



BMH Med. J. 2023;10(3):71-78. **Case Report**

Thoracic Endometriosis - Report of Two Rare Cases

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Abstract

Thoracic endometriosis syndrome (TES) is a group of diseases caused by ectopic endometrial tissue in the thoracic cavity in women of child bearing age. This extremely rare condition is difficult to diagnose, unless direct correlation with menstrual cycle is established. The syndrome consists of pleural form such as catamenial pneumothorax, non-catamenial endometriosis-related pneumothorax and haemothorax; and parenchymal form such as catamenial haemoptysis and lung nodules. Symptoms are typically cyclical and recurrent, with a right-sided predominance. Computed tomography (CT) is the first-line imaging method, but is poorly specific; therefore, its main role is to rule out other pulmonary diseases. However, in women with a typical clinical history, some key CT findings may help to confirm this syndrome. MRI can also assist with the diagnosis, by showing signal changes typical of haemorrhage within diaphragmatic or pleural lesions. Here we present two cases of pleural forms of endometriosis one presenting as haemothorax and the other as recurrent pneumothorax. Both these presentations are very rare.

Key words: Catamenial, endometriosis, haemothorax.

Introduction

Endometriosis is a common gynaecological disorder, affecting 10-15% of women of reproductive age. It is considered to be due to extrauterine growth of endometrial tissue, including endometrial glands and stroma. The ectopic tissue is typically located in the peritoneal cavity, most often in the pelvis, but endometriosis has been reported in nearly all body compartments [1]. Although rare, the thoracic involvement is the most frequent extra-abdominopelvic site of endometriosis. Up to 80% of women with TES present with concomitant pelvic endometriosis. Thoracic endometriosis syndrome (TES) is the term used to denote various clinical manifestations resulting from the presence of functional endometrial tissue in a thoracic structure. Endometrial tissue is seeded in visceral or parietal pleura, lung parenchyma, airways, or diaphragm. Clinical manifestations are more likely to

occur during menses, because of the hormonal responsiveness of ectopic endometrial tissue [2]. TES includes five well-recognized clinical entities grouped into two forms, namely the pleural form with catamenial pneumothorax (CP), non-catamenial endometriosis-related pneumothorax (NCP), and catamenial haemothorax (CHt); and the pulmonary form with catamenial haemoptysis (CH), and lung nodules [2,3]. Approximately 90% of patients with TES experience catamenial thoracic pain and different entities may be associated. The right hemithorax is involved in more than 90% of all forms, with the exception of nodules [2]. Videothoracoscopy is the preferred surgical technique. Intraoperative evaluation has shown the presence of diaphragmatic anomalies in up to 84% of cases.

Case Reports

Case 1

A middle aged lady presented with right sided pneumothorax. During the last 3 years she had 4 such episodes. She was investigated earlier for right sided pneumothorax and treated with intercostal drainage (**Figure 1**). There was no history of fever cough, loss of appetite or loss of weight. She had no contact with pulmonary tuberculosis. Following this she was symptom free for 7 months and developed first ipsilateral recurrence. She was advised intercostal drainage, but preferred conservative approach with breathing exercises and symptomatic measures. Within a span of one year she again had 2 recurrences which were managed conservatively. The present episode was the 4th recurrence and was severely symptomatic. X-ray showed 80% pneumothorax (**Figure 2**) and decided for intercostal drainage. Since it was a fourth recurrence, medical thoracoscopy was done. On thoracoscopic evaluation diaphragmatic pleura and mediastinal pleura showed brownish black indurated lesions suggestive of endometriosis (**Figure 3**). Talc pleurodesis was done and drain removed after 48 hours. There was no reported recurrence for the last 5 months and the patient is on regular follow up. The final diagnosis was recurrent pneumothorax due to pleural endometriosis.

