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From Pulmonary Cystic Lesions to Tuberous Sclerosis Syndrome

S. Allioui, L. Belkouchi, N. Moatassimbillah and I. Nassar

Department of radiology. Ibn Sina Hospital. Mohammed V University. Rabat. Morrocco.

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ABSTRACT

LAM or Pulmonary lymphangioleiomyomatosis is an uncommon disease that predominantly touches women in their reproductive years. The disease is usually discovered in an incidentally way and detected in a high resolution thoracic CT scan. Its progress could lead to respiratory failure. This article is about a patient in our ward that got a chest CT scan in which the final diagnosis was a lymphangioleiomyomatosis in a case of tuberous sclerosis complex.

Introduction

Lymphangioleiomyomatosis is a rare disease that embodies a proliferation of smooth muscle-like cells leading to cystic destruction of the lung and at times on life respiratory collapse. LAM affects sporadically or with patients with tuberous sclerosis complex.

Case Description

Of no medical history record, a 29 year old female, was introduced showing a progressive dyspnea and productive cough. The carried chest CT scan showed; multiple round shaped well defined, thin walled cysts, diffusely distributed through the lungs, their diameters varied between 2 to 18mm with absence of any other pulmonary lesion.

In the upper abdominal area viewed on the CT scan uncharacterized kidney lesions were noted, thus we completed with a performed portal phase abdominal CT scan that showed bilateral fat containing kidney lesions defined as angiomyolipoma and the existence of osteosclerosis in the bone window.

While discussing the case with her doctors, they revealed the presence of achromic skin lesions and neurological symptoms, which required a brain MRI that showed milimetric lesions with high signal intensity in T1, T2 and Flair sequence, known as tubers, residing in the right parasagittal and insular cortex, and in the left frontal, parietal, rolandic and occipital areas.

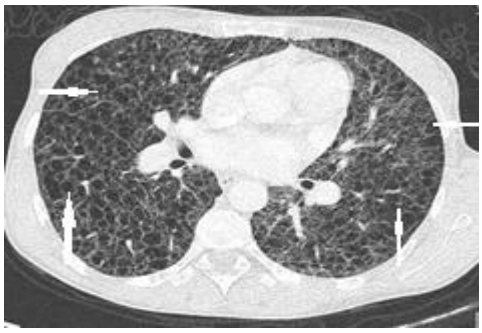

Figure1. A high resolution thoracic CT scan showed multiple well defined, thin walled, cystic lesions, with a size range of 2-5mm diffusely distributed throughout the lungs, with no preferential area or any other pulmonary lesions noted.

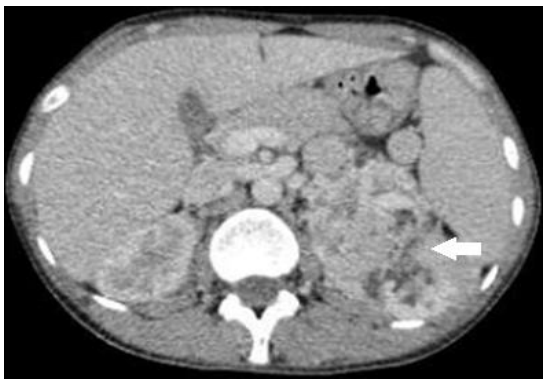

Figure 2 . An axial view of the abdomen in a portal venous phase CT showed kidney lesions containing fat that corresponds to angiomyolipoma.

Tele:

E-mail address: allioui.sou@gmail.com

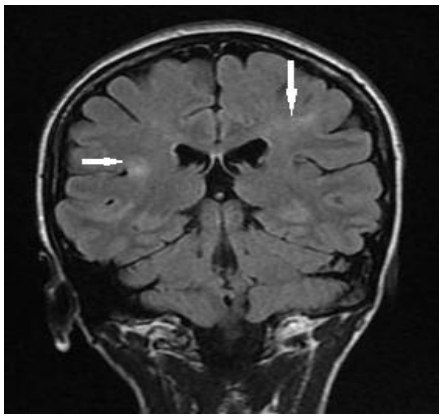

Figure 3 . A Brain MRI showed milimetric lesions with increased signal intensity in FLAIR known as tubers, residing in the right insular area, and the left parietal area.

