

**IDIOPATHIC PULMONARY HEMOSIDEROSIS PROGRESSING
TO INTERSTITIAL LUNG DISEASE.****RAMYAA RAJENDIRAN*, SHANTHI RAMESH, VIJAYASEKARAN.D
AND JOHN SOLOMON***Sree Balaji Medical College and Hospital, Department of Paediatrics, Chrompet, Chennai. 44***ABSTRACT**

Idiopathic pulmonary hemosiderosis (IPH) is a rare disorder first described in 1931, the incidence of which is around 0.24 to 1.23 per million. Pulmonary hemosiderosis can occur as a primary disease of the lung or can be secondary to systemic disease. The classical triad of IPH includes anemia, hemoptysis and pulmonary infiltrates, of which anyone could be the initial presentation. In some IPH may progress to interstitial lung disease. Interstitial lung disease (ILD) is a broad group of lung diseases comprising of both idiopathic and secondary causes such as IPH. Interstitial lung disease is characterized by inflammation and or fibrosis of the lungs. A systematic approach including a high index of suspicion, a thorough clinical evaluation, HRCT and BAL may suffice to make an early diagnosis of ILD. In a child with secondary ILD early diagnosis and appropriate treatment with immunosuppressive drugs will result in long term survival.

KEYWORDS: Idiopathic Pulmonary Hemosiderosis, Interstitial Lung Disease, Broncho Alveolar Lavage (BAL), High resolution CT scan (HRCT).



RAMYAA RAJENDIRAN
Sree Balaji Medical College and Hospital,
Department of Paediatrics, Chrompet, Chennai. 44

*Corresponding author

INTRODUCTION

Idiopathic Pulmonary Hemosiderosis (IPH) is a rare disorder which was first described in 1931. The incidence of IPH is estimated to be around 0.24 to 1.23 cases per million¹. Pulmonary hemosiderosis can occur as a primary disease of the lung or can be secondary to systemic disease. The classical triad of anemia, hemoptysis and pulmonary infiltrates is a distinguishing characteristic of this disease of which anyone could be the initial presentation¹. In some, IPH may progress to interstitial lung disease. Interstitial lung disease (ILD) is a broad group of lung diseases comprising of both idiopathic and secondary causes and is characterized by inflammation and or fibrosis of the lungs. Idiopathic Pulmonary Hemosiderosis is one of the secondary causes of ILD. Interstitial lung disease (ILD) in children presents with progressive respiratory symptoms like cough, tachypnea (usually of > 3 months), diffuse infiltrates on chest radiograph, abnormal pulmonary function tests (restrictive ventilatory defect) and impaired gas exchange². However, the clinical manifestations vary from asymptomatic presentation with radiological features suggestive of ILD to acute onset of symptoms like pneumonia (also known as Hamman Rich syndrome). High resolution CT scan (HRCT) and broncho alveolar lavage (BAL) may be helpful to confirm the diagnosis of ILD. Biopsy is indicated whenever there is clinical uncertainty/ atypical clinical presentation. In a resource poor setting, a systematic approach integrating a strong suspicion, a thorough clinical evaluation and HRCT may be suffice for the diagnosis and biopsy is rarely indicated as the specimen obtained may not always be a representative sample³.

MATERIALS AND METHODS

It is a case report on a male child with Idiopathic pulmonary hemosiderosis progressing to interstitial lung disease.

Study setting : Sree Balaji Medical College And Hospital.

CASE REPORT

A 11year old male child was admitted with history of dyspnea, chest pain, exercise intolerance and failure to thrive associated with loss of weight since infancy. At 1year and 8 months of age child was admitted in the PICU with respiratory distress and impending respiratory failure for which he was ventilated for 1 week He was also hospitalized for recurrent pneumonia. He had received blood transfusions thrice for anemia, for which the exact details were not known. Child was treated for seizure disorder. Child was investigated, treated and followed up in a tertiary care hospital from infancy to 6 years of age .The investigations done in the past showed a normal total leucocyte count, differential count and platelet counts. Peripheral smear showed microcytic, hypochromic anaemia. Hemoglobin levels were consistently less than 8 gm/dl and the Hb electrophoresis was normal. Mantoux was negative. Resting Gastric Juice for mycobacterium tuberculosis was negative. Chest X-ray showed bilateral reticular /granular pattern at both lower zones. CT Chest showed diffusely distributed nodular opacities in both lower lung fields with interlobular septal thickening. Sweat chloride test was not suggestive of cystic fibrosis. Renal and liver function tests and urine examination were normal. Abdominal and renal ultrasound did not reveal any abnormality. Duodenal biopsy revealed no significant lesion (done as a work up for failure to thrive). ABG was within normal limits. As the family shifted their residence the child was lost to follow up for the past 5 years and now at 11years of age he was admitted to our centre.



