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RESPIRATORY CASE REPORTS

Catamenial Pneumothorax and Skin Endometriosis: A Case Report

Katamenial Pnömotoraks ve Cilt Endometriozisi: Olgu Sunumu

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Abstract

Catamenial pneumothorax (CP) is an uncommon disease that is challenging to diagnose and manage. It is characterized by recurrent episodes of spontaneous pneumothorax occurring periodically within 72 hours of menses in women of childbearing age. Treatment typically includes video-assisted thoracic surgery (VATS) and hormonal treatment. This case report is of a 37-year-old female who was presented with her fourth episode of recurrent right-sided pneumothorax, her two-year status post-VATS bullectomy, chemical and mechanical pleurodesis, and dienogest treatment. This case report details the natural history of a CP presentation with a high index of suspicion to diagnose, meticulous surgical exploration of the hemithorax, looking particularly for bullae and diaphragmatic pleural endometriosis or fenestrations, and application of both mechanical and chemical pleurodesis to prevent a recurrence. This report should encourage the combined adoption of chemical and mechanical pleurodesis during VATS in CP patients suffering from repeated recurrence.

Keywords: *Catamenial pneumothorax (CP), Endometriosis, Talc pleurodesis, VATS.*

Öz

Katemenial pnömotoraks, periyodik olarak menstürasyonu takip eden 72 saat içinde meydana gelen ve tekrarlayan spontan pnömotoraks atakları ile karakterize, teşhis ve tedavisi zor olan nadir görülen bir durumdur. Burada, VATS büllektomi, kimyasal ve mekanik plörodezis, dienogest tedavileri sonrası iki yıl içerisinde dördüncü rekürren sağ pnömotoraks atağı ile başvuran 37 yaşında bir kadın hasta sunulmaktadır. Tedavisi, tüp torakostomi, sağ vats ile alt ve orta lob büllektomi, diafragma açılması, ektopik diafragmatik endometrial dokunun rezeksiyonu, apikal wedge rezeksiyon, parietal plevra abrazyonu ve talk ile plörodezisden oluşmaktadır. Bu olgu sunumda, tanıda yüksek şüphe, titiz cerrahi eksplorasyon, özellikle bülleri ve diafragmatik plevral endometrium dokusunun araştırılmasını, hem mekanik hem de kimyasal plörodezis uygulamalarını gerektiren bir katemenial pnömotoraks olgusunun doğal seyrini detayları ile ortaya koymuştur. Bu sunum, yineleyen nükslere mazur kalan katemenial pnömotorakslı hastalarda VATS ile birlikte kimyasal ve mekanik plörodezisin kombine kullanılması konusunda yol gösterici olabilir.

Anahtar Kelimeler: *Katamenial pnömotoraks, Endometriozis, Talk plöredez, VATS.*

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Catamenial pneumothorax is a feminine disease that affects young women during their reproductive age, is characterized by recurrent episodes of spontaneous pneumothorax, and has a direct relationship with menses due to ectopic endometrial tissue in the chest. Here, we report a rare case of coexisting catamenial pneumothorax and skin endometriosis with a literature review and management.

CASE

A 37-year-old female health worker presented to the emergency room (ER) with gradual onset, right-sided pleuritic chest pain and shortness of breath. She had no cough but a low-grade fever. She was hemodynamically stable, and the clinical examination was unremarkable, apart from diminished air entry into her right chest. Her chest X-ray revealed the presence of a large right pneumothorax, which was subsequently managed with a chest tube. The COVID-19 PCR test was negative.

High-resolution computerized tomography (HRCT) revealed multifocal consolidative patches on the right lung field, mainly located on the upper right lobe and likely infectious. No bullae or blebs were found (Figure 1).

Following bronchoscopy and bronchoalveolar lavage (BAL), the patient tested positive for a Pseudomonas infection. After starting a course of antibiotics, she improved clinically and radiologically. Her chest tube was removed, and she was discharged after five days of hospitalization.

