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Pulmonary Alveolar Microlithiasis in Siblings

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Pulmonary alveolar microlithiasis is a rare disease of unknown etiology, in which calcium phosphate crystals are deposited throughout the lungs. We report 2 cases of pulmonary alveolar microlithiasis occurring in siblings.

Case Reports

Case 1: A 11-year-old male child presented with symptoms of increasing weight gain for the past 6 months. A routine chest *X*-ray (*Fig. 1*) showed bilateral diffuse miliary like mottling of both lung fields, almost obliterating the cardiac shilhouette. The radiological picture was suggestive of a diffuse interstitial lung disease and possibilities of miliary tuberculosis, pulmonary alveolar proteinosis, fibrosing alveolitis, pulmonary hemosiderosis and pulmonary alveolar microlithiasis were considered. Careful examination of the cardiovascular, respiratory and other systems did not reveal any significant findings. There was no family history of any respiratory problems except that the child's grandfather had died of 'asthma' at 60 years of age.

On investigations hemogram and routine investigations were normal; serum Na^+ was 138 m Eq/L, K^+ 4.9 mEq/L, calcium 7 mg/dl and Cl^- 100 mEq/L. Gastric lavage for acid fast bacilli was negative, Arterial blood gas showed evidence of hypoxia (pH 7.37, PO_2 64.7 mm Hg, PCO_2 37.7 mm Hg; TCO_2 22.9 mmol/L;) O_2 sat 91.6%; HCO_3 21.7 m mol/L). Pulmonary function tests showed a restrictive airways

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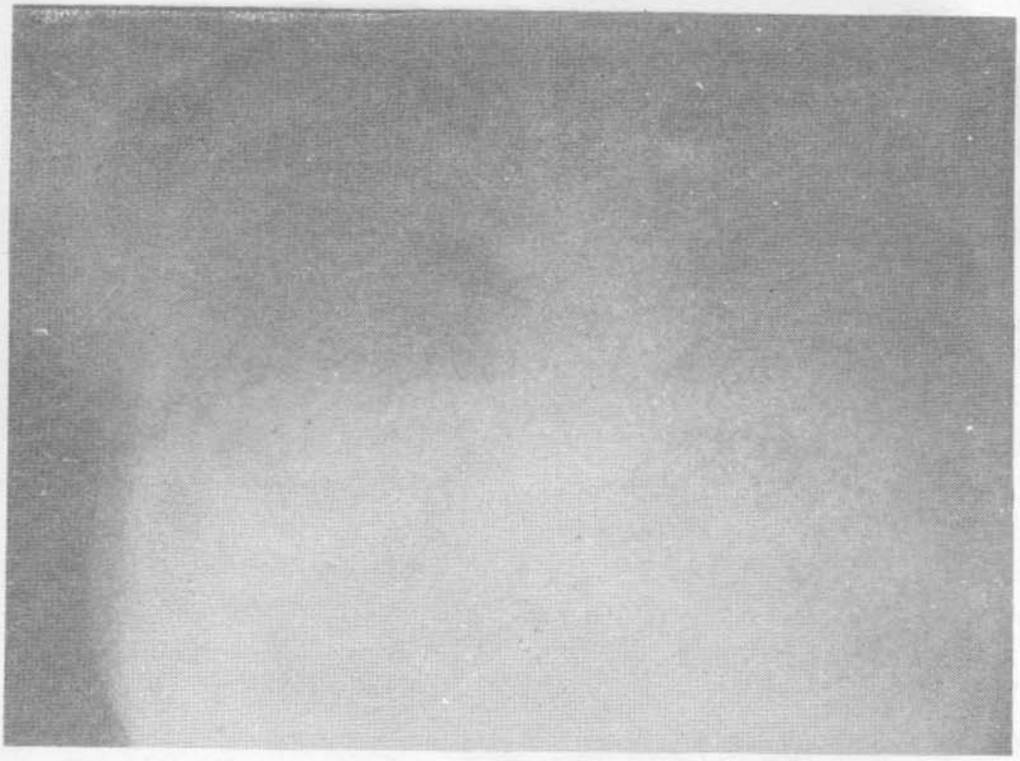

Fig. 1. X-ray showing diffuse calcific densities.

