

Catamenial Hemoptysis Secondary to Giant Mediastinal Endometriosis with Pericardial Involvement - Case Report of Thoracic Endometriosis Syndrome

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Abstract

Catamenial hemoptysis with mediastinal and pericardial involvement is a rare presentation of Thoracic Endometriosis Syndrome (TES) which is seldom proven via histopathology. TES is a clinical manifestation of the presence of endometrial tissue within the thorax cavity. Typically, it is being described as consisted of 4 components including catamenial pneumothorax, haemothorax, haemoptysis and pulmonary nodules¹. Some even suggested to include catamenial chest pain, pleural effusion and even diaphragmatic hernia². The pathophysiology of TES still is being debated and often considered as multifactorial³. The basis of the theory behind pathophysiology of TES is based on their clinical presentation and predominance characteristics and hence the description of variety presentation of TES is important to be documented as it will help us to develop more understanding regarding this syndrome. We reported a case of catamenial hemoptysis secondary to large anterior mediastinal endometriosis with radiological, operative imaging and histopathological evidence of mediastinal and pericardial involvement.

Keywords

Catamenial hemoptysis, endometriosis, precardial

Introduction

There are four main theories describing the pathophysiology of Thoracic Endometriosis Syndrome (TES), this may include the retrograde menstruation theory, coelomic metaplasia theory, lymphatic and hematogenous dissemination theory, and prostaglandin theory. None of this theory can solely explain the whole variety of presentation of TES and hence why it is widely accepted to be multifactorial.

Observation of the presentation of TES is important because each presentation will provide us with a clue of the actual pathophysiological process behind. Some atypical presentation is worth to be documented and shared as it provides us more understanding of how endometrial glands and stroma can behave in the thorax cavity.

We report a case of histologically proven pericardial involvement of mediastinal endometriosis. This is an unusual presentation of TES where the patient presented with catamenial hemoptysis due to large anterior mediastinal endometriosis which is densely adhered to pericardium and part of middle lobe of the right lung.

Case Report

A previously well 45 years old lady, para 2, with normal regular menses presented with 2 years history of intermittent hemoptysis. It occurred usually during the onset of her menses and lasted for about 2 to 3 days. She had frequent visit to the general practitioners every time she developed the hemoptysis; however, no cause can be ascertained, and she frequently discharged with a course of antibiotic and cough medications. Eight years earlier, she underwent gynaecological procedure - evacuation of retained product of conception. After 2 years of having the episode of intermittent hemoptysis, she had a chest X-ray which revealed a significant large opacity over right middle zone large. She was then referred to tertiary care center. On examination, she was medium built, well nourished, air entry was equal with no added sound over both lungs, and respiratory and cardiovascular system examination was unremarkable.

Blood investigation and electrocardiogram was normal. Blood and sputum cultures including workup for Mycobacterium Tuberculosis (which is predominant in patient's local setting) were negative. Echocardiogram shows no cardiac abnormalities. Chest X-ray shows a large centrally located, lobulated right perihilar opacity (Figure 1). Contrast enhanced Computed Tomography (CECT) scan shows a well-defined, lobulated and encapsulated heterogeneously enhancing mass, with irregular border seen at the right anterior aspect of mediastinum. Radiological measurement of the mass was 5.5 x 9.2 x 8.8cm (AP x W x CC) with local infiltration (Figure 2). Soft tissue density was visualized within right middle lobe bronchus suspicious of extension of the lesion. Computed tomography guided biopsy of the mass confirmed the presence of endometrial glands and stroma.

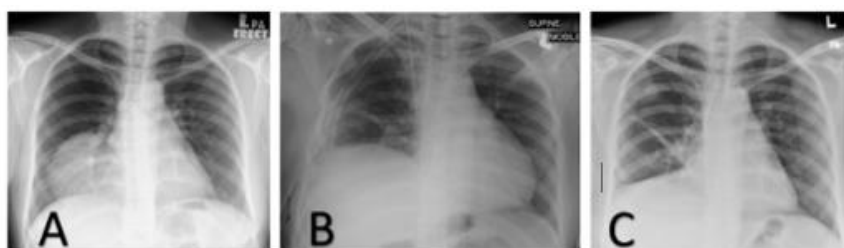


Figure 1. A: Chest x-ray upon presentation, B: Immediate post-operation, C: Reassessment

