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## Catamenial pneumothorax due to solitary localization of diaphragmatic endometriosis



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### ABSTRACT

**INTRODUCTION:** Catamenial pneumothorax (CP) is a spontaneous recurrent pneumothorax occurring in women in reproductive age. The etiology of CP has been associated with thoracic endometriosis and is its most common presentation.

**PRESENTATION OF CASE:** A case of right catamenial pneumothorax in a 38 year old woman is presented in which three episodes of CP occurred within 72 h of menses in a 6 month period. The patient underwent videothoracoscopy that revealed a solitary localization of diaphragmatic endometriosis. After surgical pleurodesis and based on final pathology of resected lesion, hormonal treatment was started. The outcome was uneventful and the patients is symptom-free at 6 months.

**DISCUSSION:** Catamenial pneumothorax (CP) is a rare clinical entity characterized by lung collapse during menstruation, believed to be secondary to pleural endometriosis. Nearly all catamenial pneumothorax occur on the right side as pleural lesions are almost exclusively right-sided. Diagnostic imaging is based on high resolution computed tomography (HRCT) and, preferably, magnetic resonance imaging (MRI) since it is able to detect the blood products in the endometrial deposits. However the lack of macroscopic findings at surgery makes this condition still under-diagnosed. Based on the solitary diaphragmatic localization of endometriosis in our case we preferred to limit surgery to videothoracoscopic pleurodesis and start hormonal treatment with successful outcome.

**CONCLUSION:** Catamenial pneumothorax is the most common presentation of thoracic endometriosis syndrome and should always be suspected in women in childbearing age. Treatment option are still debated but best results are achieved by videothoracoscopic pleurodesis combined with hormonal therapy.

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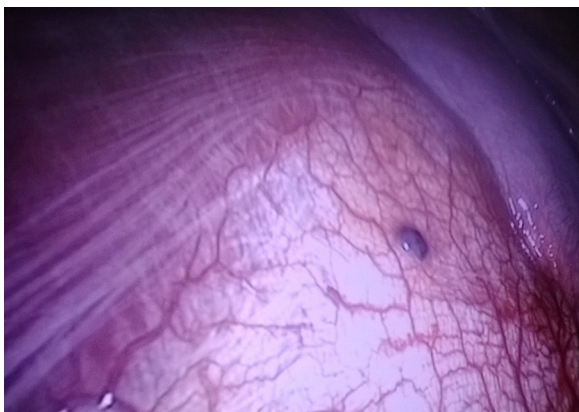
### 1. Introduction

Catamenial pneumothorax (CP) is a spontaneous recurrent pneumothorax occurring in women in reproductive age. The etiology of CP has been associated with thoracic endometriosis namely the presence of endometrial-like tissue in the thoracic cavity [1,2]. Thoracic endometriosis is considered a rare condition, usually underdiagnosed, which consists of 4 different clinical entities: catamenial pneumothorax, catamenial hemothorax, hemoptysis and pulmonary nodules [3,4]. About 60% of pulmonary endometriosis cases are associated with pelvic endometriosis [1,5]. Etiopathogenesis of thoracic endometriosis is not clear and the most accredited theory explains the retrograde implantation of endometrial tissue

by lymphatic and haematogenous dissemination or presence of diaphragmatic defects [2]. CP is the most common presentation of thoracic endometriosis syndrome (TES), with a reported incidence of 2.8–5.6% [2,6]. CP accounts for >30% of all cases of spontaneous pneumothorax in young women with a peak incidence between 30 and 35 years [3,4]. CP typically occurs within 24 h before and 72 h after the onset of menses and appears almost exclusively in the right hemithorax [1].

A combined surgical and hormonal treatment [7] is considered the best management of CP, as postoperative recurrences are common and hormonal therapy is useful to prevent them [3,6]. We present a case of recurrent catamenial pneumothorax due to a single diaphragmatic endometriosis localization which was successfully treated with combined surgical and hormonal therapy.

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**Fig. 1.** Typical purple–red spot on the right posterior diaphragmatic surface (black arrow) strongly suggestive of endometriosis localization.

## 2. Case presentation

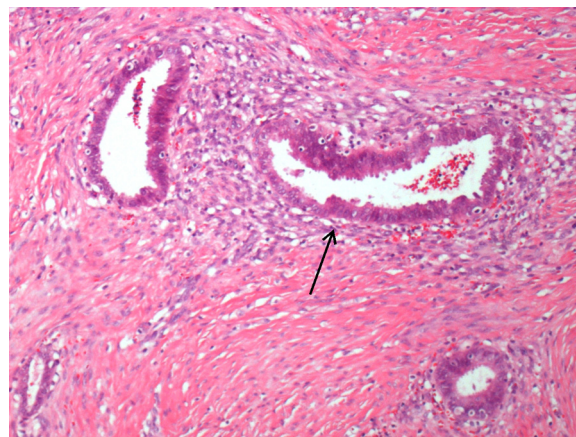
A 38-year-old otherwise healthy woman had 3 episodes of recurrent spontaneous right pneumothorax in the past six months. The 1st episode resolved spontaneously after 10 days. Two months later the patient was hospitalized because of shortness of breath and chest pain and a 2nd right pneumothorax was diagnosed which also resolved spontaneously after 3 weeks. After two more months a 3rd episode happened always on the right side, and a better analyzed clinical history revealed that all episodes were related to peri-menstruation period with the 3rd one occurring during menses. All episodes of pneumothorax were evaluated by chest roentgenogram and computed tomography (CT) which did not show any blebs or bullae. Although the patient did not have any history of pelvic or abdominal pain, her serum Ca125 level was normal (22  $\mu\text{g}/\text{mL}$ ), and further evaluation by pelvic ultrasonography and magnetic resonance imaging (MRI) excluded the presence of pelvic endometriosis, the hypothesis of catamenial pneumothorax was taken into account.

