Thoracic endometriosis syndrome manifesting as atraumatic haemothorax causing difficult ventilation under general anaesthesia

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SUMMARY

A 31-year-old woman underwent elective hysteroscopy and laparoscopy for investigation of primary infertility. The procedure was abandoned owing to intraoperative desaturation and reduced right-sided chest movement. Postoperative chest x-ray showed an opacified right hemithorax with tracheal deviation to the left. Retrospectively on direct questioning, she admitted to dry cough, mild dyspnoea and significant weight loss. Diagnostic thoracocentesis and contrast CT scan confirmed heavily blood-stained exudative pleural effusion but were otherwise non-diagnostic. After chest drain insertion, 4.6 litre drained but a hydropneumothorax developed upon drain removal, with no improvement on repeat drain insertion. Right video-assisted thoracic surgery was performed, converted to thoracotomy, with repair of diaphragmatic fenestrations, pleurectomy and decortication of trapped lung undertaken. Histological examination of samples of pleura and lung cortex was pathognomic of endometriosis. She was started on norethisterone with no recurrence to date.

BACKGROUND

Unexplained isolated unilateral pleural effusion is a common and important diagnostic problem. It can be the presenting feature of metastatic and local (lung or pleural) malignancy and, in areas of high incidence, tuberculosis (TB). Endometriosis is a common cause of primary infertility but only uncommonly causes thoracic endometriosis syndrome (TES), of which catamenial haemothorax is an unusual manifestation. This is a true multidisciplinary case bringing together gynaecology, anaesthetics, chest medicine, thoracic surgery, radiology and histopathology. It offers us the opportunity to highlight that: 1) although catamenial haemothorax clearly represented a potential underlying diagnosis, common presentations such as unilateral pleural effusion should be investigated in a standardised way to exclude the most probable causes first; 2) the highest yield diagnostic procedure for suspected tuberculous pleuritis, thoracoscopic pleural biopsy, was overtaken by the more pressing development of ex vacuo hydropneumothorax in the setting of trapped lung. The patient therefore developed an indication for a therapeutic procedure that outweighed the need for a simply diagnostic one; and 3) failure of several pre-operative consultations to detect the gross respiratory pathology was followed by the unexpected development of respiratory failure on the operating table. This represents a near-miss and reinforces the need for a full history

and examination even where positive findings are thought unlikely.

CASE PRESENTATION

A 31-year-old woman with a history of dysmenorrhoea for 5 years, judged to be American Society of Anaesthesiologists grade I, underwent elective hysteroscopy and laparoscopy under general anaesthesia for investigation of primary infertility. Pneumoperitoneum was achieved and grade IV endometriosis involving the ovaries, fallopian tubes, bowel, liver and diaphragmatic adhesions were noted. A short while into the procedure, the patient desaturated and reduced right-sided chest movement with reduced breath sounds was noted. She responded to an increased inspired oxygen concentration and the procedure was abandoned. In recovery, she reported slight dyspnoea. Peripheral oxygen saturations (SaO₂) were normal at rest but fell to 92% with exertion. Postoperative chest x-ray demonstrated an opacified right hemithorax with tracheal deviation to the left (figure 1). Referral to the respiratory team was made.

On review, she admitted to occasional dry cough, gradually progressive mild dyspnoea over 6 weeks and unintentional weight loss, dropping four dress sizes over 4 months. Review of clinical notes confirmed body weight of 63 kg currently, having been 78 kg 16 months previously. She denied haemoptysis, chest pain, fever and exposure to TB or asbestos. She had regular periods every 28–30 days lasting 7 days with severe dysmenorrhoea. She had not travelled since arriving in the UK from Nigeria 1 year previously. She had no significant medical history, had never smoked, took no medication and lived with her husband as a housewife.

On examination, she looked well and was haemodynamically stable with a respiratory rate of 18, SaO₂ of 98% on air and a temperature of 36°C. There was dullness to percussion and reduced breath sounds throughout the right hemithorax and multiple subcentimetre cervical nodes bilaterally. The rest of the examination was normal.

INVESTIGATIONS

Tests requested in gynaecology outpatients had revealed normal hormone profile and normal high vaginal and endocervical swabs. Pelvic ultrasound had found a normal anteverted uterus, multiple small fibroids, a normal left ovary and a simple cyst in the right ovary.