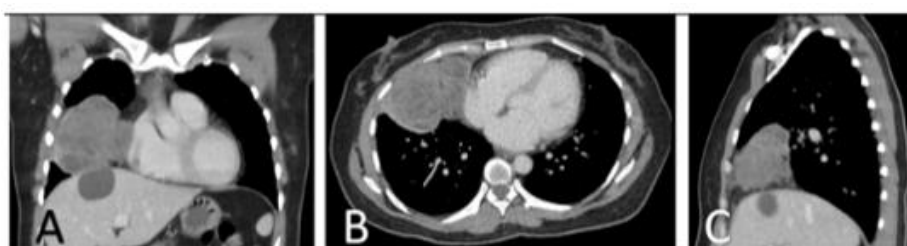


Figure 2. (Preoperative CT Thorax images) Demonstrating well-defined, lobulated, and encapsulated heterogeneously enhancing mass 5.5 x 9.2 x 8.8cm (AP x W x CC) with irregular border seen at the right anterior aspect of mediastinum with local infiltration. A: Coronal view. B: Axial view. C: Sagittal view.

Patient underwent video-assisted thoracoscopic surgery (VATS) which then converted to right thoracotomy and resection of anterior mediastinal mass with concomitant partial pericardiectomy. During surgery, there was a large pedunculated solid tumor, well encapsulated with whitish fibrous tissue (Figure 3). It appeared to have invaded into the right middle lobe as blood was seen from the middle lobe bronchus. The tumor is densely adhered to mediastinal pleura, pericardium and part of middle lobe of the right lung. The tumor was then resected with half of the middle lobe and part of pericardium due to the dense adhesion. Histopathological assessment showed presence of endometrial glands and stroma at the peripheral aspect of resected pericardium. Red blood cells were seen within the alveolar spaces. The alveoli, bronchi, and bronchiolar tissues were normal. The resected anterior mediastinal tumor composed of endometrial glands and stroma. Most of the glands were cystically dilated containing secretion. Post operatively, she was given 6 months course of intramuscular gonadotrophin releasing hormone agonist. On follow up she was well with no recurrence of hemoptysis or any other catamenial symptoms.

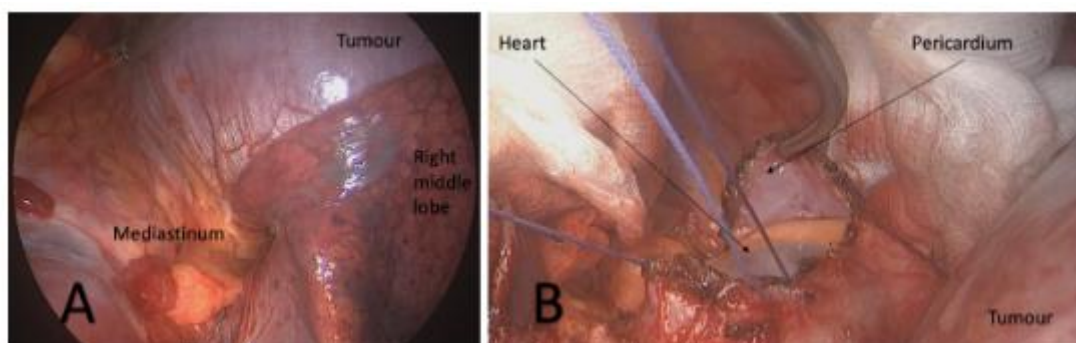


Figure 3. (Intraoperative pictures) A: Large solid tumour, well encapsulated by whitish fibrous tissue, attached to mediastinal pleura and pericardium and middle lobe of the lung. B: Opening of pericardium with heart exposed, tumour densely adhered to pericardium requiring partial pericardiectomy.

Discussion

Thoracic endometriosis syndrome (TES) is the clinical manifestation of the presence of endometrial tissue in thoracic cavity. The symptoms and signs depend on the site of implantation of the tissue. Variety of presentations has been described including catamenial pneumothorax, hemothorax, hemoptysis, pleural effusion, and pulmonary nodules.

There are four main theories of pathophysiology of TES⁴. The reason of the many theories behind TES is because none of the single theory can adequately explain the variable presentation of TES, hence it is mostly accepted as being multifactorial. Retrograde menstruation theory is supported by the high percentage of patient with TES having concomitant pelvic endometriosis. It is further strengthened by the fact that the most common findings of surgical Video Assisted Thoracoscopic Surgery (VATS) are diaphragmatic defect⁴. The defect could be from congenital or acquired fenestration. Most of TES are right sided predominance and about 79% endometriosis in TES found to have deposit on diaphragm, 14% on pleura and about 4% on the lung⁵.

