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Pulmonary Lymphangioleiomyomatosis and Extra-Pulmonary Lymphangioleiomyomas in Postmenopausal Women

Orozco Maira [α], Garay M. Florencia [σ] & Mattar Daniel [ρ]

Abstract- Lymphangioleiomyomatosis (LAM) is a rare disorder of unknown cause characterized by progressive cystic destruction of the lung, lymphatic abnormalities and abdominal tumors. It occurs almost exclusively in women of fertile age due to a hormonal influence, for this reason it is extremely rare in post-menopausal patients. While LAM is a systemic disease, the main manifestations are pulmonary. Lymphangioleiomyomas are larger cystic masses; these most commonly occur in the abdomen, retroperitoneum and pelvis but occasionally in the mediastinum and neck. In the cases of older women the disease presents a similar clinical manifestation than younger's female, with the exception that the clinical course is benign and longer.

Resumen- La linfangioleiomiomatosis es una enfermedad poco común de causa desconocida que se caracteriza por la destrucción quística progresiva del pulmón, anomalías linfáticas y tumores abdominales. Ocurre casi exclusivamente en mujeres en edad fértil debido a influencia hormonal, por esta razón es extremadamente raro en pacientes posmenopáusicas. Si bien la LAM es una enfermedad sistémica, las principales manifestaciones son pulmonares. Los linfangioleiomiomas son masas quísticas grandes; se encuentran con mayor frecuencia en el abdomen, retroperitoneo y pelvis, pero ocasionalmente en el mediastino y el cuello. En los casos de mujeres mayores la enfermedad presenta una manifestación clínica similar a la de las mujeres más jóvenes, con la excepción de que el curso clínico es más benigno y prolongado.

Resumo- Linfangioleiomiomatose é uma doença rara de causa desconhecida, caracterizada por destruição cística progressiva do pulmão, anormalidades linfáticas e tumores abdominais. Ocorre quase exclusivamente em mulheres em idade fértil devido a uma influência hormonal, por isso é extremamente raro em pacientes pós-menopáusicas. Embora a LAM seja uma doença sistêmica, as principais manifestações são pulmonares. Linfangioleiomiomas são massas císticas maiores; estes ocorrem mais comumente no abdômen, retroperitônio e pelve, mas ocasionalmente no mediastino e pescoço. Nos casos de mulheres mais velhas, a doença apresenta manifestação clínica semelhante à das mulheres mais jovens, exceto que o curso clínico é benigno e mais longo.

Keywords: Lymphangioleimyomatosis, mediastinal lymphangioleiomyomas, postmenopausal women, cystics lungs.

I. Introduction

We report a case of 72 years-old women, former smoker, denies hormone therapy replacement. She presents a history of dyspnea grade 2, rhonchi and wheezing symptoms diagnosed 4 year prior as Chronic Obstructive Pulmonary Diseases (COPD) and treated with inhaled corticoids and bronchodilators. The physical exam shows diffuse hypoventilation. The spirometry presents a reversible obstructive pattern. The Echocardiography reveals normal pulmonary arterial pressure.



Fig.1: High-resolution CT scan demonstrates multiples bullae of emphysema and some cysts (yellow arrows) characterized by the absence of intralesional vessels with thin walls.

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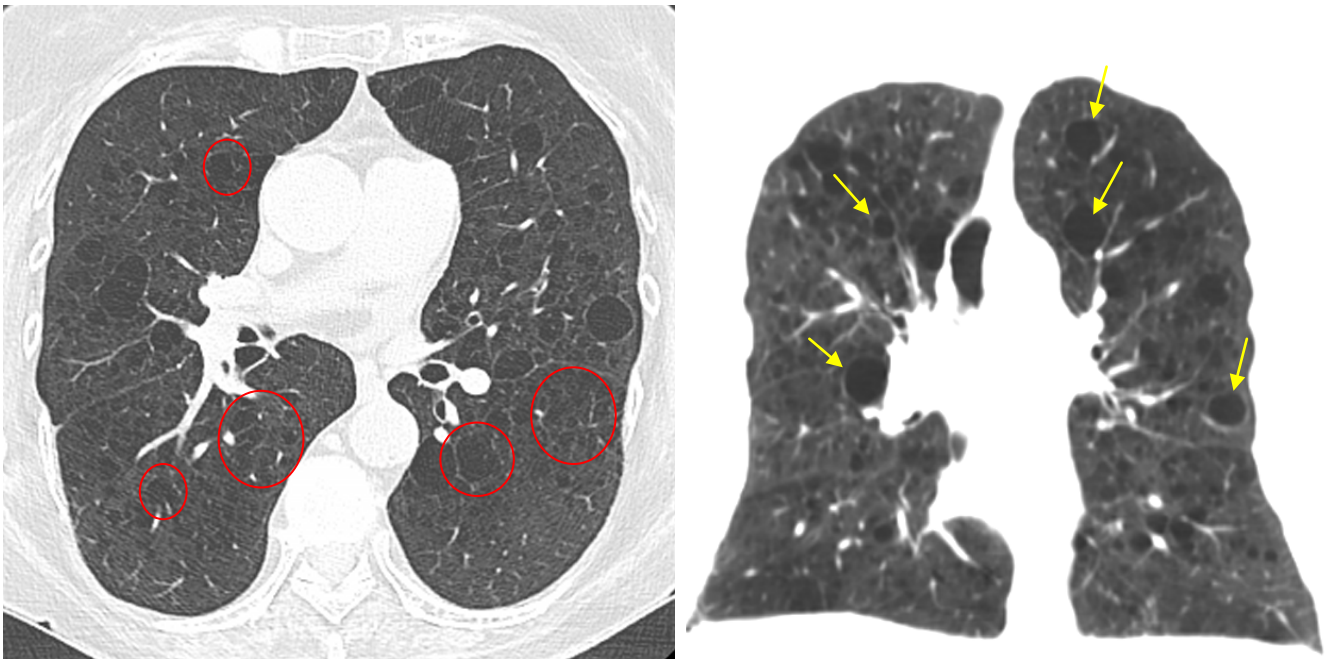


