

Case Report

Catamenial Pneumothorax: Role of Serial Chest Radiographs in the Diagnosis of a Rare Entity

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ABSTRACT

Catamenial Pneumothorax is generally considered to be a rare entity. The lack of a clear cause means that diagnosis and treatment is challenging. The objective of this case report is to increase awareness and highlight the role of serial chest radiographs in improving the diagnosis of the pathology. A 33-year-old mother of two presented at the radiology department with a history of recurrent right-sided chest pain that corresponded with her menstrual period. Previous chest radiographs performed in the preceding six months all showed the presence of a right sided pneumothorax, with associated pleural effusion. In considering the patient's age, spontaneous pneumothorax occurring in conjunction with the menstrual cycle, and in the absence of other respiratory disease, a diagnosis of catamenial pneumothorax was made. The patient was commenced on oral contraceptive pills (estrogen-progestogen) and did not present with similar symptoms in the succeeding 6-months.

INTRODUCTION

Catamenial pneumothorax (CPX) is considered to be an unusual entity¹. It is defined as the recurrent (at least two episodes in total) build-up of air in the pleural cavity in reproductive age women without associated respiratory disease occurring within 24 hours before or 72 hours after the onset of menses. The spontaneous pneumothorax occurs at the same time as menstruation, and the average age of onset is between 19 and 45 years old². Catamenial pneumothorax is still considered to be misunderstood, as is endometriosis, which is considered to be underdiagnosed. The decreased disease awareness plus lack of a clear cause means that diagnosis and treatment are challenging. The disease is believed to be largely undiagnosed or misdiagnosed, leaving the true frequency unknown in the general population³. We herein report a case of CPX, diagnosed by means of serial chest radiographs. Use of chest radiographs could be cost-effective, especially in low-resource settings where other imaging modalities are not available. Furthermore, this report further seeks to increase awareness of the rare disease and alert clinicians so that diagnosis can be improved.

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PATIENT HISTORY

A 33-year-old mother of two children presented at the radiology department from a General Practitioner with a history of right sided chest pain, dry cough and shortness of breath. This was not the first time that the patient had shown up at the radiology department with similar symptoms albeit from different doctors. The episodes of the pneumothorax coincided with the menstrual cycle of the patient, usually occurring a day before the start of the menstrual cycle. These recurrent symptoms worried her family causing them to also consult a spiritual healer but to no avail. The patient's doctor had found out that there was reduced air entry into the right lung and thus requested for the chest radiograph. A chest radiograph demonstrated a right sided pneumothorax (Figure 1). The results were then compared to previous radiographic findings retrieved from the Picture Archiving and Communication System (PACS) (Figure 2 and Figure 3) that had done in the preceding 6 months. It was discovered that this pneumothorax was recurring and it was always in sync with the patient's menstrual cycle. In all the previous images, the pneumothorax was right sided, with an associated air/fluid level which could have been pleural effusion/hemothorax. The left lung was unaffected and there was no mediastinal shift and no airspace infiltrates to suggest lung disease. Comparison of these chest radiographs revealed a common pattern in the occurrence of the spontaneous pneumothorax. Considering the patient's age, spontaneous pneumothorax occurring within 24 hours of the start of the menstrual cycle, a diagnosis of catamenial pneumothorax was made. She was commenced on oral contraceptive pills (oestrogen-progestogen) did not present with similar symptoms in the succeeding 6-months.