Mantoux test showed 7 mm induration but her BCG vaccination status was unclear. With the exception of mild microcytic anaemia (Hb 10.9 g/dl,

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Figure 1 The presenting chest x-ray prompting referral, 1/10/2011.

mean corpuscular volume (MCV) 75 fl), standard haematology and biochemistry were normal including a normal C-reactive protein of 7 mg/l. HIV testing was negative.

Upon diagnostic thoracentesis, pleural fluid appeared heavily blood stained, with protein 57 g/l (serum 80 g/l) and glucose 4.2 mmol/l (no paired serum). pH was not performed as the fluid was heavily blood stained. Microscopy for white cells, culture and alcohol and acid fast bacilli (AAFB) smear were negative. Cytology was reported as 'Moderately cellular, many red blood cells, some lymphocytes, macrophages, occasional mesothelial cells. No malignant cells seen'.

CT chest, abdomen and pelvis with contrast demonstrated a large right pleural effusion with associated collapse and mediastinal shift with unremarkable pleurae, small volume free fluid in the abdomen, a cystic pancreatic lesion (48×52 mm), a right adnexal cystic mass (42 mm), a single uterine fibroid, normal liver, spleen and kidneys and no abdominal lymphadenopathy.

DIFFERENTIAL DIAGNOSIS

The differential of a heavily blood-stained effusion includes malignancy, TB, pulmonary embolism and infarction, benign asbestos effusion, cardiac injury or surgery and traumatic haemothorax. However, following thoracentesis and CT, there were no potentially helpful diagnostic pointers such as pleural fluid lymphocytosis or acidosis, mediastinal or abdominal adenopathy

Figure 3 Chest radiograph after first drain removed, 10/10/2011.

or pleural thickening. At this point, the differential diagnosis remained broad with no single diagnosis confirmed.

TREATMENT

Although pleural biopsy for histology and AAFB culture¹ would have been the most sensitive test for pleural TB, thoracic drainage was performed with the aim of symptomatic relief (figure 2). Despite apparent radiographic resolution of the effusion, hydropneumothorax became apparent upon drain removal (figure 3). A repeat drain was inserted, but the lung now behaved as if trapped with no improvement (figure 4). We hypothesised that in the setting of trapped lung underlying long-standing pleural effusion, air had been drawn in after removal of pleural fluid and ex vacuo hydropneumothorax had developed. She was referred for right-sided video-assisted thoracic surgery (VATS), which upon confirming trapped lung, was converted to thoracotomy at the seventh interspace, followed by repair of diaphragmatic fenestrations, parietal pleurectomy and decortication. The operation note commented that 'The lower lobe, middle lobe and anterior segment of the upper lobe were trapped with a cortex. There was a cherry red lesion on the diaphragm adjacent to a cluster of fenestrations over the central tendon, through which serosanguineous fluid oozed through when the diaphragm was pressed'. She made a good recovery and was discharged 5 days postoperatively.

Figure 2 Chest radiograph after first drain inserted, 7/10/2011.

Figure 4 Chest radiograph showing trapped lung in the setting of a second drain and no air leak, 13/10/2011.

Figure 5 Low-power image showing endometrial stromal cells with inflammatory cells and haemosiderin-laden macrophages.

OUTCOME AND FOLLOW-UP

Histopathology of pleura and lung reported foci of haemosiderin pigment deposition, reactive mesothelial proliferation, foci of dense endometrial type stroma and positivity for CD10 with no definite endometrial glands and no evidence of malignancy. The findings were reported as 'entirely in keeping with endometriosis' (figures 5 and 6). When further questioned by the thoracic surgeons, the patient admitted to a previous history of episodic breathlessness on menstruation with associated chest pain. Follow-up chest radiographs up to 1 month remained stable (figures 7–9). She was commenced on norethisterone with no recurrence at 6 months follow-up.

In this case the clinical consultations and pre-operative assessment failed to elicit any chest symptoms or signs. The apparent mismatch between the severity of chest disease and the patient's clinical symptoms is explained by the gradual build up of haemothorax over many menstrual cycles. It is regrettable that no chest signs were documented. The size of the pleural effusion and the pneumoperitoneum required for laparoscopy would have reduced her capacity for compensation, possibly exacerbated by the presence of multiple diaphragmatic fenestrations, resulting in the abrupt presentation of respiratory failure.