Coelomic metaplasia theory explained that the mesothelial cell of pleural and peritoneum are subjected to influence by estrogen and progesterone, and they can develop into endometrial tissue. This is supported by the finding of endometrial tissue on the pleura in Mayer-Rokitansky-Kuster-Hauser syndrome patients despite the absence of normal uterus.

Lymphatic and hematogenous dissemination theory are supported by the findings of deposition of endometrial tissue either locally (scar tissue, intrapelvic endometriosis) or in distant like bronchopulmonary system, nervous system including brain or even bone. There have been multiple reported cases of different sites of endometrial tissue deposition. The other theory is the prostaglandin theory which is more focused on describing the catamenial pneumothorax which is the most common presentation of TES. It describes the prostaglandin F2a which is present in plasma of women during menses. It may cause constriction of blood vessels and bronchioles causing alveolar rupture especially if there's previously formed subpleural blebs or bullae. This is supported by the negative findings of endometrial tissue in some patients with catamenial pneumothorax.

This case is interesting because rarely we get to prove pericardial involvement of endometriosis in TES. To the best of the writer's knowledge, no other cases in the published literature that demonstrate pericardial involvement via the VATS approach. The other two cases available in the literature was through laparoscopic surgery approach⁶. This case also demonstrates histological pericardial involvement without concomitant diaphragmatic involvement, which challenges the theory of retrograde menstruation through a defect in the diaphragm.

Lesion found on the right thoracic cavity of this case consistent with around 80% of the TES reported⁵. Patient had hemoptysis which only happened in around 3.3% of TES cases⁷. Bronchoscopy finding is frequently negative in many TES cases unlike this case which shows some finding of blood oozing from right middle bronchus⁴.

Getting the diagnosis for TES is challenging because of the cyclical behavior of the endometriosis lesion, some lesion may resolve spontaneously at the time of imaging or operation. To get a tissue sample or histopathological diagnosis is challenging. Some lesions may be very small but still can cause significant symptoms at the time of menses. Ideally, to manage this kind of cases, one should opt for non-invasive approach before invasive management. Despite that, some physician may be cautious in starting medical treatment for endometriosis for TES due to lack of evidence. The author however agrees that we should have a high index of clinical suspicion when detailed and accurate history from patient about the symptoms can be catamenial in nature. Patient then should be counselled in comprehensive manner as to what treatment options are available.

Conclusion

In dealing with cases suspected of TES, we should take detailed history of the presenting symptoms to establish the catamenial characteristic of the symptoms. Look for any history of gynaecological procedure as it has been reported that many of those patients with catamenial hemoptysis had obstetrics or gynaecological procedures in the past⁸. Imaging and investigations should be done with consideration of the patient's cyclical menses to increase the chances in getting the diagnosis.

We learned that cause of TES could be multifactorial as highlighted by this case with atypical presentation of TES. This case finding showed the absence of diaphragmatic fenestration despite occurrence on the right side of the thorax, no pleural involvement and no pneumothorax. The most likely explanation of the pathophysiological development for this case could be the lymphatic and hematogenous dissemination theory due to the location of the lesion which is within the mediastinum. The patient also had history of evacuation of retained product of conception 8 years earlier which could trigger the endometrium tissue dissemination. Another lesson that we learned is that, although endometriosis lesion not behaving like an aggressive malignant lesion, however it can be locally infiltrative as shown by the presence of blood within

alveolar tissue. It is also densely adhered to the pericardium with the presence of the endometrial tissue and stroma within peripheral part of the pericardial tissue.

Management should be patient-centric multi-disciplinary approach with focused on patient's preference of trial of medical treatment, followed by more invasive investigation and surgery if conservative treatment failed. Patient's own family planning should be addressed as well. Clinician should have high index of clinical suspicion if the chest pain, pneumothorax, hemoptysis or hemothorax is catamenial in nature, and we should be vigilant that there could be a dearth in findings despite our best attempt in getting the diagnosis.

Conflict of interest

Authors don't have any conflict of interest.

Patient's consent

Obtained.

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Department of Pathology, Hospital Queen Elizabeth and Department of Radiology, Hospital Queen Elizabeth 2.

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