ORIGINAL RESEARCH PAPER

Pulmonology

UNUSUAL CAUSE OF SECONDARY PNEUMOTHORAX IN A PREMENOPAUSAL WOMAN: CASE REPORT

KEY WORDS:

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ABSTRACT

Lymphangioleiomyomatosis (LAM) is a rare lung disease predominantly affecting women of reproductive age, often presenting with pneumothorax. We report a 42-year-old female presenting with one week of shortness of breath, left-sided chest pain, and dry cough. Examination revealed absent breath sounds on the left hemithorax. Investigations showed exudative lymphocytic pleural effusion with low ADA, obstructive spirometry pattern, and normal blood tests. Imaging confirmed left-sided pneumothorax with cystic changes, and immunohistochemistry was positive for smooth muscle actin, confirming LAM. She was managed with intercostal drainage, oral tamoxifen 10 mg twice daily, and inhaled beta-2 agonist therapy. At three months follow-up, she showed clinical improvement without radiological progression. Early recognition of LAM is essential in women presenting with secondary pneumothorax.

INTRODUCTION

Lymphangioleiomyomatosis (LAM) is a rare interstitial lung disease with a prevalence of 1–2 per million women. It is characterized by proliferation of smooth muscle–like cells causing cystic lung destruction, lymphatic obstruction, and recurrent pneumothorax. The disease predominantly affects women of reproductive age, but cases have been reported in premenopausal women in their 40s. Clinical manifestations include dyspnea, cough, and recurrent pneumothorax. Diagnosis is confirmed by imaging, histopathology, and immunohistochemistry with smooth muscle actin or HMB-45 positivity. Early recognition and treatment are crucial to prevent progression.

Case Presentation

A 42-year-old female homemaker presented with a one-week history of shortness of breath, left-sided chest pain, and dry cough. She denied fever, hemoptysis, wheezing, pedal edema, or any history of trauma. The patient had no history of smoking, alcohol intake, or drug use. On examination, her heart rate was 130 beats per minute, blood pressure 110/70 mmHg, respiratory rate 28 breaths per minute, and oxygen saturation was 97% on room air. General examination revealed no pallor, icterus, cyanosis, clubbing, or lymphadenopathy. Respiratory system examination showed absent breath sounds over the left hemithorax, while cardiovascular, neurological, and abdominal examinations were unremarkable.

Laboratory investigations, including complete blood counts, liver and renal function tests, were within normal limits. Pleural fluid analysis revealed an exudative, lymphocyte-predominant effusion with low adenosine deaminase (ADA), and smear for acid-fast bacilli was negative. Serum alpha-1 antitrypsin, angiotensin-converting enzyme (ACE), rheumatoid factor, and antinuclear antibody (ANA) titres were all within normal ranges. Spirometry demonstrated an obstructive pattern, and arterial blood gas analysis showed mild respiratory alkalosis with pH 7.54, PO_2 95.2 mmHg, PCO_2 33 mmHg, and oxygen saturation of 97%. Echocardiography and ultrasonography of the abdomen and pelvis were unremarkable.

Chest Xray(figure 1) shows left side hydropneumothorax ,CT thorax (figure 2) demonstrates a large left-sided pneumothorax, evident by the complete collapse of the left lung. The left hemithorax shows multiple thin-walled cystic lesions of varying size, predominantly in the upper and mid zones, giving a "honeycomb-like" appearance. The cysts are well-circumscribed, rounded, and scattered, consistent with cystic lung disease. The contralateral right lung shows relatively preserved parenchyma with only minor scattered cysts. No significant pleural effusion or mediastinal shift is noted, although the left lung is markedly compressed. These imaging features, along with clinical presentation, strongly suggest a cystic lung process causing secondary spontaneous pneumothorax.

DISCUSSION

LAM is a rare disease primarily affecting women of reproductive age, often presenting with recurrent pneumothorax. Diagnosis relies on imaging and histopathology with immunohistochemistry confirmation. Management includes drainage of pneumothorax, antiestrogen therapy such as tamoxifen, and mTOR inhibitors in select cases. Lung transplantation is reserved for advanced disease.

This case is notable because the patient was 42 years old, slightly older than typical reproductive age, and presented with secondary pneumothorax as the first manifestation. Laboratory evaluation showed exudative lymphocytic pleural effusion with low ADA, normal blood counts, and normal organ function. Spirometry demonstrated an obstructive pattern, and arterial blood gas showed mild respiratory alkalosis. Management with intercostal drainage and tamoxifen 10 mg twice daily led to clinical improvement at three months follow-up

Clinicians should maintain a high index of suspicion for LAM in women presenting with secondary pneumothorax, regardless of age. Early recognition and pharmacologic intervention can prevent disease progression and improve patient outcomes.

CONCLUSION

LAM is a rare disease of women of reproductive age that can also present in premenopausal women. Secondary pneumothorax may be the initial manifestation. High clinical suspicion, timely diagnosis with imaging and histopathology, and early initiation of antiestrogen therapy are essential for favorable outcomes.

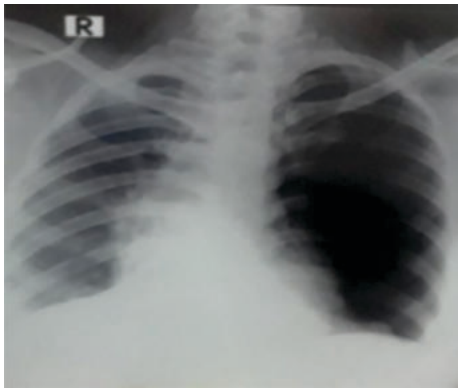

Figure 1: Chest Xray

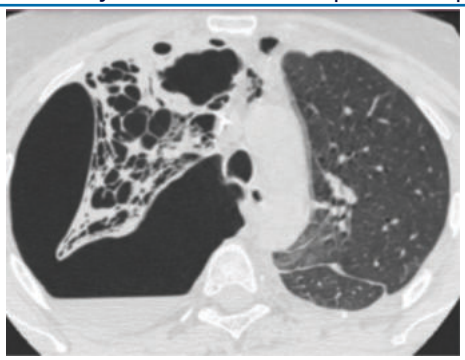

Figure 2: Chest CT Thorax

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