

# Breathlessly bleeding: A case report of catamenial pneumothorax\*

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## ABSTRACT

Catamenial pneumothorax is the monthly manifestation of air in the lungs that relates to menstruation. Main objective is to impart knowledge on the mechanism and management of catamenial pneumothorax in a patient with recurrent pneumothorax. We describe a case of catamenial pneumothorax with description, images and histopathologic evidence of the diagnosis. Standard treatment plan should be made for cases of catamenial pneumothorax.

*Keywords: catamenial, endometriosis, pneumothorax*

## INTRODUCTION

Spontaneous pneumothorax is the collapse of the lung without any occurrence of trauma or obvious precipitating cause. There are two types of spontaneous pneumothorax. Primary spontaneous pneumothorax occurs with no underlying lung disease while there is an underlying lung disease for the secondary spontaneous pneumothorax. Under primary spontaneous pneumothorax is recurrent spontaneous pneumothorax. Recurrent spontaneous pneumothorax can occur in a patient with a history of endometriosis. Endometriosis is the presence of endometrial tissue outside the uterine cavity. It can occur in almost all organs systems namely gastrointestinal, muscular, central nervous system, and pulmonary system.

Recurrent spontaneous pneumothorax was first described by Maurer et al in 1958 and this was further studied and categorized as catamenial pneumothorax by Lillington et al in 1972.<sup>1,2</sup> Catamenial pneumothorax is the monthly manifestation of air in the lungs that is connected with menstruation. It usually occurs the day before or three days after menstruation.<sup>3</sup> Patients with catamenial pneumothorax are more commonly noted to have pleural endometriosis. Clinical manifestations of catamenial pneumothorax are shortness of breath, chest pain dyspnea, and rarely hemoptysis. These symptoms may be explained by the route of spread of the endometriosis from the uterus to the lungs.

There are multiple hypothesis that may explain the occurrence of catamenial pneumothorax: transperitoneal, hematogenous, diaphragmatic stromal endometriosis. In the transperitoneal hypothesis, presence of diaphragmatic pores causes catamenial pneumothorax. Hematogenous hypothesis explains that there are deposits of endometriotic tissue noted at the pleural parenchyma. According to Makhija and Marrinan, although diaphragmatic fenestrations have been described in the past, it was not proven that a pulmonary air leak caused the catamenial pneumothorax.<sup>4</sup> On the other hand, Fukunaga described that the cause of catamenial pneumothorax was due to diaphragmatic stromal implants.<sup>5</sup> Despite knowing multiple hypotheses that can cause catamenial pneumothorax, it is difficult to decide on which treatment that can ultimately benefit the patient in the long term. Different studies reveal that patients with this disease can either be treated medically, surgically or both.

Medically, the use of GnRH, danazol and dienogest has been shown to treat catamenial pneumothorax. GnRH agonists acts by desensitizing the pituitary gland which in turn reduces the secretion of luteinizing hormone (LH) and follicular stimulating hormone (FSH). This causes the body to undergo “pseudomenopause” or “medical oophorectomy”. This in turn decreases the production of estrogen, inactivates and degenerates endometriotic implants. Danazol is a synthetic androgen, and the increasing levels of androgen in the body decreases estrogen production. Dienogest, on the other hand, is an oral progestin which also decreases the production of estrogen.

There have been few reports so far describing multiple etiologies of catamenial pneumothorax seen in a patient and how treatment was done. We report a case of catamenial pneumothorax with diaphragmatic

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fenestrations, diaphragmatic stromal endometriosis and how it was diagnosed and treated thereafter.

