Catamenial pneumothorax: A neglected diagnosis

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Background. Catamenial pneumothorax (CP) is an under-reported and misunderstood condition commonly defined as a recurrent, spontaneous pneumothorax occurring from the day before menstruation until 72 hours after its onset. It is the most common clinical manifestation of thoracic endometriosis.
Case report. We describe the case of a 36-year-old woman who presented twice with significant shortness of breath during her menses at a regional hospital in Gauteng Province, South Africa. Chest radiographs showed bilateral spontaneous pneumothoraces on both occasions, indicating a very rare presentation of CP, which is more commonly a right-sided pathology and a rare presentation on its own. Video-assisted thoracoscopic surgery was performed at a tertiary institution. Significant thoracic endometriosis and a diaphragmatic hernia were found, confirming the diagnosis of endometriosis-related CP. Pleurectomy and closure of the diaphragmatic hernia were performed, and the patient was initiated on hormonal therapy.
Conclusion. A high index of clinical suspicion is required for diagnosing CP and its associated conditions. This case highlights the need for awareness of this condition and the importance of long-term follow-up, especially in an overburdened and resource-limited healthcare system.
Keywords. Thoracic endometriosis, catamenial pneumothorax, bilateral pneumothorax, resource limitations.

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A catamenial pneumothorax (CP) is defined as a recurrent, spontaneous pneumothorax occurring from the day before menstruation until 72 hours after its onset.[1] It remains a diagnostic and therapeutic challenge. We describe a rare case of CP in a patient presenting with recurrent bilateral pneumothoraces. Available resources included an affiliated university and several of its departments.

African relevance

- Catamenial pneumothorax (CP) is a rare condition affecting women.
- Bilateral CPs are exceptionally rare.
- Delays in diagnosis result in multiple admissions and avoidable distress.
- A high index of clinical suspicion is needed for early diagnosis and appropriate care.
- A multidisciplinary team approach is essential in an overburdened healthcare system where access to other specialty disciplines is challenging.

Case report

A 36-year-old woman with no known comorbidities presented to a regional hospital in Gauteng Province, South Africa, with sudden onset of shortness of breath in December 2021. No history of trauma or preceding illness was noted. She had tachycardia (147 bpm), a respiratory rate of 45 breaths per minute, and oxygen saturation of 83% on room air (RA).

Chest examination revealed decreased air entry bilaterally. A chest radiograph confirmed bilateral pneumothoraces, for which intercostal drains (ICDs) were inserted. The oxygen saturation improved to 99% on RA.

The patient was admitted for further work-up. She was a non-smoker and tested negative for COVID-19, HIV and pulmonary tuberculosis. She spent 23 days in the medical ward and was discharged with an outpatient date for a computed tomography (CT) scan of the chest. The CT scan was done 37 days after presentation and showed bilateral hydropneumothorax and pleural thickening with nodules. No further management was initiated.

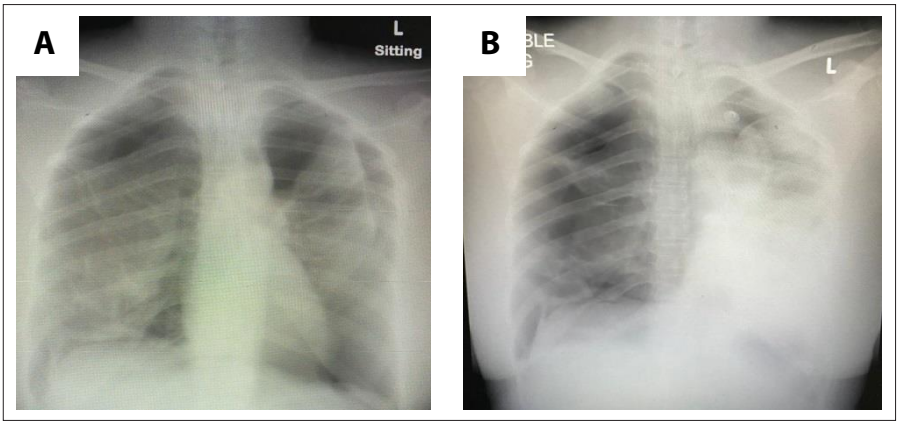

Fig. 1. Chest radiographs at first (A) and second (B) presentation.

The patient presented again to the same facility 2 months later with similar complaints. She was noted to have an increased respiratory rate and an oxygen saturation of 85% on RA. Bilateral pneumothoraces were noted on the chest radiograph, and bilateral ICDs were inserted again (Fig. 1).

Following recurrence of spontaneous pneumothoraces in an otherwise well patient, the emergency medicine consultant suggested a working diagnosis of CP. A

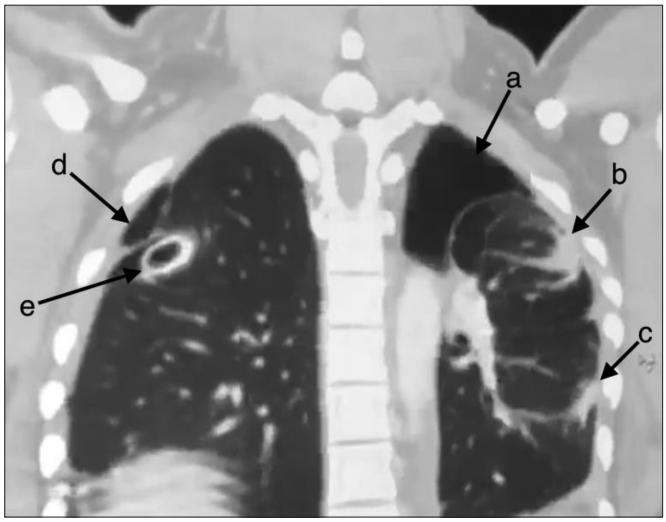

Fig. 2. Computed tomography scan after the patient's second presentation. (A) left haemopneumothorax; (B) round atelectasis; (C) round atelectasis; (D) small right haemopneumothorax; (E) right intercostal drain in situ.

comprehensive gynaecological history revealed that the patient was menstruating (between days 2 and 3 of the cycle) on both presentations and had been struggling with infertility. She had heavy/irregular menstrual cycles for which she had never sought medical help.

