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Pulmonary Lymphangioleiomyomatosis on a Post-Menopausal Woman with Chronic Lymphocytic Leukaemia

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Abstract

A 62 years old, post-menopausal female was admitted to the Internal Medicine Ward due to dyspnoea, cough and sputum of at least 2 months. Shortness of breath, cough and hypoxaemia persisted and the patient was submitted to a pulmonary angiogram ct which revealed numerous thin-walled air cysts affecting upper and medial zones of both lungs, typical images of pulmonary lymphangioleiomyomatosis. After discharge to Internal Medicine Consultation Service with Metilprednisolone, the patient was no longer hypoxaemic and remained asymptomatic, even after withdrawal of oral corticosteroid to inhalatory formulation. Further surveillance in short time was scheduled in order to implement rapid imunossupressive treatment when necessary.

Keywords

Pulmonary Lymphangioleiomyomatosis, Chronic Lymphocitic Leukaemia, Post-Menopausal

1. Introduction

Lymphangioleiomyomatosis (LAM) is a rare systemic disease of unknown aetiology which results from the proliferation in various organs of neoplastic-acting smooth muscle-like cells and occurs primarily in pre-menopausal women, leading to the hypothesis of female hormones being involved in its pathogenesis [1].

The disorder is typically characterized by the formation of thin-walled pulmonary cysts resulting in destruction of pulmonary tissue with progressive lung dysfunction and the growth of abdominal tumours such as angiomyolipomas and lymphangioleiomyomas [2]. Its key clinical features include dyspnoea, haemoptysis, recurrent pneumothorax, chylothorax, and chylous ascites [3] [4].

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LAM is frequently but not always associated with an autosomal dominant disease called Tuberous Sclerosis Complex, mainly resulting from mutations in *TSC*1 and *TSC*2 genes [5].

2. Case Presentation

A 62 years old, post-menopausal female, was admitted to the Internal Medicine Ward due to dyspnoea, cough and sputum.

At admission, being feverish and due to information of remissive chronic lymphocytic leukaemia, was medicated with Ceftriaxone and Azithromycin.

Despite becoming apyretic, shortness of breath, cough and hypoxaemia persisted and the patient was submitted to a pulmonary angiogram ct which revealed numerous thin-walled air cysts affecting upper and medial zones of both lungs (**Figure 1**), typical images of pulmonary lymphangioleiomyomatosis (**Figure 2**) [6].

Latter work-up revealed normal pulmonary function tests and seric VEGF within range values [7].

A more detailed review of her past medical history revealed a history of long-standing cough (of at least 2 months) associated with orthopnoea, not related to any known interstitial lung disease.

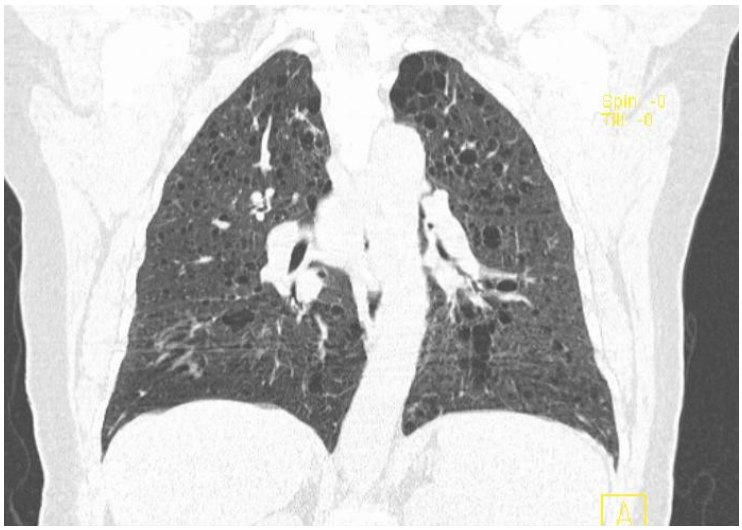

Figure 1. Multiple thin-walled round well-defined air-filled cysts throughout both lungs.

