Pulmonary Alveolar Microlithiasis

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ABSTRACT

Pulmonary alveolar microlithiasis (PAM) is a rare disorder. It is characterized by accumulation of calcium phosphate (microlith) within the alveoli. Knowledge of this condition is important because radiologically it mimics miliary tuberculosis that can lead to the wrong treatment.

Key words: Microlithiasis, crazy pavement, calcispherytes.

INTRODUCTION

Pulmonary alveolar microlithiasis (PAM) is an autosomal recessive disorder, for which mutation in the SLC34A2 gene was recently found to be responsible for the disease.[1] It has some peculiar characteristics including the formation of numerous tiny stone-like structures "CALCISPHERYTES" within the alveoli. Five hundred seventy six cases have been reported up to 2004[2], most of them came from Europe (42.7%) and Asia (40.6%). The countries involved were fifty-one and twelve of them were attributed with at least ten cases each (Bulgaria, France, Germany, India, Italy, Poland, Spain, Russia, Japan, Turkey, USA and ex-Yugoslavia). Only few cases have been reported from Pakistan.[3-5]

CASE REPORT:

A 16 years old boy was admitted in the hospital with complaints of chest pain and shortness of breath for two years. Initially he noticed these symptoms while playing cricket. The shortness of breath has progressed slightly and now he is breathless on walking one flight of stair. He was comfortable while at rest. He was prescribed anti-tuberculosis treatment, which he took for nine months without any improvement. His clinical examination was unremarkable. The chest radiograph [Fig1] showed diffuse bilateral micronodular, calcified shadows involving whole lung fields sparing the extreme apices. These nodular opacities were sharply defined and discrete. This picture is characteristically called "SAND STORM" pattern or "DESERT LUNG". HRCT was done [Fig 2], which showed presence of septal thickening and calcification along with intra-alveolar calcifications called calcispherytes. This pattern is called as "CRAZY PAVEMENT". His spirometry revealed mild restrictive pattern. Lung biopsy was done using trucut biopsy needle, which showed presence of multiple laminated structures within the alveoli called calcispherytes [Fig.3]. As part of treatment we did the whole Lung Lavage followed by Alendronate sodium 70 mg weekly. At three month follow-up, patient was feeling better subjectively though there was no improvement in spirometric values.

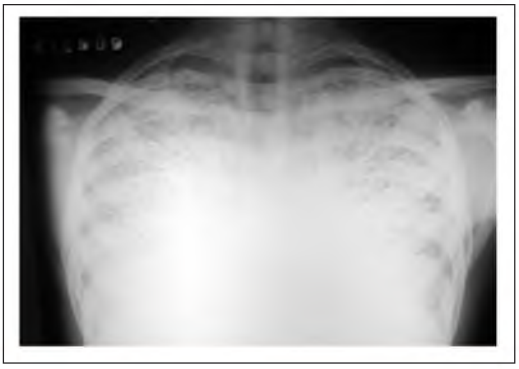

Figure – 1 : Sand storm pattern showing nodular opacities involving the whole lung field relatively sparing extreme apices.

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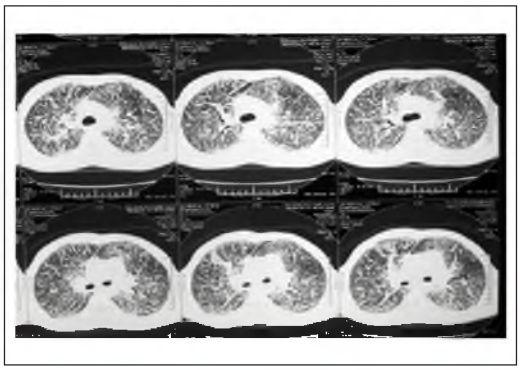

Figure – 2 : HRCT showing CRAZY PAVEMENT pattern with septal thickeningand calcifications.

