

A very rare cause of hemoptysis and chest pain lung endometriosis. A case report

Abstract

Hemoptysis can be caused by multiple conditions like tuberculosis, malignancies, bronchiectasis or pulmonary embolism. Sometimes, hemoptysis can be caused by other diseases, like vasculitis, lymphangioleiomyomatosis or other rare diseases. We present a clinical case of an 18 years old woman with recurrent mild hemoptysis. Thoracic computer tomography (CT) showed ground glass opacity and small micronodules in the right upper lobe. A thorough anamnesis revealed that the patient had catamenial hemoptysis that raised the hypothesis of pulmonary endometriosis, which is a challenge for every clinician. Spontaneous regression after one month and the repeated hemoptysis after five months linked to menstruation along with the same aspect on CT raised the diagnosis of thoracic endometriosis syndrome. The thoracic endometriosis is a rare and complex condition affecting women at childbearing age. The diagnosis is frequently difficult and histopathological confirmation is often required for positive diagnosis. Moreover, if the patient has typical clinical history and other differential diagnosis are ruled out, a presumptive diagnosis can be achieved.

Keywords: hemoptysis, endometriosis, pulmonary

Introduction

Endometriosis represents the presence of functioning endometrial tissue in extrauterine locations. Endometriosis most commonly is confined to the pelvis. However extrapelvic sites like thorax, abdomen, brain and skin may be found⁽¹⁾. Thoracic involvement is one of the most common extra-pelvic location. Pulmonary endometriosis is a rare condition and can involve the tracheobronchial tree, the lung parenchyma and lung pleura. Pulmonary endometriosis has four main clinical conditions: catamenial pneumothorax, catamenial hemothorax, catamenial hemoptysis and pulmonary nodules. The most frequent symptoms like chest pain, hemoptysis, shortness of breath happening related with the menstrual period⁽²⁾. Due to its rarity the suspicion of diagnosis is often delayed, with possible life-threatening complications. The present case aim is to raise awareness amongst clinicians, particularly pulmonologists, thoracic surgery and gynecologists, about this particular condition⁽³⁾.

Case Report

We present the case of an 18 years old woman who came to the emergency room of the hospital for hemoptysis, approximately 5 ml per day, for 3 consecutive days. The patient had no history of fever, weight loss or cough before this episode. She had no other significant comorbidities. The patient denies dyspnea or thoracic pain. Physical examination was normal, with body mass index of 18.3 kg/m². Routine laboratory tests and bleeding parameters were in normal ranges.

Chest X-ray showed an infiltrative opacity in the right upper lobe (Figure 1A). Sputum examination for acid fast

bacilli (AFB) was negative, and also negative for common bacteria. A thoracic computed tomography (CT) scan was performed which revealed ground glass opacity and small micronodules confined to the right upper lobe (Figure 1B). Bronchoscopy revealed neither active bleeding and nor endobronchial abnormalities. Bronchoalveolar lavage showed a moderate alveolar hemorrhage syndrome. Ziehl Neelsen stain was negative for AFB in bronchoalveolar lavage (BAL). No tumor cells were detected on cytology form BAL which had a normal cell count. Hemoptysis in a young and slim patient, living in a country with high incidence of tuberculosis, with a chest X-ray with an infiltrative opacity in the right upper lobe, raised the suspicion of pulmonary tuberculosis. Negative sputum and BAL for AFB and the fact that hemoptysis stopped spontaneously after three days, postponed the diagnosis. The patient did not repeat the hemoptysis in the next weeks. A thoracic CT scan was repeated one month later and a significant regression of the lesions was seen. Only a small (13 mm) area of ground glass opacity was observed, without any other abnormalities (Figure 2, A and B). At the thorough anamnesis made at follow-up, we found out that hemoptysis started with menstruation, lasted for 3 days and ended spontaneously. CT scan showed ground glass opacity with tiny micronodules with a significant spontaneous regression after one month. Bronchoalveolar lavage revealed an alveolar hemorrhagic syndrome at first hospitalization. Sputum cultures for M. tuberculosis were negative. A presumptive diagnosis of pulmonary endometriosis was made. The patient refused lung biopsy or hormonal therapy. She denied induced abortion, and she had no previous pelvic interventions. Clinical examination, which included gynecology