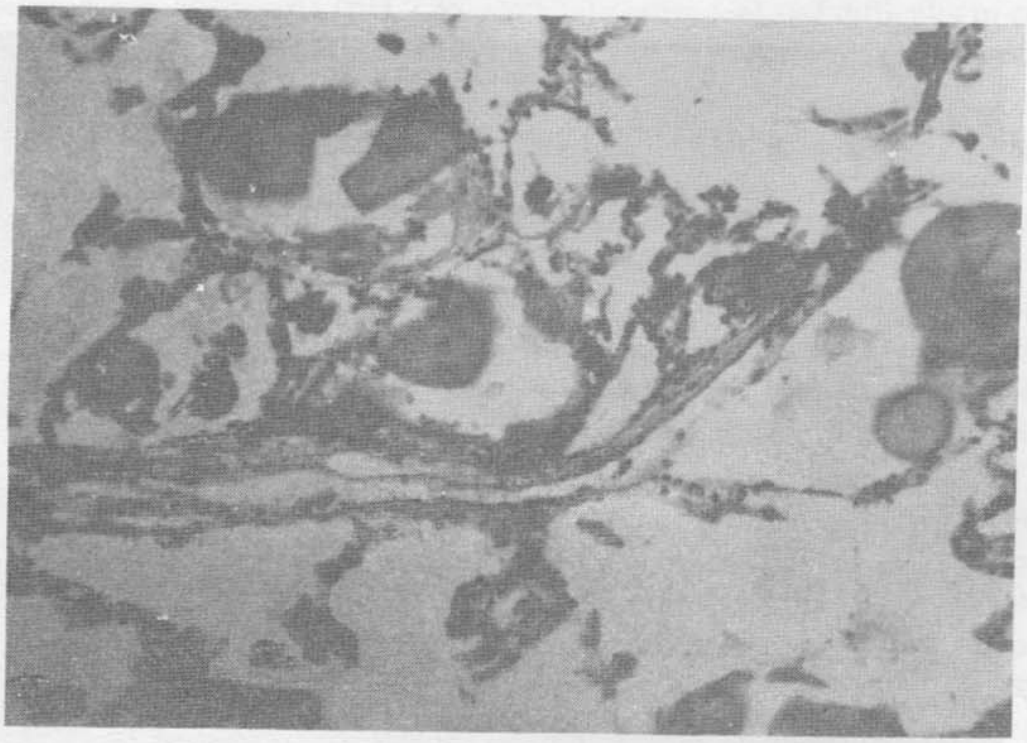

Fig. 2a. Photomicrograph showing lung with many alveoli with many microlithis of varying sizes. (H & E × 60).

