Thoracic endometriosis syndrome – case study

Endometriografia opłucnej – studium przypadku

TES (-thoracic endometriosis syndrome) is a rare disease characterized by the presence of functioning endometrial tissue in pleura, lung parenchyma, airways and/or diaphragm. TES may present with catamenial pneumothorax which occur within 72 hours before or after menstruation [1,2]. The authors present a case of a 51 year old female who was presenting cyclical recurrent episodes of chest pain, shortness of breath and right side pneumothorax. Histopathological examination after resection of the top of the upper lobe of the right lung revealed clusters of endometrial cells staining positive for estrogen receptor ES (+).

Introduction
Endometriosis affects between 5% and 15% of women in reproductive age, whereas no more than 12% of patients have ectopic tissue outside lower pelvis [1,2]. Transposed fragments of active endometrial tissue may localize in distant areas of the peritoneal cavity, in the umbilical region, lymph nodes, lungs and even bones and heart. Endometriosis is a frequent cause of infertility, painful periods and pelvic pain. The first case of pleural endometriosis was described by Schwarz in 1938 [3]. TES is a constellation of four clinical presentations, such as catamenial pneumothorax, hemoptysis, hemorrhax and pulmonary nodules [4]. TES may present with catamenial pneumothorax which occurs within 72 hours before or after menstruation [5,1]. The first case of catamenial pneumothorax was described in 1958 by Mauer et al. [6], and the term “catamenial pneumothorax” was first introduced into the literature in 1972 by Lillington et al. [7].

Case report
51-year-old patient presented to her GP for routine health check. She complained of decreased exercisetolerance and effort dyspnea. Physical examination did not show any abnormalities. She had a history numerous admissions to pulmonology wards and menstrual problems (copious and painful menstruations). She had regular periods and was not treated for any chronic condition.

At the age of 43 she had an emergency admission to pulmonology ward because of weakness, dyspnea and right-sided chest pain caused by spontaneous pneumothorax, diagnosed 5 days earlier on routine plain chest X-ray.

On admission, she presented with decreased oxygen saturation-SpO2, 91% and tachypnea-22 per minute. Laboratory tests and ECG did not show any abnormalities. A series of chest x-ray films in antero-lateral projection revealed 5 mm air deposit in the right apical region. The 3 consecutive imaging performed every 2 days showed regression of the pneumothorax. Computerized tomography (CT) of chest without contrast performed on the day 7 did not show presence of any air in the right pleural cavity, no emphysematous bullae and no focal lesions in the lung parenchyma. Pleural recesses and diaphragm were normal. Ultrasound of abdominal cavity and thyroid were normal.

The treatment included oxygen therapy (10 L/min) and estazolam. Patient was discharged home in good general condition and referred to outpatient chest clinic for follow-up.

Next month, the patient was re-admitted as an emergency to the thoracic surgery ward because of dyspnea, weakness and pain in the region of the right apex. Standard laboratory tests and ECG results were normal. X-ray examination revealed recurrent spontaneous right-sided pneumothorax (Fig. 1). Right pleural cavity.
cause of shortness of breath, weakness, and chest pain within the right apical region. Standard blood test and urine tests and ECG did not show any abnormalities. Chest x-ray showed recurrent spontaneous pneumothorax. It was treated conservatively with benzodiazepines. Control chest X-ray showed spontaneous re-absorption. The patient was discharged in good general condition and referred to outpatient chest clinic for follow up.

Because of abundant and painful menstruation and suspicion of endometriosis on abdominal ultrasound examination, the patient was admitted to gynecology ward. Exploratory laparoscopy confirmed foci of endometriosis in the left ovary. During the procedure, a 15 mm biopsy sample of the left ovary was obtained and fallopian tubes were ligated subject to patient’s consent. The histopathological diagnosis was: *Endometriosis externa*.

The patient was discharged in good general condition, referred for follow up in the outpatient gynae clinic and prescribed triptorelin as a continuation of treatment of endometriosis. Control chest X-ray performed during the treatment did not show any abnormalities.

The treatment with triptorelin was discontinued after 1 month because of adverse effects—heatwaves, vaginal dryness, head and breast pain, nausea and weakness.

After interruption of the treatment, respiratory tract symptoms with recurrent pneumothorax recurred. A few months later, the patient was admitted to the thoracic surgery ward, where, following the video-thoracoscopy and pleuradesis, the patient was qualified for apical resection of the right lung. Three samples of lung tissue and pleura showed pulmonary and nodal pneumoniosis, pleural thickening and fibrosis, focal emphysematous changes and congestion, subpleural fibrosis with eosinophilic infiltrate (Fig. 2).