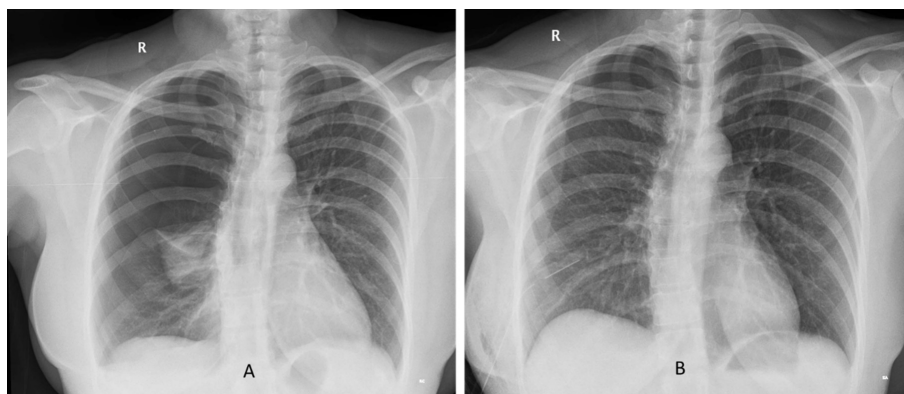


Figure 1A: Showing complete right sided pneumothorax. **1B:** Showing complete expansion on intercostal drainage.

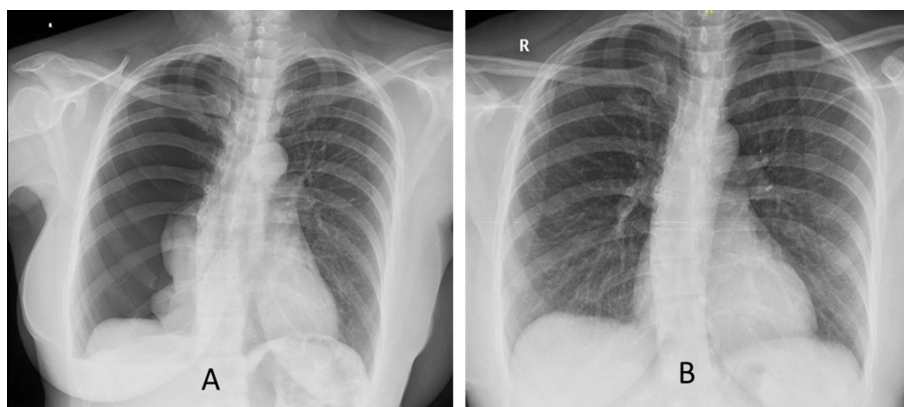


Figure 2A: Right sided pneumothorax (4th recurrence) and **2B:** X-Ray taken after medical thoracoscopy and talc pleurodesis.

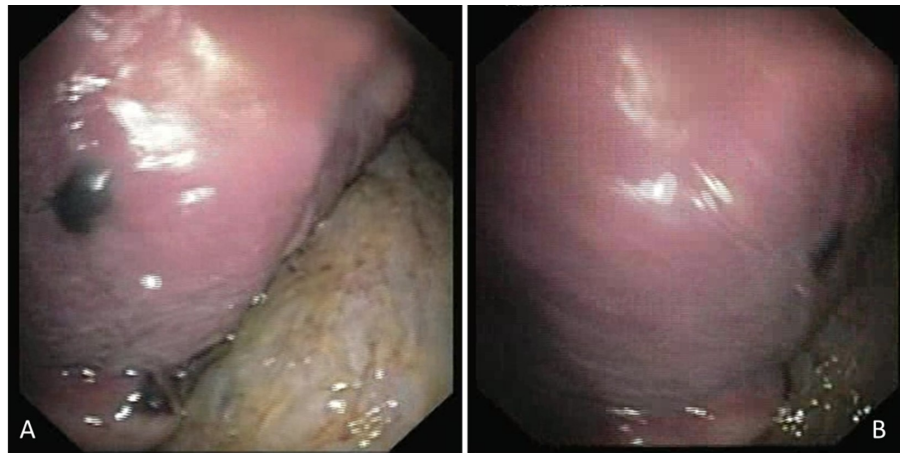


Figure 3: A & B: Thoroscopic view mediastinal pleura showing dark brown nodules suggestive of pleural endometriosis.

Case 2

Another middle aged female presented to the ED with severe breathlessness for 7 days. She gave history of intermittent, right sided chest pain associated with chest tightness and breathlessness during the last 3 years. She was evaluated in a local hospital and based on the X-ray findings she was referred to our hospital. She used to have severe bleeding during her periods for the last 3 years. Each time bleeding lasted for 5-7 days. She developed anaemia and pelvic work up revealed a left ovarian cyst.

