Case Report

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Pulmonary Alveolar Microlithiasis

Radiology Section

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ABSTRACT

Pulmonary alveolar microlithiasis (PAM) is a rare chronic disease characterized by the presence of small calculi in the alveolar space. We report a case of a 35-year-old man with a 4-days history of cough with expectoration and shortness of breath on exertion. Auscultation revealed random wheezes and fine & coarse crackles. Pulmonary function tests were normal. The chest radiograph demonstrated discrete dense nodular opacities scattered throughout the both lung fields, predominantly involving middle and lower lung zones, with pleural calcification. HRCT demonstrated discrete dense nodular opacities involving all the segments of both lung parenchymas with thin walled bullae in the apical segment of left upper lobe.

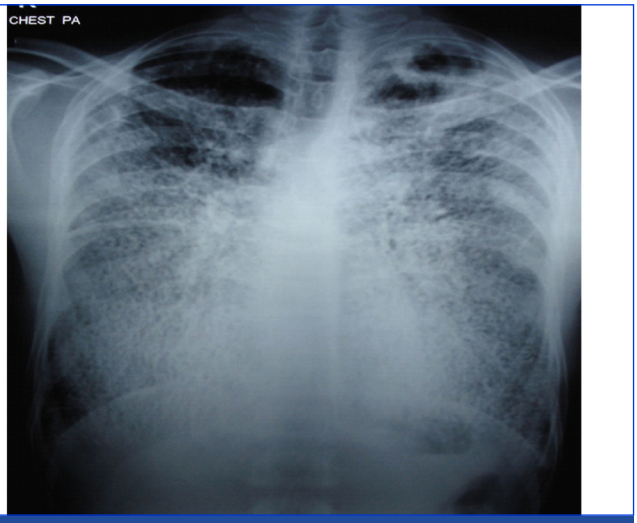
Key Words: Coeliac trunk, Left gastric artery, Cystic artery, Accessory hepatic artery, Inferior phrenic artery,

INTRODUCTION

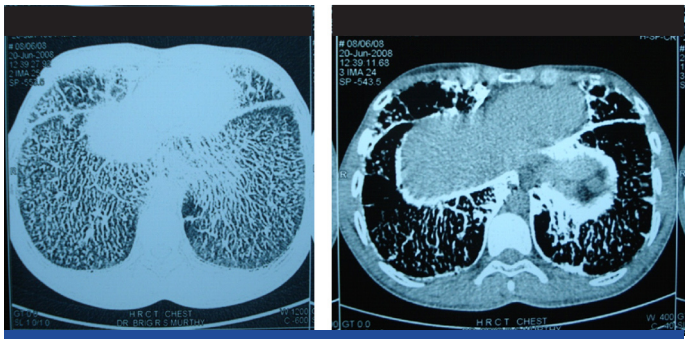
Pulmonary alveolar microlithiasis is a rare disease entity that is characterized by innumerable diffuse calcospherites (microliths which are composed of calcium and phosphorus) within the alveolar space [1], and a paucity of symptoms, which are in contrast to the imaging findings. It occurs sporadically and it is regarded as an autosomal recessive lung disease [2].

CASE REPORT

We are presenting here a 35-year-old male, a driver by occupation, who came to us with the complaints of cough with expectoration and shortness of breath on exertion, of 4 days duration. The lung auscultation revealed random wheezes and fine and coarse crackles. The pulmonary function tests were normal. The chest radiograph demonstrated discrete dense nodular opacities which were scattered throughout both the lung fields, which predominantly involved the middle and the lower lung zones, with pleural calcification. HRCT demonstrated discrete, dense, nodular opacities which involved all the segments of both the lung parenchymas, with thin walled bullae in the apical segment of the left upper lobe.



[Table/Fig-1]: Plain chest radiograph reveals diffuse, scattered, bilateral areas of micro nodular calcifications ("sand storm") that predominate in the middle and lower lung areas. The heart borders and the diaphragm are obliterated.



[Table/Fig-2]: CT scan thorax reveals diffuse and confluent calcified nodules with septal thickening and subpleural calcification. Pleural calcification and small calcispherites within the thickened pleura.

DISCUSSION

Pulmonary alveolar microlithiasis is characterized by the presence of widespread laminated calcospherites in the alveolar spaces, in the absence of any known disorder of calcium metabolism [3]. The patients may remain asymptomatic for many years and they may usually become symptomatic between the third and fourth decades of life [4]. The clinical presentation may usually demonstrate a lung disorder with a restrictive pattern [5]. A pneumothorax can be observed in the early course of the disease. The adult patients commonly show a progressive deterioration of the pulmonary functions and death usually occurs in mid-life because of respiratory failure which is associated with corpulmonale.

The plain chest radiographs usually reveal diffuse, scattered, bilateral areas of micronodular calcifications ("sand storm"), that predominate in the middle and the lower lung areas [6]. The lung bases appear to be increased in density, owing to the greater thickness of the lung tissue in these areas, as well as the increased surface densities. The distribution of the calcified nodules can also be explained by the relatively higher blood supply to this area. The heart borders and the diaphragm are usually obliterated. The 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 [4]. The chest radiograph of our patient showed diffuse, symmetric lung lesions with a dense micronodular pattern, which correlated with the pattern which has been mentioned in the literature.

CT scan usually reveals diffuse ground-glass opacities throughout both the lungs, which are associated with confluent and diffuse calcified nodules. This imaging technique confirms the predominance of symmetric abnormalities at the middle and the lower zones. Calcifications can also be seen along the bronchovascular bundles and at the central region of the bronchovascular tree [2]. High resolution CT scans may reveal small cysts in the subpleural lung parenchyma, pleural calcification and small calcospherites within the thickened pleura. Some of these findings were observed in our case, whose CT demonstrated a septal thickening with calcified nodules, as well as a subpleural calcification.

The clinical consideration of a differential diagnosis is essential, as nodular calcifications can also be found in other diseases like tuberculosis, metastatic osteosarcoma, amyloidosis and silicoproteinosis.

This disease takes a chronic, progressively deteriorating course. It is resistant to all the therapeutic measures and no effective treatments are available as yet.

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