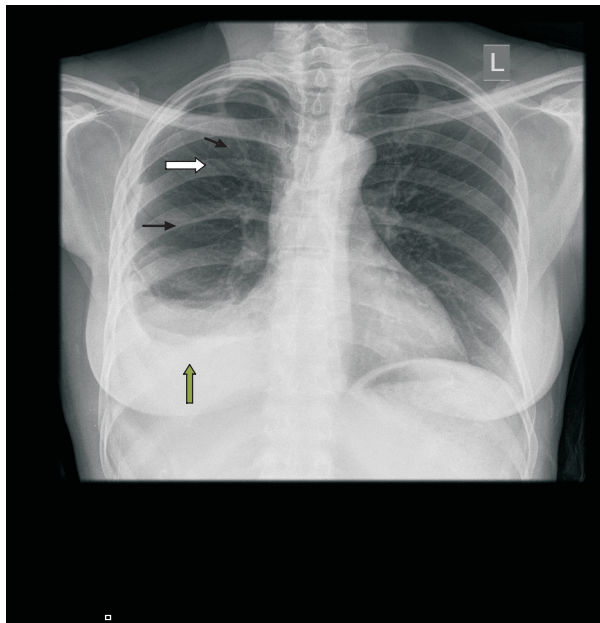


Figure 1: Third chest x-ray done 6 months after the first chest x-ray shows a recurrence of the pneumothorax (white arrow) with visible visceral pleural line (small black arrows) and an air-fluid level (green arrow) indicating a pleural effusion/hemothorax. This was when the diagnosis of CPX was made.

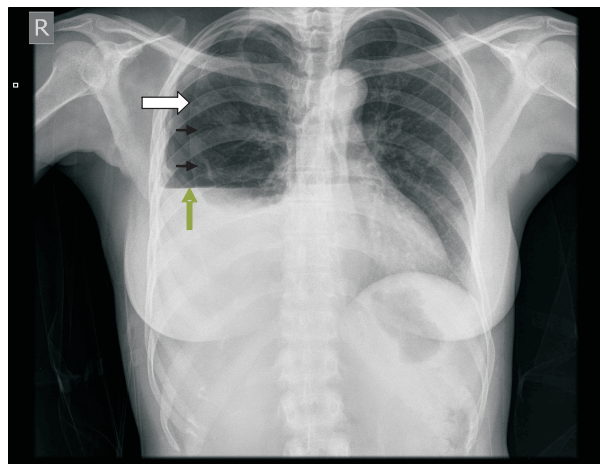


Figure 2: First chest x-ray done shows a right pneumothorax (white arrow) with an associated air fluid level extending up to the mid thorax (green arrow). It can be seen that the air fluid level crosses the midline but the trachea is not deviated. The visceral pleural line can be seen (small black arrows)

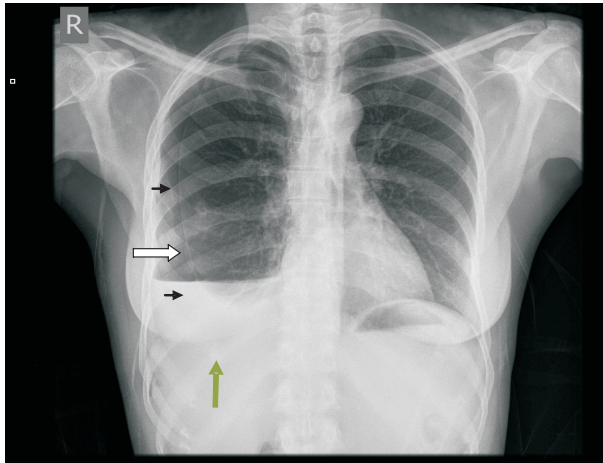


Figure 3: Second chest x-ray done 2 months after the first one. Compared to the first x-ray, it shows a much larger right lung volume with air-fluid level (green arrow) within the lower zone as compared to the previous one which was in the mid zone. The pneumothorax was still as large as in the first x-ray (white arrow) with a visible visceral pleural line (small black arrows). Still there is no mediastinal shift and the left lung is preserved. There is no airspace disease.

