within the omentum. Perhaps clearance mechanisms allowed for the diffuse spread of the tissue, which then spread to thoracic structures. The question of whether the endometrial tissue reached the pleura via lymphovascular embolization or a transabdominal-transdiaphragmatic migration is unclear because neither has been confirmed in our patient. However, we believe the right-sided unilateral catamenial pneumothorax supports the theory of a transabodominal-transdiaphragmatic migration. In conclusion the purpose of our report is to provide a very rare encounter of a patient with concurrent pelvic, omental and pleural endometriosis, with the intent of offering insight into the pathogenesis of this rare and mysterious disease.

TEACHING POINT

Catamenial pneumothorax is a complication of pleural endometriosis, of which the pathogenesis is not completely understood. The most accepted theory is Sampson's theory of retrograde menstruation. Though plain radiographs are usually the initial best test and can identify nonspecific pneumothoraces, CT imaging is far more sensitive at showing pleural and omental nodularity. Biopsy and pathologic confirmation can be made via video assisted thoracoscopy (VATS), which will show characteristic red-brown lesions that can further characterized as endometrial tissue upon close pathological examination.

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FIGURES

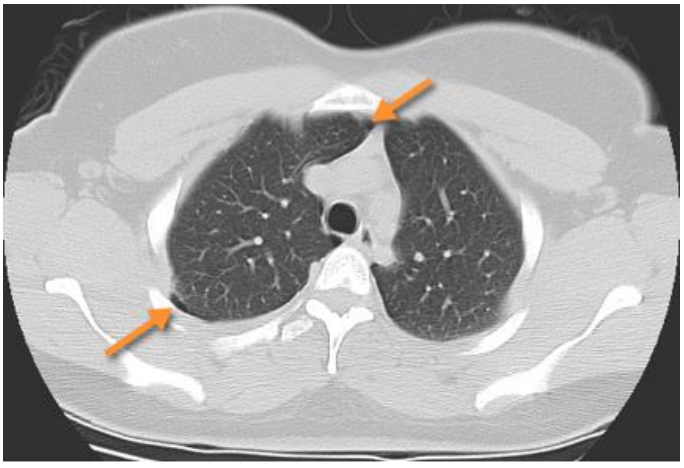

Figure 1: 37 year old female with catamenial penumothorax. Initial axial non contrast enhanced CT of the chest with lung windows demonstrating a non specific right lobe pneumothorax (yellow arrows).
Axial image of CT Chest without contrast in lung window. KV=120. Slice thickness: 3mm

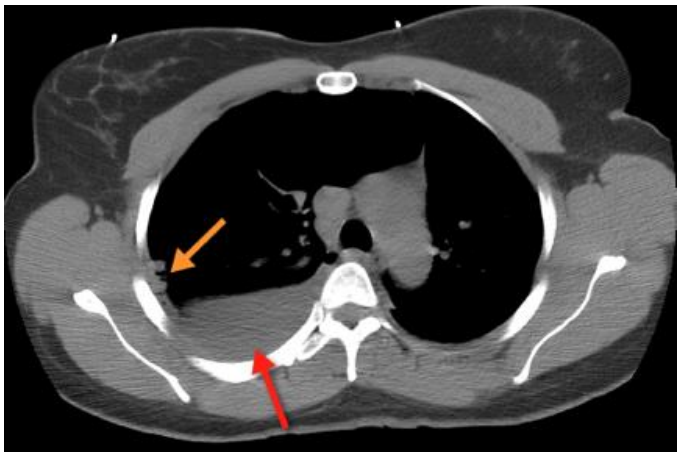

Figure 2: 37 year old female with catamenial pneumothorax. Follow up axial CT in soft tissue windows demonstrates pleural nodules (yellow arrow) which were biopsy proven endometrial implants. Red arrow demonstrates a unilateral right sided pleural effusion, which measured 35 hounsfield units.
Axial image of CT Chest without contrast in mediastinum window. KV=120. Slice thickness: 3mm

Figure 3: 37 year old female with catamenial pneumothorax. Axial CT of the abdomen shows multiple nodular densities located within the omentum (yellow arrows). These were biopsy proven endometrial implants.
Axial image of CT Abdomen without IV contrast, with oral contrast (1000cc of Gastrograffin) in abdominal window. KV=120. Slice thickness: 5mm

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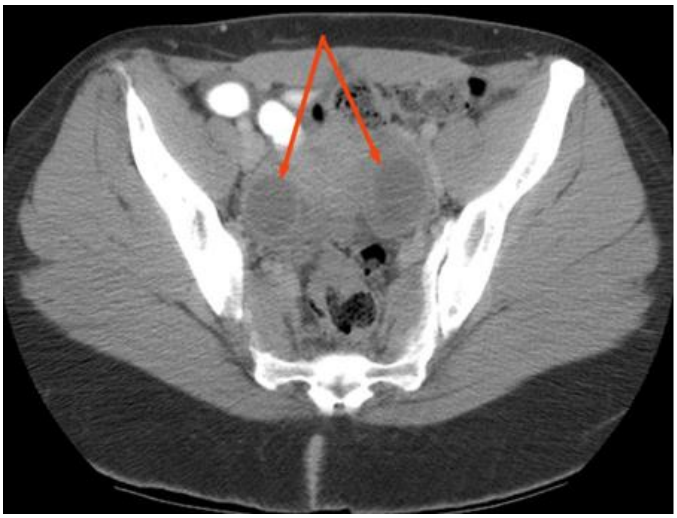