Histopathological slides and paraffin blocks were reviewed in Histopathological

Figure 1
_X-ray photo of right-sided pneumothorax, about 2 cm._
_Zdjęcie Rtg z prawostronną odmą opłucnową około 2 cm._

Figure 2
_Areas of congestion of the lungs, inflammatory foci of eosinophils (→). Features of emphysema (100 x zoom)._  
_Miejsca przekrwienia płuc, naciek eozynofilowy (→). Cechy rozdęty płuc (powiększenie 100 x)._  

Figure 3
_Staining for nuclear estrogen receptor. ER(+) (→) (200 x zoom)._  
_Pozytywne barwienie dla jądrowego receptora estrogenowego ER(+) (→) (powiększenie 200 x)._
Diagnosis Laboratory of the Department of Pathology of the Medical University of Silesia. Examinations of routine serial H&E stained sections and immunohistochemical studies for the presence of estrogen receptor (ER) were performed. Apart from the changes reported in the original examination, also clusters of cells with positive ER reaction were found below the pleura (Fig. 3). A diagnosis of pulmonary endometriosis (endometriosis pulmonum) was given.

At present, the patient is under observation of chest physician, GP and gynecologist, who implemented combined contraception. The patient has no respiratory tract symptoms as well any abnormalities on imaging.

Discussion

Distant complications of cesarean section include increased risk of endometriosis. Therefore, constantly growing number of cesarean sections (an increase from 26% to 36.5% in years 2003-2009 in the USA [9]) will be the main factor of increasing incidence of pelvic endometriosis as well as distant endometriosis, e.g. pleural and chest.

Population studies by Haga at al. enabled development of point system risk assessment of pleural endometriosis. Predictive factors include: right sided pneumothorax, pelvic endometriosis, age above 31 years, non-smoking (sensitivity 93.5% and specificity 89.4% with the result of 12 points or more) [1]. In our case, the patient had 18/18 Hagascore points, which puts her into a high risk group.

In the majority of cases, pleural endometriosis is accompanied by chest discomfort and pain, which sometimes may be neglected by patients [9]. Our patient confirmed the early onset of respiratory tract symptoms but of lower intensity. Numerous hospital admissions to pulmonology and thoracic surgery wards and diagnoses emphasize repetitiveness and cyclic nature. As discovered retrospective ly, dyspnea preceded admissions, which supports the diagnosis of catamenial pneumothorax.

Numerous epidemiological studies indicate cause-effect relationship between cesarean section and the incidence of endometriosis [3]. Therefore, it is not surprising that ectopic foci of endometrial tissue were found. Fallopian tube ligation is believed to be preventive of further dissemination of endometrial tissue via fallopian tube, and then through the fenestrations in the diaphragm into the chest [10].

Patient’s induction into artificial menopause was correlated with regression of respiratory tract symptoms and lack of pneumothorax recurrence. This observation is an indirect indication of the presence of pulmonary endometriosis, which should be taken into account in the differential diagnosis. One should emphasize that restoration of natural menstrual cycle resulted in recurrence of dyspnea and pneumothorax. It was caused by activation of ectopic endometrium within the lungs.

As in a case presented by Zeena Makhlja et al., video-thoracoscopy (gold standard in diagnostics and treatment of pleural endometriosis) was performed with lobectomy and pleuradecision (creation of adhesions between visceral and parietal pleura) [11]. The procedure brought a satisfactory therapeutic effect. VATS procedure included close inspection of chest cavity, including diaphragmatic aspect, which did not show any abnormalities. Presence of visible fenestrations in diaphragm may explain the pathway of endometrial tissue as it was in case described above [11]. However, invisible micro-fenestrations may also be seen sites for the ectopic tissue [3]. The hypothesis about hematogenous dissemination is also possible considering the fact of surgical intervention within the uterus (cesarean section) in the past [7]. Another hypothesis regarding the etiology of endometriosis relies on the possibility of metaplasia body cavity lining into endometrial cells [7].

The ultimate diagnosis and confirmation is based on the histopathological examination. That is why in thoracic endometriosis lobectomy is preferred than CO2 laser ablation or electroablation which could be performed in patients with ovarian endometriosis [12]. H&E staining usually does not allow to detect foci of endometriosis. The recommended method to detect ectopic endometrial tissue is immunohistochemistry (cytokeratin-7, BER-EP4, and estrogen and progesterone receptors) [13]. In the present case, routine histopathological examination did not confirm the presence of endometrial tissue within the pulmonary parenchyma. A characteristic finding was presence of eosinophilic inflammatory infiltrate and numerous macrophages. Similar histopathological results were obtained in case reported by Yoshinobu Ichiki et al. [9]. In a case of pleural endometriosis reported by Mesbah Raham, routine H&E stain did not reveal endometrial structures but additional immunohistochemical examinations detected endometrial cells [14]. TES is a complex disease causing numerous diagnostic problems among clinicians, as well as among pathologists. The Permanente J. 2014; 18: 61-65.

M. Klimek et al.