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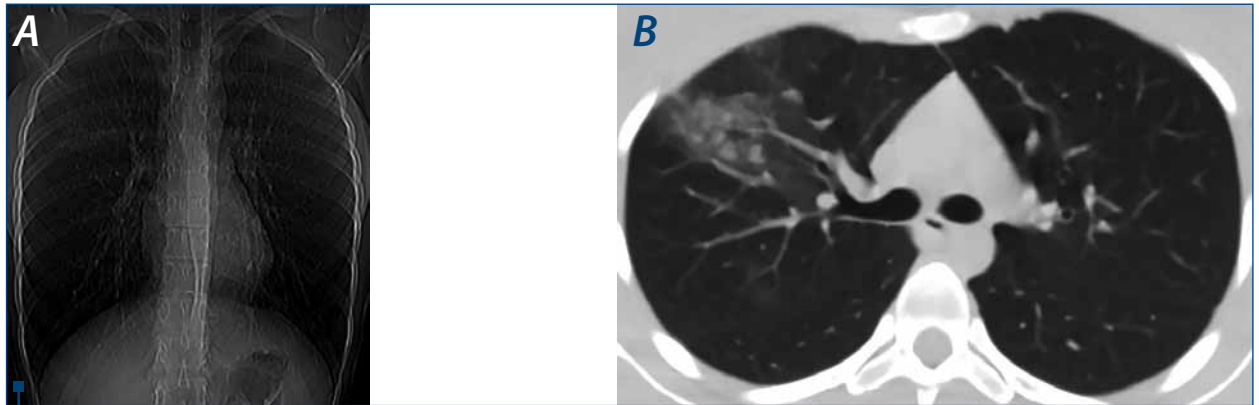


Figure 1. A. Chest X-ray of the patient with infiltrative opacities in the right upper lobe; B. Thoracic CT with ground glass and small micronodules in the right upper lobe

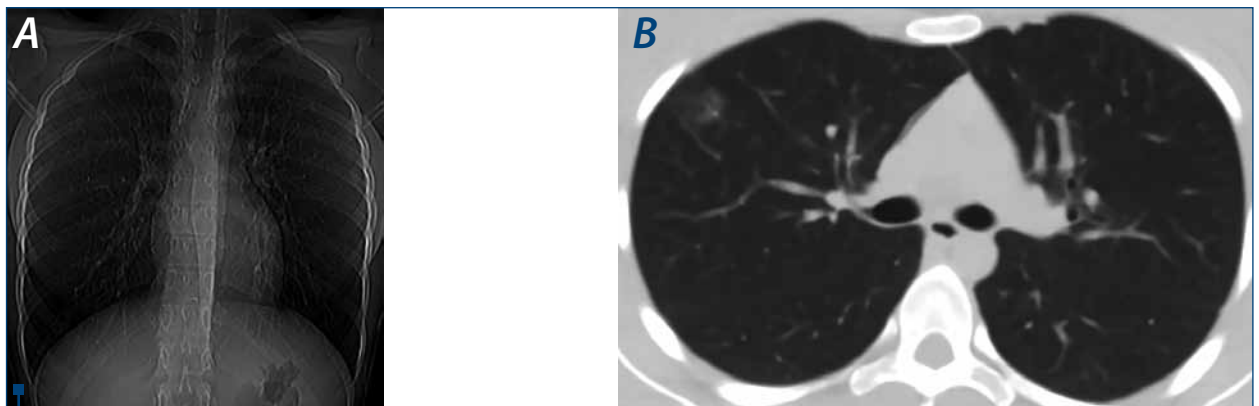


Figure 2. A. Chest X-Ray after one month –normal image; B. CT with 13 mm area of ground glass opacity in the right upper lobe (arrow)