The patient underwent a videothoroscopic exploration of the right pleural cavity under general anesthesia and single-lung ventilation by a first posterolateral access. The examination of pleural cavity showed a single typical purple–red spot on the right posterior pleural diaphragmatic surface strongly suggestive of endometriosis (Fig. 1). No other anomalies nor holes on the overall diaphragmatic extension were detected. A second incision was made to perform pleurodesis by mechanical abrasion and surgical resection of diaphragmatic lesion. After control of hemostasis a 24 Ch drain was placed in pleural space and full lung re-expansion achieved. The chest tube was left in place until postoperative day 3 when the patient was discharged. Final pathology confirmed an endometriosis-related catamenial pneumothorax (ER-CP) according the most updated classification by Legras et al. [8] (Fig. 2).

Soon after surgery the patient started a medical treatment with  $17\alpha$ -etinin testosterone 800 mg daily which was shifted to dienogest 2 mg daily due to marked intolerable hypoestrogenic side effects following gynecologist's outpatient follow up. At 6 months from surgery no recurrence has been observed.

## 3. Discussion

Catamenial pneumothorax (CP) is a rare clinical entity characterized by lung collapse during menstruation (catamenial is a derivation of the Greek term *katamēnios* that refers to monthly), believed to be secondary to endometriosis within the pleural cavity and causing 1/3 of spontaneous pneumothorax in women of child-bearing age [1,8]. Although the progressive improvement in



**Fig. 2.** Histology of resected specimen: diaphragm muscle tissue with endometriosis spot in which endometrial glands and stromal cells without atypia can be recognized (black arrow, hematoxylin-eosine 10 $\times$ ).

imaging techniques and minimally invasive procedures, the lack of macroscopic findings at surgery makes this condition still under-diagnosed with a reported incidence of 3–5% [1,3,9]. The disease more frequently occurs in premenopausal women aged 30–50 who have a history of infertility, severe endometriosis and recurrent spontaneous pneumothorax within 72 h of the onset of menses [6,9].

Endometriosis predominantly affects 5–10% of women in reproductive age and may be extragenital. CP is considered a manifestation of thoracic endometriosis, rarely present in endometriosis [10]. Pleural endometriosis presents as catamenial pneumothorax, catamenial hemothorax, catamenial pneumomediastinum and chest pain. The pulmonary form presents as catamenial hemoptysis and pulmonary nodules. Both forms are part of the so called thoracic endometriosis syndrome (TES) [11]. Three theories explain the etiology of CP. The metastatic theory assumes the transdiaphragmatic passage of endometrial tissue by lympho-hematogenous dissemination or congenital fenestration. The anatomical theory calls for dissolution of cervical mucus plug during the menses with migration of cells through vagina, uterus, fallopian tubes to peritoneum and air reaching the chest by diaphragmatic congenital fenestrations. In the hormonal theory prostaglandin F<sub>2</sub> mediated pulmonary vasospasm during ovulation leads to ischemic injury and alveolar rupture [4,6]. Congenital diaphragmatic defects are more common on the right side and this can explain the higher prevalence of right-sided CP. The prevalence of diaphragmatic fenestrations varies from 23% to 88%, and diaphragmatic fenestrations can be associated with endometrial implants [12]. The diaphragmatic defect(s) can be single or multiple, usually located at the central tendon, and are described as perforations, holes, fenestrations, pores, porosities, and stomata. There can be “invisible” holes proven only by diagnostic pneumoperitoneum, tiny holes (1–3 mm), or larger defects (>10 mm) [13]. As seen in this case and in most reports, nearly in all cases catamenial pneumothorax occurs on the right side as pleural lesions are almost exclusively right-sided. This predilection is probably due to the more extensive diaphragmatic lymphatic drainage on the right side and the clockwise peritoneal circulation that sweeps endometrial implants to the right diaphragm [11,12]. Diagnostic imaging is based on high resolution computed tomography (HRCT) to detect endometrial deposits in the lung and pleura and pelvic ultrasound due to coexistence of pulmonary and pelvic endometriosis in 50–80% of cases [14,15]. Both thoracic and pelvic MRI are considered superior to CT due to the blood products in the endometrial deposits. Typical MRI pattern shows a centrally or peripherally located low-intensity area

