

Case Report

Catamenial Pneumothorax due to Suspected Thoracic Endometriosis Syndrome in a Woman with Adenomyosis and Cystoma Ovarii

Pneumotoraks Katamenial dengan Kecurigaan Sindrom Endometriosis Toraks pada Wanita dengan Adenomyosis dan Cystoma Ovarii

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ABSTRACT

Catamenial pneumothorax (CP) is a recurrent spontaneous pneumothorax during the menstruation period and is often associated with thoracic-pelvic endometriosis. This case study reported a 39-year-old woman who presented with recurrent shortness of breath, particularly during menstruation. The patient also experienced severe dysmenorrhea and difficulty getting pregnant. Despite a negative sputum test, a history of chest tube insertion and tuberculosis treatment were identified. Elevated levels of the CA-125 marker were observed in a previous examination. Physical examination revealed decreased fremitus sounds on the mediobasal side, chest expansion, and right lung breath sounds. The plain radiograph displayed a visceral pleural line, air-fluid level appearance, and the collapse of the right hemithorax accompanied by fibrosis. The patient underwent inpatient treatment and had a chest tube inserted. A decortication thoracotomy procedure was performed, and anatomical pathology microscopic examination of the right lung tissue revealed a non-specific chronic inflammatory process accompanied by fibrosis.

Keywords: *Catamenial pneumothorax, thoracic endometriosis syndrome*

ABSTRAK

Pneumotoraks katamenial (CP) adalah gambaran pneumotoraks spontan berulang yang berhubungan dengan waktu menstruasi dan dikaitkan dengan endometriosis toraks dan panggul. Penulis melaporkan kasus wanita berusia 39 tahun dengan keluhan utama sesak berulang yang muncul terutama ketika pasien menstruasi. Pasien juga mengeluhkan nyeri berat saat haid dan kesulitan hamil. Pasien memiliki riwayat dipasang selang dada setahun yang lalu dan menjalani pengobatan tuberculosis walau dengan tes dahak negatif. Pemeriksaan marker CA-125 pasien terdahulu menunjukkan peningkatan ekstrem. Pemeriksaan fisik ditemukan penurunan suara fremitus sisi mediobasal, penurunan pengembangan dada dan suara nafas paru kanan. Pemeriksaan foto polos menunjukkan gambaran visceral pleural line dan air-fluid level serta kolaps pada hemitoraks kanan disertai fibrosis. Pasien dilakukan tatalaksana rawat inap dan pemasangan selang dada. Pasien telah menjalani prosedur torakotomi dekortikasi dan pemeriksaan patologi anatomi pada jaringan paru kanan menunjukkan gambaran mikroskopik proses radang kronis non spesifik disertai fibrosis.

Kata Kunci: Pneumotoraks katamenial, sindrom endometriosis toraks

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INTRODUCTION

Catamenial pneumothorax is a condition characterized by spontaneous and recurring lung collapse occurring between 24 hours before and 72 hours after menstruation begins (1). Catamenial pneumothorax is the most common form of thoracic endometriosis syndrome (TES) (1), a group of symptoms caused by endometrial tissue in or around the lungs (2). Hemoptysis, pulmonary nodules, catamenial pneumothorax, and catamenial hemothorax are the four distinctive clinical entities composing TES (3). Generally perceived as an infrequent event, catamenial pneumothorax has a low incidence, as evidenced by large epidemiological studies reporting rates not exceeding 3-6% (4). Among all cases of pneumothorax in women of childbearing age, the prevalence of catamenial pneumothorax ranges from 7.3% to 36.7% (5). Until now, thoracic endometriosis syndrome is still a rarely encountered disease and is still often underdiagnosed or misdiagnosed. This case report explored a case of catamenial pneumothorax in a female patient of childbearing age, presenting with suspected thoracic endometriosis syndrome alongside additional gynecological findings, including adenomyosis and cystoma ovarii. Intending to avert potential complications, the report sought to emphasize the significance of early diagnosis in both thoracic endometriosis syndrome and catamenial pneumothorax, aiming to increase awareness about thoracic endometriosis syndrome and catamenial pneumothorax.

CASE REPORT

A 39-year-old woman presented to the emergency room of Dr. Saiful Anwar General Hospital with a chief complaint of dyspnea for three hours before admission. The onset of shortness of breath was first noted a year ago, occurring intermittently, and worsened notably one week before seeking medical attention. The dyspnea was associated with a feeling of pressure, particularly during the patient's menstrual cycle, and remained unaffected by changes in position. The patient had experienced similar tightness in the past, leading to the insertion of a chest tube a year prior. Although the patient had been recommended tuberculosis treatment, the sputum test did not reveal the presence of *Mycobacterium tuberculosis* bacteria.

