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Massive Catamenial Hemothorax: Rare Case of Thoracic Endometriosis Syndrome

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ABSTRACT

Spontaneous hemothorax is only rarely due to thoracic endometriosis (TE). TE is presence of ectopic endometrial tissue in thoracic. It is rare phenomena seen in women of childbearing age with predominantly right-sided hemothorax and temporal relationship to menses. We report a thirty -seven -year-old Omani lady who was admitted in Sultan Qaboos hospital, Salalah with Spontaneous hemothorax and dysmenorrhea. She had past history of primary infertility due to endometriosis and she lost follow up. Therefore, TE was suspected and was introduced hormone therapy after chest drain inserted. She had recurrent hemothorax after discharge subsequently, she underwent VATS which help visualization of endometrial lesions with pleurectomy of the involved areas and mechanical decortication and pleurodesis using scratch pad were done. Histopathology confirms diagnosis of TE. She continued hormonal therapy and repeated image showed no recurrence.

Keywords: Spontaneous hemothorax; Thoracic endometriosis; thoracic endometriosis syndrome; Video-assisted thoracoscopic surgery.

INTRODUCTION

Hemothorax is collection of blood with hematocrit >50% in the space between the visceral and parietal pleura. Although traumatic or iatrogenic in a vast majority of cases, it may rarely be spontaneous. Spontaneous hemothorax (SH) is most commonly due to pneumothorax but may be related to malignancy, vascular causes or coagulopathy besides various other etiologies.[1] Thoracic endometriosis (TE) is a rare cause of SH and should be highly suspected in females of reproductive age, especially with pelvic endometriosis or history of pelvic surgery. [1] Such etiology for TE is termed Catamenial Hemothorax.

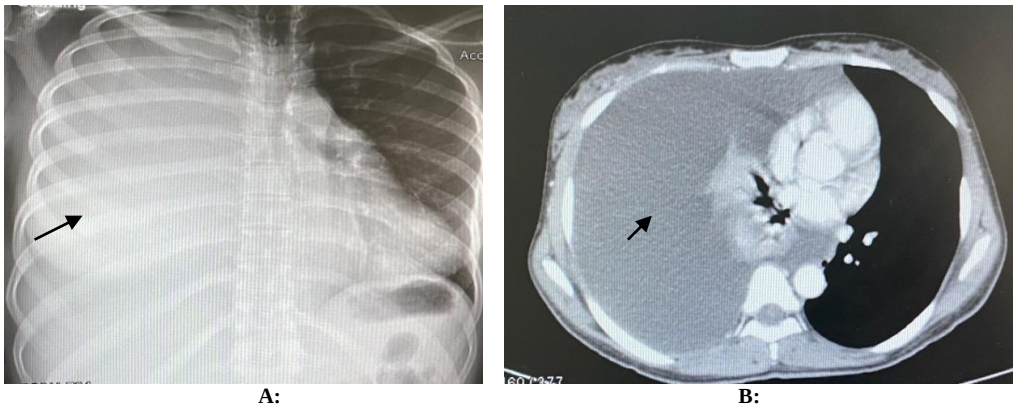
We here report a case of a 37 years old female with massive catamenial hemothorax due to TE.

CASE REPORT

A 37 years old nulliparous female presented to emergency department with dysmenorrhea and shortness of breath. Her dyspnea started one month before while menstruating and was worsening. She also complained of abdominal distension, nausea and poor appetite. She denied any cough, hemoptysis, chest pain or fever. Two years ago, she was investigated for primary infertility and was found to have severe pelvic endometriosis. Surgical treatment was advised, but she declined, and was lost from follow up.

On clinical examination, her vitals were stable, whole right side of chest was dull on percussion with diminished breath sounds. Abdomen revealed signs of mild ascites.

The chest radiograph (CXR) demonstrated a large right-sided pleural effusion with shift of mediastinum to left (Figure 1). Following chest ultrasound, pleural tap was carried out to relieve the distress and one liter of dark hemorrhagic fluid was removed. Similar fluid was obtained on abdominal paracentesis.



A: **B:**

Figure1: Massive right sided hemothorax on imaging. (A)Chest radiograph shows complete opacification of right side hemithorax (arrows) and shifting of heart to left side. (B) CT chest showing massive right sided effusion (arrow).

Figure 2: Grossly hemorrhagic pleural fluid aspirated through the chest drain.

Further evaluation with CT scan of the chest, abdomen and pelvis confirmed massive right-sided pleural effusion (Figure 1) and gross ascites as well as a complex cystic mass in pelvis. Chest drain was inserted on right, and 3.5 liters of grossly hemorrhagic fluid was removed over 48 hours (Figure 2).

The pleural aspirate was grossly hemorrhagic, exudative, negative for mycobacterium PCR and sterile on culture. Cytological examination of the pleural effusion and ascitic fluid showed hemosiderin-laden macrophage and mesothelial cells, no evidence of malignancy or infection or acid-fast bacilli (AFB).

