

Recurrent catamenial pneumothorax caused by diaphragmatic fenestration*

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Introduction

Catamenial pneumothorax (CP), which is a rare cause of spontaneous pneumothorax, is defined as a pneumothorax which starts at the onset or within the first 72 hours of the menstruation period in women.¹ CP was first described by Maurer and Schaal in 1958.² The reported incidence of CP among the spontaneous pneumothorax between 3% and 6%. But the actual incidence is higher than those reported (25-30%), because CP is usually underdiagnosed and the incidence is often underestimated.^{1,3-6} Although CP is clinically similar to the other primary spontaneous pneumothorax cases, it is distinguished by being observed in women of reproductive age and by the recurrence in menstrual periods.

Pathophysiology of CP is uncertain. However, several hypotheses have been defined. Thoracic endometriosis or subpleural endometrial implants seem to be the most realistic explanation for CP. If thoracic endometriosis does not detect, diaphragmatic fenestrations should be considered. In this situation, air enters peritoneal cavity through genital tract during menstruation, then, passes into the right hemithorax through the fenestrations on that side of the diaphragm and causes pneumothorax.^{4,5}

In this article, we aimed to present a CP case who underwent rethoracotomy due to multiple episodes of recurrent pneumothorax, and revealed diaphragmatic fenestrations at the operation.

Case Summary

A 33-year-old woman presented with the complaint of chest pain and a history of recurrent pneumothorax episodes. Chest X-ray revealed pneumothorax on the right side,

and the patient was referred to our clinic for further evaluation and treatment (*Figure 1*). According to her history, first pneumothorax attack occurred on the right side five years ago and she was only observed without placement of chest tube. Two months previously, she had had another pneumothorax attack in the right hemithorax. A chest tube had been placed at another hospital, and she had been followed for 15 days with chest tube due to prolonged air leak. Then, she had undergone partial decortication + apical wedge resection via right axillary thoracotomy. During the operation, no bullae, blebs and endometrial tissue was observed and the patient had been discharged with full recovery. Histopathological examination of the decorticated pleura and wedge resected parenchyma revealed no occurrence of bullae, blebs and endometrial tissue also. Twenty days after the operation, her complaints recurred. Her chest X-ray revealed a right pneumothorax and she was referred to our clinic.

According to medical history, she had no comorbidities except for a fibroadenoma in her right breast and infertility. She had a smoking habit of 10 pack years. When the history was inquired for the suspicion of CP, it was found out that she had been in the menstrual cycle in every pneumothorax attack. In the physical examination; she had an asthenic body type, breath sound was decreased in the right hemithorax, and scars due to a previous thoracotomy and chest tube incisions were identified in the right axillary region. Her body temperature was 36.9° C, pulse rate was 74 per minute, blood pressure was 100/60 mmHg and respiration rate was 18 per minute. Laboratory and biochemical parameters were normal. Thorax CT of the patient revealed that the pneumothorax in the right hemithorax had caused the lower lobe to collapse (*Figure 1*).

The patient underwent lateral thoracotomy with the diagnosis of recurrent pneumothorax. Thickening parietal pleura and 5-6 defects in the tendinous part of the

diaphragm were observed at the exploration. The defects were about 1-2 mm in diameter (**Figure 2**). No intrathoracic endometriosis, blebs, or bullae were visualized. Because of the suspicion of thoracic endometriosis, the diaphragm was partially excised together with one of the defects (**Figure 3**). Parietal pleura was decorticated, and the defects in the diaphragm were repaired interrupted silk sutures. The patient was discharged on the postoperative 6th day. Histopathological examination revealed no endometrial tissue in the diaphragm and pleura (**Figure 4**). After being discharged, she was referred to an obstetrics and gynecology clinic for infertility. Her pelvic ultrasonography revealed myoma uteri and endometrial polyp. She underwent laparoscopic myomectomy. No pelvic endometriosis was identified during the procedure. The patient was in the postoperative 12th month, and no problems or recurrent pneumothorax occurred.