A month later, she is back in the ER with the same complaints—right-sided pleuritic chest pain and periumbilical painful skin nodules. Her symptoms coincided with menses. Clinical examination and a chest X-ray confirmed the diagnosis of right-sided recurrent pneumothorax. A tube was placed in the patient's right chest, and she was admitted to the hospital to manage her condition further.

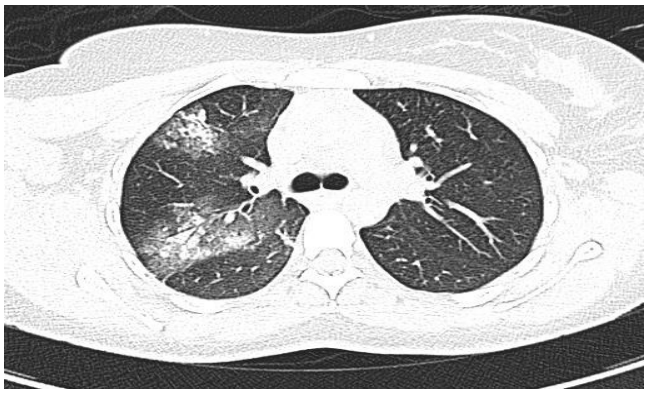

Figure 1: *Thorax computerized tomography reveals multifocal consolidative patches in the right lung field, mainly in the right upper lobe, and probably infectious. No bullae or blebs were found*

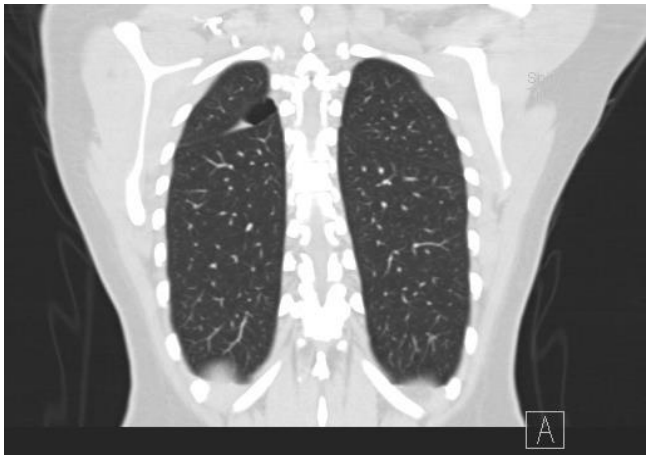

Figure 2: *Computerized tomography of the thorax reveals a lower lobe bulla*

The patient underwent right video-assisted thoracoscopic surgery (VATS) for recurrent spontaneous pneumothorax; these showed hyperemic parietal and diaphragmatic pleurae, but no bullae or blebs were found. Upper lobe apical wedge resection and pleural abrasions were performed.

In addition, a periumbilical skin nodule excisional biopsy was carried out. The histopathology report of the umbilical nodule confirmed the presence of endometrial tissue in the specimen. The examined skin tissue showed subepithelial cystic glands with periglandular stromal cells, and adequately controlled immunohistochemical staining showed that the glands and stromal cells were ER-positive. Accordingly, the patient was diagnosed with skin endometriosis.

Postoperative lung expansion was achieved, although with prolonged air leakage. The chest tube was attached to a one-way valve device (PNEUMOSTAT®), and the patient started progestogen hormonal therapy (Dienogest 2.5 mg daily) for endometriosis. She was discharged on the tenth day of post-surgery, and her chest tube was removed from the OPD after two weeks.

Three months later, a routine chest X-ray showed a third recurrence of 20% on the right pneumothorax during regular OPD follow-up. Meanwhile, the patient was asymptomatic except for mild and tolerable chest pain.

No surgical treatment was given, but the patient was told to go to the ER if her symptoms worsened. Fortunately, her symptoms improved, and the right pneumothorax was resolved entirely.

The patient remained in remission for two weeks but presented for the fourth time with severe right chest pain and progressive shortness of breath. She was hemodynamically stable with absent breath sounds on the right side; her chest X-ray showed a large, 50% right-sided pneumothorax. A chest tube was placed on the right, and preoperative chest computed tomography (CT) revealed new middle and lower lobe bullae and a right diaphragmatic

nodule (Figures 2 and 3). These were contrasting findings compared with the previous CT, attributed to the periodic nature of endometriosis.