DISCUSSION

Catamenial pneumothorax was originally defined by Maurer et al. in 1958⁴. The term “catamenial” was then created in 1972 by Lillington. It is derived from the Greek words pertaining to and monthly⁵. According to Visouli et al.⁶, the typical clinical presentation of CPX is a spontaneous pneumothorax preceding or occurring concurrently with menstruation, accompanied by pain, dyspnoea, and cough. Other symptoms that might be present include: scapular and/or thoracic pain preceding or in sync with menses; previous episodes of spontaneous pneumothorax, with or without prior operation(s) or intervention(s); primary or secondary infertility; previous history of uterine surgical procedure or uterine scraping; symptoms and/or diagnosis of pelvic endometriosis and previous history of catamenial haemoptysis or catamenial haemothorax. This was partly consistent

with our case, who was 33 years old and exhibited chest symptoms mostly a day before (24 hours) the start of the menstrual cycle. Our patient neither reported any fertility problems nor was she diagnosed with pelvic endometriosis at the time. Nevertheless, a study by Kyama et al.⁷, revealed that pelvic endometriosis remains under-diagnosed in our African settings. Additionally, the patient showed a right pneumothorax and pleural effusion/hemothorax which is also consistent with other cases found in the literature. In a study by Marshall et al.⁸, 100% of the participants with CPX had a predominant right sided occurrence. The preference for the right hemi-diaphragm is attributed by some to the well-described 'piston effect' exerted by the liver⁹. This case emphasises that whenever a female patient of reproductive age presents with chest pain or recurrent pneumothorax; it is advisable to record an elaborate mensuration history and evaluate for CPX.

The aetiopathology of CPX remains generally unknown. However, several theories have been postulated. Firstly, there is the metastatic or lymphovascular microembolization theory, which postulates that CPX is caused by the anomalous movement of uterine endometrial tissue to the diaphragm or the pleural space. This endometrial tissue can cause fenestrations in the diaphragm, which would allow air and fluid to traverse into the pleural space⁶. Secondly, in the hormonal model, scholars hypothesize that prostaglandin F₂, which is elevated during ovulation, causes constriction of the bronchioles. Bronchiolar narrowing may cause the alveoli of the lungs to rupture, allowing air to become trapped in the pleural cavity. The anatomical model, suggests that the lack of the cervical mucous plug, a normal occurrence during the menstrual cycle, permits air to pass from the genital tract into the pleural space through fenestrations in the diaphragm¹. As much as there is no general consensus on which of the aforementioned theories best describe the pathogenesis of CPX, it is generally thought that CPX is a result of endometrial deposits from

endometriosis seeding through a diaphragmatic defect or fenestration, thus a manifestation of thoracic endometriosis⁹.

The diagnosis of CPX traditionally relied on clinical signs¹⁰. Chest radiography is now usually the first line of diagnostic imaging in patients presenting with chest pain. However, the difficulty in the diagnosis of the condition is also due to a lack of suggestive imaging signs. In general, no specific diagnostic imaging criteria exist^{4,11}. In our case, diagnosis was established based on the serial chest radiography findings and treatment was initiated without any further imaging. This case therefore highlights the important role that serial chest radiographs can play in the diagnosis of CPX, especially in low resource settings. With a good PACS and Radiology Information System (RIS), previous images can be easily accessed for comparison, thereby improving the accuracy of diagnosing CPX. However, Computed Tomography (CT) and Magnetic Resonance Imaging (MRI) can aid in the identification of pleural lesions, which are not visible on plain radiographs but are a common finding in surgery¹². These modalities are, nevertheless, expensive and may not be readily available in most low resource settings.

There are currently two modalities to treat CPX, namely hormonal and surgical. Hormonal treatment mainly includes the use of oral contraceptive pills, danazol, progestational agents, and gonadotropin-releasing hormone (GnRH) analogs⁸. This case recovered after oral contraceptive pills (oestrogen-progesterone) which were used continuously without inducing menses. There were no occurrences of CPX during a 6-month use of the oral contraceptive pills. However, hormonal therapy seems to be effective in sustaining the effects of surgical treatment⁸. There are numerous surgical options to treat CPX, such as Video-assisted Thoracoscopic Surgery (VATS) with thoracotomy as an alternative, tubal ligation, and total abdominal hysterectomy with bilateral oophorectomy. Joint surgical and hormonal therapy is thought by most to be the best therapeutic modality for patients with CPX¹³.