Figure 6 High-power image showing endometrial stromal cells, lymphocytes and haemosiderin-laden macrophages consistent with endometriosis.

Figure 7 Chest radiograph post-thoracotomy, repair of diaphragmatic fenestrations, pleurectomy and decortication of trapped lung. 32F and 28F drains in situ, 14/10/2011.

DISCUSSION

Endometriosis is the presence of endometrial glands and stroma outside of the uterine cavity. It usually occurs in the pelvis and peritoneal cavity, but has been described throughout the body.² It is common, affecting 5–15% of the general population.³ The most accepted explanation is that of retrograde menstruation with a disordered immune response, allowing implantation and proliferation of endometrial tissue within the peritoneal cavity.² It most commonly presents with dysmenorrhoea, dyspareunia and subfertility, although it may be an incidental finding at laparoscopy. The American Society for Reproductive Medicine classifies endometriosis as stage I (minimal), II (mild), III (moderate) or IV (severe), based on number, location and depth of deposits and the presence of adhesions found at laparoscopy.⁴ Severity of endometriosis based on such scoring systems does not necessarily reflect symptoms or prognosis.

TES is a rare presentation of endometriosis. It may be classified as pleural, comprising catamenial pneumothorax, catamenial haemothorax, catamenial pneumomediastinum and catamenial chest pain, and parenchymal, comprising catamenial haemoptysis and pulmonary nodules.^{2 5 6} In a case series of 110 cases, its

Figure 8 Chest radiograph at discharge, 19/10/2011.

Figure 9 Chest radiograph 1 month post-thoracotomy showing stable appearances, 14/11/2011.

most common manifestations in descending order of frequency were catamenial pneumothorax (73%), catamenial haemothorax (14%), catamenial haemoptysis (7%) and pulmonary nodules (6%).⁷ Catamenial pneumothorax is usually right-sided and coexists with ipsilateral diaphragmatic defects.⁸ The aetiology of TES is poorly understood. Hypotheses include peritoneal-pleural migration of endometrial tissue through diaphragmatic defects, coelomic metaplasia, where cells within pleura and peritoneum may differentiate into endometrial tissue, and lymphatic or haematogenous embolisation of endometrial tissue to distant sites including lung parenchyma. Whatever the presentation of TES, it usually coexists with pelvic endometriosis.⁷

The investigation of suspected endometriosis starts with pelvic ultrasound, which may find or exclude ovarian endometriomas or suggest pelvic adhesions.⁹ Pelvic laparoscopy is the gold standard for diagnosis of endometriosis.³⁻⁹ MRI is considered the non-invasive imaging modality of choice for TES, where thoracic lesions are described as increasing in size during menses, being hyperintense on T2-weighted spin-echo images, and enhancing with intravenous contrast.⁵ VATS, the gold standard investigation allowing both direct visualisation and targeted biopsy. Alternatively, tissue diagnosis may be acquired by broncho-alveolar lavage or imaging-guided biopsy.

Management of endometriosis attempts to address symptoms and reproductive needs.⁴⁻⁹ Medical approaches aim to suppress ovarian oestrogen output and include combined oral contraceptives, high-dose progestogens, danazol, gestrinone and GnRH (gonadotrophin-releasing hormone) agonists. Medical therapy

alone can reduce pain, as can the levonorgestrel-releasing intra-uterine system, but does not address fertility and there is a high recurrence rate with cessation of therapy. Surgical approaches range from removal of foci of endometrial tissue and adhesiolysis to total abdominal hysterectomy with bilateral salpingo-oophorectomy. Management of infertility depends on the extent of disease, and may involve a combination of conservative surgery with assisted reproductive techniques.

Management of TES is tailored to the presentation and aims to address the acute problem and prevent ongoing recurrences.⁶ Medical approaches involve ovarian suppression and may include chemical pleurodesis. When these fail or are not tolerated, surgical approaches aim to localise and remove the ectopic endometrial tissue and close diaphragmatic defects. This may involve resection of diaphragm or lung parenchyma, or parietal pleurectomy and pleurodesis as in our case.

Learning points

- ▶ Examination is essential even in 'simple' cases.
- ▶ Thoracic endometriosis syndrome most commonly presents with pneumothorax.
- ▶ Case series suggest thoracic MRI is the investigation of choice for suspected thoracic endometriosis syndrome, but standard work-up is key to exclude more probable causes first.

Competing interests None.

Patient consent Obtained.

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