in the hyperintense cyst on T2 weighted images (shading). Thoracic MRI lesions are hyperintense on T2-weighted spin-echo, increase in size at the time of menstruation and show pronounced uptake contrast agent as compared with the intermenstrual period [11,16]. CA125 (Cancer Antigen 125) is a tumor marker that is widely used to monitor ovarian cancer because it is overexpressed in ovarian cancer cells and secreted into the blood. Serum CA125 is used in monitoring treatment and detecting recurrence in ovarian cancer. CA-125 levels may be elevated in a variety of benign ovarian conditions, such as pelvic inflammatory disease, endometriosis, and benign neoplasms. Endometriosis has also been associated with increased levels of CA-125, and although it is not considered a specific marker, it can assist in early diagnosis of endometriosis-related pneumothorax [13,17]. Bagan et al. suggest CA-125 measurement be included in females of reproductive age presenting with primary spontaneous pneumothorax without gynaecological orientation and without clinical signs of malignant and nonmalignant conditions that injure mesothelial cells. They conclude that this biological marker may favor indication of videothoracoscopy and hormonal therapy at an early stage in the prevention of catamenial pneumothorax recurrences, and should be evaluated prospectively in a larger-scale study [18].

Moreover in a study conducted by Pickhardt and Hanson [19] to determine the prevalence, work-up, and outcomes of indeterminate adnexal masses in asymptomatic women, the only case of elevated CA-125 was associated with endometriosis. However, CA-125 levels only reflect the amount of endometrial tissue, and it is currently believed that the usefulness of such measurements is low. As a matter of fact in our case Ca-125 levels were in normal range.

Management of pulmonary endometriosis can be either medical or surgical. Medical treatment is based on the use of drugs that can induce endometrial hypotrophy. First choice treatment is represented by continuative progestinic administration. Medical treatment using 17 $\alpha$ -etinil testosterone a derivative of the synthetic steroid ethisterone has been advised since it suppresses the production of gonadotropins, thus leading to a condition of pseudomenopause and finally a remission of the disease. The patient will be hypoestrogenic due to suppression of the ovarian steroidogenesis [14]. A synthetic oral progestin having progestational and antigonadotrophic effects, but no androgenic, glucocorticoid or mineralocorticoid activity is dienogest. It is believed to induce both an antiproliferative effect on endometriotic cells and a reduction of estrogen levels due to the inhibition of ovarian function. A regimen with 2 mg a day is considered as the optimal dose for provision of efficacy and safety in endometriosis studies. In fact this dosage moderately suppresses estradiol levels, and has high oral bioavailability and a half-life suitable for once-daily administration. Compared to GnRH analogs dienogest seems to cause less hypoestrogenic side effects and little changes in bone markers and bone mineral density offering better advantages in safety and tolerability [20]. However because of endometriosis is an hormone dependent disease, when the medical treatment is stopped there is a relapse of the disease and hormonal therapy alone is associated with a recurrence rate exceeding 50% within 6 months after treatment is stopped [20]. The surgical approach has been suggested to be superior to hormonal therapy alone since it can address most of the intrathoracic pathologic features. The association of surgical treatment followed by approximately 6 months of hormone therapy may achieve up to 45 month follow up without recurrence and is presently the preferred approach for treatment and prevention [4,21]. Surgical treatment should be timed during menstruation for optimal visualization of pleurodiaphragmatic endometriosis as we did in our case. For such a purpose videothoracoscopy is a very useful tool since it assesses both the diaphragmatic surface and the pleural cavity and is able to detect typical blueberry-like spots or purple-red foci, although it has been reported that

in 8.5% of explored cases no pathologic findings were demonstrated. [1,3]. Suturing the defect, endoscopic resection of the area, covering the diaphragmatic surface with polyglactin mesh are among the commonly reported surgical procedures which, however, do not prevent the risk of CP to recur. Therefore, in patients with endometriosis-related catamenial pneumothorax, pleurodesis plays a major role, when microscopic endometrial foci, or newly implanted lesions cannot be detected [2,12]. In our case we found a single typical purple-red spot on the right posterior diaphragmatic surface but no other deposits nor fenestrations were detected. This is why we preferred to limit our surgical procedure to resection of the lesion and mechanical pleurodesis, the only successful reported procedure, and combine it with the hormonal treatment to achieve the best outcome.

#### 4. Conclusion

Catamenial pneumothorax is the most common presentation of thoracic endometriosis syndrome and should always be suspected in women with a suggestive history of recurrent pneumothorax and pelvic/abdominal endometriosis [3,15]. Although the number of reported cases with catamenial pneumothorax is limited, conservative treatment is insufficient. Videothoracoscopy provides magnification and better visualization of the pleural cavity and diaphragm. Among surgical options videothoracoscopic resection of detected lesions combined with pleurodesis is the most successful one and, associated with hormonal therapy, reduces the risk of recurrence.

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