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RESEARCH ARTICLE

PULMONARY ALVEOLAR MICROLITHIASIS- A CASE REPORT

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ABSTRACT

Pulmonary Alveolar Microlithiasis is a rare and chronic disorder of respiratory system of unknown etiology which is characterised by widespread calcifications within the alveoli. We present a case of 65 year female who presented with cough with expectoration since 2 months and Grade III breathlessness. On auscultation there was presence of random wheezes and crackles predominantly in lower lung zones bilaterally. On chest radiograph finding were diffuse, discrete calcifications in both lung fields along with pleural calcification bilaterally. High Resolution Computed Tomography (HRCT) findings were extensive calcification in bilateral lung fields, thickening of pleura and fissures, multiple small nodules along thickened interlobular septa, ground glass opacity with interstitial septal thickening (crazy paving pattern) and few subpleural cysts in bilateral lung fields.

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INTRODUCTION

Pulmonary Alveolar Microlithiasis is a rare and chronic lung disorder (VoIle and Kaufmann, 1987) which is characterised with calcium deposition within the alveoli of both lungs. The etiology and pathology are still clearly not known (Spencer, 1984) . Some authors have described role of mutation in type IIb sodium-phosphate cotransporter gene (SCL34A2 gene) in its pathogenesis (Corut *et al*., 2006). The disease is an autosomal recessive derivation predominantly affecting females (VoIle and Kaufmann, 1987; Spencer, 1984). We herein, report a case of 65 year old housewife female who had chest radiograph findings with suspicion towards Pulmonary alveolar microlithiasis which was confirmed by High Resolution Computed Tomography and histopathological correlation.

CASE REPORT

A 65 years old female came with complaints of cough with expectoration since 2 months and grade III breathlessness. On auscultation the respirator sounds were reduced bilaterally along with presence of wheezes and crackles predominantly in lower lung zones bilaterally.

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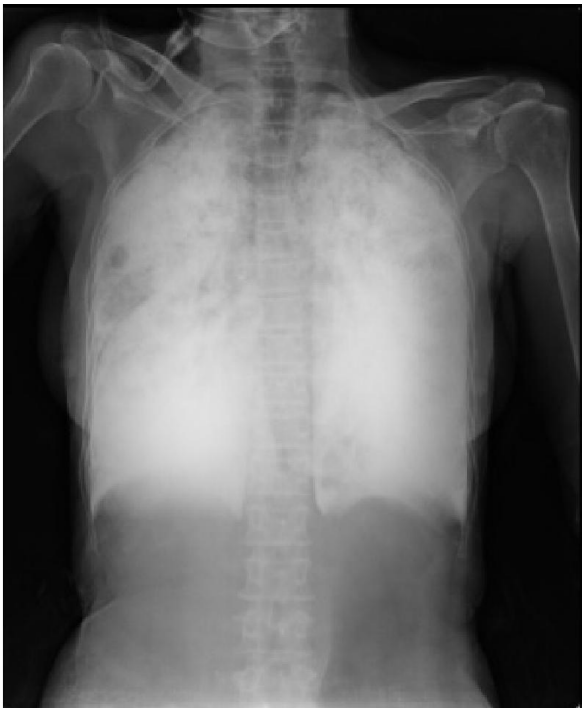
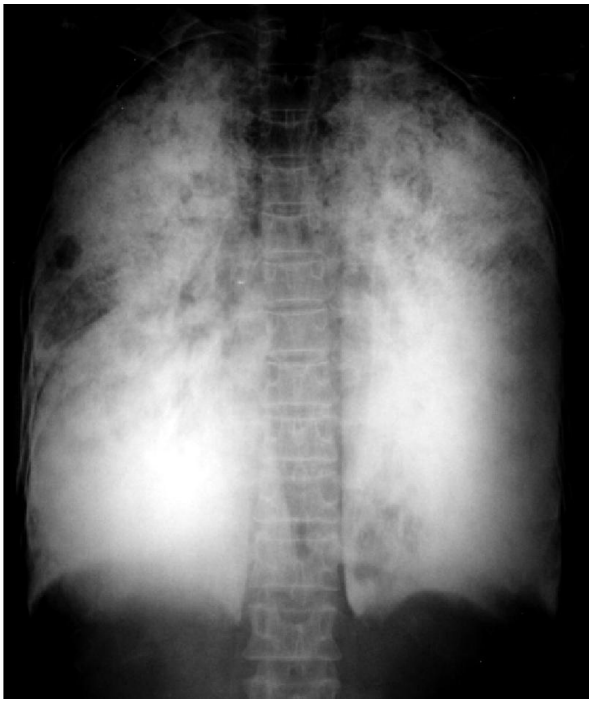
On chest radiograph findings were diffuse, discrete calcifications in both lung fields along with pleural calcification bilaterally. On High Resolution Computed Tomography findings were extensive calcification in bilateral lung fields, thickening of pleura and fissures, multiple small nodules along thickened interlobular septa, ground glass opacity with interstitial septal thickening (crazy paving pattern) and few subpleural cysts in bilateral lung fields. Biopsy was taken from apical part of lung and histopathological findings showed presence of concentrically laminated, partially calcified spheroid bodies known as calcipherites within the alveoli of lung.

