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CASE STUDY

PULMONARY ALVEOLAR PROTEINOSIS- A RARE PATHOLOGICAL CASE REPORT

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ABSTRACT

Pulmonary alveolar proteinosis (PAP) is a rare unusual lung disorder of unknown etiology characterized by the accumulation of large amounts of a phospholipoproteinaceous material in the alveoli due to impaired alveolar macrophage function caused by neutralizing anti-granulocyte-macrophage colony-stimulating autoantibodies that stains positive by using the periodic acid-Schiff (PAS) method. Diagnosis may be difficult with clinical signs and symptoms only. We present a rare case of 22-years old male having idiopathic PAP presenting with dyspnoea on exertion, productive cough and fever.

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INTRODUCTION

Case Study

A 22-year-old male, wrestler by profession, presented with dyspnoea on exertion and productive cough with sputum having lacy consistency, from last four years. He also complained of fever since two months. Family history revealed that his brother died of tuberculosis two years back. Drug history revealed that our patient was on anti-tubercular treatment (ATT) since 2 months but had no relief in symptoms. Personal history revealed that he was a non smoker and a wrestler by profession with history of chronic exposure to mud/dust. The physical examination revealed bilaterally inspiratory fine crackles on auscultation. High-resolution CT scan of the chest showed patchy ground-glass opacification with interlobular thickening (crazy-paving appearance). Flexible bronchoscopy with bronchoalveolar lavage (BAL) was performed and the appearance of BAL fluid was milky, lacy in consistency. Cytological examination of this specimen revealed large plaques of granular eosinophilic proteinous material which was PAS positive (Figure 1) (Figure 2). Transbronchial lung biopsy (TBLB) was performed and the histological examination revealed the alveolar spaces filled with amorphous eosinophilic granular material (Figure 3) consistent with PAS (Figure 4). Other possible diseases were excluded with clinical, radiological and laboratory investigations.

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DISCUSSION

Pulmonary alveolar proteinosis (PAP) is a rare disease characterised by impaired surfactant metabolism that leads to accumulation of an amorphous, largely cell-free, lipoproteinaceous material in the alveoli (Khan, 2011). The lungs become stiff with restricted ventilatory function. Secondary infection may occur. Two forms are recognised: (Olade, 2011). Primary: idiopathic and secondary: due to lung infections, haematological malignancies, inhalation of mineral dusts (eg silica, titanium oxide, aluminum) and insecticides. It is thought that impairment of surfactant clearance by alveolar macrophages, by autoantibody inhibition of the action of granulocyte-macrophage colony-stimulating factor (GM-CSF), may underlie many acquired cases, whereas congenital disease is most commonly attributable to mutations in surfactant protein genes, but may also be caused by GM-CSF receptor defects. Therapy with GM-CSF has shown promise in acquired cases of pulmonary alveolar proteinosis (Seymour, 2002). The estimated prevalence is 1 case per 100,000 population (Olade, 2011). Pulmonary alveolar proteinosis is probably underdiagnosed.It is more common in males than females and typically presents in those aged 20-50 years (Olade, 2011).

Incidence is increased in patients with haematological malignancies and AIDS, suggesting a relationship with immune dysfunction. Causes include: (Olade, 2011) inhalation of silica dust, exposure to insecticides, aluminum dust, titanium dioxide and other inorganic dusts, haematological malignancies, HIV infection.