Discussion

Catamenial pneumothorax is a type of spontaneous pneumothorax which clinical presentation involves recurrent pneumothorax during or preceding menstruation. The presenting symptoms are chest pain, shortness of breath and rarely hemoptysis. More than one pneumothorax episodes are observed before diagnosis. Pneumothorax is usually right-sided, and recurrence disposition during menses is high. Symptoms of pelvic endometriosis have been observed in most patients.^{1,4}

Endometriosis is defined as the location of endometrial tissue outside the uterus. The disease is usually observed in women of reproductive age, and causes pelvic pain and infertility. Catamenial pneumothorax (73%), catamenial hemothorax (14%), catamenial hemoptysis (7%) and parenchymal lesions (6%) are the most common presentation of

thoracic endometriosis^{4,5} CP is investigated in 2 groups; endometriosis-related and nonendometriosis-related. Alifano et al reported that the incidence of nonendometriosis-related CP, which developed due to the diaphragmatic defects, was 3.5% (4/114).⁵ Our case is also in this group.

Although the exact pathophysiology of catamenial pneumothorax is still unknown, there are three main theories. According to the microembolization theory, endometrial cells reach to the pleural cavity through the venous system or the transdiaphragmatic lymphatics. Most of the pleural lesions are right-sided and cyclical necrosis of the pulmonary foci may cause air leaks. In the physiologic hypothesis, the suggestion is that the high levels of prostaglandin F₂ during ovulation causes vasoconstriction and ischemia in the lung tissue. The occurrence of bronchospasm causes alveolar rupture and pneumothorax. The transgenital-transdiaphragmatic passage of air theory, it is suggested that the diaphragmatic fenestrations which have developed related or unrelated with the endometrial tissue allow the air to pass from abdominal cavity into pleural cavity in the absence of cervical mucus plug.^{4,6} This last theory explains the nonendometriosis-related CP better. However, it fails to explain why pneumothorax does not develop in every menstrual cycle.⁵

Characteristic lesions of CP are diaphragmatic defects and/or nodules on the diaphragm, and/or on the visceral and/or parietal pleura, even one of these is enough for the diagnosis. Thorax CT and MRI are frequently employed in the diagnosis of CP. But thorax CT is insufficient for the identification at diaphragmatic implants and fenestrations. MRI is frequently employed for patients whose medical history is suspicions for CP.³⁻⁶

The endometrial implants can be single or multiple and are found usually on the diaphragm. The diaphragmatic defects can be single or multiple and mainly located at the tendinous part of the diaphragm. They are usually identified intraoperatively, and thus transdiaphragmatic passage of the air is proven.⁶⁻⁸ Alifano et al reported that the rates of pleural endometriosis and diaphragmatic defects in patients with CP had been quite low in the previous publications, and they attributed this to the treatment methods employed in the past and to the widespread use of thoracoscopy in recent years.⁵

In cases of suspected CP, visceral and parietal pleura should be examined for endometrial implants, bullae and blebs, and the diaphragm should be examined for defects. Among women who underwent routine surgical treatment for recurrent pneumothorax, the rate of CP diagnosis reaches up to 25%. The rate of apical bullae and blebs in patients with CP is low (30%).^{4,5}

Treatment options for CP include tube thoracostomy in every episode, suppression of ovulation through oral contraceptives or low-dose androgens, chemical pleurodesis, hysterectomy and bilateral oophorectomy, and pleural abrasion or pleurectomy. Today, VATS has an important role in the diagnosis and treatment of CP, however, surgical techniques differ. These techniques are; resection of the diaphragm or plication of the fenestrations identified during thoracoscopy, placement of mesh over the fenestrations or glued with BioGlue surgical adhesive, removal of the endometrial foci, and pleurodesis.^{3-6,9}

Postoperative recurrence rate has been reported to be 30-40%. To prevent the recurrence, diaphragmatic defects should certainly be closed, when necessary, the diaphragm should be excised, and pleurodesis or pleurectomy should certainly be added

into the surgical procedure. Furthermore, the combination of surgical and hormonal treatment decreases the recurrence rate.⁴⁻⁶ In their series, Ciriaco et al identified recurrence in 4 patients out of 10. Just as in our case, two of these patients had undergone operations in other medical centers, and diaphragmatic defects were identified during the second operations.¹⁰

Teaching Points

Catamenial pneumothorax is observed in women of reproductive age and by the recurrence in menstrual periods. CP should be considered in mind for a young female presenting with recurrent spontaneous pneumothorax.

