Spontaneous/Catamenial Pneumothorax due to Thoracic Endometriosis Syndrome a case series

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Abstract

Introduction: The presence of endometrial deposits in the thorax can lead to Thoracic Endometriosis Syndrome. These patients can present with a spontaneous or catamenial pneumothorax, and less likely with haemoptysis.

Case Presentation: We report three cases of women presenting with spontaneous pneumothoraces (two of which presented with catamenial pneumothoraces, as they were menstruating at the time of presentation). Cross sectional imaging with CT, confirmed the presence of pneumothoraces and/or haemothoraces on the right side, with cystic or soft-tissue density pleural deposits noted on the right hemidiaphragm. Diagnosis was confirmed by various methods including cytology of the pleural fluid, CT guided biopsy of the endometrial deposit and VATS.

Conclusion: Thoracic Endometriosis Syndrome is an uncommon condition, but is an important consideration for women presenting with spontaneous or catamenial pneumothorax. CT chest is useful in its diagnosis, especially when faced with the typical clinical scenario.

Keywords: Pneumothorax, Spontaneous, Catamenial, Endometriosis

Abbreviations: CT – Computed Tomography, VATS - video assisted thoracoscopic surgery, GnRH - Gonadotropin Releasing Hormone.

Introduction:

Thoracic Endometrial Syndrome is the clinical manifestation of endometriosis in the thorax, and can present with a spontaneous or catamenial pneumothorax. The latter is defined as spontaneous pneumothorax occurring just before or during menstruation. It is an uncommon entity, which usually occurs in women during their fourth decade of life. The typical presentation includes pneumothorax, haemothorax and haemoptysis, if there is bronchopulmonary involvement.

This case series reviews three female patients presenting with spontaneous/catamenial pneumothoraces, eventually diagnosed with Thoracic Endometriosis Syndrome. These patients were managed at the same institution over a one-year period.

Case Presentation:

Case 1
A 40-year-old female presented with shortness of breath and a syncopal episode. On evaluation, she was found to have anaemia and a spontaneous right pneumothorax, with no pre-existing pulmonary conditions. There was a history of menorrhagia at the time of presentation. An intercostal drain was placed and small amount of bloody-brown fluid drained. Computed Tomography (CT) of the Chest showed a cystic density, pleural-based lesion on the right hemidiaphragm (figure 1). A CT guided biopsy of this lesion confirmed endometrial tissue. Treatment consisted of resection of endometrial tissue via thoracotomy and pleural adhesiolysis. Medical treatment with Gonadotropin Releasing Hormone (GnRH) agonist and chest physiotherapy were started. Significant improvement of symptoms were noted post-intervention.
Figure 1: Axial (a,b) and Coronal (c,d) contrast enhanced CT Chest images in mediastinal and pulmonary windows showing a cystic-density pleural lesion on the right hemidiaphragm, and right pneumothorax. Axial (e) CT Chest images at time of CT guided biopsy.
Case 2
A 41-year-old female complained of chest pain and shortness of breath at time of menses. The patient had no other pulmonary or gynaecological symptoms, apart from a history of menorrhagia and dysmenorrhoea. Chest radiograph showed a right pneumothorax, for which an intercostal drain was placed (figure 2). CT Chest showed two rounded soft tissue pleural-based deposits on the right hemidiaphragm and a small volume hydropneumothorax (figure 3). A clinical and radiological diagnosis of Thoracic Endometrial Syndrome was made. Patient was started on the oral contraceptive pill and had symptomatic relief.

Figure 2: Chest radiographs showing pneumothorax and a mass on the right hemidiaphragm (a), and re-expansion of the right lung after insertion of intercostal chest drain, with persistence of the mass above the right hemidiaphragm.