Figure 4 (left): 37 year old female with catamenial pneumothorax. Axial CT of the abdomen/pelvis with oral and IV contrast shows bilateral cystic adnexal masses (orange arrows). These were biopsy proven to be "chocolate cysts" compatible with endometriosis of the bilateral ovaries.
Axial image of CT pelvis without IV contrast, with oral contrast (1000cc of Gastrograffin) in abdominal window. KV=120. Slice thickness: 5mm

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ETIOLOGY	Three working theories of endometriosis within the pleura: 1) Coelomic metaplasia 2) Lymphatic or hematogeneous embolization from the uterus or pelvis 3) Retrograde menstruation with subsequent transperitoneal-transdiaphgragmatic migration of endometrial tissue The third theory of retrograde menstruation is currently the most widely accepted
INCIDENCE	3-5% , however there is a rising incidence secondary to improved imaging
GENDER RATIO	Female only
AGE PREDILECTION	Late premenopausal (30-50)
RISK FACTORS	History of pelvic endometriosis History of infertility
SYMPTOMS	Chest pain, shortness of breath, cough, pain radiating to neck (diaphragmatic pain). Symptoms in temporal relationship with onset of menses
TREATMENT	Multidisciplinary: ➔ Surgery- Video-assisted thoracoscopy (VATS) for disease assessment and resection of endometriosis along with talc pleurodesis. Diaphragmatic involvement should be treated with resection ➔ Medical- Surgery should be followed by 6-month period of hormonal treatment by GnRH agonists to achieve ovarian rest, blocking hormonal support to existing implants and preventing further seeding.
PROGNOSIS	High recurrence rate (30-40%)
IMAGING	Plain radiograph- Identify pneumothorax (right sided 90% of the time) CT- Small pleural nodules Video-assisted thoracoscopy to identify characteristic nodular brown lesions

Table 1: Summary table of Catamenial Pneumothorax

Disease	Clinical	Plain X-Ray	CT	Nuclear Imaging
Catamenial Pneumothorax	Chest pain, shortness of breath. Temporal relationship with onset of menses. Frequently associated with diaphragmatic defects.	Recurrent pneumothorax (usually right sided 90% of time)	More sensitive to show small pleural nodules. Often associated with omental nodularity seen on abdominal CT.	
Metastatic tumor (e.g. Adenocarcinoma)	Advanced staged carcinoma (History of lymphoma, breast or lung cancer), unexplained pleural effusions, weight loss	Uncommon to see pleural thickening or masses on chest radiograph	Nodular pleural thickening >1cm, pleural rind, mediastinal pleural involvement	PET could provide evaluation of location and number of endometrial implants, however can also lead to false positives for malignancy in the setting of endometriosis. Bone scan would be useful in differentiating metastatic disease to bone from endometriosis
Primary pleural tumor (Mesothelioma, leiomyosarcoma)	History of asbestos exposure, weight loss, unexplained pleural effusions	Uncommon to see pleural thickening or masses on chest radiograph	Nodular pleural thickening >1cm , pleural rind, mediastinal pleural involvement	
Empyema	Cough, febrile patient	Pleural effusion with loculation, no change on decubitus films	CT is nonspecific for diagnosis, irregular or smooth pleural thickening usually <1cm, loculations. Some associated subpleural soft tissue hypertrophy (fat tissue density) suggesting inflammatory changes,	
Tuberculosis	Recent history of travel, cough, fever , weight loss		Irregular or smooth pleural thickening usually <1cm.	
Prior hemothorax	Chest pain, shortness of breath, history of prior trauma		CT is nonspecific for diagnosis, irregular or smooth pleural thickening usually <1cm, Some associated subpleural soft tissue hypertrophy (fat tissue density) suggesting inflammatory changes,	
Pleural Plaques	History of asbestos exposure	Nodularity of parietal pleural, 10-15% of cases depict calcification	More sensitive and specific to show discrete raised parietal pleural thickening. Characteristic locations include diaphragmatic dome and undersurface of the lower posterolateral ribs.	
Thoracic Splenosis	History of trauma, more commonly occurs with diaphragmatic defect. Usually asymptomatic, but can cause chest pain or hemoptysis.		Multiple nodules in pleural space	Scintigraphy with technetium 99m tagged red blood cells confirms the diagnosis with radiotracer accumulation in the autotransplanted splenic tissue

Table 2: Differential diagnosis table of Catamenial Pneumothorax

ABBREVIATIONS

AMA = Against Medical Advice
CT = Computed Tomography
GnRH = Gonadotropin Releasing Hormone
MRI = Magnetic Resonance Imaging
TES = Thoracic endometriosis Syndrome
VATS = Video-Assisted Thoracoscopy

KEYWORDS

Endometriosis; catamenial pneumothorax; pneumothorax; abdominal pain; chest pain; chocolate cysts

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