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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.

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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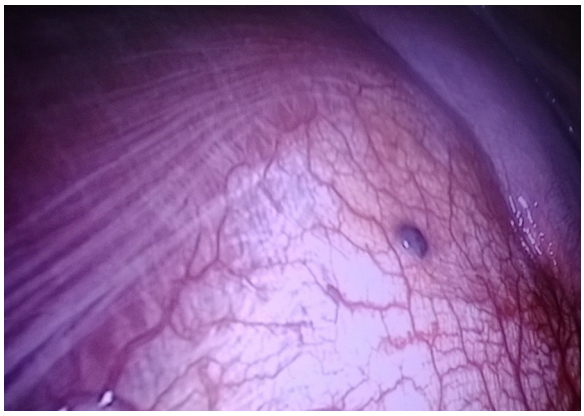

Fig. 1. Typical purple–red spot on the right posterior diaphragmatic surface (black arrow) strongly suggestive of endometriosis localization.

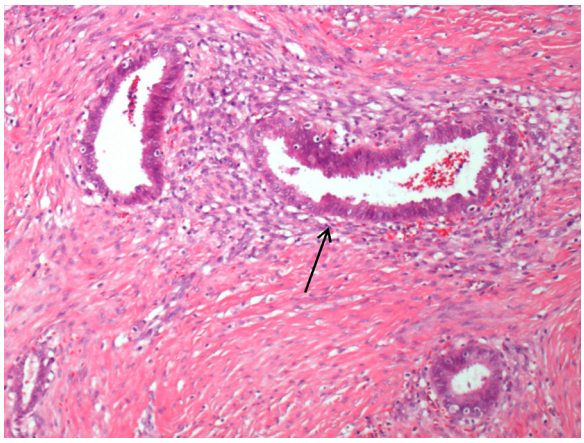

Fig. 2. Histology of resected specimen: diaphragm muscle tissue with endometriosis spot in which endometrial glands and stromal cells without atypia can be recognized (black arrow, hematoxylin-eosine 10×).

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 μg/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 videothoracoscopic 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 17α-etinil testosterone 800 mg daily which was shifted to dienogest 2 mg daily due to marked untolerable 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 imaging techniques and minimally invasive procedures, the lack of macroscopic findings at surgery makes this condition still underdiagnosed 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 F2 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