

CASE REPORT

Pleural endometriosis - A true enigma!!

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ABSTRACT

A case of a Burkinabe woman who was 41 years old admitted to ER with right sided chest pain on & off, and shortness of breath with increasing intensity is described here. she revealed that she had been experiencing similar episodes of recurrent ride sided chest pain during menstruation for a few months. She had no menstrual problems. but decreased breath sounds on the right side, lower zone. Impaired percussion on the right lower lobe and hyper-resonance in the right axilla. On evaluation her Chest X-ray showed pleural effusion. When the CT was done it was evident of a pleural honeycombing, right sided moderate hydropneumothorax with mild contralateral mediastinal shift. Pleural biopsy showed that HPE revealed content of hemorrhage and fibrin deposits. Infiltration by mononuclear inflammatory cells, occasional neutrophils and hemosiderin laden macrophages with endometrial epithelium. The immunohistochemical study showed CD10, P63, CA125- positive concluded Pleural endometriosis.

Key words: Pleural endometriosis, pleural effusion, thoracic endometriosis syndrome, endometriosis-related pleural effusion.

INTRODUCTION

Common causes of bloody pleural effusion include trauma, iatrogenesis, and malignancy. Thoracic endometriosis syndrome (TES) is a rare disorder characterized by the presence of functioning endometrial tissue in pleural, lung parenchyma, and airway (1). A majority of patients with TES present catamenial pneumothorax (73%), while ~14% of the cases show hemothorax (2). Although endometriosis-related pleural effusion (PE) is a benign and treatable disease, it is important to take it into consideration especially in women of childbearing age. The clinical characteristics of TES have been reviewed previously (2, 3). These reviews primarily focused on catamenial pneumothorax or catamenial chest pain. The clinical features of endometriosis-related PE are limited. Diagnosis of endometriosis-related PE is challenging and depends on cytological and/or histopathological examinations demonstrating endometrial cells in pleural fluids (PF) or tissue. However, there were no data examining the diagnostic yield of diagnostic options for endometriosis-related PE. Currently, there is no standard treatment for endometriosis-related PE. Treatment options include hormonal therapy [progestational agents, danazol, and gonadotropin-releasing hormone

(GnRH) analogs], thoracic surgery [removal of ectopic endometrial tissue, closing diaphragmatic defects, pleurectomy and pleurodesis through video-assisted thoracoscopic surgery (VATS) or thoracotomy], hysterectomy and bilateral salpingoophorectomy (HBSO), and combined therapies. Evaluations of the outcomes of PE treatments are lacking. Here, a case of pleural endometriosis is discussed. Endometriosis in this case was confirmed by PF cytological examinations.

CASE REPORT

CLINICAL PRESENTATION

A 41 year old female of Burkinabe nationality presented to ER with right sided chest pain on & off, and shortness of breath with increasing intensity. She was a Non-smoker with a BMI of 20.94. She has a 17year old daughter and had a normal delivery. She gave a history of Open myomectomy done in 3 years before. On deeper probation, she revealed that she had been experiencing similar episodes of recurrent ride sided chest pain during menstruation for a few months. She had no menstrual problems.

When she was examined there were no other medical issues. She had no cardiovascular problems, symmetrical chest expansion, but decreased breath sounds on the right side, lower zone. Impaired percussion on the right lower lobe and hyper-resonance in the right axilla was observed. On evaluation her Chest X-ray showed pleural effusion. When the CT was done it was evident of a pleural honeycombing, right sided moderate hydropneumothorax with mild contralateral mediastinal shift.

MANAGEMENT

Subject was admitted to ICU, she was administered antibiotics. Later Video-assisted thoracoscopic surgery was done and sent for pleural biopsy. Pleurodesis was done. She recovered fully, and the ICD removed after 1 week.

Pleural biopsy showed that HPE revealed content of hemorrhage and fibrin deposits.

HISTOPATHOLOGY

Infiltration by mononuclear inflammatory cells, occasional neutrophils and hemosiderin laden macrophages with endometrial epithelium.

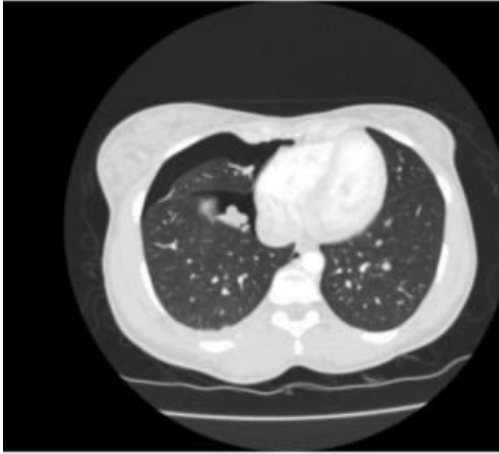
IHC MARKERS

The immunohistochemical study showed CD10, P63, CA125 - positive. Impression - Pleural endometriosis.

