A cyclic bleeding conundrum: A case of recurrent catamenial haemothorax

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SUMMARY

Catamenial haemothorax (CtH) is a rare manifestation of endometriosis and falls under one of the manifestations of thoracic endometriosis syndrome (TES). It is often difficult to diagnose in its initial presentation unless an accurate history of symptoms coincides with menstruation. We report a case of a nulliparous lady with a history of endometriosis who presented with a sudden onset of shortness of breath. She was initially treated as pneumonia, then presented with recurrent episodes of catamenial haemothorax. Diagnosis can be aided by computed tomography (CT) thorax, which can identify features of TES; however, the gold standard is direct visualisation with video-assisted thoracoscopic surgery (VATS) and histopathological evidence of endometrial cells in the thoracic cavity. CT Thorax revealed pleural nodules at the right hemidiaphragm, while biopsy of the pleural tissue via pleuroscopy was consistent with endometriosis. The intra-operative VATS findings included two litres of blood in the pleural cavity, as well as pleural nodules and fenestrations on the diaphragm. We discuss the difficulty in diagnosis during the initial stage, the possible pathophysiology of this condition and its treatment.

INTRODUCTION

Endometriosis is a benign gynaecological disorder defined as endometrial tissue that lies outside of the uterine cavity. It is estimated that it affects 6-10% of women in the reproductive age,[1] although the actual numbers may be higher, as many go undiagnosed. The most common site affected by endometriosis outside of the pelvic region is the thoracic cavity, which includes the lung parenchyma, diaphragm and pleura, with the right hemithorax being more affected. In the thoracic cavity, endometriosis can manifest as catamenial pneumothorax (73%) or haemothorax (14%), catamenial haemoptysis (7%) and/or pulmonary nodules (6%),[2] and falls under the umbrella term of Thoracic Endometriosis Syndrome (TES).[2-4]

CASE PRESENTATION

We present a case of a 40-year-old nulliparous woman with underlying endometriosis. She has had a chronic history of severe dysmenorrhea since her early teens, before seeking medical attention and eventually diagnosed with endometriosis in her late 20s. She underwent laparoscopic removal of endometrioma and laparoscopic cystectomy for bilateral ovarian cysts in 2009. She has been married for more than 15 years, however, she suffers from primary subfertility as a result of underlying endometriosis. Besides that, she also has a strong family history of breast cancer, leukaemia, and oral, cervical and uterine malignancy.

She initially presented with a sudden onset of breathlessness and right-sided chest pain and was treated for community-acquired pneumonia with a course of intravenous antibiotics and discharged home. However, her dyspnoea did not completely resolve, and she presented a week later with worsening shortness of breath. Chest x-ray showed she had a spontaneous right-sided pneumothorax and thus was managed with chest tube insertion. Once the pneumothorax resolved, the chest tube was removed, and she was discharged home. Outpatient contrast-enhanced computed tomography (CECT) Thorax was done to investigate the cause of spontaneous pneumothorax, and it showed two pleural-based nodules at the right hemidiaphragm. A radioconference was held, and it was noted that close correlation to menstruation history was needed.

The patient presented again with recurrent hydropneumothorax at intervals of 6 months, 3 months and 1 month before undergoing a pleuroscopy. Biopsy of the parietal pleura was consistent with endometriosis; other pleural fluid investigations were negative for malignancy and bacterial growth. She had another two episodes of recurrent haemothorax, which brings the total to seven incidences of catamenial pneumo- and haemothorax over a period of 18 months. Retrospectively, a detailed history was taken, and it was found that each episode had coincided within 1-2 days of her menstruation. Thus, she was diagnosed with recurrent catamenial haemothorax (CtH).