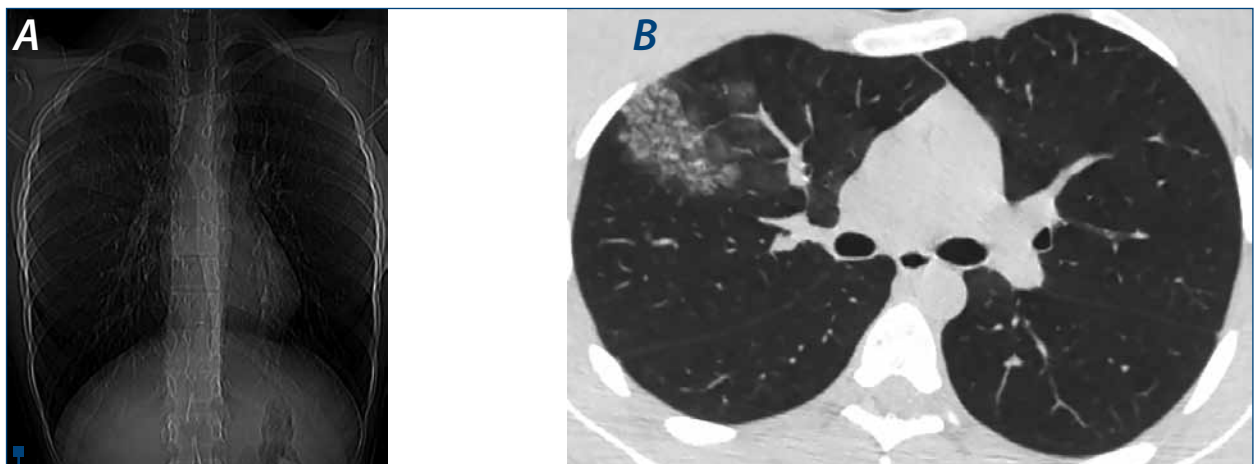


Figure 3. A. Chest X-ray after 5 month with upper right lobe infiltrate; B. CT scan with ground glass opacity and tiny micronodules in the same region

logic examination, and pelvic ultrasonography were normal. The catamenial hemoptysis repeated after 5 month and the thoracic CT revealed the same lesions as the first one (i.e. ground glass opacities and micronodule, Figure 3, A and B). The second time the hemoptysis stopped after 3 days and the radiological lesions showed a spontaneous regression.

The patient refused further investigation and treatment, but the hemoptysis did not recur for almost two years.

Discussion

Endometriosis is a benign gynecological condition. It is more frequent encountered in pelvis: ovaries, uterine

ligaments, peritoneum, cervix, labia, and vagina⁽¹⁾. It was described in 1860, by Carl von Rokitansky, and named “adenomyoma”⁽²⁾. He found endometrial glands in the myometrium and named this condition “cystosarcoma adenoids uterinum”. Today it is defined as “pelvic endometriosis”. Its major complain is pelvic pain, often intense, but dysmenorrhea, dyspareunia, and infertility are also possible. Malignant transformation is an exceptionally rare complication of endometriosis, most frequent in the ovaries^(1,4).

Although endometriosis can affect up to 15% of women of childbearing age, pulmonary endometriosis is a very rare finding. The etiological mechanisms of pulmonary endometriosis are still not well known. The most accepted theories include peritoneal-pleural movement of endometrial tissue through diaphragmatic defects and microembolization through pelvic veins or lymphatic circulation⁽³⁾. The theory of trans-diaphragmatic passage explains the presence of endometriotic lesions on the right pleura in over 90% of cases and the more common presentation of catamenial pneumothorax versus other less common entities of thoracic endometriosis syndrome (TES). The last theory can explain parenchymal disease. It was assumed that obstetrical and gynecological procedures that disrupt endometrial blood vessels and lymphatics (e.g., hysteroscopy, curettage) may allow endometrial tissue to enter and to embolize in lung parenchyma⁽⁴⁾. There is a significant relation between the presence of pelvic and pulmonary endometriosis. Fifty to 84% of women diagnosed with TES have concomitant pelvic endometriosis⁽⁴⁾. In a retrospective study of 110 patients with TES from 1996⁽³⁾ was found that the peak incidence for TES was between 30 and 34 years, whereas the peak incidence of pelvic endometriosis was between 24 and 29 years, approximatively 5 years earlier.

