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Pulmonary Alveolar Microlithiasis: A Case Report

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Abstract: Pulmonary alveolar microlithiasis is a rare diffuse lung disease characterized by deposition of calcium phosphate within the alveolar airspaces. The disease is usually discovered from birth up to 40 yrs of age and is often diagnosed incidentally during radiography of the chest for other reasons. Many patients are asymptomatic and the majority of patients either have normal or restrictive pulmonary function. The clinical course of the disease varies. While it remains static in some patients, it progresses into pulmonary fibrosis, respiratory failure and cor pulmonale in others With the exception of lung transplantation, there is no known effective treatment for the disease. Although the etiology remains unclear, mutations of the solute carrier family 34 (sodium phosphate) member 2 gene (the SLC34A2 gene), which encodes a sodium/phosphate cotransporter, are considered to be the cause of the disease. We present a case report of pulmonary alveolar microlithiasis and its clinical, radiological and histological features.
Keywords: Alveolar microlithiasis, genetic polymorphisms, type IIb sodium dependent phosphate co-transporter

INTRODUCTION

Pulmonary alveolar microlithiasis (PAM) is a rare autosomal recessive lung disease characterized by calcifications within the alveoli in the absence of known calcium metabolism disorders. Most of the patients are asymptomatic with female sex preponderance, usually detected incidentally during routine examinations [1]. The Mutations in the SLC34A2 gene, which encodes a type IIb sodium phosphate co-transporter, are responsible for this disease, leading to intra-alveolar accumulation of phosphate that favors the formation of microliths. The hallmark of this disorder is clinical-radiological dissociation, with typical imaging findings that correlate well with specific pathological findings. The long-term prognosis is poor and no treatment has been discovered to date [2].

The aim of this review is to describe the main pathological, clinical, and imaging aspects of PAM.

CASE REPORT

A 65- year old female patient presented with the chronic history of breathlessness on exertion since 9 months which was insidious in onset and progressive in nature associated with cough with white scanty sputum. Breathlessness was initially of grade 2(NYHA) now progressed to grade 4(NYHA) due to which she became bed ridden for past one month. There is no history of fever or chest pain.

On examination

The patient is tachypneic (30 cycles/min) with the use of accessory respiratory muscles.

Pulse – 102 bpm	pallor -absent
BP- 116/70 mmHg	icterus -absent
RR – 30 cycles/min	cyanosis -absent
Temp- 98.6 F	clubbing -grade 3
Spo2 90%	at room air.

JVP raised 5cm from sternal angle
Weight 48kg

Systemic examination shows signs of right heart failure with pulmonary artery hypertension are present with diffuse inspiratory crackles on auscultation.

Chest x ray

A Plain chest radiograph reveals diffuse, scattered, bilateral areas of micronodular calcifications, producing a "sandstorm" appearance (Fig. 2) that first involves the inferior portions and then the middle and upper portions of the lungs. The distribution of calcified nodules can also be explained by the increased blood supply to these areas. The lung bases show increased density due to thicker lung tissue and increased surface densities. The calcifications may be so dense that may obliterate the cardiac borders, diaphragm [3].

High-resolution computed tomography (HRCT)

Extensive calcification is noted in bilateral lung fields predominantly along subpleural region,

fissures and parenchyma of bilateral lung fields predominantly in apical segment of bilateral upper lobes and in all segments of lower lobes. Thickening of pleura and fissures with calcification is noted. Ground glass opacity with interstitial septal thickening (crazy paving pattern) is noted in bilateral lung fields. Multiple small nodules are noted along thickened interlobular septa in bilateral lung fields. There is extensive calcification in bilateral lung fields with thickening of pleura and fissures. Subpleural cysts/emphysematous changes in bilateral lung fields. Bronchiectasis in bilateral apical segments is seen.

Bronchoalveolar lavage (BAL) fluid examination showed no microliths. Histopathological examination of a transbronchial lung biopsy revealed numerous concentric calcified concretions. The histopathology and the typical radiological picture on HRCT confirmed the final diagnosis of PAM.

