CASE REPORT

Rare case of thoracic endometriosis presenting with lung nodules and pneumothorax

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SUMMARY

We present the case of a 34-year-old Nigerian woman who was referred to the Respiratory team with a 12-month history of breathlessness. She was concurrently being investigated for an abdominal mass and rectal and vaginal bleeding. Consequently, she underwent cross-sectional imaging of her chest, abdomen and pelvis, revealing a small right-sided pneumothorax and right lower lobe pleural-based lesion. Shortly thereafter, she was admitted to the hospital with chest pain and required chest drain insertion. This partially treated her pneumothorax but she required referral to a cardiothoracic centre for definitive diagnosis and to manage her non-resolving pneumothorax. Biopsies from the video-assisted thoracoscopic surgery confirmed the very rare diagnosis of thoracic endometriosis.

BACKGROUND

This case is important as it not only revealed a very rare diagnosis, but also highlights that this diagnosis should be considered in women of a menstruating age with recurrent pneumothoraces and pleural effusions with or without an established diagnosis of systemic endometriosis.

CASE PRESENTATION

A 34-year-old Nigerian woman, who works as a solicitor, was originally referred to the gynaecologists with weight loss, rectal bleeding and a palpable mass thought to be in her uterus. She had urgent imaging of her abdomen and pelvis which showed an enlarged uterus with fibroids. Her Cancer Antigen 125 (CA-125) was found to be raised and she subsequently underwent MRI of her pelvis, ruling out uterine malignancy. She was

reviewed by the General Surgical team who investigated her for rectal bleeding, with no findings on colonoscopy or biopsy, before being referred to the Respiratory team for an incidental pleural effusion which had been seen at the base of the lungs when scanning the abdomen.

The patient was a lifelong non-smoker. She had lived in the UK since 2008 and had no other medical problems. On review by the Respiratory physicians, she complained of 12 months of breathlessness, worse on exertion, which was limiting her activities such as partaking in the church choir. She had no other respiratory symptoms of cough, wheeze or haemoptysis. She did, however, complain of some right-sided shoulder pain which coincided with the onset of the breathlessness.

INVESTIGATIONS

A CT thorax scan to complete the staging and pulmonary function tests were arranged to be discussed in the lung Multidisciplinary Team Meeting (MDT). Her CT revealed a right-sided pleural-based cystic nodule with an incidental pneumothorax (figure 1). The MDT concluded that this could represent infection and/or malignancy. An incidental pulmonary embolism was identified by the radiologist which was treated with enoxaparin. Further imaging with a positron emission tomography scan was performed in preference to CT-guided biopsy due to the presence of a pneumothorax. This demonstrated no avidity relating to the right-sided pleural-based abnormality. In the interim, she presented to the hospital with chest pain with presence of hydropneumothorax which was treated with a chest drain. The pneumothorax did not completely resolve; however, the chest drain was removed. The pneumothorax remained stable. She was referred to the cardiothoracic surgeons for video-assisted thoracoscopic surgery (VATS) pleural biopsy and pleurodesis in view of persistent pleural lesion and pneumothorax.

DIFFERENTIAL DIAGNOSIS

- ▶ Thoracic endometriosis
- ▶ Hydatid disease
- ▶ Pleural malignancy

TREATMENT

The patient was admitted to the Cardiothoracic centre and underwent VATS pleural biopsy with closure of the diaphragmatic fenestration and talc pleurodesis (figure 2).

Figure 1 Right lower lobe cystic nodule seen with pneumothorax.

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Figure 2 Intraoperative imaging showing diaphragmatic fenestrations and cystic endometrial deposits.

OUTCOME AND FOLLOW-UP

The biopsy results from the pleura confirmed the diagnosis of thoracic endometriosis. She was seen by the gynaecologists who commenced a gonadotropin-releasing hormone (GnRH) analogue to prevent menstruation. Her full diagnosis was multifocal endometriosis involving the pleura and rectovaginal septum.

She continues anticoagulation for the small pulmonary embolism. She has noticed a great improvement in her breathlessness, although her most recent imaging shows a small residual trapped pneumothorax (figure 3).

DISCUSSION

Thoracic endometriosis syndrome (TES) was first described as early as 1912 by Hart, and again by Schwartz in 1938 who described it as a ‘lung tumour’ which bled every month.¹ Since then, we have been able to improve the understanding of the pathogenesis, diagnosis and management of this rare condition. However, the diagnosis remains rare and there are no randomised control trials to ascertain the optimal management.

