Case Report

Catamenial Pneumohemothorax: A Rare Case of Spontaneous Recurring Cyclic Pneumohemothorax in Reproductive Age Women

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Background: Thoracic endometriosis syndromes (TES) is defined as cyclic radiological and clinical change of endometrial tissue in the thorax. Classic symptoms of catamenial pneumohemothorax (CPH) are cyclic and recurrent chest pain, chest discomfort, dyspnea and hemoptysis from spontaneous pneumothorax during menstrual period. Investigation of choice for diagnosis is computed tomography (CT) or magnetic resonance imaging (MRI). Video assisted thoracoscopy (VATS) provides visualization of lesion and treatment.

Objective: This document aims to detail the pathological findings, clinical manifestations and initial management associated to a thoracic endometriosis.

Materials and Methods: Document and follow-up CPH found in our institution.

Results: In this case, CT and VATS were done and showed TES. We prescribed two-milligram dienogest oral daily. The patient had amenorrhea, with absence of recurrent CPH.

Conclusion: Young women with cyclic or recurrent spontaneous pneumothorax suggested the presence of TES. The most essential attentiveness is a multidisciplinary team approach which combines surgical and medical treatment for individual patient.

Keywords: Pneumothorax, Hemothorax, Pneumohemothorax, Catamenial, Thoracic endometriosis

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Endometriosis is a disease characterized by having the extrauterine endometrial tissue. Endometriosis is found in up to 10 % of reproductive age women[1]. There are two types of endometriosis, namely pelvic and extra pelvic endometriosis. Pelvic endometriosis, known as endometrial tissue invading pelvic region, commonly occurs at ovary and anterior cul-de-sac[1]. Extra pelvic endometriosis such as thoracic endometriosis (TE), gastrointestinal tract and urinary tract endometriosis comprises around 8.9% of all endometriosis cases[2]. TE is described as endometrium invading thoracic region including diaphragm (38.8%), visceral pleura (29.6%), parietal pleura, and lung[3]. Thoracic endometriosis syndromes (TES) is defined as cyclic radiological and clinical change of endometrial tissue in thorax. TES comprises of catamenial pneumothorax (CP), hemothorax, hemoptysis, and lung nodules at 73, 14, 7 and 6%, respectively. Up to 90% of TE patients also experience catamenial thoracic pain counting of chest pain, shoulder pain, scapular pain, dyspnea, cough and hemoptysis. The symptoms usually manifest around 24 hours before and 72 hours after the onset of menstruation[4,5]. CP is a rare entity that is responsible for only 2.5 to 5.0% of spontaneous pneumothorax (SP) and yet accounts up to 31.4% in surgically treated of SP in women[6]. This document aims to detail the pathological findings, clinical manifestations and initial management associated to a TE found in our institute.

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Case Report

A 35-year old, nulligravida, Thai female, non-smoking without underlying disease presented to Thammasat Hospital due to recurrent pneumohemothorax. She had been complaining about dry cough and pleuritic chest pain for roughly four months prior to this visit. Her chest x-ray showed right pneumothorax and pleural effusion (Figure 1). Pleural paracentesis was performed and revealed serosanguinous fluid. Intercostal chest drainage was inserted to remove the air. A computed tomography (CT) imaging showed lung

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blebs and diaphragmatic nodules (Figure 2). Video assisted thoracoscopic surgery (VATS) was performed. Lung bullae at the right upper and middle lobes, dark spots, and bleeding nodules were found at the right diaphragm. The resection was done at the right pleura, right middle, lower lobes lung bleb (blebectomy), and diaphragmatic nodules. Pleurodesis diaphragmatic repair was done as a treatment. All tissues were sent for pathologic findings. Pathology report showed a focus of an endometrial implant of the visceral pleura (Figure 3). Pelvic examination was performed with noted enlarged uterus with nodularity at posterior fornix. Transvaginal ultrasonography demonstrated an enlarged uterus with a 5 cm intramural leiomyoma at its posterior wall. No abnormality was found at both ovaries. After surgery, the physician prescribed 150 mg depot medroxyprogesterone acetate intramuscularly. She later suffered from vaginal spotting. Three months later, she developed another episode of right pneumohemothorax. To reduce long-term side effects from leuprolide acetate, the oral progestin 2 mg (Dienogest) had been prescribed once daily to this patient. Four months after recovery, there was no recurrence of pneumothorax or pleural effusion. The patient no longer had any spotting with amenorrhea, no pelvic pain, no dyspnea or chest pain.