Figure 3: Axial CT slices through the lower thorax showing soft tissue nodules on the right hemidiaphragm (a,b) and haemopneumothorax, with intercostal drain in situ (c).
Case 3
A 39-year-old female complained of shortness of breath and complained of a heaviness in the chest. Patient gave gynaecological history of having regular periods and is multiparous. Patient was not menstruating at the time of presentation. X-ray and CT chest confirmed a right haemopneumothorax (figures 4, 5), and a right intercostal drain was inserted. Cytology of the pleural fluid showed features that supported the diagnosis of thoracic endometriosis. Subsequent right VATS (video assisted thoracoscopic surgery) and decortication were performed. Patient also had total abdominal hysterectomy and bilateral salpingo-oophorectomy. She was also started on an oral contraceptive pill and a Gonadotropin Releasing Hormone (GnRH) agonist. The patient noted significant improvement after treatment.

Figure 4: Chest radiograph, showing right haemopneumothorax

Figure 5: Interval CT Chest axial slices in pulmonary windows confirms a large right pneumothorax (a), and mediastinal windows show two soft tissue right hemidiaphragm deposits in the axial (b) and coronal slices (c).
Discussion

Endometriosis is defined as the presence of ectopic endometrial tissue outside the uterine cavity. The common locations for endometrial deposits can be broadly classified into abdominal and extra abdominal sites. Gynaecologically, different syndromes have been described, and symptomology is primarily related to the presence of endometrial deposits which are scattered across the abdominal cavity, including subperitoneal endometrial infiltrates.

Pulmonary endometriosis is a rare site for which the prevalence has not been determined. Over 150 cases have been published since the first described case of pulmonary endometriosis, which is further subdivided into pleural and bronchopulmonary (parenchymal) endometriosis. The differentiation between the two can be aided by the symptomology. Haemoptysis occurs secondary to endometrial deposits in the bronchial tree, and spontaneous pneumothorax or haemothorax occurs where the endometrial deposits are localized to the pleura.

Types of pneumothoraces related to pulmonary endometriosis include spontaneous pneumothorax and catamenial pneumothorax. The latter is defined as a pneumothorax that occurs within 24 hours prior to the onset of menses to 72 hours post-commencement of the menses. Thoracic Endometriosis Syndrome is an uncommon condition that encompasses the pulmonary manifestations of endometriosis with or without coexisting pelvic endometriosis.

The pathogenesis of endometriosis has not been confirmed, but the regurgitation theory has been sited at the most plausible explanation for thoracic endometriosis. This theory postulates that peritoneal endometrial deposits originate from retrograde menstruation via the fallopian tubes, which then migrate into the pleural cavity (transabdominal-transpleural migration), through diaphragmatic defects known as fenestrations or porosities which may be acquired or congenital in nature.

Diagnosis of thoracic endometriosis is aided by the patient’s clinical history and symptoms, particularly in relation to the menstrual cycle. Initial diagnosis is suspected in the presence of a chest radiograph with a spontaneous pneumothorax.

Confirmation requires histological correlation, that can be obtained via biopsy, usually by video assisted thoracoscopic surgery (VATS), whereby the endometrial deposits are visualized and biopsy specimens obtained. VATS exploration is not routinely performed for diagnostic purposes.

Cross-sectional imaging techniques such as CT are helpful in aiding diagnosis when pulmonary or pleural soft tissue density lesions are corroborated with the relevant clinical scenario. MRI imaging is commonly employed in cases of pelvic endometriosis, however has not been widely described for investigation of thoracic endometriosis. According to Paolo Claudio et al, MRI and CT scans demonstrated comparable accuracies for identifying endometrial deposits within the thoracic cavity.

Medical therapeutic options are available which focuses primarily on hormonal therapy, including the use of oral contraceptive pills and Gonadotropin Releasing Hormone (GnRH) agonists, which cause degeneration of the pleural endometrial implants. Upon persistence of disease despite medical therapy, surgical options can be explored which consists of thoracoscopic with resection of abnormal lung parenchyma, repair of diaphragmatic defects and pleurodesis, if necessary.

Conclusion

Thoracic Endometriosis Syndrome, although an uncommon condition, is an important consideration for women presenting with spontaneous or catamenial pneumothorax, with or without a known diagnosis of pelvic endometriosis. Cross sectional imaging has an important role to play in the diagnosis of Thoracic Endometriosis Syndrome, and can avoid the need for histological confirmation in the typical clinical scenario.

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