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# Idiopathic Pulmonary Hemosiderosis in a 8 Year Old Male Child: A Case Report

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**Abstract:** ***Idiopathic Pulmonary Hemosiderosis (IPH) is a rare disorder of unknown etiology characterized by Hemoptysis, iron deficiency anemia and diffuse pulmonary infiltrates. The diagnosis of Pulmonary Hemosiderosis refers to the subset of patients with Diffuse Alveolar Hemorrhage (DAH). Due to lack of pathognomonic findings, IPH diagnosis is established upon exclusion of all other possible causes of DAHin combination with specific pathologic findings revealing bland alveolar hemorrhage with absence of vasculitis and/or accumulation of immune complexes within lung parenchyma. We present a case of a 8 year old male child who presented to us in view of iron deficiency anemia requiring blood transfusions since the age of 6 years and had first episode of hemoptysis at the age of 4 years. His iron deficiency anemia coupled with chest high-resolution computed tomography scan, revealed findings compatible with diffuse alveolar hemorrhage. After excluding all other sources of bleeding through extensive gastrointestinal workup and thorough immunologic profile, video-assisted thoracic lung biopsy was performed and the diagnosis of Idiopathic Pulmonary Hemosiderosis was established.***

**Keywords:** IPH, idiopathic pulmonary hemosiderosis

## 1. Introduction

Idiopathic pulmonary haemosiderosis (IPH) is a rare disease characterized by recurrent episodes of intrapulmonary bleeding, chronic iron deficiency anemia and pulmonary fibrosis. IPH is a diagnosis made by exclusion of other causes. It occurs in both adults and children. Other conditions than IPH can cause pulmonary haemosiderosis. The etiology is unknown, but might be an immunological mechanism causinga defect in the basement membrane of the pulmonary capillary. IPH should be suspected in patients with recurrent episodes of coughing, hemoptysis, dyspnoea and anemia (1).

## 2. Case Report

An 8 year old male child from a poor socioeconomic status family, born of a non-consanguineous marriage presented with cough, respiratory difficulty, recurrent hemoptysis and pallor with easy fatigability beginning at the age of 4 years. There was no history of recurrent fever, skin lesions, bleeding from any other site, arthritis, bone pain and significant drug intake. His appetite, bladder & bowel habits were normal. He had received blood transfusions at the age of 6 and 7 years. At the time of admission, thechild had marked pallor, respiratory distress, tachypnea (respiratory rate = 44/min) and tachycardia (pulse = 130/min). Other vital parameters were normal. His weight was 17 kg and height was 110 cm. Examination of chest revealed crepitations bilaterally. Other examination findings were normal. On investigating, hemoglobin was 4.2gm% with hypochromic microcytic anemia, total leukocyte count was 1200/cu mm (polymorph 39%, lymphocyte 41%, eosinophil 07%, monocyte 03%), platelets were adequate, serum ferritin level was low (4mg/l), serum iron was 19mcg/dl suggestive of iron deficiency anemia. Clotting time, prothrombin time, activated partial thromboplastin time, liver function test, renal function test were within normal limits. Antinuclear antibody (ANA), anti-neutrophil cytoplasmic antibody (ANCA) and Mantoux test were negative. Chest radiograph revealed bilateral pulmonary infiltrate (Figure 1a). CT scan showed bilateral ground glass opacity (Figure 2). As no cause was identified, Lung Biopsy was done suggestive of idiopathic pulmonary hemosiderosis. The patient was given blood transfusion and oral prednisolone (2 mg/kg) along with iron. His condition improved after 7 days of therapy. Child has been undergoing periodic follow ups and has not required blood transfusion after the treatment was started.