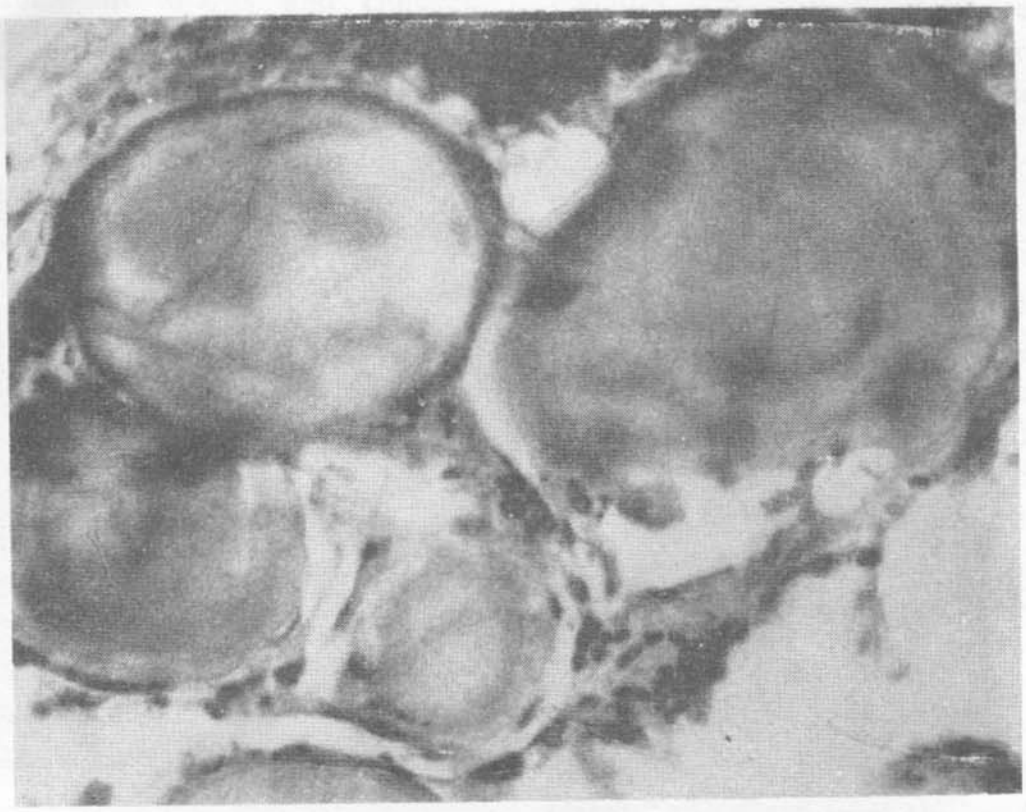

Fig. 2b. Photomicrograph showing lung with alveolar septa and concentrically laminated calcified bodies calcosphenitis within the alveoli (H & E ×180).

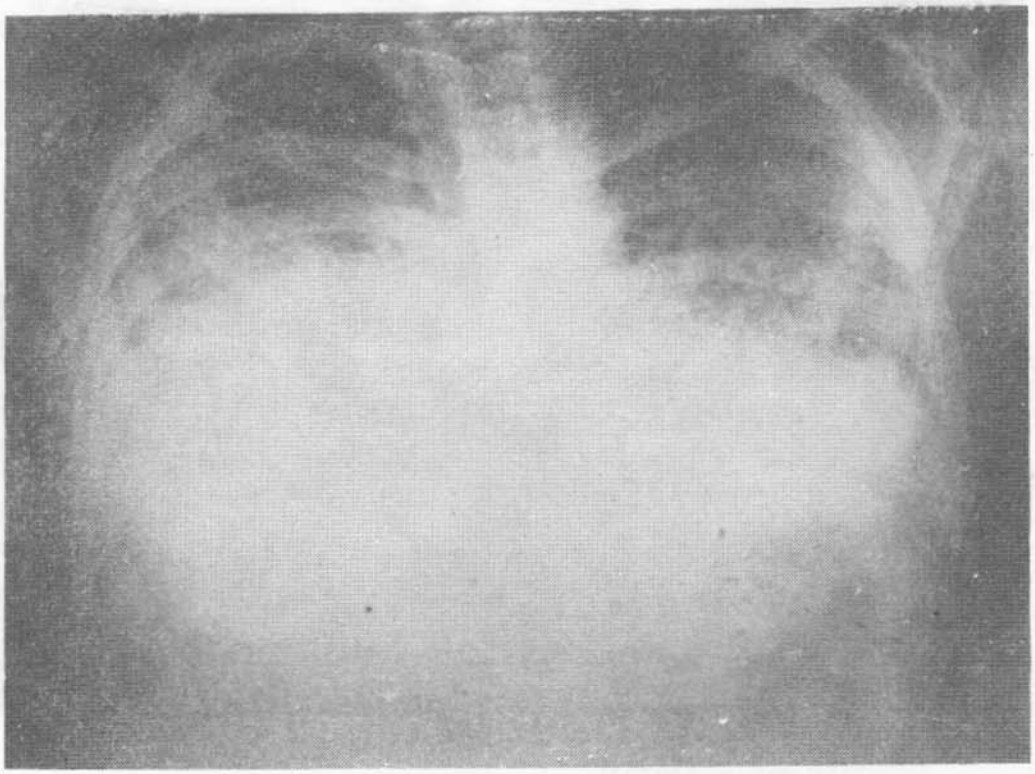

Fig. 3. X-ray of sibling showing diffuse bilateral opacities.

pattern and ultrasound examination of the abdomen showed evidence of gall stones.