Fig. 2: High-resolution CT scan. A axial. B. coronal. Multiples bulles of emphysema and cysts (yellow arrows) with random distribution.

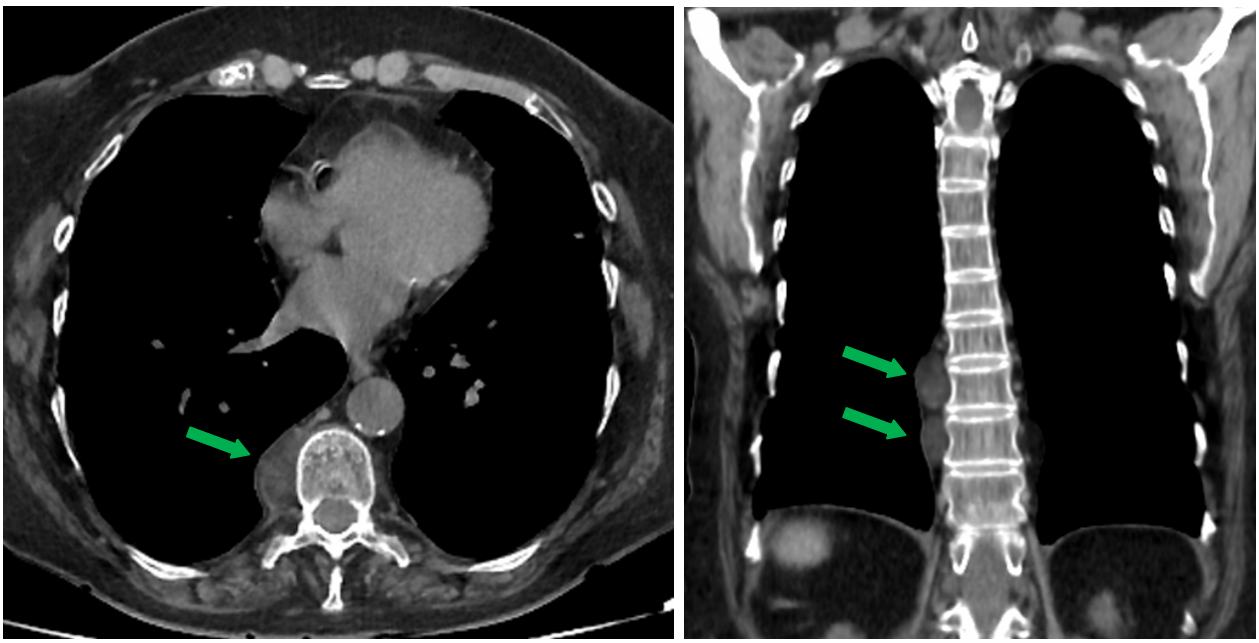


Fig. 3: High-resolution CT scan. A axial. B. coronal. Paravertebral masses, well circunscripte, with fat density, compatible with lymphangioleiomyomas (green arrows).

II. Discussion

Lymphangioleiomyomatosis (LAM) is a rare disorder of unknown cause characterized by peribronchial, perivascular, and perilymphatic proliferation of abnormal smooth muscle cells (1) leading to progressive cystic destruction of the lung, lymphatic abnormalities and abdominal tumors (2).

It occurs almost exclusively in women of fertile age due to a hormonal influence, for this reason it is extremely rare in post-menopausal patients. In these cases it is usually associated with hormone replacement therapies. It is known that this disease is strongly associated with other conditions, such as tuberous sclerosis and renal angiomyolipomas (3).