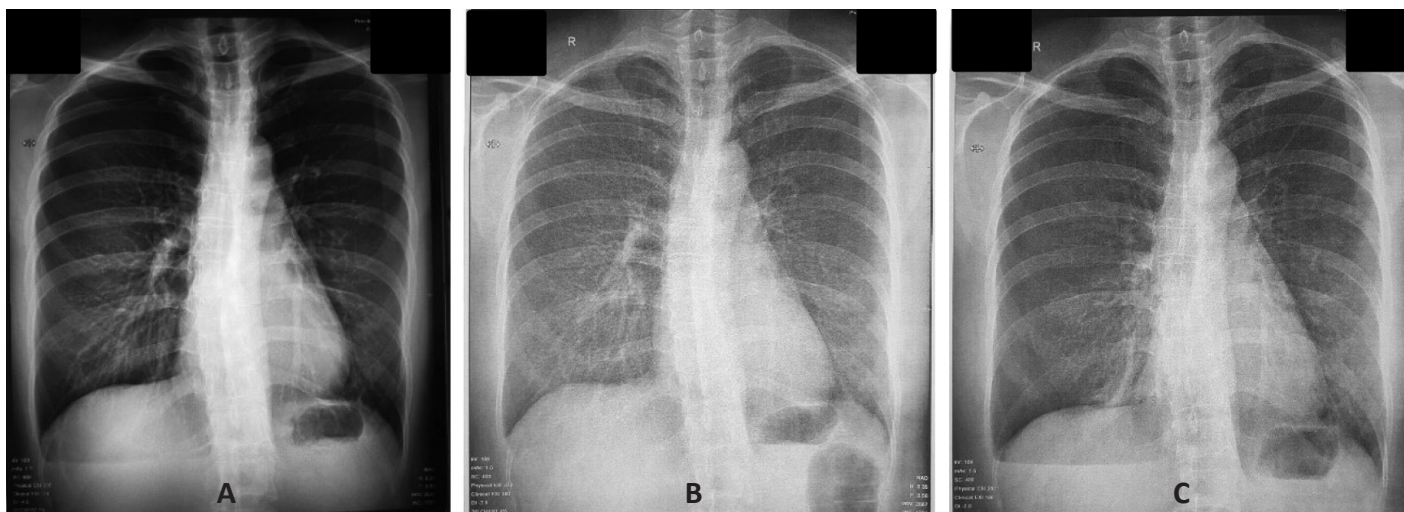
## CASE REPORT

A 42-year-old patient was referred to our service due to recurrent episodes of shortness of breath. The patient has no history of any cardiac or thyroid disease. Patient was noted to have recurrent episodes of pneumothorax on the right since October 2017 to January 2018 (Figure 1 A-C). Upon further examination, sputum exams revealed that she has pulmonary tuberculosis. Patient was treated with isoniazid, rifampin, pyrazinamide, and ethambutol. Three weeks after treatment, patient again had trouble breathing. Chest radiograph revealed pneumothorax. Chest tube insertion was then done to relieve the symptoms. Interval history revealed no recurrence until February 2018, where patient was again noted to have pneumothorax. Due to the multiple recurrence of the pneumothorax, it was decided that the reason for the disease was probably not only pulmonary. Patient was then advised admission to undergo right video-assisted thoracic surgery (VATS). Upon opening the pleural space was explored for any type

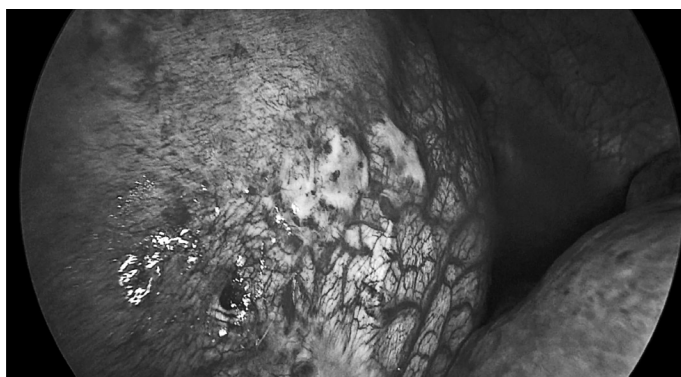
of lesions that could explain the recurrent pneumothorax. Surgery revealed multiple endometriotic implants on the diaphragm with diaphragmatic fenestrations (Figure 2). All of the implants noted on the diaphragm were removed and electrocauterized. After the excision of the endometriotic implants the diaphragm was repaired with Vicryl 2-0. Histopathologic results revealed endometriotic implants over the diaphragm (Figure 3). One week after the operation, patient followed up with her obstetrician. She was started on GnRH agonist every month. After 4 months of treatment, the patient did not have recurrence of pneumothorax.

## CASE DISCUSSION

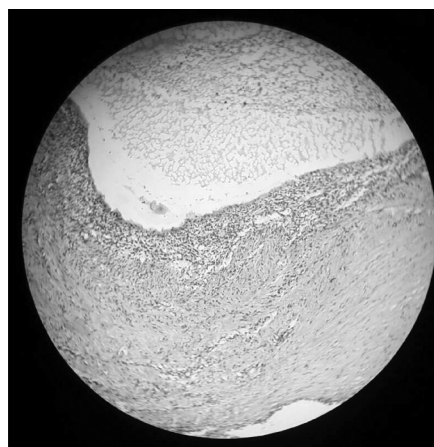
Catamenial pneumothorax occurs in the third to fourth decade of life which coincides with that of endometriosis. It is apparent 48 to 72 hours after the onset of menstruation. Studies by Maurer and Crutcher revealed that congenital anomalies can cause fenestrations on the diaphragm and that the endometrial implants to the lungs can originate from the extrusion of the mucus plug during menstruation.<sup>6,7</sup> The fenestrations can also be due



**Figure 1.** Right sided pneumothorax, (A) November 2017, (B) December 2017, (C) January 2018.



**Figure 2.** Diaphragmatic fenestrations with diaphragmatic stromal endometriosis seen on VATS.



**Figure 3.** Endometrial glands and stroma seen on specimen obtained in VATS.

to the diaphragmatic stromal endometriosis which can cause holes on the diaphragm. These holes may then be a passageway for air from the genital tract to the abdomen into the pleural cavity during menstruation. Meyers and Holm-Nielsen explained the predominance of right sidedness may be due to the preferential flow of peritoneal fluid and endometrial tissue from the pelvis along the right paracolic gutter up to the subphrenic space.<sup>8</sup> These hypotheses was demonstrated by our case above.