Mantoux test read after 48 hours was 10 mm. He was then started on two antitubercular drugs (rifampicin and isoniazid) and after a course of 1 month of treatment, repeat *X*-ray remained unchanged. At this stage an open lung biopsy was done and the histopathology was reported as psommama like bodies (Calcispheres) within the alveoli with preservation of alveolar architecture consistent with diagnosis of pulmonary alveolar microlithiasis (*Figs. 2a & 2b*).

Case 2: A 14-year-old healthy female, elder sibling of the above patient was identified on family screening for pulmonary alveolar microlithiasis. She was a healthy well nourished young adolescent. On enquiry she had no respiratory symptoms. Her routine investigations were normal. Serum calcium was 8 mg/dl. Serum electrolytes were Na^+ 130 mEq/L, K^+ 2.5 mEq/L, and Cl^- 98 mEq. An arterial blood gas revealed evidence of hypoxia (pH 7.38, PO_2 70 mm Hg, PCO_2 35.2 mmHg, TCO_2 23.4 mmol/L; HCO_3 22 mmol/L; O_2 sat 98%). Pulmonary function showed restrictive airways pattern. Chest *X*-ray showed bilateral diffuse opacities with obliteration of cardiac silhouette (*Fig. 3*). As the radiological picture was identical to *Case 1* lung biopsy was not performed. Chest *X*-ray of the other members of the family (father, mother and another sibling) were normal.

Discussion

Pulmonary alveolar microlithiasis is a rare disease of unknown cause. The age of patients reported ranges from newborn to 80 years. There is no sexual predominance and in about half of the reported cases a familial pattern has been noted. The familial incidence is almost always restricted to siblings(1-3) as is also seen in our cases. The striking features of this condition is that the patients are absolutely asymptomatic and most of the cases are diagnosed accidently while investigating for some other condition(3,4). Pulmonary alveloar microlithiasis cases have been reported in the Indian literature also(3,5,6).

The diagnosis is based on typical radiological appearances, described as, fine sand like particles of calcific density spread throughout both the lung fields, with slight variation in size and some time with accentuation along the pleural surfaces(2). Jirayr *et al.*(7) described some more radiological features like fine ground glass appearance, streaky radiation, inter alveloar calcific deposit and the 'vanishing heart' phenomenon. Both the cases described by us had these radiological features.

The definitive diagnosis of pulmonary alveloar microlithiasis can be established by open lung biopsy. The gross features include gritty feel and difficulty in cutting the gross specimen. Under light microscopy, the alveloar architecture is well preserved and microliths are seen within the alveoli as irregularly concentric, laminates substance prganized around an amorphous nidus a pattern that gives rise to the characteristic 'onion skin' appearance(8). Similar histologic features has been demonstrated in our case also. Calcification in other areas of the body like renal calculi and calcification of the prostrate has been described in patients with pulmonary alveolar microlithiasis(2), ours is the first case reported with gallstones. Additional features like abnormality in pulmonary function tests have been seen in about 30% of patients. A restrictive type of respiratory dysfunction is the major finding in these patients(8).

The disease is slowly progressive(2)

and despite the severe radiological and pathologic changes, patients remain relatively asymptomatic till about 4th or 5th decade when they develop features of chronic cor pulmonale and right heart failure. No known therapy is available for pulmonary alveolar microlithiasis. The disease is reported to be uninfluenced by steroids and chelating agents(9). Bronchpulmonary lavage has been tried but has had no effect on the progression of the disease(10).

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