We postulated catamenial pneumothorax (CP), and the patient underwent the right VATS exploration. There were middle and lower lobe bullae and islands of dark red soft tissue nodularity over the diaphragmatic pleura, with fenestrations and a 2 cm defect in the central tendon of the diaphragm, through which the liver was bulging (Figure 4).

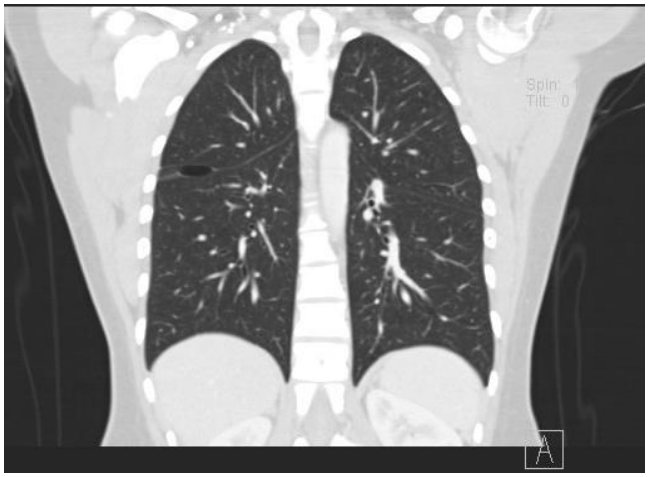

Figure 3: *Thorax computerized tomography reveals the middle lobe bulla*

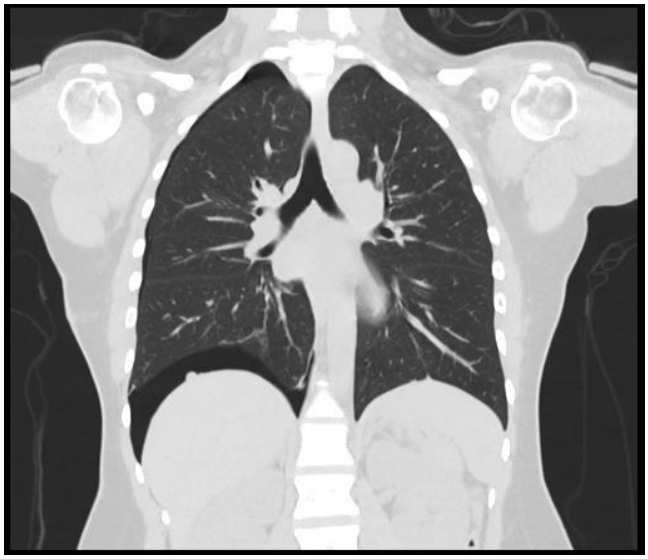

Figure 4: *Thorax computerized tomography reveals a 2 cm defect in the central tendon of the diaphragm, through which the liver bulged*

