中文題目：與月經週期相關的罕見反覆性氣胸

英文題目：A rare type of recurrent pneumothorax related to menstrual cycle

作　　者：劉舜傑[1]，王喬弘[2]，徐永勳[3]

服務單位：台北市立聯合醫院仁愛院區一般內科[1]；胸腔內科[2]；腎臟科[3]

Background:

Catamenial pneumothorax is a disorder bearing a temporal relationship with menstruation. It is a relatively rare condition but a treatable one. Although the symptoms are usually of mild intensity, the high recurrent rate can severely lower the quality of life. Several theories have been developed to explain the underlying mechanisms. Accordingly, there are different approaches to control such intractable disorder.

Case presentation:

A 39-year-old woman without underlying medical illness presented with sudden shortness of breath. Physical examination revealed no breathing sound over the right chest. Laboratory had no significant findings. Chest X-ray showed obvious right sided pneumothorax. She mentioned that she had been having similar symptoms within one day of menstrual bleeding every month during recent 6 months. She had never smoked and had no asthma history. She had never had a child; however, no history of infertility had been discovered, nor had she undergo any pelvic surgery. She was admitted for pneumothorax decompression with pigtail catheter first and then received pleurodesis under the diagnosis of catamenial pneumothorax. After surgical treatment, the symptoms subsided and she continued to do well on hormonal therapy.

Discussion:

Pneumothorax is a common medical emergency in both hospitals and out-of-hospital settings. While primary spontaneous pneumothorax occurs without a precipitating event, secondary pneumothorax requires a workup for the underlying etiology. One particular type deserving special attention is linked to menstruation, namely catamenial pneumothorax. The better terminology is thoracic endometriosis. Chest was the second most common involved organ after the pelvis in endometriosis. While the patient can present with hemoptysis or hemothorax, pneumothorax is the most common presenting symptom. It usually starts just before or within 72 hours after the onset of monthly bleeding. However, the definite correlation between menstruation and catamenial pneumothorax has not been unequivocally determined.　Four theories have been proposed for the development of thoracic endometriosis:

1. <u>Physiological</u>

   High concentration of circulating prostaglandin F2 was found during menses, which leads to vasoconstriction and bronchospasm, thereby causing alveolar rupture.

2. <u>Coelomic metaplasia</u>

Due to the shared embryologic origin of endometrium and mesothelium, it was proposed that the pluripotent cells undergo transformation into endometrial cells in the pleura at some point during differentiation.

3. Lympho-vascular embolization

Endometrial tissue can “metastasize” to the lung parenchyma, pleura, bronchus and diaphragm through the venous or the lymphatic system. This theory is especially responsible for the occurrence of bilateral parenchymal lesions.

4. Diaphragmatic defects

Retrograde menstruation is the most favored mechanism responsible for pelvic endometriosis. With the air trapped in the peritoneal cavity through fallopian tubes by uterine contraction, it travels to the pleural cavity by negative intrathoracic pressure and, most importantly, diaphragmatic defects.

Histopathological specimens (hormone receptor-positive endometrial stroma and glands) are necessary to make a definitive diagnosis of “thoracic endometriosis”, which aids for predicting the risk of recurrence and for the planning of the hormonal suppressive therapy. More often than not, a diagnosis is made clinically and treatment should not be delayed while obtaining tissue.

Treatment usually consists of surgery and postoperative hormone therapy. Video-assisted thoracoscopic surgery (VATS) is usually performed for both diagnostic and therapeutic purposes should there be suspicion for the diagnosis. One should always look for endometrial implants and diaphragmatic defects to repair. Because of the high recurrence rate, an adjunctive hormonal suppressive therapy for 6 to 12 months is usually recommended. Pharmacological treatment is aimed to induce hypoestrogenism, resulting in temporary menopause. Thus treatment plan should always balance possible benefits of reduced recurrence against the acute symptoms of menopause and risks of osteoporosis. The most commonly used drug is gonadotropin-releasing hormone (GnRH) agonists such as leuprolide.

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Conclusion:

Recurrent pneumothorax occuring in women of reproductive age should prompt an investigation of thoracic endometriosis especially when they present perimenstrually. We reviewed a clinical case regarding a woman visiting ER due to repeated dyspnea on a monthly basis. We also discussed the pathomechanisms, clinical manifestations as well as the management of the disesase. Through this discussion, we hope to raise the awareness of this rare disease.