A year before the current admission, the patient had a history of severe abdominal pain during menstruation, which could only be relieved by taking anti-pain medication. A family history of malignancy was obtained during the medical history assessment. A month before the intense dyspnea occurred, the patient sought consultation with a gynecologist due to persistent menstrual pain and difficulty getting pregnant. The initial ultrasound revealed a solid cystic septation lesion with an irregular shape, unclear boundaries, and irregular edges measuring approximately 7.6 x 4.2 cm. The conclusion was a complicated ovarian cyst on the left side, assessed with a low risk of malignancy (O-RAD Score 3). A CA-125 tumor marker examination yielded a result of 619.5 (normal value <35 U/ml). The patient was advised to follow up at the gynecology clinic at Dr. Saiful Anwar General Hospital and returned for evaluation two weeks later. The second ultrasound result which was performed at the gynecology clinic can be seen in Figure 1.

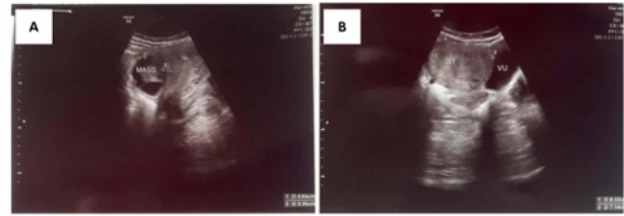


Figure 1. Abdominal ultrasound results

The result revealed an enlarged uterus measuring 8.23 x 7.14 cm with homogeneous density. Additionally, a pathological lesion measuring 4.65 x 3.36 cm was identified in the right adnexa of the uterus, appearing attached to the uterus. The diagnosis included ovarian cystoma and uterine myoma.

The physical examination conducted in the emergency room revealed an elevated heart rate and respiratory rate attributed to dyspnea. The lung examination identified reduced vocal fremitus on the medial and basal sides of the patient's right lung, restricted expansion of the right chest, and diminished breath sounds upon auscultation of the right lung. The plain chest radiograph result can be seen in Figure 2.

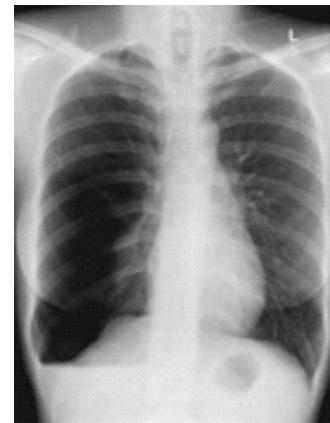


Figure 2. Plain chest radiograph result

The chest radiograph displayed a distinct clear space with a visceral pleural line, indicating the collapse of the right lung within the right hemithorax, accompanied by partial fibrosis. An air-fluid level was evident at the right hemidiaphragm, exerting pressure on the inferior hemidiaphragm and widening the surrounding intercostal spaces.

The patient was diagnosed with recurrent secondary spontaneous hydropneumothorax dextra along with catamenial pneumothorax, ovarian cyst, and adenomyosis. The patient received inpatient care and management for pneumothorax. Pharmacological medication were given to our patient including the administration of normal saline 0.9% at a rate of 1000cc/24 hours, IV Ranitidine twice daily at 50mg each, PO Paracetamol three times daily at 500mg each, and PO codeine three times daily at 10mg each. Upon referral, the patient had a chest tube installed from the previous

hospital. The chest tube remained in place and was connected to an active water-seal drainage (WSD) suction set at 10cmH₂O. On the fifth day of treatment, a cardiothoracic surgeon performed a decortication thoracotomy. Concurrently, the right lung tissue underwent anatomical pathology analysis. The macroscopic sample, measuring 5.5 x 5 x 3cm, revealed an alveolar pattern. The microscopic image result can be seen in Figures 3A, 3B, and 3C.

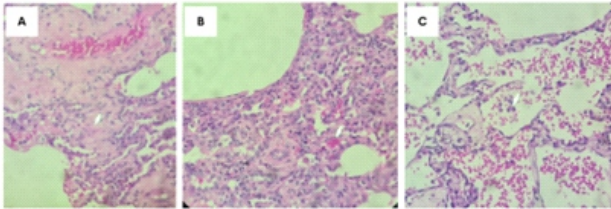


Figure 3. Histopathological results from the right lung specimen

In Figure 3A, the arrow exhibits a non-specific chronic inflammatory process accompanied by extensive fibrosis. There is also an increased appearance of the interstitium cell, surrounded by histiocytes, lymphocytes, and inflammatory cells within the same figure. In Figure 3B, the arrow shows an intraalveolar hemorrhage found without the presence of endothelial cells nearby, indicating that the hemorrhage occurred abnormally and didn't come from the surrounding small vessels. In Figure 3C, the arrow points out one of the active macrophages. From the same picture, plenty of macrophages appearance (more than 10 macrophages) could suggest the presence of necrotic debris.