Upon elective admission, she was asymptomatic. However, physical examination revealed reduced breath sounds in the right basal zone, and the chest x-ray showed right pleural effusion. She underwent right video-assisted thoracoscopic surgery (VATS). Intra-operatively, there was 2L of blood in the pleural cavity with pleural nodules and fenestrations on the diaphragm, mainly at the tendinous part. Diaphragm fenestrations were resected using a stapler device, the pleural layer at the lower hemithorax was excised (pleurectomy), as well as pleural nodules, and pleurodesis was done.

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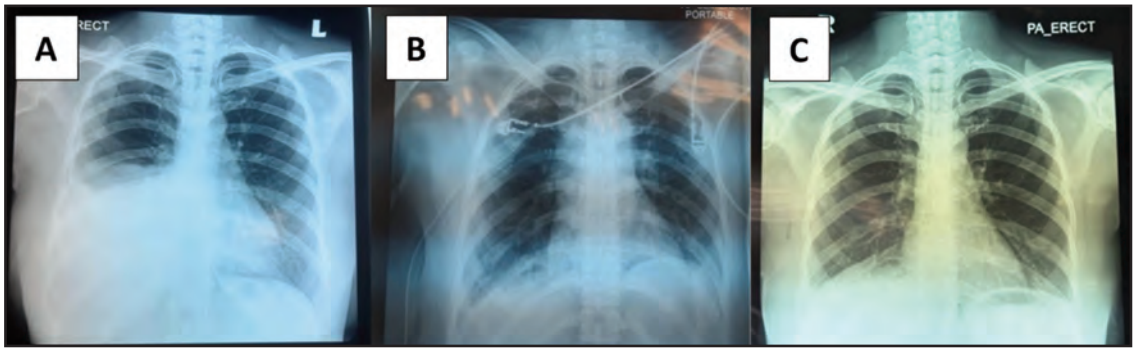

Fig. 1: (A) Pre-operative chest X-ray illustrating right-sided pleural effusion with a blunted right costophrenic angle. (B) Immediate postoperative chest X-ray with right chest drain in situ, showing resolved right haemothorax. (C) Post-operative Day 4 chest X-ray with resolved haemothorax and lungs well expanded following right chest drain removal

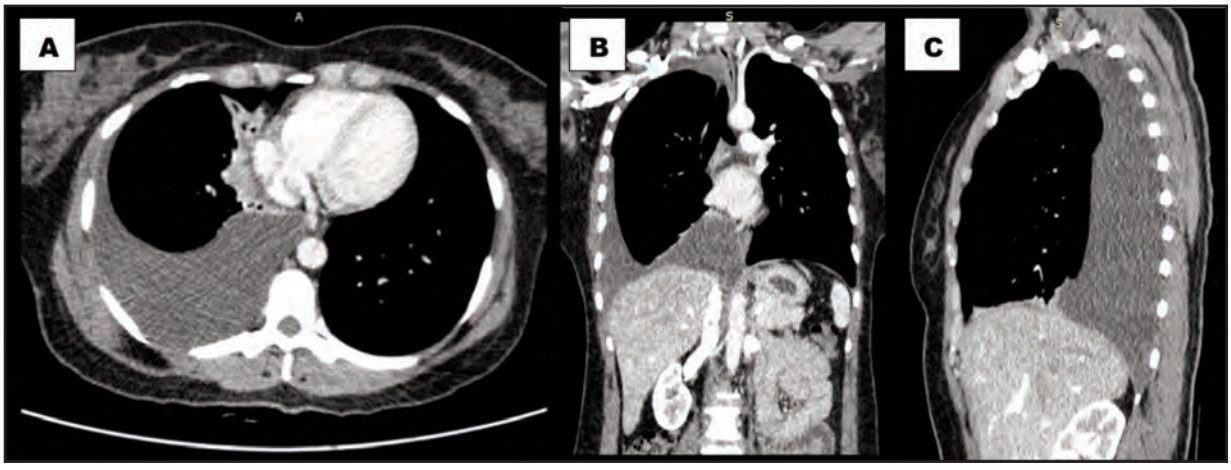

Fig. 2: Multi-density right pleural effusion on axial (A), coronal (B) and sagittal (C) cuts of CT Thorax, consistent with right haemothorax