Discussion

Endometriosis is commonly found in reproductive age. It mostly affects pelvic organs. Several theories were proposed in order to explain pathogenesis of the pelvic and extra pelvic endometriosis, namely the metaplasia theory, the endometrium transplantation, and the induction theory[7]. The presence of cyclic endometrial tissue shredding in the thorax is called the TES. Majority of patients with TES develop CP followed by hemothorax and hemoptysis. These symptoms may be presented with dyspnea, chest pain, chest discomfort, shoulder pain, scapular pain, hemoptysis and chronic cough. The patient in this report presented with dry cough, dyspnea and chest tightness for 4 months. Chest radiography showed right pneumothorax which can be easily misdiagnosed for SP. There were no tangible criteria for diagnosing CP. The mean age of CP was 32 to 35 years old[4,8]. The disease is the condition that is defined by monthly pneumothorax that is usually synchronized with mensural cycle. TE especially pleural form is the most likely cause of pneumothorax. Approximately 50 to 84% of patient with CP has pelvic endometriosis, with or without infertility[9]. These symptoms match the information in this case since the patient has a history of dysmenorrhea, infertility, chest pain and pneumothorax on the second day of her period. All above

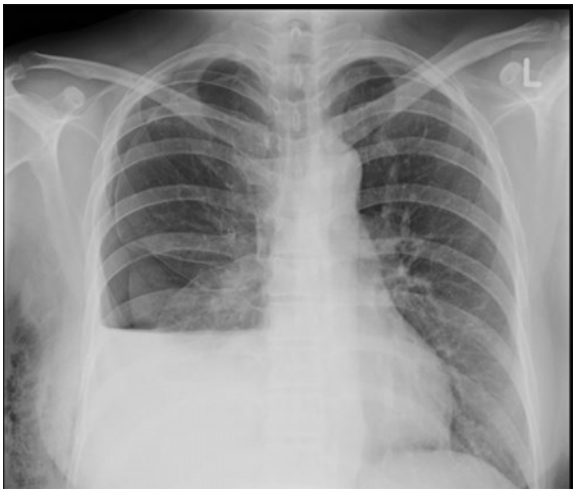

Figure 1. Chest x-ray PA upright: Visible right visceral pleural edge is seen as a very thin, sharp white line. No lung markings are seen peripheral to this line. Peripheral space is radiolucent compared to the adjacent lung. The right costophrenic angle is blunt. The diagnosis are right pneumothorax and pleural effusion.

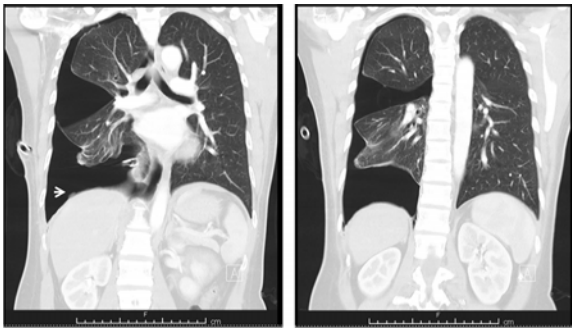

Figure 2. CT-chest: Coronal view evidence with arrow sign showing a right diaphragmatic nodule. Visceral pleura are seen. Right upper lung bleb was seen between right upper and middle lung.