There was no history of cough or haemoptysis. Clinically she had moderate pleural effusion on the right side which was confirmed by chest X-ray (**Figure 4A**). Routine blood examination revealed anaemia (Hb 8.3 gm/dL), normal total ($7.600/\text{mm}^3$) and differential WBC counts (N-71, E-04, B-00, L-22, M-03), and mild increase of ESR (28 mm/h). She underwent therapeutic as well as diagnostic thoracentesis. Physical observation of pleural fluid revealed a deep brownish-red haemorrhagic fluid (**Figure 1B**), closely resembling blood. Haematocrit of pleural fluid was more than 50% of blood. Total cell count of pleural fluid was $650/\text{mm}^3$, and 60 % were lymphocytes, with plenty of RBCs. It was a low ADA lymphocytic effusion. There were no malignant cell or AFB detected in the fluid. Contrast enhanced CT thorax and abdomen was taken. CT thorax showed moderate right sided pleural effusion without any evident parenchymal lesions (**Figure 2A**), pleural lesions or mediastinal lymphadenopathy. CT abdomen revealed a large cystic lesion in the left ovary (27 x 24 mm) and a left adenexal mass of 37 x 29 mm abutting left ovary and uterus. There was moderate ascites. (**Figure 2B**) Estimations of CEA of pleural fluid (5 $\mu\text{g/dL}$) and CA-125 level of blood (55.1 U/L) were performed. Both CEA and CA 125 were increased. Medical thoracoscopy with semirigid thoracoscope was performed which showed moderate hemorrhagic pleural fluid in the pleural cavity. Diaphragmatic pleura and mediastinal pleura showed multiple haemorrhagic cysts (**Figure-3A**), brownish nodules and gunshot lesions (**Figure-3B**) suggestive of pleural endometriosis. All the cysts were broken and biopsy taken from the nodules. Chest was closed after putting intercostal drainage tube. Histopathological examination of the biopsy specimen showed foci of endometrial glands lined by pseudostratified columnar epithelium with surrounding cellular endometrial stroma (**Figure 4A & B**). Considering the clinical features, results of various investigations and histopathology, diagnosis of pleural endometriosis was confirmed. Gynaecology opinion was sought and they suggested hysterectomy with bilateral salpingo-oophorectomy as an elective procedure. Till then menstruation will be suppressed with hormonal therapy.

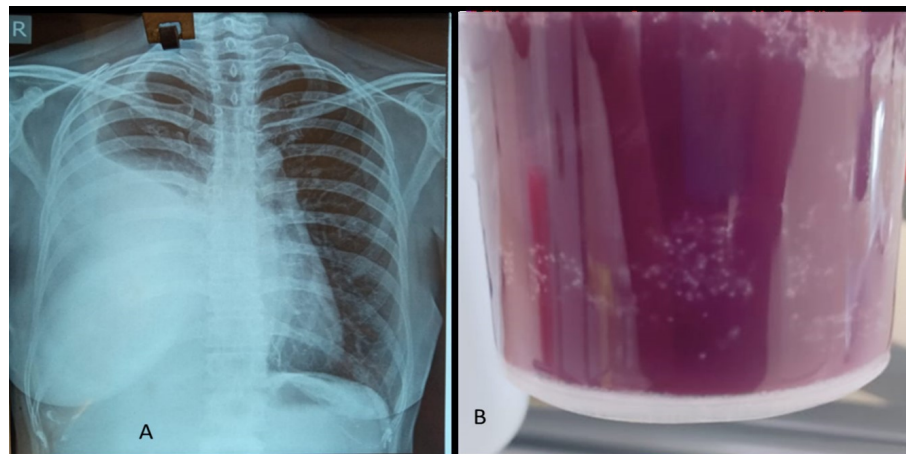


Figure 4A: X Ray Chest showing right sided moderate pleural effusion. **4B:** Showing aspirated reddish pleural fluid suggestive of haemothorax.

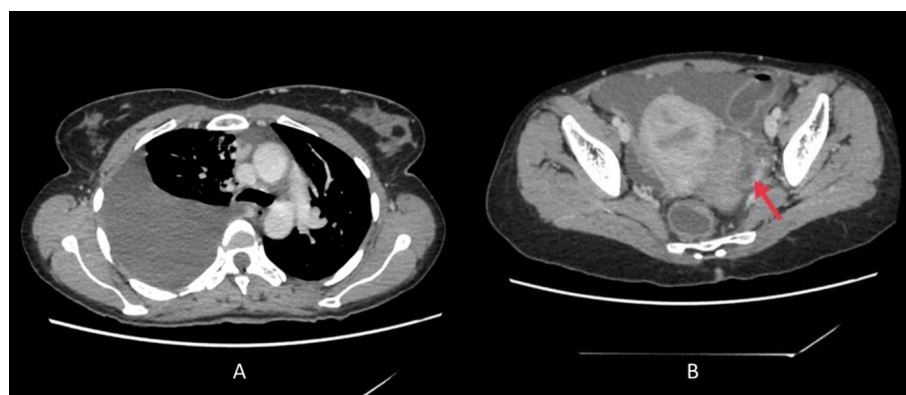


Figure 5A: CT Thorax showing right sided pleural effusion. **5B:** CT abdomen showing left ovarian lesion and an adnexal mass on left side (Red arrow).