The typical clinical presentation includes respiratory symptoms related to menstruation: dyspnea, hemoptysis, and chest scapular pain. The most common presentation of TES is catamenial pneumothorax, almost 70% of cases. Other clinical entities include catamenial hemothorax (14%), catamenial hemoptysis (14%) and lung nodules (2%)⁽⁵⁾.

Catamenial pneumothorax is defined as recurrent pneumothorax occurring within 72 hours from the onset of menstruation (i.e. rarely 96 hours). It is interesting to know that pneumothorax due to endometriosis also can be non-catamenial (less than 10%). The correct diagnosis of endometriosis-related pneumothoraces without a temporal relationship with menses is often made during surgical interventions for recurrent pneumothorax. The symptoms of pneumothorax are chest or scapular pain, and dyspnea.

Catamenial hemoptysis is produced by parenchymal or endobronchial endometriosis. The mean age of patients with hemoptysis appears to be lower than patients with pneumothorax or hemothorax⁽⁵⁾. Usually, hemoptysis does not occur with every menstrual cycle but can frequently occur throughout patient's life. In a small retrospective study from 2010, from 19 patients with catamenial hemoptysis 16 patients (84%) had a history of gynecological or obstetric procedures before developing hemoptysis⁽⁶⁾.

Pulmonary endometriosis is rare but can be life-threatening. Complications can vary from mild symptoms to major

complications. Massive pneumothoraces can lead to lung collapse, respiratory failure, or even death. A catastrophic pulmonary hemorrhage can produce cardiovascular shock and subsequent death. Repeated pleurodesis can raise the risk of infection, pachypleuritis⁽⁷⁾. A presumptive diagnosis of thoracic endometriosis can be made with typical clinical history. Although histopathologic confirmation is preferred, it is not always necessary. TES should be suspected in women of childbearing age with pneumothorax, hemothorax or hemoptysis, especially with perimenstrual appearance, with previous pelvic surgery or infertility. Diagnosis is made by combinations of typical symptoms, with suggestive imaging features and histopathological confirmation⁽⁷⁾.

Treatment for thoracic endometriosis can be expectant, medical or surgical depending on symptoms severity or patient desire. No large-scale randomized trial has been made. Medical treatment is the usual first step in the management of TES. Usually are used Danazol, gonadotropin releasing hormone agonists, oral contraceptive pills, and other progestational agents. Hormonal therapy is expensive, with frequent side effects, and the recurrence can appear when the therapy is ceased⁽⁷⁾. Due to its benign course, in the particular case of patients with hemoptysis treatment may be conservative or with hormonal agents⁽⁶⁾.

Surgical treatment should be considered in cases of medical therapy being ineffective or with side effects. Surgical options are pleurodesis, pleurectomy and segmental resection⁽⁸⁾. Video-assisted thoracoscopy (VATS) allows visualization of pleural lesions with histopathological confirmation and the possibility to resect lesions, apical blebs, and to repair diaphragmatic fenestration. There is the possibility of a combined protocol with VATS and video-assisted laparoscopy for severe patients, which address both abdominal and thoracic disease in a single operation⁽⁷⁾.

Conclusions

TES is a rare and often complex syndrome. For the correct diagnosis, it requires a high index of suspicion in any woman of childbearing age with recurrent chest pain, hemoptysis or dyspnea. Once the diagnosis is made, these women with TES need proper gynecologic follow-up considered the concurrent pelvic endometriosis with potential evolution to infertility. ■

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