Serum CA-125 was markedly high (102.2 µ/mL; normal < 35.2 µ/mL).

An MRI was carried out to further explore the pelvic mass which demonstrated extensive deep pelvic endometriosis, pelvic adhesions and bilateral adnexal cystic areas engulfing both ovaries. Follow-up CXR showed resolving hemothorax and chest drain was removed.

Because pleura effusion cytology analysis and abdomen image, thoracic endometriosis was highly suspected.

Therefore, started hormone suppression therapy, levonorgestrel 0.15 mg and ethinylestradiol 0.03 mg combination pill once daily. Later she developed mild vaginal bleeding and was started on Triptorelin injection 3.75mg (Gonadotropin-releasing hormone agonist) once per month.

On a follow-up visit in chest clinic after 2 months, her chest image showed re-accumulation of large right-sided pleural effusion with mild mediastinal shift to left. Repeat diagnostic pleural aspirate showed hemorrhagic fluid, no malignant cells or acid -fast bacilli and was negative for bacterial culture.

As such, it was decided that she has failed medical therapy and warranted a diagnostic and therapeutic intervention. She

underwent video-assisted thoracoscopy (VATS). There were reddish black deposits on the parietal pleura, in the mid and lower zones, consistent with endometriosis and 1.7 L of tea colored pleural fluid were seen (Figure 3). There was no diaphragmatic defect or fenestrations seen. Pleurectomy of the involved areas and mechanical decortication and pleurodesis using scratch pad were done.

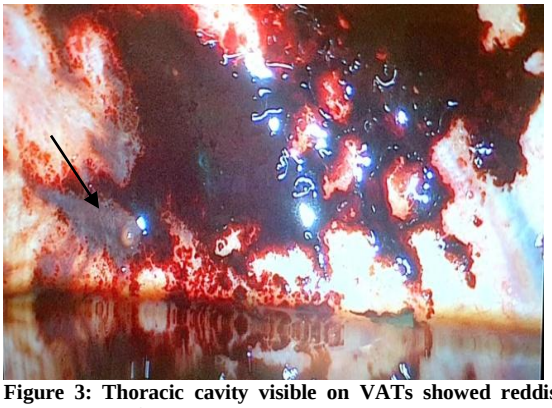
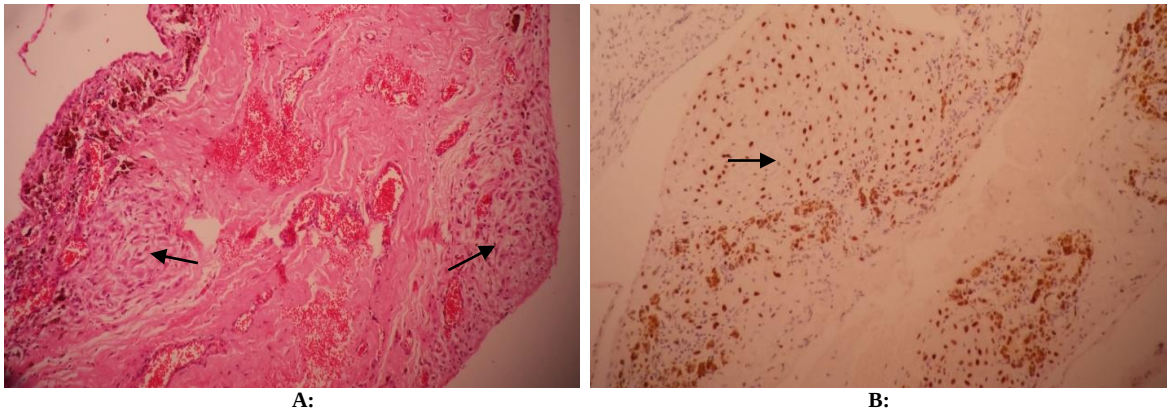


Figure 3: Thoracic cavity visible on VATs showed reddish-black lesions of ectopic endometrial tissue in pleura (arrows).



A: **B:**

Figure 4: Pleura tissue biopsy :(A) Pleural tissue with mesothelial lining on one surface with congestion, hemorrhage and deposition of hemosiderin laden macrophages. Two foci of decidual tissue are seen (arrows). (H&E x 100).
(B) Immunohistochemistry. Decidual cells show nuclear positivity for progesterone receptors (arrows). The cells were also positive for estrogen receptors (X 100)

Pleural biopsy showed evidence of old and recent hemorrhage with hemosiderin-laden macrophages but endometrial glands and stroma were not seen (Figure 4A). There were foci of inflammatory cell infiltration, fibrosis and calcification. Small islands of decidual reaction were also seen. The nuclei of the decidual cells where positive for estrogen and progesterone receptors (Figure 4B). She discharged home with hormonal therapy. At follow up chest imaging showed no recurrence of pleural effusion.

