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Thoracic endometriosis syndrome: A case report and literature review

Olalere FD Haleemah [1], Kuku-Kuye TO [2], Akinlusi FM [3], Adeyeye FA [4]
[1-3] Department of Obstetrics and Gynecology, Lagos State University College of Medicine, Ikeja, Lagos, Nigeria
[4] Registrar, Department of Obstetrics and Gynecology, Lagos State University Teaching Hospital, Ikeja, Lagos, Nigeria
Corresponding Author: **Olalere FD Haleemah**

Abstract
Endometriosis is defined as the presence of functional endometrial glandular tissue outside the uterine lining. Presence of functional endometrial tissue in the thoracic cavity (Thoracic Endometriosis Syndrome) which before now was thought to be a very rare occurrence is now suspected to occur with much higher frequency as cases are misdiagnosed and underreported. Delay in diagnosis or missed diagnosis can be minimized when diagnostic tools are easily accessible but in a resource poor setting where access to such advanced diagnostic tools as Video Assisted Thoracoscopy (VATS) is not easily accessible and/or affordable, early diagnosis of Thoracic endometriosis could be a daunting task. However, this is attainable if a high index of suspicion is maintained when reviewing any woman of reproductive age group presenting with cyclical or recurrent chest symptoms.

Keywords: Endometriosis, Thoracic endometriosis, CTTD

Introduction
Endometriosis is defined as the presence of functional endometrial stroma and glandular tissue outside the uterine cavity [1]. It is an important disease entity affecting about 5-10% of women of reproductive age group [1]. Endometriosis within the lung parenchyma or on the diaphragm and pleural surfaces produces a clinical entity called Thoracic endometriosis syndrome, comprising of catamenial pneumothorax, hemothorax, hemoptysis, and pulmonary nodules [2]. It is very rare, often misdiagnosed and had an enormous delayed in diagnosis [3]. It has a significant social, economic and emotional impact on the affected women, as well as their fecundity [4]. The quality of life as regards to pain and infertility are key features of endometriosis in general, while cyclical cough and/or haemoptysis, temporal relationship with menses, predominant right-sided symptoms, young age, presence of recurrent disease, and a history of infertility are significant findings in Thoracic endometriosis [2]. There are, however, varying presentations of this disease entity and this underscores the importance of high index of suspicion for early diagnosis and treatment and our reason for making this case report.

Patient concerns
A 35 years old P0+0 married Nigerian nurse was referred to our facility on the 26th of October 2020 on account of recurrent chest pain and cough of 2 years duration and chest x-ray findings of massive pleural effusion. The chest pain was of gradual onset and progressively worsened, more on the right side, worse at the onset of her menses and subsides toward the end of her monthly cycle, radiates to the right shoulder, has no known aggravating factor but relieved slightly with the use of analgesics. There was an associated history of non-productive cough which was worse during her menstruation and history of dyspnea which was worse on lying down. No history of fever, night sweats, haemoptysis or haematemesis, no significant weight loss or contact with persons with chronic cough. She had maintained her usual 5 days flow in a regular 28days cycle since attaining menarche at 12 years. There was a positive history of menorrhagia and severe dysmenorrhoea which dates to 4years and history of Infertility of 8yrs duration prior to presentation.She was receiving care at different general hospitals where she was managed as a case of upper and/or lower respiratory tract infections and administered antibiotics and antitussives. A chest X-ray requested prior to referral however showed massive right sided pleural effusion and she was referred on the suspicion of pulmonary tuberculosis. Significant findings at presentation, was tachypnoea, with reduced air entry in the right middle and lower lung zones.

Diagnosis/Intervention

Following an assessment of Thoracic endometriosis, she had a multidisciplinary team management involving the gynaecologists, cardiothoracic surgeons, and chest physicians. She had right closed thoracostomy tube drainage (CTTD) which drained frank blood. FBC done was normal, ESR was raised (64mm/hr), serum E/U/Cr was normal, Gene expert was negative, Mantoux test was negative, Pleural fluid analysis was suspicious but probably reactive. Serum CA requested was mildly elevated at 60iu/ml. Abdominopelvic MRI requested was not done due to financial constraints while transvaginal Ultrasound was normal.

She was placed on antibiotics, commenced on gonadotropin releasing hormone analogue (subcutaneous buserelin) and had pleurodesis done after chest tube removal. She has been on follow up care and her subsequent menstruation has been free of chest symptoms since discharge.She presented for follow up with resolution of symptoms.

Conclusion

This case report highlights the challenges to early diagnosis and prompt treatment of patients with Thoracic endometriosis syndrome and the need for better awareness especially among general practitioners who are the first ports of call.

Discussion

Pelvic endometriosis is the commonest presentation of endometriosis, accounting for over 90% of cases. Although thoracic endometriosis is reported in the literature to be rare, its true incidence is unknown and is said to have been underreported as most cases are misdiagnosed as spontaneous pneumothorax for a protracted period before clinching the appropriate diagnosis [5, 6]. This is a similar finding in the index case in which our patient was being managed as a case of recurrent respiratory tract infection despite the catamenial nature of her symptoms. Thoracic Endometriosis Syndrome (TES), however, is the most common presentation of endometriosis outside the abdominopelvic area and most cases occur in the 3rd and 4th decades of life [5, 7], and peak at 35years as seen in the case presented.