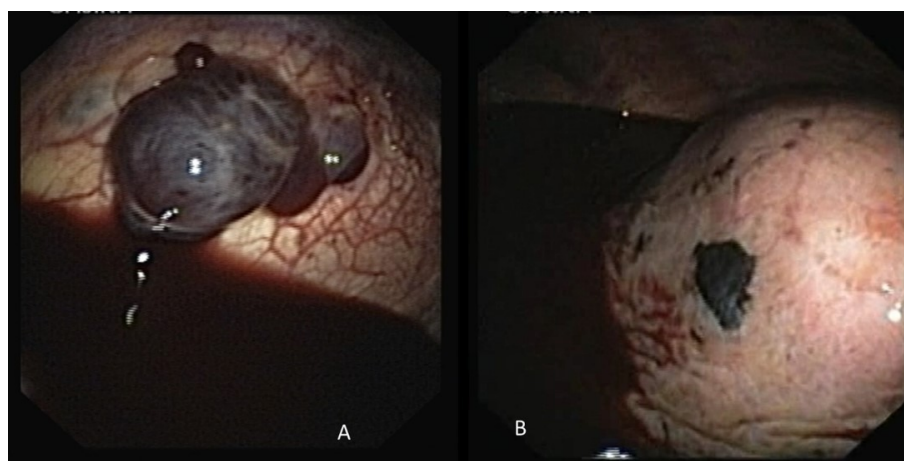


Figure 6A: Thoracoscopic view showing hemorrhagic fluid and dark brown cystic lesion on the diaphragmatic pleura. **6B:** Showing brownish black indurated lesion suggestive of "Gunshot lesion" of endometriosis.

Discussion

Pleural endometriosis is considered rare but may be under-diagnosed [2,4]. There are three theories of the pathogenesis of endometriosis: implantation, vascular or lymphatic metastasis, and coelomic metaplasia [5,6]. There were only 38 pathologically confirmed cases of thoracic endometriosis in the literature till the year 2007. A study on 110 patients showed that the mean age at presentation was 35 +/- 0.6 years, with a range from 15 to 54 years. Interestingly, the peak incidence for pelvic

endometriosis is between 24 and 29 years, whereas the peak incidence for thoracic endometriosis is approximately 5 years later [8].

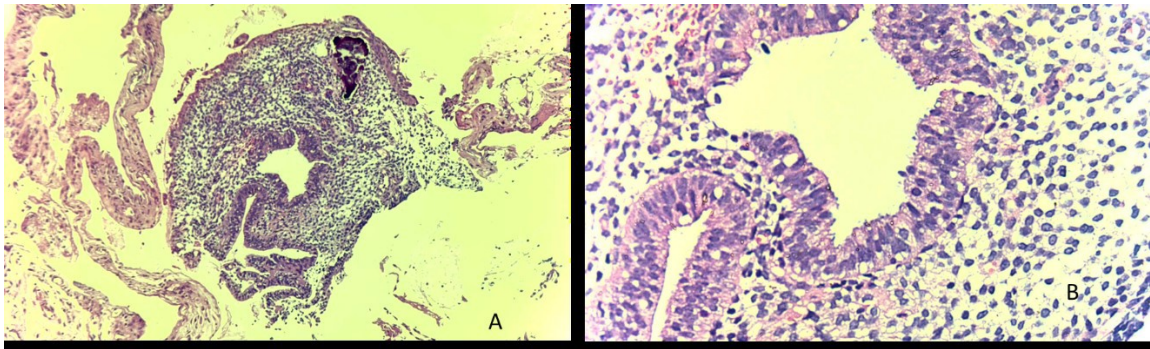


Figure 7A: Low power view and **4B:** High power view showing endometrial glands lined by pseudostratified columnar epithelium with surrounding cellular endometrial stroma.