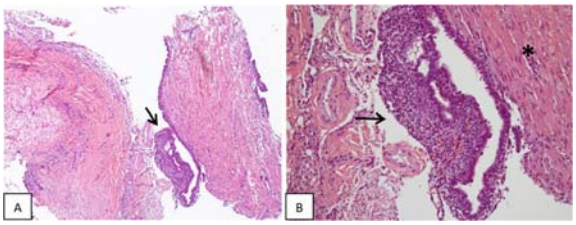

Figure 3. H&E stains: (A) Endometrial tissue dissociating pleural fibrosis. A focus of an endometrial implant of the visceral pleura (arrow). (B) The presence of atypical glandular structures and stromal component (arrow) was found beside the pleural fibrosis (black *).

favor CP. Chest radiography revealed non-specific findings such as pneumothorax and pleural effusion. CT or magnetic resonance imaging (MRI) should be performed during menstruation period to enhance imaging sensitivity. Size difference of lesion or appearance during the menstrual period was the key feature of diagnosis. CT revealed endometriosis implants as multiple nodular lesions with ground-glass/hypo-attenuating infiltration. CT result assisted the surgeon to proceed with appropriate excision during the thoracotomy[4,5]. Endometriosis usually occurs in pelvic form. As a result, additional initial management such as pelvic ultrasound, maybe helpful in the diagnosis of pelvic endometriosis[7]. Diagnosis of CP made on symptoms and imaging could be misleading since a recent retrospective study by Bobbio et al in 2017 revealed that only 8 out of 20 CP patients were presented with catamenial chest pain earlier on right side[10]. Clinical suspicion is critical in CP diagnosis. The pathological evidence of endometrium is needed for definitive diagnosis of TE. From the work of Korom et al in 2004, CP were presented on the right, left, and both lungs at 91.7, 4.8, and 3.5%, respectively[3]. Histopathology of this case is presented in Figure 3. Previous research found endometrial stroma, gland, and bullous dystrophy in 92, 62, and 37%, respectively[11]. Moreover, Legras et al in 2014 showed 100% involvement of diaphragmatic lesions with only 7% report of having parietal pleura lesion. The usage of antibodies such as estrogen, progesterone receptor, and CD10 in detecting endometrium in TE, was also reported[11]. Bronchoscopy is limited for diagnosis due to most of endometriosis lesion located at periphery of the lung[12]. Unfortunately, there is no concrete diagnosis for CP. There is obviously no current guideline for such a condition. Nonetheless, the treatment of CP could be categorized into the surgical and medical treatment. Surgical treatment is primarily VATS. Gold standard for diagnosis was direct visualization, biopsy and treatment for endometriosis lesion followed by diaphragmatic repair with or without pleurodesis[3,5]. The pleural installation with polyglycolic acid (PGA) was also successfully reported for treatment of pneumohemothorax with lesion involvement in the pleura and diaphragm[13]. Hormonal treatment utilizes suppressor endometrial implants which work by depressing the hypophyseal-gonadal axis to decrease risk of post-surgical therapy recurrence. Oral contraceptive pills, progestin, intrauterine devices, and danazol were seldom recommended due to abnormal shredding, especially those in the thorax. Gonadotropin-releasing hormone (GnRH) analog is an effective option for recurrence prevention of CP[14,15]. However, long-term use side effects from GnRH analog are vasomotor symptoms and bone mineral density change. In the present case, the author prescribed two-milligram dienogest oral daily. The patient had amenorrhea, with absence of clinical symptoms or recurrent CP after four months of treatment.

Conclusion

CPH is an extremely rare condition. It presents with critical conditions, which needed precise history taking and physical examination. Young women with cyclic chest pain or recurrent spontaneous pneumothorax with clinical of pelvic endometriosis suggest the presence of CPH. The most essential attentiveness is a multidisciplinary team approach which combines surgical and medical treatment for individual patient.

What is already known on this topic?

This case report presents a case of catamenial pneumohemothorax which extremely rare and frequent misdiagnosis. Most of previous reports had only clinical and imaging diagnosis. This case demonstrated imaging and pathological confirmed diagnosis. Gonadotropin-releasing hormone (GnRH) analog was an effective option for recurrence prevention of Catamenial pneumothorax. Though, long-term used side effect from GnRH analog were vasomotor symptoms, bone mineral density change.

What this study adds?

Gonadotropin-releasing hormone (GnRH) analog was an effective option for recurrence prevention of Catamenial pneumothorax. Though, long-term use side effects from GnRH analog were vasomotor symptoms, bone mineral density change. To prevent side effects, we used 2 mg dienogest oral daily. The patient had amenorrhea, with the absence of clinical symptoms or recurrent catamenial pneumohemothorax after 4 months of treatment. To our knowledge, this is the first report of dienogest used as a treatment for catamenial pneumohemothorax in Thailand.