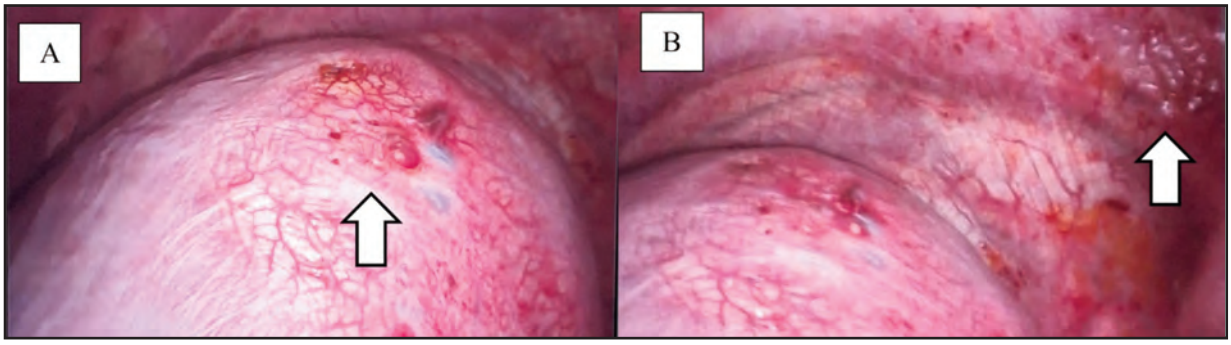

Fig. 3: Intraoperative images via video-assisted thoracoscopic surgery (VATS). (A) The arrow indicates fenestrations and nodules over the diaphragmatic surface. (B) The arrow points at multiple pleural nodules lining the pleural cavity

She recovered well in the post-operative period, the chest drain was removed on the fourth post-operative day, and she was discharged home well. Intraoperative biopsy of the pleural nodules was proven to be endometriosis. During the follow-up period, her symptoms had resolved, however, she complained of neuropathic pain. She was referred to the Gynaecology Team for hormonal treatment to manage endometriosis and the Acute Pain Service for pain optimisation.

DISCUSSION

Extra-pelvic manifestation of endometriosis is rare and is considered part of the disease progression of endometriosis, as it tends to develop after a chronic history of pelvic endometriosis, and in the later part of the reproductive age. As reported in literature,[2] the peak incidence age for endometriosis is 24 to 29 years old, while the peak incidence rate for TES is 30-34 years old. TES is difficult to diagnose, rarely during the initial presentation, as it is often missed. It

carries an interval of 8-19 months from the onset of symptoms to diagnosis and management.[5] This is quite similar to our index patient presented above, who has suffered with endometriosis since early teenage years and subsequently required surgery.

Signs and symptoms of CtH may be vague, as it can mimic a myriad of other respiratory disorders, such as infection or malignancy, and therefore is often missed in the initial stage. A high level of clinical suspicion is required to reach an accurate diagnosis. This can be achieved with thorough history-taking of symptoms that are directly linked with menstruation and/or have a close cyclical relation with menstruation. In terms of imaging, chest radiography is the first line, followed by subsequent CECT Thorax. They are both sensitive in detecting pneumo- or haemothorax, but are unable to pinpoint the underlying pathology.6 MRI, rarely done, can aid in detecting diaphragmatic lesions, but is also unable to accurately diagnose CtH. Other investigative tools include bronchoscopy or pleuroscopy, which can help obtain a biopsy in order to make a histopathological diagnosis, however, they can still be missed if lesions are located peripherally.[7] It must be noted that brush cytology may sometimes be more superior in acquiring sample for accurate diagnosis.[8] However, the gold standard for achieving a correct diagnosis of TES is via VATS, where the classical features of diaphragmatic and pleural lesions, as well as blebs, bullae and scarring, are directly visualised.[1] Our patient was initially thought to have a lung infection, then developed spontaneous pneumothorax with an unknown cause. However, after CECT revealed pleural nodules, there was clinical suspicion of endometriosis. This was later confirmed with a biopsy via pleuroscopy and reinforced during VATS.