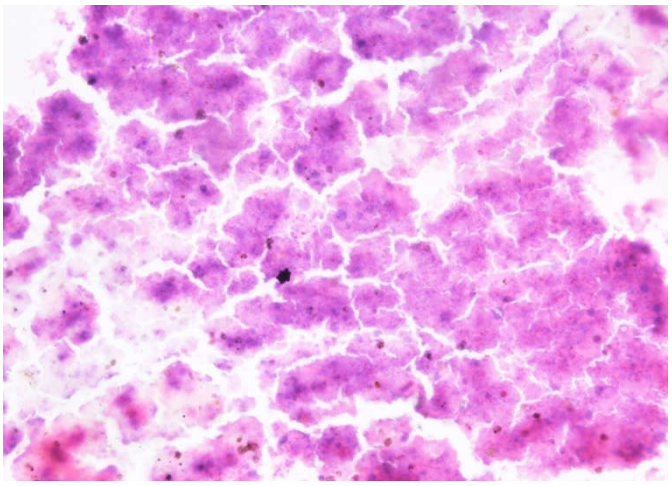

Figure 1. H&E stain 40X of BAL fluid

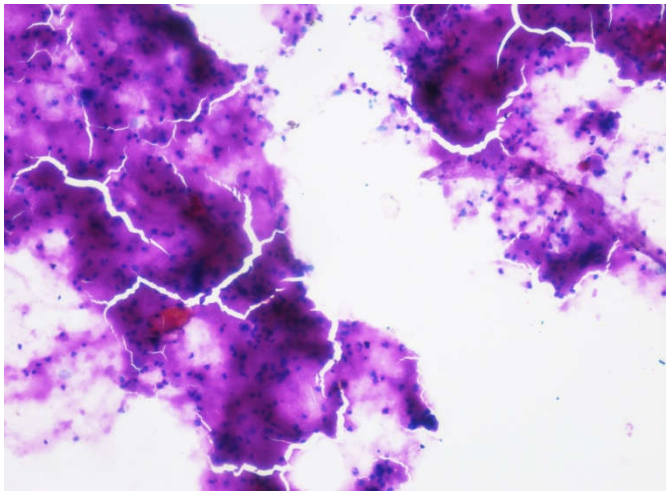

Figure 2. PAS staining 40X of BAL fluid

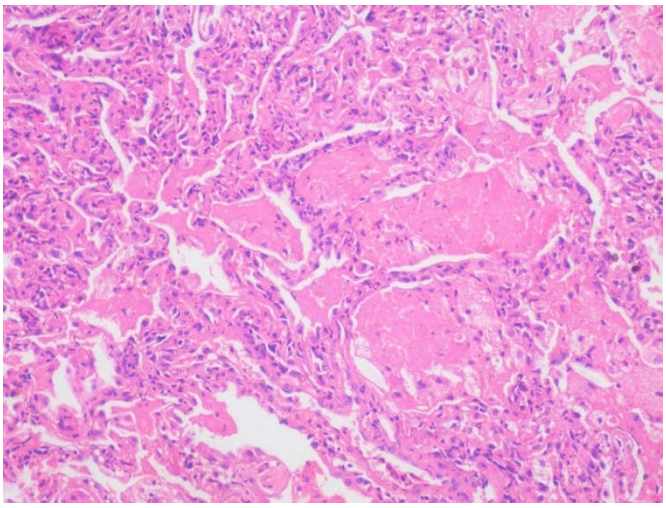

Figure 3. TBLB/H&E stain 20 X

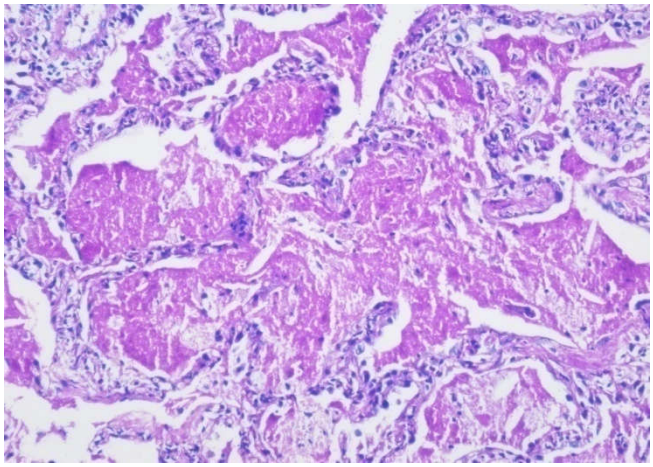

Figure 4. TBLB/PAS staining 20 X

Usually presents at age 20-50 years, with a gradual onset of nonspecific dyspnoea or dyspnoea on exertion and persistent dry cough (Das, 2010). As many as 30% of patients are asymptomatic, even with diffuse CXR abnormalities. Symptoms include: (Olade, 2011), persistent cough (dry or with sputum production); haemoptysis is rare; progressive dyspnoea, fatigue and malaise, weight loss, intermittent low-grade fever and/or night sweats, pleuritic chest pain. Signs are usually nonspecific and include: (Olade, 2011), fine end-inspiratory crackles, clubbing, cyanosis. Investigations reveal: Serum lactate dehydrogenase (LDH) is usually elevated, CXR: bilateral perihilar infiltrates with consolidation. Changes progress into a diffuse reticulogranular pattern.