In the cases of older women the disease presents a similar clinical manifestation than younger's female, with the exception that the clinical course is benign and longer after menopause (3).

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a) Physiopathology

Pathologically, LAM cells contain proteins related to melanoma, including glycoprotein 100, the HMB-45 and receptors for estrogens and progesterone. These cells have a high proliferation capacity, damaging the airway through the production of metalloproteinases with the obstructing capillaries, lymphatics and the airway. Lymphatic obstruction usually causes chylothorax and airway obstruction conditions an obstructive functional pattern. A 30% -40% of cases present pneumothorax, which can be bilateral and recurrent (4).

b) Pulmonary Manifestations

While LAM is a systemic disease, the main manifestations are pulmonary. In lungs, the atypical progressive proliferation is seen around small airways giving rise to cystic pulmonary changes and progressive respiratory failure (5).

The pulmonary symptoms tend to dominate the clinical course, with the most common features being pneumothorax, progressive dyspnoea and chylous pleural effusions .Other respiratory symptoms include cough, haemoptysis and chyloptysis. Dyspnea is suffered by the vast majority of patients with LAM and is the result of airflow obstruction and replacement of the lung parenchyma by cysts. Approximately two thirds of patients will have a pneumothorax at some point in their clinical course, which are recurrent in most and a cause of significant morbidity. Chylous pleural effusions are less common, but again can be difficult to treat conservatively and tend to recur after simple aspiration. Haemoptysis and chyloptysis are due to LAM cell obstruction of pulmonary capillaries and lymphatics, respectively, and both occur in a small number of patients (2).

The chest radiograph often appears normal in early disease, although it may show pleural effusions or pneumothorax (2) in a 40% de los pacientes (7).

The main pulmonary finding includes cysts these are a thin-walled (usually <3 mm), well-defined and circumscribed, air- or fluid-containing lesion 1 cm or more in diameter, nevertheless, some do not have walls and others have an irregular configuration (7). Cysts are scattered in a bilateral roughly symmetric pattern, without any lobar predominance. These cysts range in size from barely perceptible to several centimeters and in number from a few scattered cysts to near complete replacement of the lung parenchyma (2) with increased lung volume (7).

c) Extrapulmonary manifestations

Extrapulmonary manifestations of LAM are angiomyolipoma, which occur mostly in the kidneys, chylous ascites, abdominal lymphadenopathy and large cystic lymphatic masses termed lymphangioleiomyomas (2). Lymphangioleiomyomas are larger cystic masses; these most commonly occur in the abdomen, retroperitoneum and pelvis but occasionally in the mediastinum and neck. Symptoms associated with lymphangioleiomyomas are nausea, bloating, abdominal distension, peripheral oedema and urinary symptoms (6).

Abdominal findings may be present in more than 70% of patients with LAM, and the most common abdominal finding is renal angiomyolipoma. These are often small (<1 cm), multiple, bilateral, and asymptomatic (7) and can usually be identified by CT because of the presence of fat, which gives lesions a characteristic CT appearance. MRI may be adequate for the diagnosis when iodinated contrast is contraindicated. Diagnostic difficulty may arise in the small number of angiomyolipomas showing little evidence of fat (2).

Lymphangioleiomyomas are produced by a proliferation of smooth muscle cells in the lymph vessels with the production of cystic masses consistent with dilatations of the abdominal lymph vessels due to lymphatic obstruction (7). Lymphangioleiomyomas are usually localized in the mediastinum, retroperitoneum, and pelvis along the axial lymphatics and can appear larger in the evening due to accumulation of chyle in the cystic structures (2).

Enlarged lymph nodes are described in up to 40% of cases and Chylous Ascites is an unusual abdominal complication of LAM that can occur in the absence of pleural effusion (7).

d) Imaging Diagnosis

High-resolution CT (HRCT) is the recommended imaging technique for LAM. The diagnosis of LAM is probable when a characteristic lung CT is found in a patient with compatible clinical history or when compatible CT features are present in a patient with angiomyolipoma or chylous effusion (2).

e) Differential diagnoses

Within the main differential diagnoses we mention tuberous sclerosis, pulmonary Langerhans cell histiocytosis, emphysema, and pulmonary fibrosis (8).

f) Treatment

The treatment of this pathology is based on treating complications. Hormone treatment should be discouraged except in individual cases with rapid progression of the disease in which progesterone may be trialled (2). The first successful lung transplantation for LAM was performed in 1983. However, is unclear whether lung transplantation actually improves long-term survival in patients with LAM compared with continued medical management (8).

III. Conclusion

LAM is a systemic disease, the main manifestations are pulmonary. The extrapulmonary

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manifestations are unusual. The clinical course is benign and longer in post-menopausal women.

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