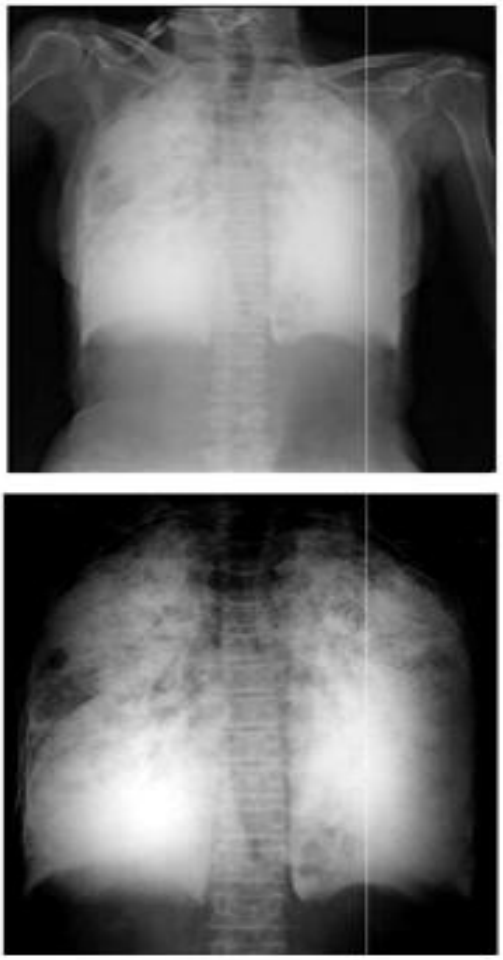

Fig-1: Diffuse, scattered, bilateral areas of micronodular calcifications, producing a "sandstorm" appearance involving the inferior portions and then the middle and upper portions of the lungs

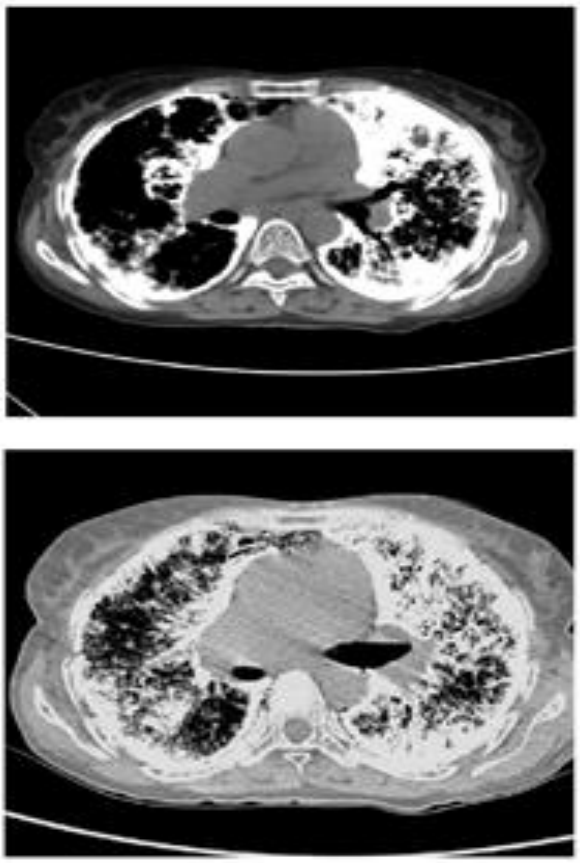

Fig-2: Extensive calcification is noted in bilateral lung fields predominantly along subpleural regions and parenchyma of bilateral lung fields predominantly in apical segment of bilateral upper lobes.Ground glass opacity with interstitial septal thickening (crazy paving pattern) is noted in bilateral lung fields

Diagnosis

PAM is usually diagnosed on the basis of a typical radiological pattern, namely a very fine, sand-like micronodulation of calcific density diffusely involving both lungs, with basal predominance. Many authors argue that this pattern precludes the need for a lung biopsy in most cases [4]. However, histological examination shows intraalveolar microliths in the alveolar spaces and can reveal an increase in chronic inflammatory cells that may induce interstitial fibrosis. Microliths may also be seen in the interstitium. Microliths can sometimes be found in sputum or bronchoalveolar lavage, which can contain alveolar macrophages with or without carbon particles [5]. Calcifications within the lung can result from a number of systemic and pulmonary conditions, and the differential diagnosis is complex, including sarcoidosis, pneumoconiosis, pulmonary hemosiderosis, amyloidosis, miliary tuberculosis, histoplasmosis, calcifications after viral pneumonia, and metastatic pulmonary calcifications associated with chronic renal failure [6]. After PAM is diagnosed in a given patient, family members should be screened by chest

radiography, and parents should be counseled that future children are also at risk of developing the disease [3].

CONCLUSION

PAM is a genetic lung disease with an autosomal-recessive trait caused by mutations of the SLC34A2 gene. Microlith formation is the result of phosphate-chelating calcium in the extracellular fluid. The disease shows an important clinical radiological dissociation. Most patients are asymptomatic and changes in the lung parenchyma are incidental findings, but other cases exhibit more severe symptoms at diagnosis. Because characteristic chest CT findings correlate well with specific pathological findings, the diagnosis of PAM can be made on the basis of the typical radiological pattern. The long-term prognosis is poor due to progressive deteriorations in pulmonary function, respiratory failure and, cor-pulmonale. No effective treatment has been established; thus, lung transplantation is currently the only effective therapy.

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