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Catamenial pneumothorax, clinical manifestations — a multidisciplinary challenge

Odma opłucnowa spowodowana endometriozą, objawy kliniczne
— problem interdyscyplinarny

Abstract

Introduction: Pleural endometriosis is a rare condition. Spontaneous, recurring pneumothorax occurring during menstruation, referred to as catamenial pneumothorax, is associated with pleural endometriosis. A multidisciplinary approach is needed for a successful result.

Material and methods: During the last five years (2005–2010), we have treated six patients with menstruation related pneumothorax at Oslo University Hospital. The surgical treatment was performed by the thoracic surgery department but the medical follow-up was carried out by the gynecological and pulmonary medicine departments.

Results: We report three of the patients treated. All three patients were premenopausal, aged 19–36, and had recurring, menstruation related, spontaneous pneumothorax, predominantly on the right side. The condition was treated by various surgical approaches, including chest tube drainage, video assisted thoracic surgery, chemical pleurodeses and thoracotomy.

Conclusion: Spontaneous, recurring pneumothorax in women with no previous history of endometriosis can be the first manifestation of pleural endometriosis. The disorder requires surgical intervention, but early diagnosis and postoperative hormonal therapy are just as important for a successful outcome.

Key words: pneumothorax, endometriosis, catamenial

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Streszczenie

Wstęp: Endometrioza opłucnej występuje bardzo rzadko. Samoistna, nawracająca odma opłucnowa występująca podczas krwawienia miesięczkowego, znana i omawiana jako odma katamenialna, związana jest z endometriozą jamy opłucnej. Do dobrego zdiagnozowania i pomyślnego leczenia, zwykle potrzebna jest współpraca i wysiłki lekarzy różnych specjalności.

Materiał i metody: W okresie ostatnich pięciu lat (2005–2010), leczylismy sześć pacjentek z odmą opłucnową spowodowaną krwawieniem z ognisk endometriozy umiejscowionej w jamie opłucnej. Leczenie chirurgiczne wykonano w klinice chirurgii klatki piersiowej Uniwersyteckiego Szpitala w Oslo, zaś postępowanie i leczenie pooperacyjne przeprowadzono w klinice pulmonologicznej i klinice ginekologicznej tego samego szpitala.

Wyniki: Wszystkie omawiane pacjentki były w wieku przedmenopauzalnym (19 do 36 lat) z nawracającą, samoistną odmą jamy opłucnej związaną z krwawieniem miesięczkowym. W leczeniu zastosowano różne metody postępowania chirurgicznego, np: drenaż klatki piersiowej (niekiedy z pleurodezą chemiczną), torakoskopię lub torakotomię.

Wnioski: Samoistna, nawracająca odma jamy opłucnej u kobiet bez poprzednio rozpoznanej endometriozy, może być pierwszym objawem endometriozy opłucnej. Zaburzenie to wymaga najczęściej leczenia chirurgicznego, ale wczesne, prawidłowe rozpoznanie i pooperacyjne leczenie hormonalne są również bardzo ważne dla pomyślnego wyleczenia.

Słowa kluczowe: odma opłucnowa, endometrioza, leczenie chirurgiczne

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Introduction

Endometriosis is a relatively common gynecological disorder that affects women in fertile age and is characterized by the manifestation of ectopic endometrial foci. These foci are typically located in the peritoneal cavity, most often in the pelvis, but endometriosis can also be found extraperitoneally [1].

Endometriosis in the lung or pleura is a rare type of ectopic endometriosis, pleural endometriosis being the more common of the two [2]. The relationship between spontaneous pneumothorax and menstruation was first described by Maurer and co-workers in 1958 [3]. The disorder is referred to as catamenial pneumothorax, recurring pneumothorax in relation to menstruation due to ectopic endometrial tissue [4]. Approximately 90% of all catamenial pneumothorax manifest on the right side, contrary to pneumothorax caused by congenital defects presenting on the left side [2]. Although being rare, catamenial pneumothorax is responsible for a higher number of spontaneous pneumothorax in fertile women than previously assumed [5].

The treatment of catamenial pneumothorax depends on a multidisciplinary approach as the presenting symptoms are diverse. Cooperation between the pulmonologist, gynecologist and thoracic surgeon is crucial. Once the disorder is suspected, laparoscopic approach for lesions below the diaphragm should be applied, while lesions above the diaphragm should be addressed by video assisted thoracic surgery (VATS) or thoracotomy. Hormonal therapy is necessary in most patients to prevent recurrence of catamenial pneumothorax.

Women with recurring pneumothorax are treated by thoracic surgeons, admission to hospital is necessary and various surgical approaches are administered. We have treated a number of fertile women with spontaneous, recurring pneumothorax at our hospital and present our experiences and therapeutic challenges.

Material and methods

During the last five years, we have treated six patients with catamenial pneumothorax at Oslo University Hospital, Ullevål. It is likely that a larger number of patients have received treatment for this disorder, but no systematic registration has been in place to identify this group of patients. The patients were admitted to the thoracic surgery department, and the gynecology and pulmonary departments were consulted for a multidisciplinary approach.

Results

All six patients were premenopausal and their age varied from 19 to 36 years at the time of diagnosis. We report three cases which illustrate the diversity of catamenial pneumothorax.