The pathophysiology of TES is uncertain. Multiple theories have been proposed, but none can be solely attributed to the disease manifestation, therefore, it is likely that it is multi-factorial or multi-causal.[1] The most popular theory is the retrograde menstruation theory, which postulates that during menstruation, endometrial cells undergo migration through the fallopian tube and into the pelvis. There, these cells will then plaster themselves onto peritoneal surfaces. After chronic and repeated retrograde migration, endometrial cells migrate further into the right paracolic gutter and towards the right hemidiaphragm. Once there, they implant themselves onto the sub-diaphragmatic surface and subsequently migrate through congenital or acquired fenestrations in the diaphragm and enter the right pleural cavity. This theory explains why TES is more common on the right hemidiaphragm (85%), as the falciform and phreno-colic ligaments prevent migration flow to the left sub-diaphragm.[5] This is the most probable diagnosis for our patient, as evidenced by diaphragmatic fenestrations and the nature of right-sided dominance of disease.

Another theory to emerge hypothesises that mesothelial cells of the diaphragm undergo metaplasia due to external stimuli such as high oestrogen exposure and transforms into endometrial cells. This can also explain the rare cases of endometriosis found in men, following a high-dose oestrogen exposure. Besides retrograde menstruation flow, endometrial cells may disseminate via the lymphatics or haematogenous route. This theory can illustrate why bronchopulmonary lesions are bilateral, and how some rare cases of endometrial cells have been found in the brain and bone. Lastly, the prostaglandin theory accounts for rare cases of TES, where blebs and bullae are the only pathological lesions observed. During menstruation, prostaglandin F2-alpha levels increase, causing bronchoconstriction and vasoconstriction, which can then cause alveolar rupture and develop into catamenial pneumothorax or haemothorax.

The treatment for TES is with multi-modal and multi-disciplinary approach, where it can be divided into medical and surgical management, with gynaecology and thoracic disciplines. The first-line medications recommended are gonadotrophin-releasing hormone (GnRH) analogues. However, this can cause menopausal-like side effects and osteoporosis, and can fail in 50% of cases, especially when not coupled with surgery.[10] Other options are oral contraceptive medication and GnRH antagonists. Hormonal medications tend to be prescribed long-term due to the chronic and recurrent nature of the disease, even after surgery, in order to prevent recurrence.

VATS is the ideal surgical approach to manage TES manifestations, as it is both diagnostic and therapeutic.[1] It is also an option to be considered for those with recurrent refractory disease. Endometrial implantation on the diaphragm and pleura can be fulgurated with bipolar diathermy or excised with energy devices, while infiltrative or larger lesions can be resected with stapler devices, and larger defects repaired using synthetic mesh. Pleurodesis, either chemical or mechanical, can also be administered for pleural lesions as well as pleurectomy. However, there is insufficient evidence to prove that it reduces the rate of recurrenceI.9 Following surgery, these patients must be monitored with hormonal suppression therapy in order to reduce the risk of recurrence.[9]

CONCLUSION

CtH is a rare manifestation of endometriosis and is difficult to diagnose correctly. Initial symptoms may resemble common thoracic pathologies, however, there should be a high clinical suspicion in these patients who have a concurrent history of endometriosis, in order to reach a timely diagnosis and to prevent morbidity and mortality for women of childbearing age. VATS is the gold standard for diagnosis and can also be therapeutic. Although surgical pleurodesis has been found to be superior to hormonal suppression, a combined surgical and medical approach to the management of TES provides the best outcome and helps in preventing recurrence.

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DECLARATION

The authors would like to declare no conflict of interest.

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