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PULMONARY ALVEOLAR PHOSPHOLIPOPROTEINOSIS

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Primary alveolar proteinosis (PAP) now phospholipoproteinosis is a syndrome of unknown etiology described by Rosen and *et al.* in 1958(1). PAP results from abnormal accumulation of surfactant phospholipids and proteins in the alveolar spaces. The disorder has been described in all the age groups from six months to 72-year-old patients, but the preponderance of those affected is between 20 and 50 yrs(2). However, the number of cases occurring in infants and children among the total cases described is small(3) and is now being recognized as a possible cause of neonatal respiratory distress(4).

Case Report

A 14-year-old school student was admitted with the complaints of reduced appetite, failure to gain weight and easy fatigability since the age of 2 years. The patient also complained of progressively increasing exertional dyspnea since the past 2 years and dry cough for the past 6 months. He was treated with antituberculous treatment for four months by a private practitioner which was omitted due to lack of clinical and radiological response. The child was a full term normal hospital delivery, with birth weight of 2.5 kg. He was fully immunized. There was no significant past and family history.

The child was undernourished, asthenic with height of 142 cm and weight of 27 kg (expected weight per ICMR chart is 48 kg). There was no clubbing or significant lymphadenopathy. Respiratory system examination revealed occasional end inspiratory rales with bilateral scattered rhonchi. Other systems' examination was essentially normal.

His complete hemogram, renal and liver function tests were normal. Serum LDH level was normal. *X*- ray chest showed bilateral fluffy alveolar shadows distributed around both the hilar and perihilar region extending into all the zones. The shadows were static when compared with the previous roent-

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Received for publication: September 15, 1992; Accepted: January 4, 1993

genogram. Sputum for acid fast bacilli on three occasions was negative and did not show any other opportunistic organisms like nocardia. Tuberculin test with 5 tuberculin units was negative. Pulmonary function test revealed a severe restriction with normal flow rates. Diffusion studies could not be performed due to low vital capacity. Arterial blood gases at rest revealed basal PaO_2 of 79 mm Hg, $PaCO_2$ of 38 mm Hg, pH of 7.393 and saturation 95%. Fall of 8 mm Hg in PaO_2 after exercise with a load of 20 watts for 4 minutes was observed. Diagnostic fibreoptic bronchoscopy with bronchoalveolar lavage and transbronchial biopsy were performed from the right middle lobe. Bronchoalveolar lavage cytology showed Periodic Acid Schiff (PAS) positive amorphous material in a background of bronchial squamous cell, and few lymphocytes and macrophages and occasional squamous cells. A transbronchial lung biopsy showed small pieces of lung tissues in which most of the alveoli contained granular eosinophilic material as well as cleft like spaces. The material was PAS positive, alcian blue negative and metachromatic with toluidine blue. The alveolar walls were of normal thickness and at spaces were lined by flattened epithelium. The immunoglobulin profile was normal.

Discussion

Alveolar type II cells secrete the complex material that line epithelial surfaces of the lung and is necessary for maintaining the expanded state. The phospholipid components are known to be critical for the effectiveness of surfactant. PAP like changes can occur in variety of clinical situations which results from exposure to chemicals, infections, silica, aluminium associated Hodgkin's lymphoma and coexisting immune disorders. This form is called as "secondary" alveolar proteinosis. In our patient no such association was found.

The clinical manifestations of PAP are extremely varied. The usual presentation is progressive dyspnea and cough which may be productive. Pleuritic chest pain, fatigue and substantial weight loss are other common presenting features(2). Children before the age of one year may present with vomiting, diarrhea and high grade fever. Hemoptysis is rare. The onset may be abrupt or an insidious one presenting with failure to thrive or growth failure as in our patient(3). Physical findings are relatively few and usually consist of only a few scattered rales and rarely clubbing of the fingers and toes. Although PAP can be diagnosed by sputum examination which may show PAS positive material, segmental lavage through fibreoptic bronchoscope or transbronchial lung biopsy or open lung biopsy may be required for definitive diagnosis(2). Elevation of the serum lactic acid dehydrogenase (LDH) in the absence of hepatic damage is reported. The LDH levels return to normal in patients who recover. The cause of the elevated LDH is not known but it may be due to an increased death of pulmonary cells(5). Immunologic examination of some children has revealed various types of defects including thymic aplasia, atrophy, low serum IgA levels and lymphopenia(3). Complicating fungal infections like nocardia are frequent and probably due to the defective clearance of inhaled organisms by abnormal macrophages. Prognosis is generally unfavorable in children, the mortality being more than 75%(5). Secondary bacterial and mycotic infections are particularly common and are frequently cause of death. It is due to an acquired functional defect of alveolar macrophages overburdened by heavy load of debris and also due to presence of a "nutritious broth" in the air space. In adult,

the disease may remain stable for considerable period of time and spontaneous improvement may occur.

Before therapeutic bronchoalveolar lavage was introduced by Ramirez RJ *et al.* in 1965(6), patients with PAP were treated with corticosteroids, heparin, antibiotics, trypsin and pancreatic enzymes with little success. The treatment of PAP consists of therapeutic bronchoalveolar lavage. The lavage can be accomplished safely in children and adults and with relative ease by using double lumen endotracheal tube or catheter to isolate and lavage one lung while ventilating the other(7). In very young children the same can be accomplished by using double lumen catheter and rigid bronchoscope. The therapeutic efficiency of bronchoalveolar lavage is primarily due to the mechanical removal of intra alveolar phospholipids. Most patients who undergo whole lung lavage for PAP that is unassociated with pulmonary fibrosis experience an improvement in pulmonary function and in exercise performance. The reported complications associated with PAP are severe pulmonary fibrosis followed by respiratory failure and death, formation of emphysematous bullae followed by pneumothorax and usual complication of sudden asphyxia and death due to flooding of the airways by thick alveolar material during anesthesia(2).

Acknowledgements

The authors wish to thank Dr. (Mrs)P.M. Pai, Dean, Seth G.S. Medical College and King Edward VII Memorial Hospital for allowing them to publish this case report.

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INFLAMMATORY LINEAR VERRUCOUS EPIDERMAL NEVUS

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Inflammatory linear verrucous epidermal nevus (ILVEN) is a rare skin disease of

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Received for publication: November 20, 1992; Accepted: January 29, 1993