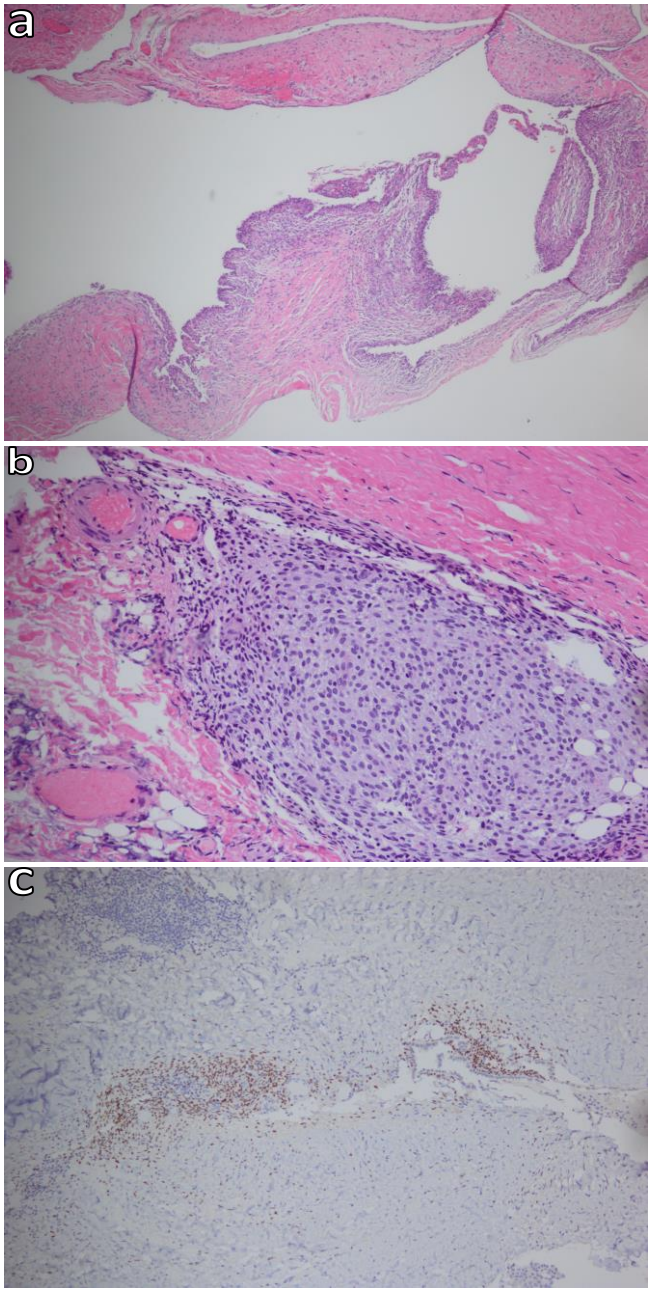

Figure 5a, b, and c: *Bulla lined by endometrial stroma (H&E X10)* ***(a)****; ER+ endometrial stroma (H&E X10)* ***(b)****; Endometrial stroma in the wall of the bulla (H&E X10)* ***(c)***

A bullectomy was performed on the middle and lower lobe using an Endo-GIA stapler. The soft tissue nests of the diaphragmatic pleura and the already existing diaphragmatic defect were excised and primarily repaired using a nonabsorbable (Prolene®) suture. Mechanical and chemical pleurodesis were used, as well as apical parietal pleurectomy and talc poudrage. The postoperative course was uneventful. On postoperative Day 3, the chest tube was removed, and the patient was discharged and put on dienogest hormonal therapy, as recommended by her gynecologist.

Histopathology (Figure 5a, b, c) showed that the right lower lobe had a bullous lesion with patchy foci of endometrial stroma, and the right middle lobe had a benign lung bleb with focal endometriosis. The diaphragm biopsy revealed a fibrous stroma with chronic inflammation and foci of the endometrial stroma. These findings confirmed the diagnosis of CP.

The patient attended regular OPD follow-up appointments, and as of the time of writing (November 2022, 24 months postoperatively), she had no recurrence of pneumothorax.

DISCUSSION

Endometriosis is characterized by the presence of endometrial-like glands and stroma outside the uterine cavity and most commonly involves the pelvis. However, endometrial tissue can be found in the abdomen, thorax, brain, and skin and is believed to affect 6–10% of reproductive-age women (1).

The chest cavity is the second most frequent site for endometriosis after the pelvis (2). Interestingly, our patient had concomitant extrapelvic, pulmonary, and skin endometriosis. To the best of our knowledge, there have only been three reported cases of concomitant umbilical endometriosis and catamenial pneumothorax in the literature (3-5), with the case presented herein being the fourth of its kind.

Catamenial pneumothorax (CP) is a spontaneous and recurrent pneumothorax occurring in women of reproductive age. Some say that CP happens simultaneously with menstruation if the person does not have lung disease, while others say it happens within 72 hours before or after menstruation (6).

CP is common in women of reproductive age, with a mean age of 34 years (ranging from 15 to 45 years). It is also commonly seen in nulliparous females (6), such as our patient.