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Potential conflicts of interest

The authors declare no conflicts of interest.

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ภาวะลมรั่วและเลือดออกในช่องเยื่อหุ้มปอดขณะมีประจำเดือน เหตุการณ์ที่พบได้ยาก การกลับเป็นซ้ำแบบเป็นรอบเดือนของภาวะลมรั่วและเลือดออกในช่องเยื่อหุ้มปอดหญิงวัยเจริญพันธุ์

อวัสดา บุณยัษเฐียร, ณัฐภัทร กาญจนวิไล, อภิษฎา ชุ่มคำ, อุรวี ลิมปีเวศน์, คมสันติ์ สุวรรณฤกษ์, อธิตา จันทเสนานนท์, กรณ์กาญจน์ ภมรประวัติธนะ

ภูมิหลัง: กลุ่มอาการภาวะเยื่อบุผนังมดลูกเจริญที่บริเวณทรวงอก คือ ภาวะที่ช่องอกมีเยื่อบุผนังมดลูกเปลี่ยนแปลงตามรอบเดือน โดยพบว่ามีอาการลมรั่วในช่องเยื่อหุ้มปอดขณะมีประจำเดือน เลือดออกในปอด ไอเป็นเลือด และตุ่มเยื่อบุผนังมดลูกเจริญในปอด อาการที่พบบ่อยของของภาวะลมรั่วและเลือดออกในช่องเยื่อหุ้มปอดขณะมีประจำเดือนคือ เจ็บหน้าอก แน่นหน้าอก หายใจไม่สะดวก และไอเป็นเลือด ซึ่งพบอาการสัมพันธ์ตามรอบประจำเดือน ซึ่งเป็นอาการปรากฏซ้ำของภาวะลมรั่วในเยื่อหุ้มปอดที่เกิดขึ้นได้เอง การสืบค้นเพิ่มเติมสำหรับวินิจฉัยคือการส่งตรวจวินิจฉัยโรคด้วยเอกซเรย์คอมพิวเตอร์ หรือการส่งตรวจด้วยการสร้างภาพด้วยสนามแม่เหล็กไฟฟ้า นอกจากนั้นสำหรับการผ่าตัดช่องทรวงอกด้วยการส่องกล้องจะทำให้เห็นรอยโรคได้ชัดเจนยิ่งขึ้นช่วยในการวินิจฉัยทั้งยังช่วยทำการรักษาได้พร้อมกัน

วัสดุและวิธีการ: เพื่อบันทึกและรายงานรายละเอียดเกี่ยวกับพยาธิวิทยาของชิ้นเนื้อที่ตรวจพบ อาการแสดง และการดูแลรักษาอาการภาวะเยื่อบุผนังมดลูกเจริญที่บริเวณทรวงอก โดยการติดตามผู้ป่วยลมรั่วและเลือดออกในช่องเยื่อหุ้มปอดขณะมีประจำเดือนที่พบ

ผลการศึกษา: สำหรับผู้ป่วยรายนี้ได้ส่งตรวจวินิจฉัยเพิ่มเติมด้วยเอกซเรย์คอมพิวเตอร์ และผ่าตัดช่องทรวงอกด้วยการส่องกล้อง พบว่าผู้ป่วยมีภาวะลมรั่ว และเลือดออกในช่องเยื่อหุ้มปอดขณะมีประจำเดือน ผู้ป่วยได้รับยา dienogest 2 มิลลิกรัมต่อวันสำหรับรักษา เมื่อติดตามการรักษาพบว่าผู้ป่วยไม่มีประจำเดือน และไม่มีอาการเลือดออกและลมรั่วในเยื่อหุ้มปอดอีก

สรุป: ผู้ป่วยหญิงอายุน้อยที่มีอาการลมรั่วในเยื่อหุ้มปอดแบบเป็นซ้ำหรือตามรอบเดือน บ่งถึงอาการเยื่อบุผนังมดลูกเจริญที่บริเวณทรวงอก การรักษาที่สำคัญคือรักษาเป็นทีมและให้การรักษาโดยการผ่าตัดและการใช้ยา