DISCUSSION

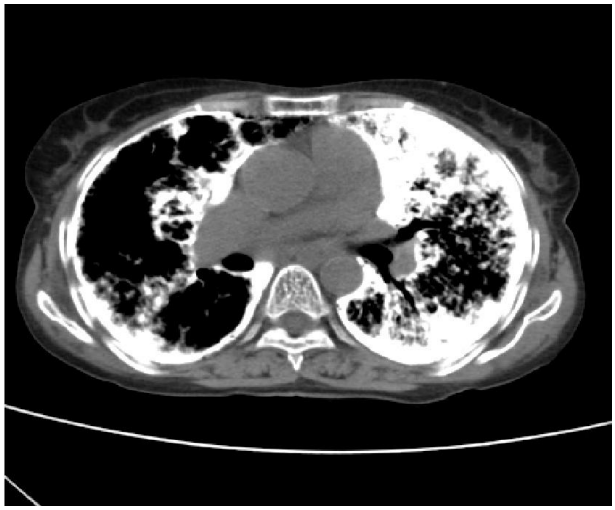
Pulmonary Alveolar Microlithiasis is uncommon disease characterised by widespread localisation of 'calcipherites' that is salts of calcium and phosphate within the alveoli without presence of any calcium metabolic disorder (Moran *et al*., 1997). Incidence of this disease is prevalent worldwide, however approximately 25% of cases reported from Turkey and having almost equal male and female sex predilection (Shah *et al*., 2003). Mostly, patients affected with this disease are asymptomatic at the initial phase of the disease and only become symptomatic as the disease advances. Cough and dyspnea on exertion are the common.

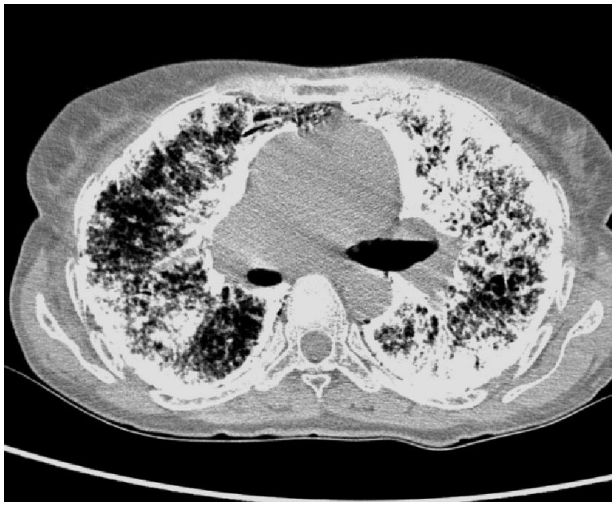




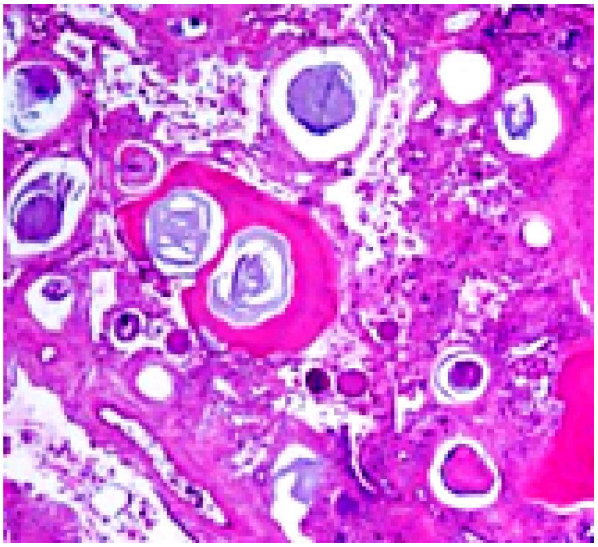
Plain chest radiograph reveals areas of microcalcifications bilateral lung fields. The cardiac borders are obliterated

Early symptoms, however in the later course of the disease - respiratory insufficiency, cor pulmonale, and even death may occur (Barbolini *et al*., 2002). On chest radiograph, numerous sand-like microliths or calcipherites are seen diffusely scattered in bilateral lung fields and may obscure the diaphragmatic, mediastinal, and cardiac borders. The propensity of the disease for the lung bases is higher due to the larger volume of the lower lobes. A 'lack pleural line' may be seen in bullae in the lung apices, a zone of hyperlucency between the lung parenchyma and the ribs. Calcifications of pleura may also be seen.





High Resolution Computed Tomography show diffuse and confluent calcified nodules with septal thickening and subpleural calcification. Pleural calcification and small calcispherites within the thickened pleura



Photograph showing multiple calcipherites within the alveoli (H & E, 100x)

Occasionally, reticulations and septal lines may be seen on chest radiograph (Marchiori *et al*., 2007). High-resolution CT with thin-section acquisitions and high-spatial-frequency reconstruction algorithms is preferred for the evaluation of pulmonary alveolar microlithiasis because it allows detection of minimal structural changes of the lung parenchyma that are not optimally evaluated with radiography or with other CT techniques (Korn *et al*., 1992).

CT scan reveals diffuse ground-glass opacities in both lung fileds. It may be associated with confluent and diffuse calcified nodules. It also confirms the predominance of affected zones being symmetrical and at the middle and the lower zones. Calcifications is also seen along the bronchovascular bundles and central bronchovascular tree (Helbich *et al*., 1997). There is no current medical treatment for Pulmonary Alveolar Microlithiasis. If the disease prolongs to advance the stages the only option left is lung transplantation (Tachibana *et al*., 2009). The important differential diagnosis to be considered for this disease includes tuberculosis, silicoproteinosis and amyloidosis as nodular calcifications may also be found in this conditions. In our case the patient was house wife and had no significant family history showing negative relationship of the disease with occupation or family history.

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