There have been two large retrospective studies on TES. They found that the mean age of onset is 35 years with the peak incidence occurring between 30 and 34 years, compared with a

Figure 3 Repeat cross-sectional imaging following video-assisted thoracoscopic surgery biopsy and pleurodesis showing residual pneumothorax.

younger peak incidence (25–29) for pelvic endometriosis.^{2,3} The most common presentation was with pneumothoraces, seen in 73% of the cohort. Interestingly, in over 90% of presentations, the right hemithorax was involved.²

In the intervening 14 years between these two studies, the use of thoracoscopy had increased and so by 2010, 85% of the cohort had undergone thoracoscopy or thoracotomy.³ They found no significant association between the presence of diaphragmatic defects and pneumothorax. However, the likelihood of haemothorax was associated with an increased burden of pleural deposits of endometrial tissue.³

Previously, the endometrial tissue had been thought to travel by retrograde menstruation back through the fallopian tubes into the peritoneal cavity, and then through diaphragmatic fenestrations into the thorax.¹ However, these thoracoscopic findings negate that theory.³

An alternative theory suggests that physiological stimuli can cause the transformation of pleural epithelium into endometrial tissue. Unfortunately, this does not explain the right-sided predominance seen in TES cases.¹

One of the latest theories is that endometrial transplantation occurs through lymphatic embolisation, explaining both the thoracic and other sites of implantation, in much the same way that a cancer metastasises.⁴ However, it is likely that the true mechanism is multifactorial and incorporates all of these theories.⁵

The most common presentation of TES is a ‘catamenial pneumothorax’. This is defined as a pneumothorax which occurs within the first 72 hours after the start of menstruation.⁶ It does not necessarily occur with every menstrual cycle. It is the underlying aetiology in 2.5%–5% of spontaneous pneumothoraces in women and makes up 80% of the presentations of TES.⁶

‘Catamenial haemothorax’ is a rarer presentation which often presents with chest pain and cough and appears as a pleural effusion on chest X-ray. Haemothoraces are associated with the presence of pelvic and peritoneal endometriosis.²

Cyclic haemoptysis is an uncommon presentation of TES, tending to occur in the younger age group.³ This will manifest as synchronous haemoptysis with menstruation. It is caused by parenchymal deposits of endometrial tissue, and the lesions can typically be seen at bronchoscopy, especially during menstruation.⁵

The rarest manifestation of TES is pulmonary nodules, as seen in our patient. The literature quotes between 2% and 6% of patients with TES are found to have pulmonary nodules.^{2,3} They are most prevalent in women over 35, compared with the other presentations occurring more frequently in younger patients.¹

Having a high index of suspicion for TES is vital for diagnosis. This should be especially heightened if the woman is known to have, or reports symptoms of, pelvic endometriosis.

The current treatment of TES is based on the suppression of ovarian oestrogen secretion, with GnRH agonists being the agents of choice in women who wish to preserve fertility.¹ However, with the increasing prevalence of VATS, this is rapidly taking over to enable rapid diagnosis of thoracic endometrial deposits and also allows immediate initial treatment. There are various procedures that can be carried out in this manner, including resection of endometrial implants, closure of the diaphragmatic fenestrations and abrasive mechanical or chemical pleurodesis.¹ If the main complaint is of haemoptysis, endobronchial laser ablation can be considered; however, there is no consensus on treatment given the paucity of data.⁵

Surgical treatment followed by hormonal therapy has been associated with no recurrence in a follow-up period as long as

45 months and, therefore, this combination therapy may be the preferred approach to prevent long-term morbidity.¹

The definitive treatment of systemic endometriosis is hysterectomy with bilateral salpingo-oophorectomy; however, it should only be considered if all other treatment modalities have failed. Interestingly, subsequent hormone replacement can worsen or cause recurrence of thoracic endometriosis.²

Learning points

- ▶ Thoracic endometriosis is a rare but important differential in menstruating women.
- ▶ A diagnosis of thoracic endometriosis does not necessarily need an established diagnosis of systemic endometriosis.
- ▶ Increasingly, video-assisted thoracoscopic surgery procedures are being used to diagnose and treat the complications of thoracic endometriosis.

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