Flexible bronchoscopy with bronchoalveolar lavage: alveolar secretions are PAS-positive but contain no organisms or any excessive cellular response. High-resolution CT scan of the chest: patchy ground-glass opacification with interlobular thickening (crazy-paving appearance). (Khan, 2011). Appearance is similar to that seen in lipoid pneumonia, sarcoidosis and acute respiratory distress syndrome. Lung biopsies: definitive diagnosis requires lung biopsy that typically shows partial or complete filling of alveoli with PAS-positive granular and eosinophilic material with preserved alveolar architecture (Khan, 2011). Adults affected by pulmonary alveolar proteinosis (PAP) have circulating neutralising anti-granulocyte-macrophage colony-stimulating factor (anti-GM-CSF) antibodies. More effective diagnostic tests (eg anti-GM-CSF antibodies) are under investigation (Huizar, 2009).

Patients with minimal symptoms require only symptomatic treatment but patients with hypoxaemia require more aggressive management. Management includes: appropriate treatment of any underlying cause, mechanical removal of the lipoproteinaceous material by whole-lung lavage (WLL), which is the most widely accepted form of therapy for symptomatic pulmonary alveolar proteinosis (PAP): (Khan, 2011 and Ioachimescu, 2006). This is performed under general anaesthesia. The lung is ventilated briefly with 100% oxygen before lavage with isotonic sodium chloride solution. Lung lavage may take several hours. WLL is usually very effective but repeated lavages are usually necessary. Correction of functional granulocyte-macrophage colony-

stimulating factor (GM-CSF) deficiency with exogenous GM-CSF has emerged as an alternative therapy (Khan, 2011), Lung transplantation for patients with congenital PAP and adult patients with end-stage interstitial fibrosis and cor pulmonale (Olade, 2011). In the past, patients have also been treated with systemic steroids and aerosol mucolytics, but without much success. In a third of patients, no appreciable disability develops and the disease remits spontaneously or fails to progress. The natural history depends on the underlying aetiology. Estimates of 5-year mortality rates vary between 10% and 30% (Olade, 2011). Whole-lung lavage (WLL) often produces a dramatic response but recurrences are common and require repeated lavages. Lung transplantation for congenital pulmonary alveolar proteinosis (PAP) often has a good outcome. Patients with alveolar proteinosis related to inhalation of inorganic dusts or insecticides should avoid further exposure (Olade, 2011).

Conclusion

PAP is a rare disease characterised by impaired surfactant metabolism that leads to accumulation of an amorphous, largely cell-free, lipoproteinaceous material in the alveoli. The diagnosis, although rare, should always be kept in mind while evaluating a case with "crazy paving" pattern on HRCT and alveoli filled with granular, eosinophilic, proteinaceous material with preservation of alveolar architecture.

REFERENCES

Das, M., Salzman, G.A. 2010. Pulmonary alveolar proteinosis: an overview for internists and hospital Hosp Pract (Minneap). Feb; 38(1):43-9.

Huizar, I., Kavuru, M.S. 2009. Alveolar proteinosis syndrome: pathogenesis, diagnosis, and management. *Curr Opin Pulm Med.* Sep;15(5):491-8.

Ioachimescu, O.C., Kavuru, M.S. 2006. Pulmonary alveolar proteinosis. *Chron Respir Dis.*, 3(3):149-59.

Khan, A, Agarwal, R. 2011. Pulmonary Alveolar Proteinosis. Respir Care. Apr 15.

Olade, R.B. et al. 2011. Pulmonary Alveolar Proteinosis, Medscape, Mar.

Seymour, J.F., Presneill, J.J. 2002. Pulmonary alveolar proteinosis: progress in the first 44 years. *Am J Respir Crit Care Med.* Jul 15; 166(2):215-35.

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