Recurrent pneumothorax in a woman with endometriosis

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Abstract

Catamenial pneumothorax is a rare cause of pneumothorax occurring in less than 5% of women with pelvic endometriosis. In this report, we present a 39-year-old woman who presented with a second episode of pneumothorax within a ten-year period of being diagnosed with endometriosis.

Introduction

Spontaneous pneumothorax in a woman with history of endometriosis is known as catamenial pneumothorax (CP). A high index of suspicion is necessary to make the diagnosis and management is often delayed due to misdiagnosis. In this report, we present a young woman with endometriosis with two episodes of pneumothorax in a 12-year period.

Case Presentation

A 39-year-old woman presents to the emergency room with new onset chest pain and shortness of breath. Physical exam had decreased breath sounds on the right lung field. She referred onset of menstruation in the past 3 days despite the strict compliance with continuous noncyclical combination oral contraceptive pills (COCPs). Her past medical history included endometriosis, infertility, scoliosis, hypertension, and ruptured brain aneurysm which required neurosurgical intervention in October 2014. Genetic test for collagen disorders and alpha 1 antitrypsin were negative. She had an episode of right-sided pneumothorax attributed to a ruptured apical bleb 12 years prior, which was treated with pleurectomy via video-assisted thoracic surgery (VATS). Chest radiography (CXR) showed a right lung pneumothorax causing mass effect upon the mediastinal structures with left shift and a subtle right diaphragmatic perforation (Figure 1A). Tube thoracostomy resulted in partial re-expansion (30% persistent pneumothorax) of the right lung field and she continued to have persistent air leak for 7 days. Two chest CT scans were performed during this period which showed residual right-sided pneumothorax involving the right upper lung field with bibasilar atelectatic changes; no bullae or lesions suggestive of endometriosis were identified. Bronchoscopy showed clear patent airways with no significant findings. Due to the persistent pneumothorax, the patient was taken for open exploration (via right posterolateral thoracotomy) on day 8 of admission. On gross inspection no bullae, blebs, or endometriosis implants were identified. However, on inspection of the diaphragm, several 2-6 mm central tendon defects with protrusion of liver were identified (Figure 1B). The defects were repaired with simple plication with plegated 3-0 monofilament absorbable suture. Mechanical pleural abrasion with limited pleurectomy was also performed. Biopsies were taken from the right middle lobe and pleural cavity. Post-op day 1 imaging demonstrated she was free of pneumothorax, she no longer had air leak, and the chest tube was removed. The pathology report found that the biopsies stained positive for both estrogen and progesterone receptors supporting the diagnosis of catamenial pneumothorax (Figure 2). Prior to discharge she was placed on a combination of a pro-gestational and GnRH analogue therapies, with depot medroxyprogesterone acetate and leuprolide, respectively. At 1 year follow up she was free of symptoms and back to her normal daily activities.

Discussion

CP classically presents within 72 hours before or after onset of menses in women with history of pelvic endometriosis[1-3]. Our patient developed CP despite strict adherence to COCPs which in most cases controls the progression of endometriosis. She had a pneumothorax 12 years prior which may have been related to her endometriosis however, this was not considered at that time. This could have been due to the young age of presentation which may have led physicians to wait for a second episode to establish a temporal relationship with menses. Her history of infertility could have been the determining risk factor which, along with a history of pelvic surgery, has been found to be the one of the strongest predictors of CP[5]. Few reports have documented CP in women on ovulatory suppression[2]. To our knowledge, this is the second case of CP reported in a Puerto Rican woman[7].

Characteristic lesions of CP include endometrial foci at the pleura, lungs, and/or diaphragm. Single or multiple diaphragm fenestrations at the central tendon have also been described[1-2].

However, CXR rarely reveals small diaphragmatic defects suggestive of perforation, as was demonstrated in our patient's CXR (Figure 1A and 1B). Interestingly, histological evidence of thoracic endometriosis (endometrial stroma and glands or stroma only plus positive staining for estrogen/progesterone receptor) is not revealed in all cases[1]. Rousset-Jablonski et al. found that histologically confirmed thoracic endometriosis occurs in 64 to 87% of surgical biopsies[1-5].

Several theories have tried to explain the pathophysiology of CP. These theories includes the anatomic model, the visceral model and the metastatic/lymphovascular micro-embolization theory. In this patient, the anatomic and/or visceral pleural models may account for both pneumothorax events as diaphragmatic fenestrae were found at the central tendon. A thorough examination of the diaphragm was not performed at her first intervention as they may have been present at that time.

Surgery and adjunct hormonal suppression constitutes the treatment of choice for CP. Surgical approach includes video-assisted thoracic surgery (VATS), video-assisted mini-thoracotomy, or open thoracotomy. We decided to explore via right thoracotomy for various reasons: 1) the recurrent nature of our patient's pneumothorax, which had been treated with VATS 12 years prior 2) pre-operative CT scans did not show a bleb or an obvious lesion as the etiology of the new recurrence, 3) persistent leak after 7 days of tube drainage, 4) right thoracotomy provided excellent visualization in a previous CP patient operated by the senior author [7]. The principles of operative management include: 1) resection of all visible intrathoracic lesions such as blebs, bullae, endometrial implants via limited wedged resection, 2) diaphragmatic plication with or without resection, 3) and mechanical or chemical pleurodesis.

Despite best efforts, CP reoccurs in 5% and 25% of surgically treated patients at 6 and 12 months, respectively[1,6,8]. Adjuvant gonadotropin-releasing hormone (GnRH) analogues reduce recurrence rate down to less than 5%[8]. Depot medroxyprogesterone acetate and leuprolide for 6 months after surgery prevents cyclical hormonal changes and induces suppression of ectopic endometrium until effective formation of pleural adhesions occur after pleurectomy. GnRH analogues are often chosen as first-line treatment due to their effectiveness at suppressing ovarian hormonal production but side effects (increased risk of osteoporosis, infertility, acute menopausal symptoms) could be a determining factor when choosing a hormonal agent. Even with combined treatment of surgery and hormonal suppression, up to 32% of women will experience recurrence[9].

In closing, we describe an unusual presentation of CP in a woman with severe endometriosis that had two episodes of pneumothorax within 12 years. Thoracic endometriosis may have accounted for the first episode but this was never confirmed. High clinical suspicion for CP is important in any woman with history of endometriosis that presents with shortness of breath or pneumothorax. This will ensure proper surgical and medical management to address this complex clinical entity.

Figures

Figure 1. A. Chest X-ray. Right pneumothorax with perforation at the right diaphragm (area magnified 2x). **B. Diaphragmatic fenestrations.** Intra-operative findings of diaphragmatic fenestrations with liver protrusions.

Figure 2. Microscopic Findings. **A**. **H&E stain**. Parietal pleura biopsy: pleural tissue with chronic inflammation, hemosiderin deposits, and focal mesothelial hyperplasia (40x). **B/C. Immunostaining (40x)**. Estrogen and progesterone receptors positive, respectively.

Conflict of Interest: The authors has declared that no conflict of interest exists.

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