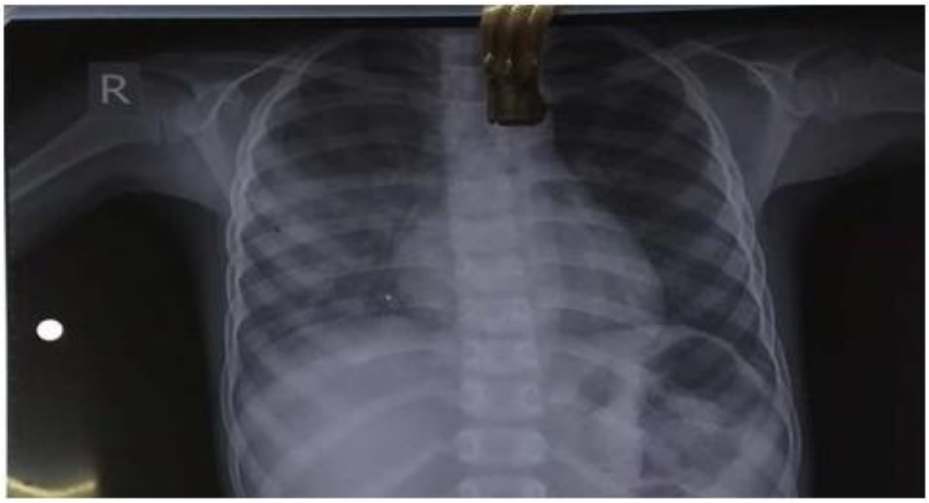


**Figure 1(a):** X ray on admission

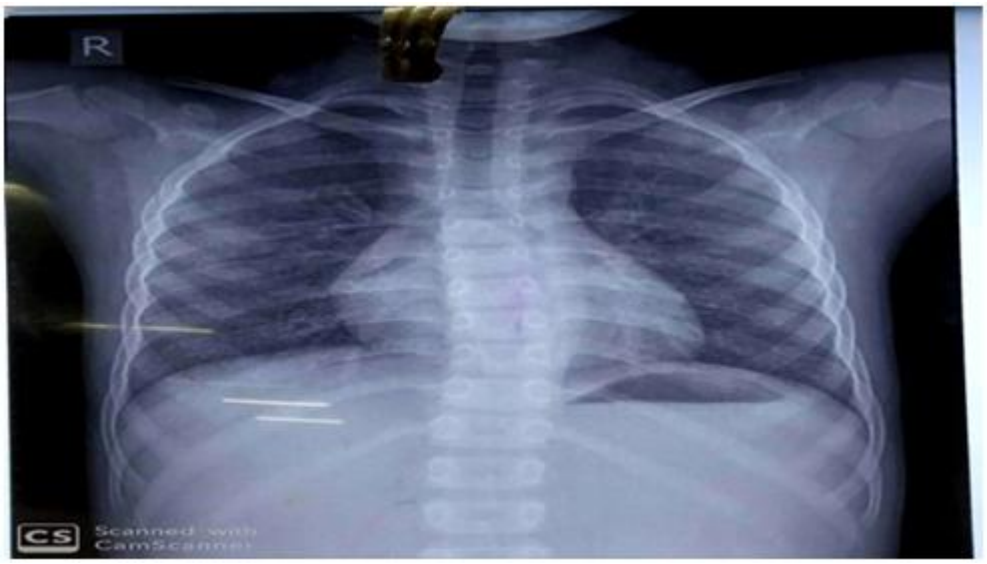


**Figure 1(b):** X ray on discharge

**Figure 2:** HRCT Chest

HRCT suggestive of diffuse and dense ground glass opacities in entire right lung. Patchy areas of ground glass opacity seen in apico-posterior and anterior segment of left upper lobe and superior segment of left lower lobe. Multiple non necrotic lymphadenopathies seen in bilateral supraclavicular, axillary, pre-tracheal, para-tracheal and right hilar region.

Lung Biopsy done for this child was suggestive of collapse of alveolar spaces with widening of alveolar septae with extensive areas of interstitial fibrosis and hemosiderin deposition. Alveolar spaces were filled with hemosiderin laden macrophages. Special stains like Prussian blue confirmed the pigment to be hemosiderin.

## 3. Discussion

Diffuse alveolar hemorrhage (DAH) is a rare and life-threatening condition characterized by hemoptysis, dyspnoea, alveolar infiltrates on chest radiograph and various degrees of anemia. It may occur either as a primary disease of the lung or a secondary condition due to cardiac, vascular, collagen or renal disease (2) (3) (4). Idiopathic pulmonary hemosiderosis (IPH) is a separate form of DAH of unknown origin, associated in some cases with celiac

disease. The estimated incidence of IPH in children is 0.24-1.23 cases per million, with a mortality rate as high as 50% (5). Only about 500 cases of this disease have beendescribed in medical literature. Our patient presented with similar findings of cough, recurrent hemoptysis along with features suggestive of iron deficiency anemia. The etiology of IPH is unknown. Some children with IPH have plasma antibodies (precipitins and IgE) against cow's milk, giving birth to the hypothesis that the disease might be caused by an allergic reaction to milk .Other authors have failed to reproduce these findings, which may be explained by an accidental coexistence of the two diseases. Apparently, there is a defect in the alveolar capillaries either in the alveolar basement membrane or in the endothelial cell .Circulating plasma immune complexes have been demonstrated in a few patients. However, immune-histochemical examination of lung tissue has not produced evidence of an immunological pathogenesis .The coexistence of coeliac disease and IPH, which has been reported in some patients, is in favour of an immunological pathogenesis. The literature reports at least 10 cases with coexistent coeliac disease and IPH, whereas our patient was investigated for coeliac disease and did not have it. In some of these patients, a gluten-free diet induced remission of the pulmonary symptoms .However, the possible pathogenic link between coeliac disease and IPH is still unclarified (6) (7) (8). He also had bilateral lung infiltrates. The diagnosis is based on the presence of iron deficiency anemia, characteristic chest radiograph and demonstration of hemosiderin-laden macrophages in Lung Biopsy as was done in our patient. Secondary causes of hemosiderosis are to be excluded. We have extensively searched for secondary causes but did not find any. Corticosteroids and/or immunosuppressive drugs may be effectiveduring an acute bleeding episode, and may in some patients improve symptoms and prognosis on the long-term, but the response to treatment displays great inter-individual variation (6). This patient responded with prednisolone and there has been no recurrence until followed up.

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