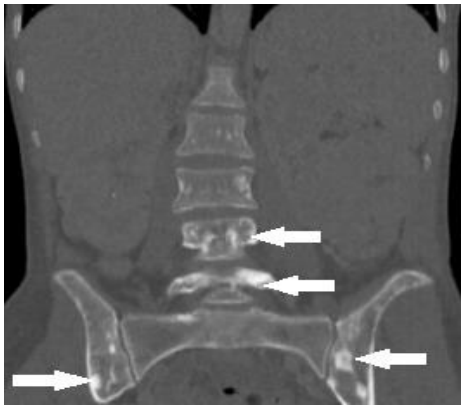

Figure 4. A coronal view of the bone window showed osteosclerosis bone lesions

Discussion

Cystic pulmonary diseases have many common differential diagnoses such as emphysema, bronchiectasis and honeycombing. The cyst has a fine wall with no centrolobular artery, on the opposite of emphysema that appears as focal licences without a wall determining them (1). Honey combing appears as clustered cystic air spaces between 3-10 mm in diameter (2). Bronchiectasis can be seen as a stretch of bronchial tree that are often subpleural in distribution (3). Concerning unique cysts, it is crucial to distinguish between cysts and cavity lesions; the latter are thick walled (over2mm) (2).

The analysis of the cystic lesions in terms of size, number, wall thickness, and distribution in the lung is substantial in differential diagnoses, which are:

Histiocytosis : Smokers are subject to langerhans cell histiocytosis. The disease is due to peribronchial epithelium and progressive dilation of bronchiales resulting in cysts that have irregular outlines. These originally are nodules that become excaved then turn into thick walled cysts, then to thin ones. It is quite unusual to find isolated nodules. They are mainly related to cysts and often are located in the upper and mid pulmonary lobes (4).

Lymphocytic interstitial pneumonia (lip) : It affects people at the age of 50 and on. It is caused by hyperplasia of peri bronchial lymphoid cells, resulting in an abnormal proliferation of smooth muscle, leading to air trapping and cystic change. It appears in a chest CT as a few cysts sized between 2 to 5 cm with high ground glass densities mostly found in the middle area of the lung (sparing sub pleural area) (5). The disease is mostly associated with systemic and autoimmune diseases such as Gougerot-Sjögren's syndrome.

Pneumocystis : It basically affects patients with immune deficiency. With the help of chest imaging, we can see isolated or multiple thin walled cysts, of varying sizes, surrounded by high ground glass densities mostly focused in the upper area of the lung (6).

Metastasis : Based on Imaging we can see the existence of thick walled cavities or fine walled cysts. The cysts can naturally be formed or changed after chemotherapy.

Male patients with ORL cancer, female genital cancer, kidney cancer or bone sarcoma are the most vulnerable. This diagnosis should not be considered unless there is a collapse of general condition or a suspicious related lesion. (7)

Lymphangioleiomyomatosis : Is a result of abnormal proliferation of smooth muscle, responsible of bronchiolar obstruction, air trapping, therefore a hyperinflated lung leading to cyst creation (8). The cysts can cause a pneumothorax if found in the pleural cavity.also, the smooth muscle proliferation can occasionally cause lymphatic obstruction resulting in chylothorax or pulmonary oedema or hemorrage if the venules are obstructed. This disease affects mainly females, during their reproductive years, and is characterized by several thin walled cysts (>10) trapping air with varied sizes (from 2-5mm to 30 mm). These cysts are diffusely distributed throughout the lungs, with no preferential or spared area. (9, 10, 11)

Our patient had similar pulmonary lesions, associated with kidney angiomyolipoma, osteosclerosis, skin lesions and brain MRI signs of tuberous sclerosis complex, which was the confirmed diagnosis.

Tuberous sclerosis complex (TSC) is a phakomatosis and a rare autosomal dominant disorder because of a mutation in the TSC 1 (hamartin) and TSC 2 (tuberin) genes. Hence we get a hamartoma formation. More clearly, we get astrocytic hamartoma as primary lesion. The percentage of TSC in the lung damage identical to LAM is 1%.

Cerebral lesions of TSC entails subependymal nodules, Cortical Dysplasia or hamartomas (tubers), Pachygyria, or finally huge cell astrocytoma that is definite but not a common ventricular tumor that takes place mainly in the foramen of Monro (12). TSC may touch other organs like the eyes (Retinal phakoma or astrocytic hamartomas), the skin (facial angiofibromas, Adenoma sebaceum of Pringle, Koenen's periungual fibroma),.the kidneys (angiomyolipoma and cystic kidneys), the liver(hamartomas and adenomas), the lungs (Lymphangioleiomyomatosis 1%), the heart (Cardiac rhabdomyoma in new borns), bone and vessels are similary subject to TSC. (13, 14, 15)

TSC's treatment is mainly mostly symptomatic like: bronchodilators, antiepileptic drugs and treating its complications. Therefore, there is no room for cure.

Conclusion

LAM should be considered in every young female patient that consults for dyspnea, hense a high resolution chest CT scan should be performed. Female patients presenting pulmonary cystic lesions must benefit of a full medical examination with an abdominal CT scan, in order to find other lesions that could be in favour of a TSC instead of a sporadic LAM. The diagnosis of STB is rare, but the characteristic imaging could help start a treatment and improve the patient's vital prognosis.

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