Catamenial pneumothorax can go undetected in many patients. That is why it is important to take a thorough history and physical examination in all patients experiencing recurrent spontaneous pneumothorax. Symptoms of endometriosis may be so mild that it can go undetected for many patients. In these cases, catamenial pneumothorax will be difficult to diagnose. As describe in the case above, it was clear that multiple etiologies can cause catamenial pneumothorax. Knowing the etiology is only the first step in treating patients with this disease. the goal of treatment is to prevent it from recurring.

Endometriosis itself is a difficult disease to treat. Adding thoracic endometriosis makes it even more challenging to treat. There are multiple studies that explain the treatment of catamenial pneumothorax. Some authors say that surgical treatment alone prevent the recurrence of catamenial pneumothorax.<sup>9</sup> Ichiki et al. described that there are multiple surgical interventions that can treat catamenial pneumothorax. Some of these procedures include plueral ablation, resection of apical blebs and parenchymal implants, excision and closure of diaphragmatic implants. In our case, the last procedure was done on the patient. Other authors claim that

medications with GnRH agonists, danazol or dienogest alone can prevent recurrence.<sup>10</sup> Lolis et al. revealed that GnRH agonists are as effective as danazol in the treatment of pelvic endometriosis with lesser side effects.

The main goal of treatment is to prevent the recurrence of endometriosis. Our patient presented with multiple etiologies namely endometriosis with diaphragmatic fenestrations and diaphragmatic stromal endometriosis. It is recommended that in patients with severe or life threatening catamenial pneumothorax, surgery be done to remove the endometriotic implants. Addition of medical treatments such as GnRH agonists will optimize the treatment in the prevention of life threatening pneumothorax as well as the pelvic endometriosis. This management was exemplified in our case presented.

## SUMMARY

Decision on whether to use surgical or medical approach depends on the patient's age, parity, co-morbidities, and financial capability. In our case, the patient is a 42yo with a completed family and with life threatening pneumothorax, hence it is advisable to do the VATS to ablate the endometrial implants and repair the diaphragmatic fenestrations. As for the use of medical treatment such as GnRH agonist, it is used to prevent the recurrence of severe pneumothorax. After the surgery, our patient was treated with GnRH agonist and has no recurrence for the past 4 months. Since this disease has not been studied thoroughly, we recommend that more studies be made to search for a standard treatment in patients with catamenial pneumothorax. ■

## REFERENCES

1. Maurer ER, Schall JA, Mendez FL. Chronic recurring spontaneous pneumothorax due to endometriosis of the diaphragm. *JAMA*. 1958; 168.
2. Lillington GA, Mitchell SP, Wood GA. Catamenial pneumothorax. *JAMA*. 1972; 219:1328-32.
3. Fukuoka, M, et al. Clinical Characteristics of Catamenial and Non-Catamenial Thoracic Endometriosis-Related Pneumothorax. *Respirology*. 2015; 20(8):1272-1276.
4. Makhija, Z, and Michael M. A Case of Catamenial Pneumothorax with Diaphragmatic Fenestrations. *The Journal of Emergency Medicine*. 2012; 43(1):e1-3.
5. Fukunaga, M. Catamenial Pneumothorax Caused by Diaphragmatic Stromal Endometriosis. *Apmis*. 1999; 107:7-12.
6. Maurer ER, Schaal JA, Mendez FL Jr. Chronic recurring spontaneous pneumothorax due to endometriosis of the diaphragm. *JAMA*. 1958; 168:2013-4.
7. Crutcher RR, Walter TL, Blue ME. Recurrent spontaneous pneumothorax associated with menstruation. *J Thorac Cardiovas Surg*. 1967; 54:599-602.
8. Meyers MA. Distribution of intra-abdominal malignant seeding: dependency on dynamics of flow of ascitic fluid. *Am J Roentgenol Radium Ther Nucl Med*. 1973; 119:198-206.
9. Ichiki, Y, et al. Surgical Treatment of Catamenial Pneumothorax: Report of Three Cases. *Asian Journal of Surgery*. 2015; 38(3):180-185.
10. Lolis, D., et al. Successful Conservative Treatment of Catamenial Pneumothorax with GnRH Agonist. *Archives of Gynecology and Obstetrics*. 1995; 256(3):163-166.