ALVEOLAR PROTEINOSIS

(A Case Report)

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ABSTRACT

A case of pulmonary alveolar proteinosis diagnosed by open lung biopsy is being reported and relevant literature is discussed.

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KEY WORDS : Alveolar proteinosis; Pulmonary alveolar lipoproteinosis.

Introduction

Pulmonary alveolar proteinosis (PAP) or pulmonary alveolar lipoproteinosis is a rare entity of unknown etiology caused by deposition of lipid-rich proteinaceous material in the alveoli [1,2]. Histologically the material appears granular and eosinophilic and is strongly positive to periodic-acid-Schiff (PAS) stain. This is a relatively new disease which was first reported by Rosen *et al* in 1958 [3] and by 1980 only 260 cases were reported in world literature [2].

CASE REPORT

LS, a 45 year old serving Junior Commissioned Officer was referred for evaluation from a peripheral hospital. The individual had been symptomatic for a year with progressive dyspnoea, dry cough and weight loss. There was no history of fever, haemoptysis or chest pain. He was a nonsmoker. There was no occupational dust exposure or past history of illness. He has been treated with antibiotics for one month and antitubercular treatment for 3 months before he was transferred to our centre but it was without relief. The clinical evaluation was remarkable by the absence of positive findings. ESR was 08 mm fall in the first hour, Hb 14.6 g/dl, AEC 430 cells/mm³, Mantoux 05 mm, sputum smear for AFB negative. Chest X-ray showed central acinar shadows with confluence, without any evidence of cardiomegaly or pulmonary artery prominence nor any Kerley B lines (Fig. 1). CT scan showed no hilar or mediastinal lymphadenopathy. It confirmed a predominant alveolar filling process (Fig. 2).

Biochemical and serum immunoglobulin profiles were normal. Rheumatoid factor and LE cells were negative. The pulmonary function tests showed normal FVC, FEV₁, but DLCO (16.97 ml/mmHg) was 62% of predicted and KCO (3.73) was 72.3% of predicted. Arterial blood gases showed hypoxia (PaO₂ 58.5 mmHg) worsening on

Fig. 1 X-ray chest PA showing central acinar shadows with confluence. There is no cardiomegaly.

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Fig. 2 CT scan chest showing predominantly alveolar filling process.

Fig. 3 Photomicrograph (x 400) showing alveoli filled with granular, eosinophilic material.

12 minutes walking on level ground (PaO_2 48.9 mmHg). An open lung biopsy was performed which revealed areas of alveolar exudates, which were granular, eosinophilic and strongly positive for periodic acid-Schiff stain (Fig. 3). The alveolar septa showed few inflammatory cells. There were focal areas of proliferation of pneumocytes and lipophages were also seen. AFB stain gave a negative result. This favoured a diagnosis of alveolar proteinosis. The patient was offered treatment by repeated lung lavage. However, he declined the same for the present due to personal reasons.

Discussion

PAP has a peak age of onset between 30-50 years, and is three times more common in males [3,5]. It is believed to be due to disorder of surfactant homeostasis where surfactants cannot be removed by defective alveolar

macrophages [3-5]. In many cases no predisposing factors are found and they are termed "primary". In some cases various predisposing factors like consanguinity in marriage, pulmonary alveolar proteinosis in siblings, previous infective processes like tuberculosis, fungal diseases, HIV infection, connective tissue diseases and even use of some drugs are present and this group is called secondary [2,4,5,7].

Our patient was a 45 years old male and presented with progressively increasing dyspnoea, dry cough and weight loss. Earlier, from our centre another case was reported [6] where patient was totally asymptomatic and accidentally detected to be having bilateral hilar haze on a routine chest skiagram. However, scanty sputum production, minimal haemoptysis, finger clubbing, lung crackles and even cyanosis have been reported [3,7]. Most important radiological findings in these cases are alveolar type of opacity with air bronchogram, perihilar in distribution, without any cardiomegaly, in a subacute or chronic setting. This type of presentation can exclude acute cardio-respiratory problems like pulmonary oedema or adult respiratory distress syndrome. Pulmonary function studies may show restrictive defect with arterial hypoxaemia and diffusion abnormality which was present in our case. Prognosis of these patients is variable [6,7] with 25% showing spontaneous remission. Another 20% may have a fulminant course leading to respiratory failure. Treatment of this disease includes lavage by using 10-15 litre of normal saline. One lung at a time is lavaged by using a double lumen endotracheal tube [3,7]. With this modality of therapy remission can be achieved in 80% of cases or even more [3,5,7]. However, no treatment is needed if patient is asymptomatic and if disease is non progressive.

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