

Review

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Pulmonary Endometriosis: A Review

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INTRODUCTION

Endometriosis is a benign gynaecological condition whereby endometrial tissue exists outside the uterus in women of reproductive age group. It occurs mainly in the pelvis and rarely extra-pelvic areas such as the lungs. Pulmonary endometriosis is a rare but can be life-threatening. There is active endometrial tissue in the tracheobronchial tree, lung parenchyma and lung pleura.¹ Pulmonary endometriosis has four main clinical conditions namely catamenial pneumothorax, catamenial haemothorax, catamenial haemoptysis and endometrial nodules in the lung. Catamenial pneumothorax is the most common manifestation.² Pulmonary endometriosis is associated with pelvic endometriosis and subfertility. Because of its rare phenomenon, there may be delayed diagnosis leading to serious life-threatening complications. This article aims to raise awareness amongst clinicians particularly gynaecologists about this rare but life-threatening condition. It is a benign, treatable condition and no women should die from it.

AETIOLOGY

The aetiological mechanisms of pulmonary endometriosis are not well known.³ There are no predisposing factors.⁴ A possible explanation for the pathogenesis may involve peritoneal-pleural movement of endometrial tissue through diaphragmatic defects and microembolisation through pelvic veins.⁵ Endometriotic deposits can be found in the diaphragm, pleura, lung parenchyma and tracheobronchial tree. The preferred sites are the diaphragm in keeping with the embryological suspected peritoneal-pleural migration route.

Clinical Presentation

Classically there are chest symptoms associated with menstruation. These include dyspnoea,⁶ intermittent productive coughing with blood-tinged sputum, chronic anemia, loss of appetite, generalised weakness⁷ and chest pains.⁸ Patients can also present with catamenial haemoptysis.^{9,10} All these symptoms can be found in patients with pulmonary tuberculosis. Some patients may be asymptomatic.

Complications

The complications of pulmonary endometriosis can be repeated mild symptoms to massive pulmonary complications. Catamenial pneumothoraces can be recurrent needing repeated pleurodesis. If massive they can lead to lung collapse, respiratory compromise and death. Catamenial haemothoraces can lead to chronic pulmonary bleeding and chronic anemia. If they are massive, catastrophic pulmonary haemorrhage can occur leading to cardiovascular shock and death. Repeated pleurodesis runs the risks of infection, lung punch and fibrosis.

Investigations

The cycle of pulmonary symptoms associated with menstruation can lead to a clinical¹¹ diagnosis of pulmonary endometriosis. Many diagnostic methods both clinical and laboratory have been used but none of them is the golden standard.¹² Investigations can be done during and after menses to compare appearances of lesions. Imaging techniques may be non-specific.¹³ The diagnosis can be difficult to make.⁴ A chest x-ray or computed tomography (CT) can reveal multiple lung nodules,¹⁴ if these are present. A computed tomography during and after menstruation can be useful in precise location of paranchymal pulmonary endometriotic lesions.¹⁵ Magnetic resonance imaging is now increasingly being used for assessment of lung conditions such as endometriosis.¹⁶ It is good at characterization of pleural endometriotic nodules and haemorrhagic pleural effusions.¹⁷ Bronchial angiography can demonstrate prominent vasculature.¹⁸

Rigid bronchoscopy can allow bronchial samples to be obtained from tracheobronchial lesions⁶ and samples sent for histological examination. Through the bronchoscope hyperaemic tissue in the tracheobronchial tree can be seen.¹ Fibre-optic bronchoscopy can also reveal lesions such as diffuse erythema and also allows bronchial washings to be obtained for histological testing.^{8,19} Video-assisted thoracoscopic surgery can reveal endometrial tissue embedded in the diaphragmatic pleura.^{9,20}

Tumour markers CA125⁴ and CA19-9⁷ can be elevated causing fears of the existence of malignancy. Pulmonary tuberculosis²¹ is another differential diagnosis that may be considered during the investigations for pulmonary endometriosis. Lesions found during investigations can cause confusion with lung cancer.⁶

Management

The treatment of pulmonary endometriosis can be commenced based on the clinical history alone of cyclical chest symptoms associated with menses with complete resolution of symptoms.¹¹ The management of pulmonary endometriosis calls for a multi-disciplinary approach involving the anaesthetist, gynaecologist, pneumologist and thoracic surgeon.^{22,23}

Medical therapy involves the use of oral contraceptives¹¹ or medroxyprogesterone acetate for 3 to 6 months and patients can be asymptomatic 12 months after treatment.¹⁹ Danazol therapy was previously extensively used to treat endometriosis²⁴ but it has fallen off due to its side effects profile. Currently GnRH analogues^{4,8,25} are the ones widely used. Courses of 3 to 6 months produce good outcomes. The GnRH analogues cause a reversible hypo-estrogenic state that starves endometrial tissue of oestrogen hence growth and the tissue dies out. Urgent tube thoracostomy for patients in respiratory distress relieves pneumothoraces and haemothoraces while awaiting definitive treatment. Thoracotomy²⁶ with lobectomy,²⁷ parietal pleurectomy and partial diaphragmatic excision²⁸ can be done in

life-threatening endometriosis. This is life-saving surgery.

Immediately after surgical treatment, medical therapy must be started^{4,13} as recurrence is common since the endometrial tissue will be responsive to ovarian hormones.

Prognosis

Following treatment complete cure is possible.^{1,8,29} Fertility returns to normal after treatment.

CONCLUSION

This benign gynaecological disorder that is common in the pelvis but rare in the lungs can have life-threatening consequences. Clinicians must be made aware of this condition so that they have a high index of clinical suspicion³⁰ to prevent deaths in young women. Prompt treatment of pneumothoraces and haemothoraces by a multidisciplinary team will save lives.

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