Case Report

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Case Report of Idiopathic Pulmonary Haemosiderosis in a Child with Recurrent Chest Infections

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Abstract

Idiopathic Pulmonary Hemosiderosis (IPH) is a rare condition, usually occurring in children and teenagers. The classic presentation is of haemoptysis, iron deficiency anaemia and diffuse pulmonary infiltrates. We report a case of IPH in an 8 year old boy, who presented with recurrent episodes of fever, cough and respiratory infections without any trigger since childhood.

Keywords: *Hemosiderosis, Idiopathic Pulmonary Hemosiderosis*

Introduction

Idiopathic pulmonary hemosiderosis (IPH) is a rare chronic pulmonary disease characterized by recurrent diffuse alveolar haemorrhage (DAH) without any cause. It was first described as “brown lung induration” by Virchow in 1846. It is characterized by triad of haemoptysis, bilateral diffuse pulmonary infiltrates and iron-deficiency anaemia. Common clinical presentation is of cough, haemoptysis, and dyspnoea. Etiologic mechanisms described are -Genetic, autoimmune, allergic, environmental, and metabolic; however, none has been confirmed to date. The diagnosis is made by demonstrating hemosiderin laden macrophages in the sputum, broncho-alveolar or gastric lavage, or on lung biopsy.

Case report

8-year-old boy presented to the hospital with history recurrent episodes of fever and cough and characteristic complaint of significant drop in his haemoglobin with hypochromic microcytic anaemia with each episode. In the present episode, she was very pale with extreme tiredness, febrile and had persistent cough. Chest X-ray showed diffuse pulmonary infiltrates and High resolution computed tomogram (HRCT) showed ground glass opacities (Figure-1). Broncho-alveolar fluid examination revealed numerous hemosiderin laden macrophages in the sediment. Complete blood count revealed hypochromic microcytic anaemia.

Gross Examination

The specimen was received in formalin container labelled with patient data & right lung tissue. It included 0.9x0.9x0.9 cm lung tissue. The sample was entirely processed for histopathological analysis.

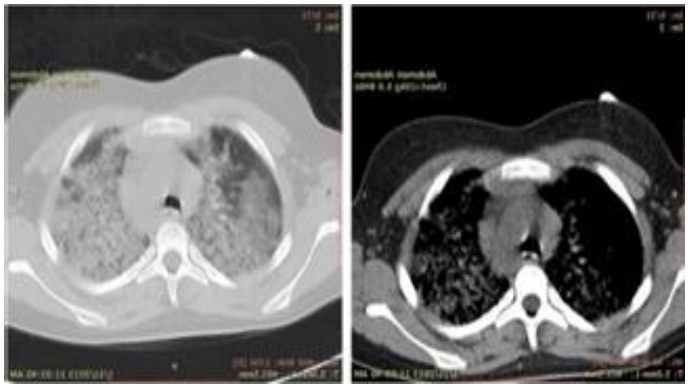

Figure 1: CT scan of the thorax showed diffuse alveolar interstitial infiltration. CT guided Trans bronchial lung biopsy was sent to the laboratory.

Microscopic Examination

Microscopy of the broncho-alveolar lavage stained with Prussian blue stain revealed hemosiderin laden macrophages (Figure - 2).

Haematoxylin-Eosin stained paraffin sections revealed interstitial fibrosis and abundant accumulation of hemosiderin laden macrophages within the alveolar spaces and interstitium. Parenchymal remodeling with alveolar architectural simplification and possibly cystic change (Figures 3, 4, 6).

Prussian blue staining highlighted hemosiderin laden macrophages (Figure - 5).

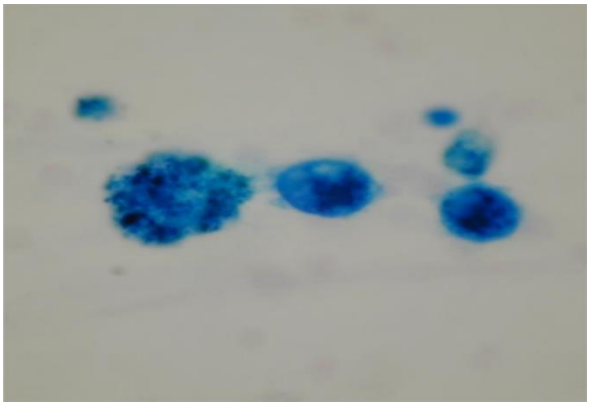

Figure 2: Bronchoalveolar Lavage fluid – Prussian blue stain 40 x Original magnification

