

A pregnant mother with pulmonary alveolar microlithiasis

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Introduction

Pulmonary alveolar microlithiasis (PAM) is a rare disease of unknown pathogenesis, characterized by widespread laminated calciphretites in alveolar spaces in the absence of any known disorder of Calcium metabolism. In Sri Lanka, very few cases have been documented [1].

Case report

A 37 year old, previously healthy pregnant mother in her fourth pregnancy presented at 20 weeks of gestation with progressive breathlessness and cough for 2 weeks and fever for 2 days. Cough was productive with whitish sputum. She is a product of consanguineous marriage. Two of her siblings have died of an unknown chest disease.

On examination, she had diffuse end inspiratory crackles, symmetrically distributed over both lung fields in middle and lower zones which did not disappear after coughing.

Her full blood count, ESR and serum calcium were normal. Arterial blood gases were normal. The chest radiograph revealed diffuse high density micronodules and reticular lines, obliterating bronchovascular bundles, margins of the hearts and diaphragm (Fig 1). Tuberculosis was excluded by negative sputum for acid fast bacilli, negative Mantoux test and a negative bone marrow PCR. A restrictive pattern was observed in lung function tests.

The diagnosis of PAM was made by the typical histopathological appearance of open lung biopsy which showed enormous numbers of wavy, concentrically laminated microlithiasis filling the alveoli and delicate fibrosis of alveolar walls (Fig 2). The patient was initially treated with amoxicillin 500 mg 8 hourly and intravenous metronidazole 1g twice daily for 1 week, subsequently intravenous cefuroxime 750 mg 8

hourly for 11 days while nebulising with salbutamol which resolved her symptoms.

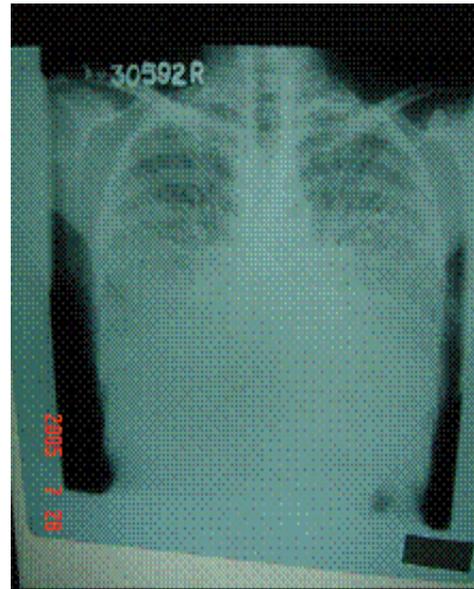


Figure 1

The chest radiograph showing high density micronodules.

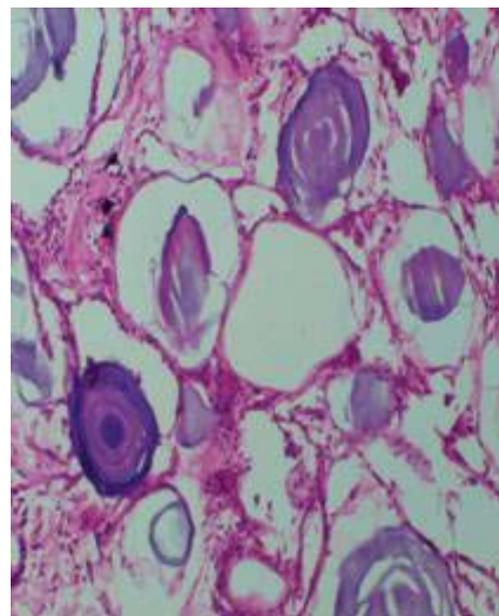


Figure 2

Open lung biopsy : decalcified specimen stained with haematoxylin and eosin .
magnification – 400

Case reports

Discussion

In English literature case reports of PAM has been reported from 51 countries where 40.6% of patients are from Asian countries and 42.7% of patients are from European countries.

In English literature, PAM has been reported from 51 countries where 40.6% are from Asia [2]. The mean age at presentation is 35 years [3]. There are two varieties described, familial and sporadic [4]. Autosomal recessive familial form has female preponderance. 50% of patients may be asymptomatic despite fluorid interstitial shadows on chest radiograph, or present with dyspnoea, prolonged cough, chest pain or haemoptysis [4]. In later stages, they may also present with features of cor pulmonale [4]. Initially there may be no functional impairment, but later there may be restrictive type of defects [4]. Diagnosis is made by bronchoalveolar lavage or open lung biopsy. Typical sandstorm appearance in high resolution computed tomography scans also have a diagnostic value. The only curative treatment is bilateral sequential lung transplantation [4].

Other pharmacological agents which have no proven benefit include disodium etidronate 10 mg/kg per day for 1 year [1]. Few cases of PAM during pregnancy have been published [5].

References

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