After a month, the patient underwent a laparoscopic biopsy and adhesiolysis. The macroscopic sample showed two pieces of grayish-white, solid, springy tissue from the uterine wall, measuring 1.5 x 1 x 1cm and 1.3 x 1 x 0.8cm. The microscopic image displayed pieces of smooth muscle tissue and revealed areas of necrosis, bleeding, and blood vessel dilatation. Between the myometrial stroma were found the endometrial stroma and the remaining endometrial glands. The biopsy specimen confirmed the diagnosis of adenomyosis with no indications of cancer found in this preparation. The patient did not have any problems or recurring complaints for five months following the decortication thoracotomy.

DISCUSSION

Catamenial pneumothorax (CP) is defined as spontaneous recurrent pneumothorax concomitant with menstruation and typically affects women of reproductive age (6). Among women of reproductive age, the prevalence of catamenial pneumothorax varies from 7.3% to 36.7% of all pneumothorax cases (5). Catamenial pneumothorax is also known to be closely associated with the presence of thoracic endometriosis syndrome (TES), a rare and severe type of extrapelvic endometriosis (7). Thoracic endometriosis syndrome manifest itself through four entities, with catamenial pneumothorax (CP) as the most common disease (73%), and the other three are catamenial hemothorax (14%), catamenial hemoptysis

(7%), and endometriotic pulmonary nodules (6%) (1, 7). Several theories have been proposed to understand the mechanism of TES, including coelomic metaplasia, lymphatic or hematogenous spread, and retrograde menstruation with subsequent transdiaphragmatic migration of endometrial cells (5). One of the theories, the coelomic metaplasia, proposes that endometriosis may develop from the metaplasia of mesothelial cells that line the pleura and peritoneal surfaces into endometrial-like glands and stroma. Furthermore, these cells may undergo transformation in response to various physiological stimuli, including estrogen (2). Compared to estradiol levels in the menstrual blood and peripheral blood of normal healthy women, estradiol levels are known to increase in the menstrual blood of women with endometriosis and adenomyosis. With this increase in estradiol levels, the conditions of endometriosis and adenomyosis are associated with excessive tissue injury and repair mechanisms. This phenomenon is further exacerbated in an estrogen-sensitive environment normally controlled by ovaries (8).

The diagnosis of CP was divided into clinical, surgical and pathological diagnoses. From clinical manifestations, CP was diagnosed in this reproductive-age patient based on menstrual-related chest pain and dyspnea, generally a day before to 72 hours after menses (5). During their menstrual cycle, female patients with CP report experiencing shortness of breath and right-sided pleuritic chest pain (1). Although CP can occur on the left side and bilaterally, the majority of CP cases (93-95%) show signs and symptoms of involvement on the right side of the lung (1,9). Thoracic endometriosis syndrome, represented by catamenial pneumothorax, is strongly associated with severe pelvic endometriosis and high rates of infertility (10). This conditions manifest as the patient's complaint of constant menstrual pain and difficulties conceiving. Endometriosis is also coexists with adenomyosis, characterized by the presence of endometrial glands outside the uterine endometrium (11). From physical examination, Catamenial pneumothorax may show reduced or diminish breath sounds on the affected lung side, indicating the presence of pleural effusion and/or pneumothorax (4,12).

Although some serum laboratory tests may lack sensitivity and specificity as diagnostic tools, several tumor markers like CA-125 and CA 19-9, show elevated results in TES patients (13,14). CA-125 concentrations were found significantly higher in women with TES than among disease-free women (3). In TES, cytological analysis of pleural fluid seldom confirms the diagnosis. Nevertheless, some physicians continue to perform pleural fluid examinations when inserting the chest tube to check the existence of endometrial cells (4). Imaging is necessary to be performed for further evaluation (13). In one study involving 131 women seeking treatment for symptomatic fibroids, 86% were found to have concurrent endometriosis, detected through supportive examinations like ultrasonography and pelvic scans (3). Basic imaging, such as chest radiography, can be performed without specific diagnostic criteria. However, it can help determine the presence, side, and location of a pneumothorax, as well as basal air-fluid levels and mediastinal shift (4). Computed Tomography (CT) and Magnetic Resonance Imaging (MRI) are the recommended imaging tools to assist the diagnostic process of thoracic endometriosis. The use of contrast can be considered to prove diaphragmatic lesions,