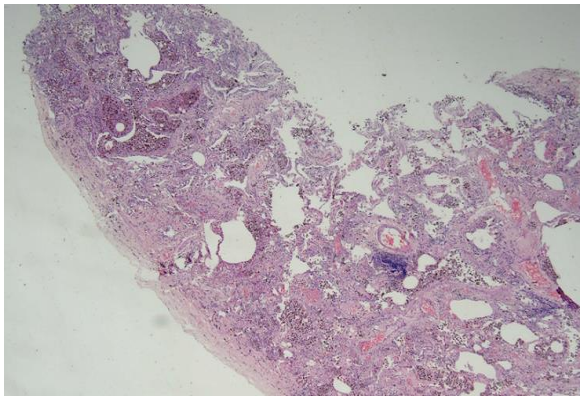

Figure 3: 2.5 X Original magnification (Haematoxylin – Eosin stain)

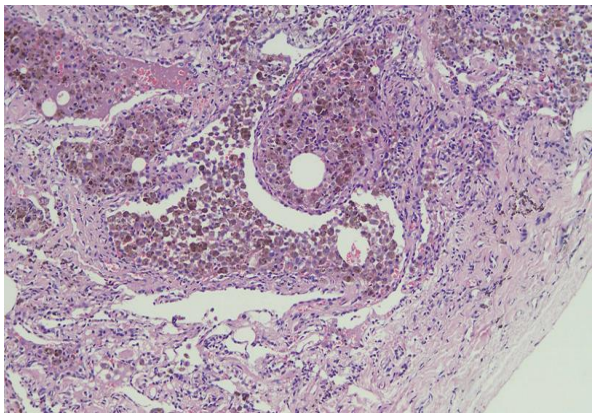

Figure 4: 10 x original magnification (Haematoxylin – Eosin stain)

Discussion

IPH is considered to be a rare disorder with an incidence of 0.24–1.23 cases per million. It was first described by Ceelen in 1931 as a triad of haemoptysis, anaemia and pulmonary infiltrates. 80% of cases occur in children with an equal sex distribution in childhood and a slight male

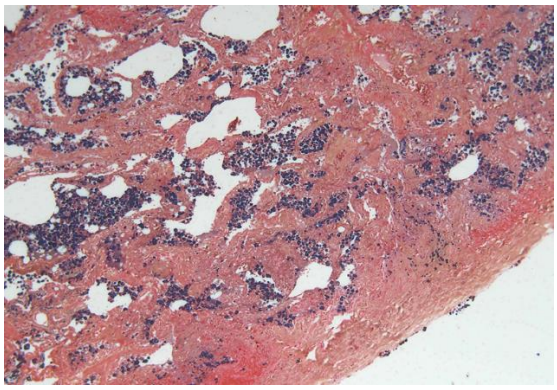

Figure 5: 5x original magnification (Prussian blue stain)

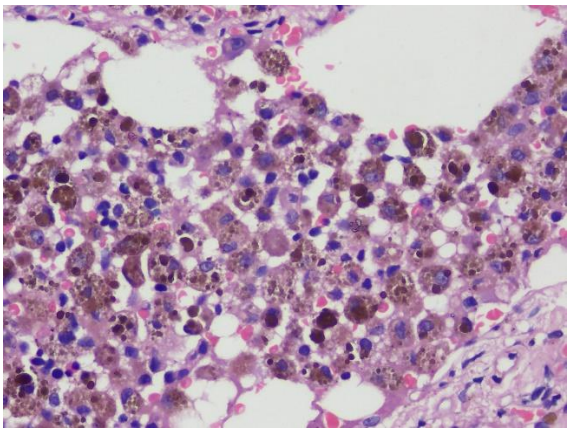

Figure 6: 40 x original magnification (Haematoxylin – Eosin stain)

The exact aetiology and pathogenesis is not yet known. When no cause for repeated episodes of diffuse alveolar haemorrhage is apparent, the entity is referred to as IPH. [1]. Consanguinity, environmental factors, autoimmunity are thought to play a role in some cases. The primary defect and clinical feature in IPH is recurrent alveolar haemorrhage. Diagnosis is by exclusion of other systemic diseases which can cause pulmonary haemorrhage such as Wegener's granulomatosis, Systemic Lupus Erythematosus, Rheumatoid arthritis, Good pasture's Syndrome, coagulopathies, platelet defects, pulmonary infections, pulmonary neoplasms, pulmonary veno-occlusive disease, pulmonary capillary haemangiomatosis and toxins such as cocaine, pesticides or insecticides. The radiological features are nonspecific and vary between interstitial changes, consolidation, and pleural effusion, pulmonary nodules with or without cavitations, lung fibrosis and ground glass opacities. Acute pulmonary haemorrhage can be fatal due to respiratory failure. Chronic haemorrhage leads to hemosiderin-laden macrophages and pulmonary fibrosis. If left untreated due to progressive pulmonary fibrosis it has

been reported that systemic vasculitis has developed in a patient with IPH, eight years from diagnosis [1, 2, and 3].

Our patient was transfused with blood to correct anaemia. He was then started on prednisolone; loading dose and the later on was put on tapering dose. He is clinically better now; though complete improvement has yet not been achieved.

Informed Consent: As identity of the patient is not disclosed; informed consent is not required.

Ethical approval: As identity of the patient is not disclosed; ethical committee approval is not required.

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