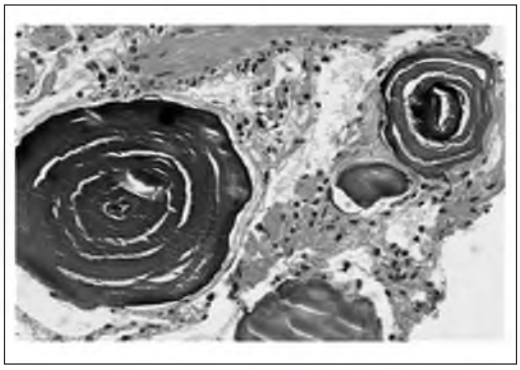

Figure – 2 :.Lung biopsy showing laminated structures (calcispherytes) within the alveoli and chronic inflammatory cells surrounding the alveoli.

DISCUSSION

Friederich[6] first described pulmonary alveolar microlithiasis (PAM) in 1856 as "Corpora-Amylacea in den lungen". Some investigators claim that Harbitz described it first in 1918.[7] Later in 1933, Puhr[8] named it and used the term 'Microlithiasis alveolaris pulmonum'. PAM is associated with formation of 'calcispherytes' (lamellar concretions of calcium/ microlith) within the alveoli. Microlith has been reported in other tissues like gonads, prostate, kidneys and sympathetic chain.[9]

The mechanism of formation of calcipherytes is not known. It is suggested that it is caused by inborn error in metabolism at alveolar level which leads to alkalinity / increased mucopolysacchride deposition, promoting local calcium accumulation. In these patients studies of calcium metabolism are normal so the deranged calcium metabolism is unlikely cause of PAM.[10,11] The patients suffering from Pulmonary alveolar microlithiasis (PAM) are usually asymptomatic for many years and are diagnosed incidentally if their chest x-rays are done for other reason. The symptoms usually develop in third or fourth decades in the form of slowly progressive dyspnea and dry cough.[12,13] These patients commonly show progressive deterioration of the pulmonary function and they die in their midlife due to respiratory failure / corpulmonale.[14] Pneumothorax can be observed in the early course of the disease.[14] Restrictive pattern is the usual pattern seen on spirometry.[11]

Plain chest radiographs usually reveal bilateral, diffuse micronodular calcifications ("sand storm"), involving predominantly mid and lower lung fields.[11, 12] The lung bases appear denser owing to the greater thickness of lung tissue in these areas, as well as the increased surface densities.[15] Regardless of the effect of superimposition or summation of shadows, individual deposits are usually identifiable particularly with magnification roentgenography. Very sharply defined, they measure less than 1 mm in diameter and are discrete.[16] The heart borders and the diaphragm are usually obliterated. Other typical findings include small apical bullae and a black pleural line, which is demonstrated as an area of increased translucence between the lung parenchyma and the ribs.[17]

The chest radiographs of our patient showed similar pattern of diffuse symmetric, dense micronodular lung lesion.

The CT scan usually demonstrates[12,15,17] diffuse micronodular calcified nodules involving predominantly middle and lower zones. They are more marked in the subpleural region and along the bronchovascular bundle. A predominance of calcifications in the medial areas when compared with the lateral portions of the lungs is also evidenced in the CT scan. High resolution CT scans may reveal small cysts in the subpleural lung parenchyma, pleural calcification and small calcispheryte, within the thickened pleura.

Murch and Carr[18] described the crazy-paving pattern (scattered or diffuse ground-glass attenuation with superimposed interlobular septal thickening and intralobular lines) as characteristic of pulmonary alveolar proteinosis but this finding has been described in several other conditions like PAM, ARDS (Acute respiratory distress syndrome), *Pneumocystis carinii* pneumonia, mucinous bronchioloalveolar carcinoma, sarcoidosis, nonspecific interstitial pneumonia, organizing pneumonia, exogenous lipoid pneumonia and pulmonary hemorrhage syndromes.[19,20]

Serum levels of surfactant proteins A and D correlate with the progression of the disease, and may be a useful monitoring tool.[21] At present, no effective treatment is available. Therapeutic modalities including systemic corticosteroids, calcium-chelating agents, and bronchopulmonary lavage have been shown to be ineffective.[22] Lung transplant is being performed in severe cases.[23]

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