especially those located in the upper posterior part of the diaphragm (15). CT results are also useful to reveal additional features, such as pneumothorax, compromised diaphragm integrity with or without lung herniation, and nodules (16). Another assessment method using video-assisted thoracoscopic surgery can be conducted to enable direct visualization of the lung and diaphragmatic surfaces, assessing perforations on the diaphragmatic surface, brown and purple endometrial deposits, and even larger masses (11). Considering that our patient also has cystoma ovarii, imaging techniques such as CT, MRI, and transvaginal ultrasonography particularly three-dimensional sonography are useful in determining whether the ovarian tumor is malignant, as tumor markers like CA-125 alone cannot be used to diagnose ovarian cancer (17).

Ectopic endometriosis is not always feasible to be confirmed histologically (18). Positive histopathologic confirmation of endometriosis ranges from 39% to 87.5% and could be attributed to the site of the biopsy (5,19). It is known that specimen from the base of the lung would yield more endometriosis than that from the apex (20). On pathological aspect, studies reported that the 'triad' classic features of endometriosis, consisting of endometrial glands, stroma, and hemosiderin-laden macrophages, were recognized in 44% of the thoracic endometriosis patients, and only stroma was present in the rest 56% cases (21,22). If the procedure is possible, catamenial pneumothorax is associated with the following findings: single or multiple fenestrations in the tendinous part of the diaphragm, as well as red/brown spots or nodules located in the diaphragm or visceral pleura. Histopathological analysis of these nodules shows endometrial glandular and stromal cells, and macrophages filled with hemosiderin, while immunohistochemical testing can show the presence of estrogen and progesterone receptors (23). Previous research stated that the diagnosis of thoracic endometriosis requires the discovery of both stromal cells and endometriotic glands (18). The presence of only stroma, pulmonary parenchymal hemorrhages, and/or hemosiderin-laden macrophages was considered probable or suggestive (24-26). An additional immunohistochemical examination of CD10 receptor, estrogen receptor (ER), and progesterone receptor (PR) are recommended to confirm the presence of endometrioid glands and stroma (24). In the case of our patient, a histopathological examination of the lung tissue showed results indicating endometriosis with the discovery of stromal cells, although no endometrial gland cells were found. The histopathology result of our patient

also revealed a non-specific chronic inflammatory process accompanied by fibrosis. The appearance of active macrophages we found on our patient's result suggested the possible existence of widespread intraalveolar hemorrhage and necrotic debris. Macrophages are responsible for debris clearance, which helps terminate inflammation and aids in lung tissue repair (27).

For the treatment option, CP with mild symptoms is managed with simple rest and thoracentesis or chest tube for symptomatic relief (5). The surgical standard treatment for CP remains controversial. Although some studies have reported that pleural abrasion alone is necessary for CP management, others have reported that apical blebs and parenchymal implants should be resected, and diaphragmatic fenestration should be closed (9). Surgery is usually performed when conservative treatment fails or in women with multiple recurrences (5) with video-assisted thoracoscopic surgery (VATS) served as the preferred procedure. Additionally, most recurrences following a prior procedure cases are indicated to undergo thoracotomy (23). Pleurodesis is the most common intervention, the majority of which is performed mechanically (abrasion or pleurectomy), proving more successful than chemical pleurodesis (28). Approximately two-thirds of CP patients receive a combination of hormonal and surgical treatments, with surgery being the most common form of treatment. For all patients with confirmed catamenial and/or endometriosis-related pneumothorax, an immediate postoperative course of oral contraceptives or gonadotrophin-releasing hormone (GnRH) analog is recommended for 6–12 months (4). Nonetheless, the decision on which hormonal treatment to use is influenced by the costs, side effects, duration of treatment, and the patient's preference especially to become pregnant (9). In our case, the administration of GnRH analog is postponed due to our patient's desire to continue trying to conceive.

In conclusion, catamenial pneumothorax is a condition related to the time of occurrence of menstruation, closely linked to TES episodes, and may manifest with additional TES symptoms. Multidisciplinary specialists in pulmonology, thoracic surgery, and gynecology must collaborate to manage TES to avoid misdiagnosis, achieve the best therapeutic outcomes, and prevent complications. Additionally, chest-abdominal CT or MRI imaging is strongly recommended for its detailed diagnostic capabilities in assessing TES. Further immunohistochemical examination is essential to confirm the diagnosis when histopathology results remain inconclusive.

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