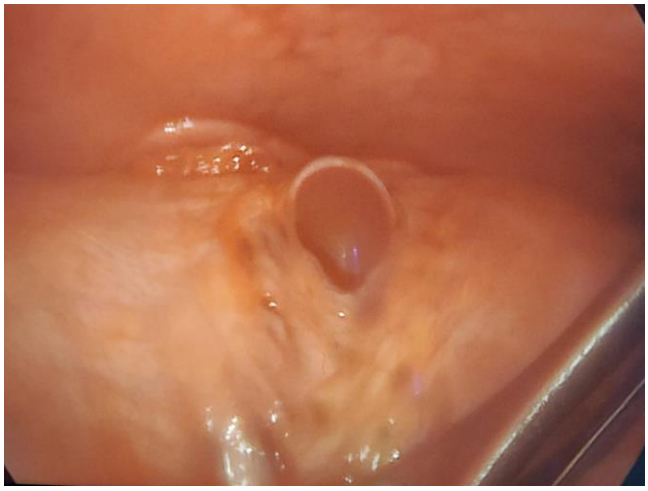

Figure 6: *Diaphragmatic fenestrations seen intraoperatively*

Although CP is not rare, it is overlooked and underdiagnosed. The prevalence of CP in all cases of pneumothorax in women of reproductive age ranges from 7.3% to 36.7%. Increased reporting of cases and outcomes has improved understanding of the condition (6, 7).

CP is the most common form of thoracic endometriosis syndrome, including catamenial hemothorax, catamenial hemoptysis, catamenial hemopneumothorax, endometriosis lung nodules, catamenial chest pain, and pneumomediastinum (8).

The pathogenesis of CP is controversial. The two main hypotheses are as follows:

(1) Autotransplantation via retrograde menstruation—Sampson's theory explains the presence of endometrial tissue in the thoracic cavity as a result of the retrograde movement of menstrual endometrium through the fallopian tube, resulting in autotransplantation of endometrial tissue and cells into the peritoneal and thoracic cavities (9).

(2) Microembolization/metastasis: the metastatic spread of endometrial tissue to the lungs via the venous or lymphatic system, resulting in tissue rupture and pneumothorax during menstruation (10).

However, we thought the retrograde menstruation theory alone could not explain why our patient had skin endometriosis.

To explain the findings of thoracic endometriosis in pneumothoraces in the intermenstrual period, Yoshioka et al. (11) suggested that non-catamenial endometriosis-related pneumothorax could be attributed to the formation of pulmonary cysts due to pulmonary endometriosis and the later rupture of those cysts occurring unrelated to the menstrual cycle. Another explanation could be the late apoptosis of visceral pleural and superficial parenchymal endometrial implants that undergo sloughing rather than acute necrosis (12). The classical clinical presentation of CP includes spontaneous pneumothorax preceding or occurring concurrently with menses. Symptoms usually begin just before or within 72 hours after the onset of menstruation (rarely within 96 hours). Chest or scapular pain is the most common symptom, noted by 90% of patients, while dyspnea develops in approximately one-third (1).

A history of recurrent spontaneous pneumothorax, primary or secondary infertility, uterine surgical procedures or scraping, pelvic endometriosis, and, rarely, catamenial hemoptysis or catamenial hemothorax may be present (8–12).

Clinical examination of the chest may reveal diminished or absent breath sounds. Pneumothoraces are typically right-sided (88–100%), small to moderate, and rarely life-threatening (13,14). Case reports of left-sided or bilateral pneumothoraces have been reported (1). Clinicians should not expect pneumothoraces or associated symp-

toms to occur monthly but rather intermittently (months to years) and close to menstruation.

Due to delayed diagnosis, which is typical in patients with thoracic endometriosis, pneumothorax is frequently recurrent. Thus, in patients with primary spontaneous pneumothorax, eliciting a history of prior episodes and timing each episode with the menstrual cycle is helpful for diagnosis.

Chest radiographs may reveal other associated features of CP, such as an effusion due to hemothorax with or without mediastinal shift depending on the size, parenchymal nodules and cavities, and a nodular appearance of the diaphragm due to abdominal viscus protrusion through diaphragmatic perforations (15,16).