Catamenial pneumothorax classified into two groups; catamenial endometriosis-related pneumothorax and catamenial nonendometriosis-related pneumothorax.

MRI is superior to computerized tomography while evaluating a patient who has suspicious for CP.

The choice of treatment for woman presenting with recurrent pneumothorax should be surgical exploration. In cases of suspected catamenial pneumothorax, it is important to perform a thorough examination of the chest cavity and lung parenchyma to check for possible endometrial implants and diaphragmatic fenestrations.

Characteristic operative findings of CP are diaphragmatic defects and/or nodules on the diaphragm, and/or on the visceral and/or parietal pleura, even one of these is enough for the diagnosis. Endometrial implants should be resected and diaphragmatic fenestrations repaired appropriately.

Depending on the case, hormonal treatment can be supplementary after the surgery. Hormonal treatment can be applied for patients whose pathologic examination confirmed with endometrial tissues. It is not sufficient for patients only with diaphragmatic fenestration.

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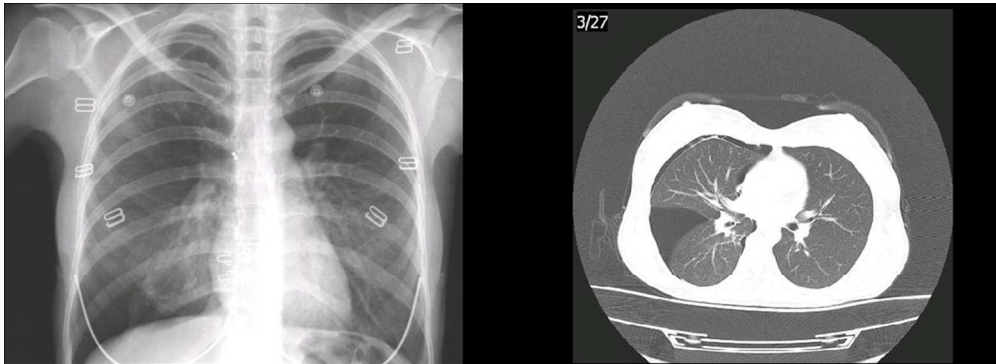
Figure Legends:

Figure 1: Chest X-ray and thorax CT revealed pneumothorax in the right hemithorax.

Figure 2: At the exploration, numerous fenestrations were detected on the tendinous part of right diaphragm (marked with arrows).

Figure 3: We can see the defect on both side (**a**, pleural surface; **b**, peritoneal surface) of partially excised diaphragm.

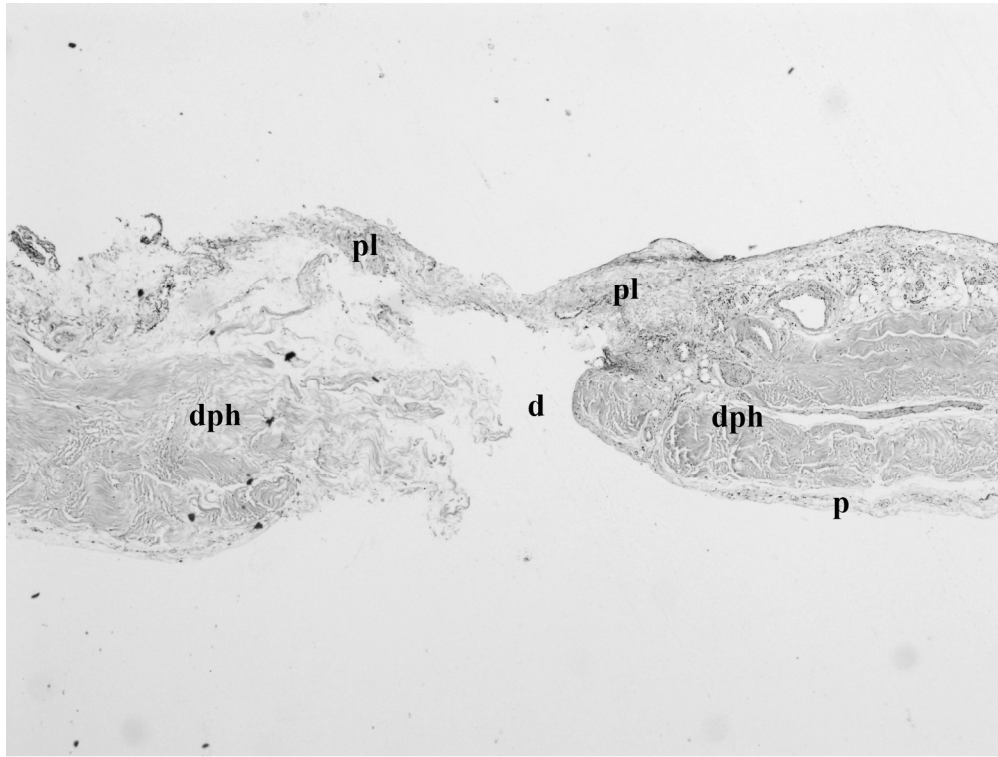
Figure 4: We can see the defect microscopically on the diaphragm (**d**: defect, **dph**: diaphragm, **pl**: pleura, **p**: periton)