DISCUSSION

Endometriosis is presence of ectopic endometrial tissue (stroma and glands) outside the uterine cavity, mostly within the pelvic cavity and rarely outside. Approximately, 6 to 10% of childbearing women are affected, and only 12% of those with endometriosis have extra-pelvic involvement, predominantly thorax called thoracic endometriosis (TE).[2] While women with pelvic endometriosis alone usually present at 24-29 years, the manifestations of TE peak 5 years later, mainly at age of 35, around 30-50% of women with endometriosis are infertile.[3,4] The symptoms are usually catamenial occurring around menstruation time.[5]

TE usually affects the pleura, catamenial pneumothorax (73%) being the most common manifestation, followed by catamenial hemothorax (14%); while catamenial hemoptysis (7%) and lung nodules (6%) due to lung parenchymal involvement are less common.[3] Besides the four well-known categories mentioned above, the spectrum of thoracic endometriosis syndrome (TES), also includes catamenial chest pain and endometriosis-related diaphragmatic hernia.[2]

Catamenial hemothorax has nonspecific symptoms of cough, chest pain

or shortness of breath that usually occur 1 day before to 2–3 days after the onset of menses.[5,6] Fifty to eighty percent of women with TES have concomitant pelvic endometriosis.[2,7] Approximately, 80 % of catamenial hemothorax involved right side.[8]

Our patient was suffering from infertility and was already diagnosed to have pelvic endometriosis. Her presentation with shortness of breath during dysmenorrhea, a massive right-sided pleural effusion and a grossly bloody aspirate were relatively straight forward for thoracic endometriosis.

The pathology of thoracic endometriosis is as yet unexplained and many theories have been proposed. The most prominent is Sampson's theory of retrograde menstruation: the endometrial tissue migrates to right side of pleural cavity through right paracolic gutter and diaphragmatic fenestrations. This theory is supported by predominant right thoracic endometriosis.[2,6,9] The coelomic metaplasia theory suggests transformation of the pleural and peritoneal mesothelial cells into endometrial tissue and is supported by the observation that some cases of TE occur in the absence of pelvic endometriosis.[2,10] Another theory hypothesizes lymphatic and haematogenous spread of endometrial tissue as a possible mechanism.[6,2]

The radiological abnormalities in TE are transient; diagnostic yield is increased when performed around menses.[2] Chest X-ray and CT images are usually the initial studies to detect pneumothorax and hemothorax; however, MRI is more sensitive in revealing the endometrial lesions (hyper-intense) than CT scan where the lesions are hypo- or iso-attenuating.[6,11]

Bronchoscopy is not of much avail in the diagnosis of TE as the pathologic changes are mostly peripheral.[11]

CA-125 may be elevated in serum and pleural fluid but is not specific, and likewise pleural fluid cytology is seldom of any help.[12]

The gold standard for diagnosis as well as therapy is, however, video-assisted thoracoscopic surgery (VATS) which help visualization of endometrial lesions, scars, bullae, blebs etc.[2]

Classically, to diagnose TE, identification of both endometrial stroma and glands is required on histological examination; it is nevertheless considered suggestive if stroma alone or pulmonary parenchymal hemorrhages or hemosiderin-laden macrophages are detected.[5]

In clinical practice, it is hard to detect intact glands and stroma with hormone receptor study because of autolysis and degradation of proliferative ectopic endometrial tissue 48 hours after menstruation.[6] Only one-third of the cases of TE have typical histopathological finding.[7]

Hence, in many cases the diagnosis is based on a compatible clinical picture, suggestive imaging studies and a bloody pleural aspirate, corroborated with characteristic findings on VATS.

In our case, she had right-sided hemothorax, and VATS showed typical blackish pleural lesions. The biopsy of lesions failed to reveal presence of endometrial glands and stroma, but the finding of hemosiderin-laden macrophages was suggestive for TE there were also foci of decidual tissue (Deciduosis).

TE treatment includes medical and surgical modalities which can be combined.[7] Medical approach (Ideal for women who wish to preserve fertility) depends on suppression of ovarian estrogen secretion by oral contraceptives, progesterone agonists, gonadotropin-releasing hormone agonists or danazol for at least 6-12 months.[6,11] However, the recurrence rate is more than 50% on stopping the treatment.[7]

Surgery is considered in patients with refractory or recurrent disease.[11] Best surgical modality is VATS which allows removal of ectopic endometrial tissue, pleurectomy and closure of diaphragmatic defects.[11] Pleurodesis is an alternative or can be considered additionally during VATS.[2,11,12]

Hysterectomy and bilateral salpingo-oophorectomy is definitive treatment but causes infertility and disease may recur if hormone replacement therapy is used.[6, 11.12]

Our patient had received both medical and surgical treatment, underwent VATS with pleurectomy and pleurodesis, and received follow-up hormonal treatment.

CONCLUSION

TE-associated catamenial hemothorax is a rare etiology for spontaneous hemothorax. It usually involves the right side. High clinical suspicion for TE is required in women of childbearing age as the relationship with menstruation may not be immediately obvious. The successful treatment of TE is also as challenging as is the diagnosis.

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