FOLLOW-UP

She had 3 more emergency visits in the following 3 months with similar complaints. She reported a right sided chest pain, shortness of breath. Each time - managed symptomatically. A Gynecology opinion was taken. Later she was given the therapy with GnRH analogues - Triptorelin 3.75mg IM once a month for 3 doses. Serial monitoring with sr CA125 levels was done and it was observed as a decreasing trend for the next 2 years. After finishing 3 months triprorelin course, she was kept on Dienogest 2mg OD for 6 months.

She had major symptomatic relief over the next 6-12 months after commencement of hormone therapy. She did not require any further admissions for pleural effusion. She was also regularly screened for any pelvic pathology - but nothing abnormal detected.

Figure 1: Right lower lobe cystic nodule seen with pneumothorax**DISCUSSION**

Hart first identified thoracic endometriosis syndrome (TES) in 1912, and Schwartz did so again in 1938, referring to it as a "lung tumour" that bled every month. 1 Since then, we've made progress in our knowledge of the pathophysiology, diagnosis, and treatment of this uncommon ailment. The diagnosis is still uncommon, and there are no randomised control trials to determine the best course of treatment. Two sizable retrospective studies on TES have been conducted. In contrast to pelvic endometriosis, which has a younger peak incidence (25-29), they discovered that the mean age of onset for this condition is 35 years, with the highest incidence occurring between 30 and 34 years of age. 2,3 Pneumothoraces were the most typical manifestation, occurring in 73% of the group. It's interesting to note that the right hemithorax was implicated in over 90% of presentations. 2 The usage of thoracoscopy had grown throughout the 14 years between these two investigations, and by 2010, 85% of the group had undergone a thoracoscopy or thoracotomy. 3 They discovered no conclusive link between diaphragmatic abnormalities and pneumothorax. Conversely, a larger load of pleural deposits of endometrial tissue was linked to a higher risk of haemothorax. 3 The endometrial tissue was formerly believed to move retrogradely into the peritoneal cavity through the fallopian tubes, into the thorax by diaphragmatic fenestrations. 1 These thoracoscopic results, however, refute that idea. 3 According to a different notion, physiologic triggers might cause pleural epithelium to change into endometrial tissue. However, this cannot account for the right-sided dominance observed in TES cases. One of the most recent theories proposes that lymphatic embolization, similar to how cancer metastasizes, is how endometrial transplantation happens, explaining both the thoracic and other sites of implantation. 4 The actual mechanism, though, is probably complex and combines all of these possibilities. 5 A "catamenial pneumothorax" is the TES's most typical manifestation. This type of pneumothorax is one that happens within the first 72 hours of the onset of menstruation. 6 That doesn't always happen during menstrual cycles. It accounts for 80% of TES manifestations and the underlying aetiology of 2.5%–5% of spontaneous pneumothoraces in females. 6 Catamenial haemothorax is a less common condition that frequently manifests as chest pain and cough and shows up on a chest X-ray as a pleural effusion. Endometriosis in the pelvis and peritoneum is linked to haemothoraces. Cyclic hemoptysis is a rare kind of TES presentation that usually affects younger age groups. 3 Synchronous hemoptysis with menstruation will be how this will show up. The lesions, which are usually visible at bronchoscopy, are brought on by parenchymal deposits of endometrial tissue, especially during menstruation. 5 Pulmonary nodules, like the ones we saw in our subject, are the most uncommon TES symptom. According to the research, lung nodules are observed in 2% to 6% of TES patients. 2 Compared to the other presentations,

which are more frequently seen in patients who are younger, they are most common in women over the age of 35. 1 For TES to be diagnosed, a high index of suspicion is necessary. This should be taken into consideration much more if the lady is known to have pelvic endometriosis or exhibits symptoms of it. GnRH agonists are the preferred treatment for TES in women who want to maintain their fertility. The current management of TES is based on the inhibition of ovarian oestrogen secretion. 1 But, as VATS becomes more common, it is quickly replacing it in order to provide quick identification of thoracic endometrial deposits and to also permit urgent starting treatment. This method can be used to perform a number of treatments, such as the removal of endometrial implants, closing of the diaphragmatic fenestrations, and abrasive mechanical or chemical pleurodesis. 1 Endobronchial laser ablation can be taken into consideration if hemoptysis is the predominant complaint, although there isn't much agreement on the best course of action given the lack of information. 5 This combination therapy may be the best strategy to avoid long-term morbidity because surgical treatment followed by hormone therapy has been linked to no recurrence in a follow-up period as long as 45 months. 1 Hysterectomy with bilateral salpingo-oophorectomy is the only effective treatment for systemic endometriosis, however it should only be used when all other treatment options have failed. It's interesting to note that thoracic endometriosis might get worse or return after a subsequent hormone replacement.

CONCLUSION

This case is extremely rare as there is evidence of significant thoracic disease without any evidence of pelvic endometriosis. Thoracic endometriosis is a rare but important differential in menstruating women. A diagnosis of thoracic endometriosis does not necessarily need an established diagnosis of systemic endometriosis. Increasingly, video-assisted thoracoscopic surgery procedures are being used to diagnose and treat the complications of thoracic endometriosis.

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