Patient no. 1 was a 35-year old, mother of one, with hypothyroidism. There was no previous history of endometriosis before she sustained her first spontaneous pneumothorax on the right side. She was treated with a chest tube and discharged after three days. During eight months, she sustained two more pneumothorax on her right side, the first treated with a chest tube and the second with VATS and pleurectomy as no bulla could be verified. One month after the operation, she experienced another pneumothorax on the right side. She was once again treated with VATS, but as there was no apparent pathology, the procedure was converted to a thoracotomy and a bulla on the dorsal part of the upper lung lobe was resected. One month after the second operation, the pneumothorax recurred. The patient was treated with a chest tube and chemical pleurodeses. Endometriosis has been never proven histologically in this patient. The patient is now on oral contraceptives and administration of a gonadotrophin-releasing hormone (GnRH) analog is being considered.

Patient no. 2 experienced her first spontaneous pneumothorax at the age of 19. She had multiple recurrences and each time in relation to menstruation. Both sides have been affected, although the right — most frequently. The patient was treated several times with a chest tube and has undergone two VATS procedures, one of which revealed brown spots on the visceral pleura and a biopsy confirmed tissue consistent with endometriosis. Following surgery, the patient underwent hormonal treatment with oral contraceptives and later progesterone. At the age of 25, six years after her pneumothorax debut, the patient underwent diagnostic laparoscopy due to infertility. Peritoneal endometriosis was revealed. The patient will be treated with in vitro fertilization (IVF), alternatively establishing amenorrhea when reproduction is not in focus.

Patient no. 3 was 32 years old when during a very short period of time she was diagnosed with hemothorax and deep endometriosis of the pelvis. She had no previous medical history, had never been pregnant but suffered from dysmenorrhea since menarche. The patient's advanced form of pelvic endometriosis was first treated with adhesiolysis and resection of endometriosis. Laparoscopic rectum resection was later performed due to

rectal involvement. The patient experienced several pneumothorax episodes on the right side that were treated with a chest tube and finally with a VATS pleurectomy. No bullae, nodules or ectopic endometrial foci were found intraoperatively. Two years after the onset of her debut symptoms, the patient underwent IVF and became pregnant on the first attempt. Due to placenta previa and hemorrhage, she delivered a healthy baby boy in the 34th week by cesarean section. Following the pregnancy, she underwent hormonal therapy with a GnRH analog and established amenorrhea. Estrogens were added to prevent adverse effects of the GnRH analog.

None of the patients suffered from a recurrent pneumothorax after their final surgical procedures, the shortest observation time being eight months and the longest 32 months.

Discussion

Catamenial pneumothorax is a seldom disorder. We have described three cases from Oslo University Hospital, Ullevål, that illustrate the diversity of this form of pneumothorax. Most of the materials published are small, the most recent being eleven patients from Japan [6] and six patients from Finland [7]. Retrospective studies have suggested that 3–6% of all spontaneous pneumothorax in women are related to endometriosis [8]. In a prospective study, Alifano and co-workers indicate that the percentage could be as high as 25–33% [5].

The diagnosis of pleural endometriosis is not always verified by histopathologic analysis, even though the patients present with a pathognomonic history, illustrated by patient nr.1. Kumakiri and co-workers did not identify endometriosis of visceral pleura lesions in seven out of eleven patients [6]. Rahman and co-workers also present a very representative patient and review, but again without histopathological verification [9].

It is not fully understood how endometrial cells enter the thoracic cavity and how the catamenial pneumothorax occurs. Three theories have been suggested, retrograde menstruation with migration of endometrial cells through defects in the diaphragm, migration of endometrial cells through blood or lymphoid vessels and metaplastic transformation of the coelom membrane both in the peritoneal- and thoracic cavity [10–12]. Regarding how pleural endometriosis causes pneumothorax, the effect of prostaglandins on alveolar tissue and weakening of the visceral pleura by endometrial tissue have been suggested [7].

The incidence of catamenial pneumothorax associated with intraperitoneal endometriosis is very low. The highest incidence of endometriosis in the pelvis is reported between the age of 24 and 29, while the highest incidence of catamenial pneumothorax can be found five years later [11]. Except for patient no. 2, all our six patients presented with catamenial pneumothorax in their fourth decade of life. Other reports suggest a higher mean age for the onset of catamenial pneumothorax, approximately 40 years [6, 10]. Complicated pelvic endometriosis is reported in 25–60% of the patients with catamenial pneumothorax [11, 13–15] and one of the six patients reported had deep pelvic endometriosis. Based upon these findings, Nezhat and co-workers have proposed a systematic procedure including both VATS and laparoscopy in patients with catamenial pneumothorax, to establish the relation between pneumothorax and endometriosis in the peritoneal cavity and pelvis [16].

Recurrence of pneumothorax is frequent in catamenial pneumothorax despite surgical intervention. Alifano and co-workers reported a 32% recurrence in patients with catamenial pneumothorax as opposed to only 5% recurrence in patients with non endometriosis related pneumothorax.

Conclusion

In this complex group of patients, it is crucial to apply a multidisciplinary approach involving pulmonologists, gynecologists and thoracic surgeons. During VATS, all suspicious areas should be resected and postoperative hormonal treatment instituted promptly. Adjuvant chemical pleurodeses must be considered. The diagnosis is not always verified by histopathologic analysis. The goal of the hormonal treatment must be amenorrhea, using GnRH analog, progesterone therapy or oral contraceptives. The treatment should be continued for at least six months, or longer if recurrence of pneumothorax is observed [17]. The patients need a close gynecological follow-up until menopause.

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