Chest CT is superior to chest radiography in delineating abnormalities not visualized in chest radiography. These include pneumothorax, pneumomediastinum, or pneumoperitoneum; bullae; pleural nodules; parenchymal nodules; small cavities; scarring; ground glass infiltrates; or pleural effusions.

When thoracic endometriosis is suspected clinically, patients should undergo contrast-enhanced CT imaging of the chest, preferably when symptoms are present (i.e., during menses). This was clear in our patient because the first CT scan taken during the first episode showed no bulla, while the second CT taken during the fourth episode showed bullae and diaphragmatic nodules caused by the liver pushing through the diaphragm defect.

Magnetic resonance imaging (MRI) may detect similar findings to CT observations. In addition, MRI can detect diaphragmatic nodules with increased resolution compared to chest CT.

Clinically, the definitive diagnosis of CP is made when diaphragmatic fenestrations are seen during surgery (Figure 6), and histopathology confirms that endometrial tissues are in the thoracic cavity (12).

- Characteristic intraoperative findings may include the following: Endometrial implants may be found on the pleural, diaphragmatic, and pericardial surfaces (14,17). They can be single or multiple, vary in size (1 mm to a few cm), and are raised and red but sometimes appear purple, gray, black, or white.
- Perioperative bronchoscopy may reveal tracheobronchial endometrial implants.
- Diaphragmatic perforations can be circular or elliptical, single or multiple, and are usually located at the central tendon. They are typically small, measuring 1-3 mm in size, but can be more extensive by up to 10 mm or more in some cases. Endometrial implants are usually found at the edges of perforations.
- Although protrusions of the liver or other organs through the diaphragm are rare, this was present in our patient (Figure 6).

Endometrial glands and stroma are essential structures used to diagnose uterine endometrium and endometriosis histologically. Both stain positively with estrogen and progesterone receptors; the only difference is that endometriosis may contain blood, cysts, and fibrous tissue (14,17). However, a small sample size and the effects of inflammation or decidualization of endometrial tissue due to hormone withdrawal often lead to the observation of endometrial stroma without hormone receptor positivity or stroma with hemosiderin-laden macrophages (from the breakdown of red blood cells), i.e., histologically "probable" thoracic endometriosis.

Similar to primary spontaneous pneumothorax, CP is primarily managed with pleural drainage (tube thoracostomy, needle aspiration), followed by secondary prevention of recurrence using surgical bullectomy, pleural abrasion or chemical pleurodesis, and diaphragmatic repair. Once the diagnosis has been confirmed by histopathology, oral progestin hormone therapy is usually given for 6 to 12 months after surgery.

Recurrence is the most common complication, occurring in 8–40% of patients at a mean follow-up period of four years (8). The recurrence rates in this population are high, which can be especially concerning to patients. As there is no consensus on the optimal type of pleurodesis, we prefer to combine both mechanical pleural abrasions and chemical pleurodesis with talc poudrage to reduce recurrence to the minimum. Our patient was free of recurrence after two years of follow-up. The rarity of the disease is the main limitation to investigating the reproducibility of the management approach.

This case report details the natural history of a CP presentation requiring a high index of suspicion for diagnosis, meticulous surgical exploration of the hemithorax, mainly focusing on bullae, diaphragmatic pleural endometriosis, and fenestrations, and the application of both mechanical and chemical pleurodesis to prevent a recurrence.

CONFLICTS OF INTEREST

None declared.

AUTHOR CONTRIBUTIONS

Concept - A.B., F.A.A., S.E., A.B., A.S., A.A.; Planning and Design - A.B., F.A.A., S.E., A.B., A.S., A.A.; Supervision - A.B., F.A.A., S.E., A.B., A.S., A.A.; Funding - A.B., F.A.A., A.A.; Materials - A.B., S.E., A.S.; Data Collection and/or Processing - A.A., F.A.A., S.E., A.B., A.S.; Analysis and/or Interpretation - A.B., F.A.A., S.E., A.B., A.S., A.A.; Literature Review - A.A., A.B., F.A.A.; Writing - A.A., A.B., F.A.A.; Critical Review - A.B., F.A.A., A.B., A.A.

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