Endometriosis is described as a disease of theories, as the exact pathogenesis is yet to be unravelled. Of the postulated theories for TES, the microembolization theory and the peritoneal-pleural theories explain the right sided predominance of pleural endometriosis as is found in our patient and reported in 85-90% of cases[8]. While the microembolization theory postulates that thoracic endometriosis occurs by transport of endometrial tissues via the lymphatics or blood vessels into the pleura or lung parenchyma (lymphatic drainage of the diaphragm being more extensive on the right) [9 ,10], the peritoneal-pleural migration theory states that endometrial tissue gains access to the pleural cavity via a transdiaphragmatic communication between the peritoneal and the pleural cavities, commonly via the right paracolic gutter [3, 8, 11, 12]. Both theories are built on the widely accepted retrograde menstruation theory [13]. The coelomic metaplasia theory suggests that pleural and peritoneal structures share a common mesothelial origin as the endometrium and endometriosis may occur because of metaplasia of these tissues. These postulations are further buttressed by the strong association between pelvic endometriosis and TES, as 50-84% of cases of endometriosis have associated thoracic endometriosis [11]. There was no radiological evidence of pelvic endometriosis in our patient despite her history of dysmenorrhoea which is the commonest presentation of pelvic endometriosis. She would have been an ideal candidate for diagnostic laparoscopy which is the gold standard in diagnosis of pelvic endometriosis.

Two forms of endometriosis are described in literature. The pleural form is localised within the thoracic cavity but outside the lung parenchyma and presents with catamenial pneumothorax, catamenial haemothorax, catamenial pnuemomediastinum and chest pain. On the other hand, the pulmonary form affects the lung tissues and may present as catamenial haemoptysis and presence of lung nodules [14, 15].

Chest pain and shoulder pain are leading symptoms in our patients, and these are the most common presenting symptoms of TES, seen in up to 90% of cases [6, 16].

Haga *et al* highlighted 4 clinical findings of high predictive value in the differentiating TES from spontaneous pneumothorax, viz right sided, age 31yrs and above, history of endometriosis and absence of smoking [17]. These are comparable findings in our patient, except for the absence of radiological evidence of pelvic endometriosis from pelvic ultrasound scan.

The diagnosis of endometriosis could prove a herculean task, as they may present with intermittent symptoms rather than the characteristic catamenial presentation [11].

Management is multidisciplinary with the gynecologist with special interest in endometriosis and subspecialisation in laparoscopy and cardiothoracic surgeons and chest physician. Video Assisted Thoracoscopic Surgery (VATS) with identification of endometrial epithelium from the Biopsy of suspicious tissue is the gold standard in the diagnosis and management of thoracic endometriosis [18, 19]. Other options include closed thoracotomy tube drainage, bronchoscopy, and open thoracotomy. However, histologic diagnosis is confirmed in only 30% of cases [20] and yield is best if the procedure is done within 48hrs of menses [21]. While MRI and High-Resolution CT scan are imaging modalities of choice, they may identify the presence of pneumothorax or haemothorax but may not necessarily detect the ground glass appearance typical of endometriosis.

While high end interventions like MRI and VATS may be desirable, they are not readily accessible to patients in resource poor settings where care is accessed essentially through the out of pocket spending. This explains why our patient, like many others in similar settings, could not afford to have an MRI done. Due diligence and high index of suspicion is therefore indispensable tools needed by practitioners at all levels if patients are to be diagnosed early and referred promptly.

Management of TES involves the relief of acute symptoms via thoracocentensis, CTTD or bronchoscopy while secondary care is the prevention of recurrence of symptoms, and this can be medical and/or surgical [22]. Suppression of oestrogen production using gonadotrophin releasing hormone agonists for six to twelve month is the first line medical management [23]. Other medical options include the use of the oral contraceptive pills, progestogen, and androgenic agents like Danazol. Our patient has been symptom free on medical management in the last 6months following removal of her CTTD.

In recurrent cases, surgical options such as VATS has been shown to be effective [24] Visible lesions are resected at surgery, and this may or may not be accompanied by pleurodesis [25]

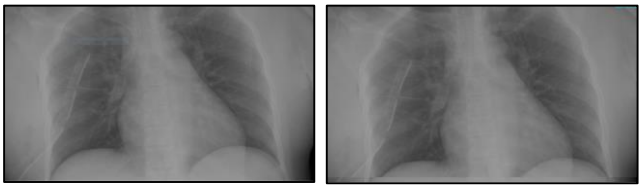

Fig 1: Showing the chest Xray of the patient with chest tube in - situ

Conclusion

This case report illustrates the challenges to early diagnosis and management of Thoracic endometriosis in low resource settings and advocates increased awareness among medical practitioners at all levels of care as well as in the general populace.

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