Figure 1A
shows the clinical picture of the patient.



Figure 1B
Chest X-ray showing bilateral diffuse reticular infiltrates in both the lower lung fields.



Figure 1C
HRCT showing diffuse reticular opacities with septal thickening.

The child was admitted at our centre for increasing respiratory distress and fever. The child was reevaluated clinically and was found to have a weight of 20 Kgs (expected Wt.36Kgs) and a height of 138 cms (expected

height 143cms) and was chronically ill looking. Physical examination revealed pallor, clubbing and mild respiratory distress. Auscultation of the chest revealed , reduced air entry with fine crackles on both the infrascapular

areas. Examination of other systems was unremarkable. Investigations revealed hemosiderin laden macrophages by pearls staining suggestive of pulmonary hemosiderosis in the broncho alveolar lavage (BAL). High resolution computed tomography (HRCT) shows diffuse reticular opacities (inter & intralobular septal) thickening in both the lung fields more prominent in bilateral bases, apices and sub pleural region. Superimposed para septal emphysematous changes predominantly in anterior segment of bilateral upper lobes, medial segment of right middle lobe and basal segments of both lower lobes. Pulmonary function test showed a very severe restrictive pattern. ECHO was normal. Antinuclear antibody (ANA) and anti neutrophilic cytoplasmic antibody (ANCA) results were negative. Based on the above clinical findings like progressive respiratory symptoms, failure to thrive, anemia and with BAL and HRCT findings a diagnosis of pulmonary hemosiderosis leading to ILD was made. The child is on hydroxyl chloroquine and is on regular follow up.

DISCUSSION

Idiopathic pulmonary hemosiderosis commonly occurs in children less than 10 years of age ¹. The classical triad of IPH is anemia, hemoptysis and pulmonary infiltrates but anyone of the them could be the initial manifestation. Iron deficiency anemia may be the first and the only manifestation of IPH, preceding other symptoms and signs by several months ⁴. This child presented initially with severe unexplained microcytic, hypochromic anemia requiring blood transfusion during infancy substantiating the diagnosis ⁵. Though hemoptysis may be one of the presentations, this child did not have any episode of hemoptysis ⁶. Some studies from India demonstrated cow's milk intolerance as one of the causes of pulmonary hemorrhage leading to IPH, however, no such

history was reported in this case ⁷. The child had no associated systemic symptoms like renal involvement, joint disease, cutaneous rashes and recurrent fever typical of collagen-vascular disorders. This was supported by the investigations like a normal renal function and normal urine examination and a negative ANA and ANCA antibodies results and hence a diagnosis of IPH was made. The long term prognosis of IPH is extremely variable. Two decades ago the mean survival was 3 years, while recent data indicates a 5-year survival rate in 86% of cases ⁶. The prognosis for patients with IPH seems to have improved over time. The early diagnosis and appropriate treatment with long term immunosuppressive treatment has resulted in the long term survival of this child. Pulmonary hemosiderosis causes recurrent episodes of subclinical diffuse alveolar hemorrhage (as evidenced by the fact that this child required repeated blood transfusions) leading to fibrosis and restrictive lung disease. A diagnosis of IPH progressing on to ILD was made as the child had a gradual progression of pulmonary symptoms with typical radiological findings on HRCT ⁸. Interstitial lung disease (ILD) appears to be under reported in India ⁹. Secondary causes of ILD can be diagnosed early if there is a high index of suspicion and a methodical approach.

CONCLUSION

The long term prognosis of IPH is extremely variable. Two decades ago the mean survival was 3 years, while recent data indicates a 5-year survival rate in 86% of cases. The prognosis for patients with IPH seems to have improved over time. The early diagnosis and appropriate treatment with long term immunosuppressive treatment has resulted in the long term survival of this child. Interstitial lung disease (ILD) appears to be under reported in India. Secondary causes of ILD can be diagnosed early if there is a high index of suspicion and a methodical approach.

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