She was transferred to the respiratory unit of the affiliated tertiary institution the next day, and a second CT scan of the chest was performed. It showed bilateral pneumothoraces, larger on the left, with bilateral loculated effusions, pleural thickening, left-sided subpleural round atelectasis, and masses with pleuroparenchymal tethering (Fig. 2).

Rigid bronchoscopy, left video-assisted thoracic surgery (VATS) including pleurectomy and pleural biopsy was performed 2.5 weeks after transfer. A fenestrated diaphragmatic hernia was found intraoperatively and repaired. A month after the left VATS, right VATS was performed, which included a pleural biopsy and apical pleurectomy. Specimens taken showed areas of haemorrhage and CD10-positive stromal cells confirming endometriosis. The patient recovered well and was discharged 1 month after the procedure for follow-up at gynaecology, infertility and cardiothoracic outpatient departments (OPDs). She received goserelin 3.6 mg subcutaneously, and abdominal laparoscopy done 3 months after discharge showed stage 4 endometriosis. She was discharged on dienogest 2 mg daily with regular follow-up at gynaecology and infertility OPDs. She continued to do well with no recurrence of CP for 6 months. No further records could be found, and the patient was possibly lost to follow-up.

Discussion

Thoracic endometriosis is the presence of endometrial tissue outside the uterus and within the thoracic cavity. This can involve the pleura and/or the pulmonary parenchyma.[1] CP is the most common manifestation of thoracic endometriosis, but the exact mechanism is poorly understood, and theories about its development are many and varied.[2]

These theories include metaplasia, embolisation, retrograde menstruation, and intraperitoneal air.[3] The two most commonly noted theories are a ruptured bullous or alveolus following vasoconstriction,[4] and alternatively the passage of air through the cervix, uterus and peritoneal cavity, and finally to the pleural space via a diaphragmatic defect.[5]

Thirty to fifty percent of patients with CP have associated pelvic endometriosis, as in our case. It is believed that this disease entity spreads beyond the abdominal cavity with associated complications, including CP.[1]

Histological confirmation of endometrial tissue on specimens obtained from the thoracic cavity is the gold standard for definitive diagnosis.[5] These specimens are retrieved during VATS, which is the preferred procedure, as done in this case.[1]

CT findings of thoracic endometriosis are nonspecific and are most sensitive during menses, which makes the diagnosis challenging. However, CT has an essential role in ruling out other differential diagnoses and imaging the lesions before surgery.[6]

Clinical awareness is crucial in diagnosing CP to avoid significant delay before initiating necessary investigations. There are well-documented clinical clues that should raise the suspicion of CP.

Firstly, the location of CP is more prevalent on the right, in 85 - 93% of cases.[1] This position is possibly explained by the fact that common congenital diaphragmatic defects most often appear on the right, or by fluid from the pelvis continuously flowing into the right upper quadrant of the abdomen.[7] Left-sided presentations are rare, and bilateral presentation is exceptionally rare, highlighting the importance of this particular case as an atypical presentation.[1]

The paucity of literature on CP cases in Africa points to lack of clinical awareness of the condition and the tendency of clinicians to diagnose more commonly encountered pathologies such as HIV and tuberculosis. A high index of suspicion is required for prompt diagnosis,[3] and any history of infertility with or without proven endometriosis in a female (particularly women around their 40s) with recurrent spontaneous pneumothorax before or during their menstrual cycle should encourage suspicion of the condition.[4] Long-term follow-up is crucial owing to the recurrent nature of this condition. Additional risk factors include those applicable to endometriosis, e.g. early menarche/late menopause, short menstrual cycles, and prolonged menses.[8]

Recent studies suggest that CP is vastly under-reported.[4] Up to 35% of all spontaneous pneumothoraces may meet the definition of CP.[1] All cases of CP should therefore be used to create awareness of this condition and the rare presentations associated with it, such as our case.

Acute-care doctors who are cognisant of CP will be less likely to misdiagnose it, thereby decreasing the number of recurrences (average 2 - 8) for the patient before definitive diagnosis.[1,9] Owing to high rates of recurrence of CP, long-term follow-up is essential. In cases where diaphragmatic defects are present, such as in our case, surgical repair together with postoperative hormonal therapy has been shown to be associated with improved outcomes.[10,11]

In an overburdened health system, each presentation usually results in lengthy hospital admissions and an increased risk of morbidity and mortality. Education and awareness are needed in limited-resource settings where access to gynaecological services is challenging. Unfortunately, endometriosis itself is often overlooked and retrospectively investigated following the diagnosis of CP.

Conclusion

This case demonstrates that the diagnosis of CP is often missed or delayed. CP should be considered in female patients of reproductive age presenting with recurrent spontaneous pneumothoraces before or during

menses. Awareness of CP and the necessity for long-term follow-up may prevent lengthy, recurrent hospitalisations. It may also improve patient outcomes, especially in a resource-limited setting.

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