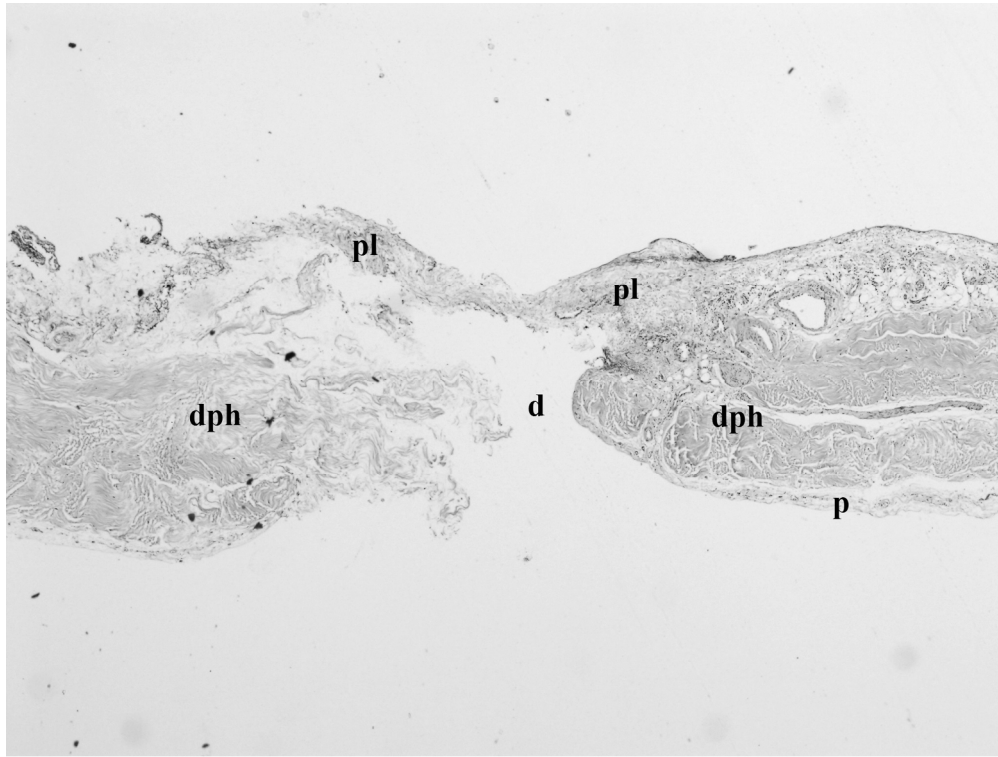
Chest X-ray and thorax CT revealed pneumothorax in the right hemithorax.
220x79mm (150 x 150 DPI)



At the exploration, numerous fenestrations were detected on the tendinous part of right diaphragm (marked with arrows).
153x115mm (75 x 75 DPI)



We can see the defect microscopically on the diaphragm (d: defect, dph: diaphragm, pl: pleura, p: periton)



We can see the defect microscopically on the diaphragm (d: defect, dph: diaphragm, pl: pleura, p: periton)