CONCLUSION

This case highlights the important role of serial chest radiographs in diagnosing CPX. A good PACS and RIS also aided this diagnosis as previous images were easily available for comparison. In the absence of higher-end modalities, this could be a cost-effective alternative. Additionally, it is also of paramount importance to suspect CPX whenever a fertile-aged female patient presents with chest pain or recurrent spontaneous pneumothorax.

LIMITATIONS

There were no follow up radiographs that were taken for comparison. This was due to the fact that she presented with no chest symptoms in the succeeding 6-months after the intervention.

ETHICAL CONSIDERATIONS

The permission to use the radiographic images were obtained from the radiology manager of our medical centre. Written and signed informed consent was obtained from the patient after assurance that her name will not be included in the report.

CONFLICT OF INTEREST.

No conflict of interest to declare from the authors.

REFERENCES

1. National Organization for Rare Diseases (NORD). <https://rarediseases.org/rare-diseases/catamenial-pneumothorax/>. (Accessed on 05 Jan 2022).
2. Marjanski T, Sowa K, Czaplak A, Rzyman W. Catamenial pneumothorax- A review of literature. *Polish Journal of Thoracic and Cardiovascular Surgery*. 2016; 13(2): 117-21. <https://doi:10.5114/kitp.2016.61044>
3. Maniglio P, Ricciardi E, Meli F, Vitale SG, Noventa M, Vitagliano A, Caserta D. Catamenial pneumothorax caused by thoracic endometriosis. *Radiology case reports*. 2018; 13(1): 81-85 <https://doi.org/10.1016/j.radcr.2017.09.003>

4. Maurer ER, Schall JA, Mendez FL. Chronic recurring spontaneous pneumothorax due to endometriosis of the diaphragm. *JAMA*. 1958; 168:2013–2014.
5. Weerakkody Y, Sharma R. Catamenial pneumothorax. Reference article, Radiopaedia.org. (Accessed on 05 Jan 2022) <https://doi.org/10.53347/rID-17512>.
6. Visouli AN, Darwiche K, Mpakas A, Zarogoulidis P, Papagiannis A, Tsakiridis K, Zarogoulidis K. Catamenial pneumothorax: a rare entity? Report of 5 cases and review of the literature. *Journal of Thoracic Disease*. 2016; 4(1): 17-31 <https://doi:10.3978/j.issn.2072-1439.2012.s006>
7. Kyama CM, Mwenda JM, Machoki J, Mihalyi A, Simsa P, Chai DC, D'Hooghe TM. Endometriosis in African Women. *Women's Health*. 3(5): 629-635.
8. Marshall MB, Ahmed Z, Kucharczuk JC, Kaiser LR, Shrager JB. Catamenial pneumothorax: Optimal hormonal and surgical management. *European Journal of Cardio-thoracic Surgery*. 27(2005): 662–666.
9. Visouli AN, Zarogoulidis K, Kougioumtzi I, Huang H, Li Q, Drylli G, Zarogoulidis P. Catamenial Pneumothorax: From definition to diagnosis and treatment. *Journal of Thoracic Disease*. 2014; 6(4): S448-S460. <https://dx.doi.org/10.3978/j.issn.2072-1439.2014.08.49>
10. Kirschner PA. Porous diaphragm syndromes. *Chest Surg Clin North Am*. 1998; 8(449-472)
11. Dong, B., Wu, CL., Sheng, YL. Catamenial pneumothorax with bubbling up on the diaphragmatic defects: a case report. *BMC Women's Health*. 2021; 21: 167 . <https://doi.org/10.1186/s12905-021-01318-0>
12. Ciudad MJ, Santamaría N, Bustos A, Ferreirós J, Cabeza B, Gómez A. Imaging findings in catamenial pneumothorax. *Radiologia*. 2007; 49(4): 263-7. doi: 10.1016/s0033-8338(07)73768-2.
13. Papafragaki D, Concannon L. Catamenial Pneumothorax: A Case Report and Review of the Literature. *Journal of Women's Health*. 2008; 17(3): 1