Pneumothorax is the most common manifestation of thoracic endometriosis (73%). Followed by haemothorax (14%) [2]. Approximately 20% of women of reproductive age who experience spontaneously recurring pneumothorax have thoracic endometriosis [3,9]. Catamenial haemothorax represents the second-most common manifestation of thoracic endometriosis, affecting the right side in about 80 % of the time. Wilkins et al. [10] reported 15 cases of thoracic endometriosis presenting with haemothorax and all of his cases were in the right hemithorax. Concomitant pelvic endometriosis was found in 100 % of cases [8].

Diagnosis

Diagnosis is frequently delayed until several episodes have occurred as patient fails to associate symptoms with menstruation. Pleural fluid cytology is usually not helpful. Level of CA-125 may be elevated in the serum and body cavity fluid of patient with endometriosis [11]. The concentration of CA-125 correlates with both the severity and the clinical course of the disease [7]. Diagnosis of thoracic endometriosis is usually based on clinical grounds, after excluding other pulmonary diseases [2]. Symptoms have a catamenial pattern, occurring between 24 h before and 72 h after the onset of menses, [4] and typically recurring [3,9,12]. CP is defined by at least two episodes of pneumothorax occurring during this time interval. The right-side predominance of symptoms represents a diagnostic clue.

Chest X-rays in cases of pleural endometriosis usually reveal a pneumothorax or occasionally a pleural effusion or pleural lesion [7]. Spiral CT may show pleural or diaphragmatic thickening in involved areas [13]. In the present case, CT revealed a right-sided moderate pleural effusion without any pleural thickening or diaphragmatic lesions. Although the CT aspect of thoracic endometriosis is poorly specific, CT remains the first-line imaging method, as it can rule out other diagnoses and map the lesions for surgery if necessary [4,14,15,16,17].

Medical Thoracoscopy

On visualization of pleura multiple, dark red or blue nodule, and cysts are seen on the diaphragmatic pleura. In our case multiple dark red cysts were seen largest measuring 2 x 1 cm. Most of them were bleeding on touch. Apart from that few blackish nodules were seen. Lesions characteristically described as gun shot lesions were also seen.

Histopathology

Microscopically, typical endometriosis consists of a presence of both endometriotic glands and

stroma. The glands usually have an endometrioid appearance ranging from inactive, proliferative to hyperplastic. The endometriotic stroma characteristically resembles eutopic inactive or proliferative endometrial stroma [18]. Immunohistochemically, in the series reported by Flieder and associates, most glands showed cytoplasmic positivity with broad spectrum cytokeratin, cytokeratin [7], and BER-EP [4], and strong nuclear staining for estrogen and progesterone receptors [9]. Estrogen and progesterone receptors are present in endometriotic glands and stroma in a lower concentration than in eutopic endometrium [19,20].

Therapy for thoracic endometriosis includes the suppression of endometrial tissue and the prevention of further pelvic seeding. Medical therapy should be considered as the first line of treatment [21]. Ghio et al., [22] reported a case of catamenial pneumothorax with chest pain and used medroxyprogesterone acetate as therapy. According to Light [23], hormonal therapy (progestational agents, danazol and leuprolide acetate, fail in at least 50 % cases. Medical treatment for endometriosis symptoms (with or without surgery) is generally needed for longer periods of time because of the chronic and recurrent nature of the disease; progestins may be an appropriate alternative for the medical management of endometriosis, given that these agents are relatively well tolerated, have a more limited metabolic impact than other agents, and are also inexpensive [24]. Treatment with GnRH analogs, such as leuprolide, is limited to only 6 months, because these agents induce a hypoestrogenic state that substantially decreases bone mineral density. Poor tolerability represents the major drawback of danazol as a treatment for endometriosis: this agent has both androgenic and anabolic properties. Pleurodesis may be considered as a means of preventing the recurrence of haemothorax [25].

Conclusion

Thoracic endometriosis is a rare, unusual lesion that may mimic various conditions. Diagnosis of thoracic endometriosis is challenging, as these women's symptoms may not immediately be attributed to endometriosis, and as some radiological abnormalities (especially pneumothorax and haemoptysis) are non-specific or may be taken for artefacts (notably endometriotic diaphragmatic implants on MRI). The key features are the temporal relation with menses, cyclical changes in radiographic abnormalities, and the distinctive posterosuperior location of diaphragmatic lesions. Thorough observation of different symptoms and signs along with appropriate investigations are essential for early diagnosis in such cases.

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