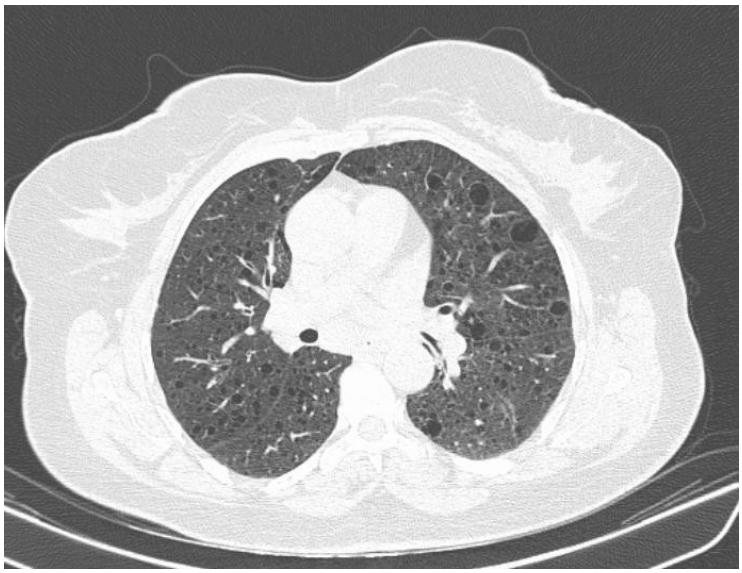

Figure 2. Axial cut from medial zone parenchyma typical of pulmonary lymphangioleiomyomatosis.

Due to facial lesions initially interpreted as herpetic but with suspicious cutaneous manifestation of tuberous sclerosis, abdominal ultrasound and high-definition ct scan were not suggestive of this diagnosis [8].

Nevertheless, the patient was instructed to seek medical assistance if any complications related to her disease occurred.

After discharge to Internal Medicine Consultation Service medicated with oral metilprednisolone, the patient was no longer hypoxaemic and remained asymptomatic, even after withdrawal of oral corticosteroid to inhalatory formulation.

Further surveillance in short time was scheduled in order to implement rapid imunossupressive treatment when or if necessary [2].

3. Discussion

The main clinical case is not atypical in any way from common lower respiratory tract infections. However, the persistance of symptoms after a first empiric antibiotic cycle suggested a pulmonary embolism (due to a procoagulant status from the patients heamatologic condition), thus the pulmonary angiogram ct.

LAM's findings were awkward regarding the patients age and menopause. Bringing the patient up to speed on her possible diagnosis, she was reluctant to any invasive diagnostic procedure (such as bronchfiberscopy or pulmonary biopsy) so we became dependant on a presuptive diagnosis.

Seric VEGF values were no help (regardless of its doubtful value in the diagnosis of LAM) but the images were interpreted by more than two experienced rediologists and were so typical that the diagnosis remained.

Our patient was fully instructed to any symptom regarding a possible complication (persistent ou sudden dyspnoea, haemoptysis or assimetric chest expansion with pleuritic pain). Surveillance for any new signs or symptoms by regular appointments (3 months apart) with pulmonary function tests (each 6 months) and abdominal ultrasound (each 12 months) could confirm or infirm our suspicion.

Due to a lack of histologic findings, according to the ERS guidelines, LAM is *probable*; the surveillance described before is important to find any angiomyolipomas, thoracic or abdominal chylous effusion or lymph-node involved by LAM, obtaining a *definite LAM* diagnosis. Nevertheless, the patient is clinically stable and healthy, which should be our prime consern.

4. Conclusions

Pulmonary lymphangioleiomyomatosis is a valid diagnosis even on post-menopausal women, regardless of the *not-so-practical* guidelines from the European Respiratory Society guidelines [6].

Patients should be alerted to signs and symptoms regarding complications in order to an early implementation of medical and chirurgical treatments [3].

It is advised to exclude the diagnosis of tuberous sclerosis [5].

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