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Pictorial Review

Thoracic endometriosis syndrome: CT and MRI features

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Thoracic endometriosis is considered to be rare, but is the most frequent form of extra-abdominopelvic endometriosis. Thoracic endometriosis syndrome affects women of reproductive age. Diagnosis is mainly based on clinical findings, which can include catamenial pneumothorax and haemothorax, non-catamenial endometriosis-related pneumothorax, catamenial haemoptysis, lung nodules, and isolated catamenial chest pain. 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 often under-diagnosed syndrome. MRI can also assist with the diagnosis, by showing signal changes typical of haemorrhage within diaphragmatic or pleural lesions.

Introduction

Endometriosis is a common gynaecological disorder, affecting 10–15% of women of reproductive age. It is defined by 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 rarely involved, the thoracic cavity is the most frequent extra-abdominopelvic site of endometriosis.[2] Thoracic endometriosis syndrome (TES) is the term used to refer to the various clinical and radiological manifestations resulting from the presence and cyclical changes of functional endometrial tissue in a thoracic structure (visceral or parietal pleura, lung parenchyma, airways, or diaphragm).[2] Clinical manifestations vary during the menstrual cycle and 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

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catamenial haemoptysis (CH), and lung nodules.[2,3] Approximately 90% of patients with TES experience catamenial thoracic pain and different entities may be associated.[2] Less frequent presentations, with isolated catamenial right scapular or cervical pain ("diaphragmatic pain") consistent with phrenic nerve irradiations are also seen in patients with diaphragmatic involvement. The right hemithorax is involved in more than 90% of all forms, with the exception of nodules (70%).[2]

The aim of this review is to summarize clinical and pathophysiological knowledge of TES, to describe key CT and magnetic resonance imaging (MRI) features, and to examine the role of imaging studies in patient management.

Epidemiology

TES is considered rare but may be under-diagnosed.[2,4] Pneumothorax is the most common manifestation of thoracic endometriosis (73%). CHt is less common (14%), followed by CH (7%) and lung nodules (6%).[2] Approximately 20% of women of reproductive age who experience spontaneously recurring pneumothorax have thoracic endometriosis.[3,5] The prevalence of catamenial haemoptysis, catamenial haemothorax, and lung nodules is unknown, as most data come from simple case reports or short series. TES affects women of reproductive age. Mean age at diagnosis is 35 years (range 19–54 years),[2] some 5 years later than pelvic endometriosis. TES is usually but not systematically associated with pelvic endometriosis.[5]

Pathogenesis

Hypothesis regarding the presence of thoracic endometrial implants

The aetiological mechanisms of TES are controversial.[4] Three hypotheses have been forwarded but none can fully explain the clinical manifestations of TES, suggesting that the disease might have a multifactorial origin.[2,6]

Transplantation theory

This theory, supported by the Sampson's theory of retrograde menstruation through the fallopian tubes, is the most widely accepted explanation for pleural and diaphragmatic endometriosis.[7] Endometrial cells in the peritoneal cavity may then follow the clockwise peritoneal circulation, through the right paracolic gutter towards the right subdiaphragmatic area[7–9]; indeed the phrenicocolic ligament on the left-hand side and the falciform ligament form barriers that prevent cells and fluid from reaching the left sub-diaphragmatic area.

The peritoneal area close to the posterosuperior tendinous portion of the right diaphragm is a preferential site of endometrial cell implantation, leading to the formation of endometrial nodules[4] on the abdominal side of the diaphragm. Their size and appearance can vary with their age and the phase of the menstrual cycle (Fig 1 and Supplementary Material Fig S1).[4] Transdiaphragmatic passage of endometrial tissue might occur through congenital or acquired diaphragmatic defects.[2] Congenital defects exist in many individuals and predominate on the right-hand side.[8] Endometrial implants traversing the entire thickness of the diaphragm, followed by cyclical necrosis, may lead to the formation of holes (Supplementary Material Fig S2), that can coalesce into larger defects (Fig 2).[10] Once viable endometrial tissue has entered the pleural space, it may colonize other parts of the diaphragm or the pleural space.[4,11] CHt is due to bleeding from endometrial implants on the pleural surface during menstruation.[2]

The lymphatic or haematogenous microembolization theory

Characteristics supporting the microembolization theory include the bilateral or left-sided location (30%) of lung nodules, and the occurrence of other extra-pelvic sites of endometriosis such as the brain.[2,12,13] In this theory, pulmonary endometrial implants are believed to result from the pulmonary network filter function, that traps endometrial particles.[1,2] Implants may take the form of solitary or multiple circumscribed nodules, often located in peripheral bronchi or, less frequently, in the lung parenchyma.[14] Haemoptysis could result from capillary rupture within the lesion, at the time of menstruation. The haematogenous dissemination theory is supported by histopathological evidence of endovascular endometrial epithelium, and uterine procedures, such as curettage and Caesarean section are considered as risk factors.[5] However, this theory does not explain the right predominance of CH and endometrial lung nodules.[2]

Coelomic metaplasia theory

The coelomic metaplasia theory is supported by the fact that the endometrium and pleural and peritoneal

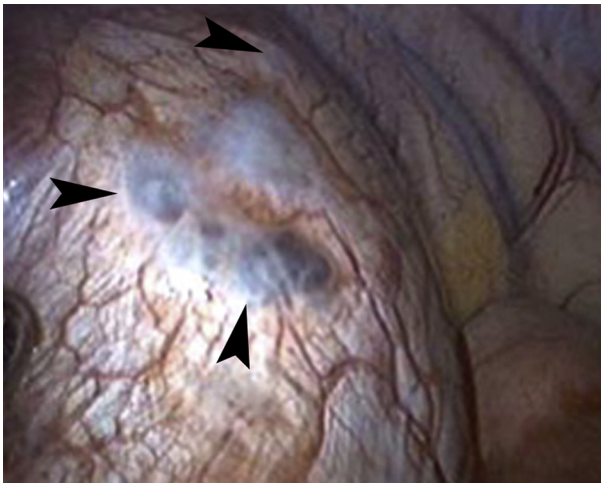

Figure 1 A 31-year-old women with recurrent right catamenial pneumothorax. Image obtained during video-assisted thoracoscopic surgery shows blue diaphragmatic endometriotic implants of different sizes (arrowheads), corresponding to haemorrhagic lesions. (For interpretation of the references to color in this figure legend, the reader is referred to the web version of this article.)