CASE REPORT

IDIOPATHIC PULMONARY HAEMOSIDEROSIS

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ABSTRACT

We report a case of idiopathic pulmonary hemosiderosis (IPH) in a 8year old female who presented with anaemia, breathlessness, puffiness of face and grade 4 clubbing. She had been investigated a number of times at multiple hospitals and had received multiple blood transfusions considering iron deficiency anaemia. Diagnosis of IPH was suspected on basis of clinical history of persistent respiratory distress, hypoxemia, repeated anaemia episodes and diffuse bilateral infiltrates on chest X ray.

Keyword: Anaemia, Haemosiderosis, Breathlesness, Haemoptysis.

INTRODUCTION

Idiopathic pulmonary hemosiderosis (IPH) is a rare disorder of unknown etiology, characterized by iron deficiency anemia, recurrent or chronic pulmonary symptoms such as cough and hemoptysis and diffuse pulmonary infiltrates. The incidence is reported from 0.24 to 1.23 cases per million in selected populations. The clinical course of IPH is exceedingly variable, anemia, coughing and radiological evidence of pulmonary infiltrates are seen in most of the affected individuals but in the absence of pulmonary problems diagnosis and treatment are delayed. We report a case with a misleading presentation of iron deficiency anemia.

CASE REPORT

A 8year old girl was admitted to our hospital suffering from fatigue, pallor, anorexia and breathlessness. There was no history of hemoptysis or bleeding from any site. She had been investigated at a number of other hospitals and diagnosed as having iron deficiency anemia. The girl had received multiple blood transfusions and iron preparations. Physical examination revealed a child with extreme pallor, respiratory rate 68, a normal precordium with a grade 2/6 systolic ejection murmur, pulmonary auscultation b/l crepitations. The liver and spleen were not enlarged. She was severly undernourished. Laboratory findings showed a severe microcytic anemia with hemoglobin (Hb) 2.7 gr/dl, hematocrit (Hct) 17.8%, mean corpuscular volume(MCV) 64 fl, leukocyte count 4.94 *10^3/cmm , platelet count 441*10^3/cmm , serum iron 42mg /dl, serum iron binding capacity 361 mg /dl, and ferritin 47.4 ng/ml.Retic count 0.2%. No abnormalities indicating hemolysis, chronic infection, malignancies, marrow dysfunction, hemoglobinopathies or coagulation disorders were found. Because of the finding of positive occult blood in feces. This led to a conclusion of iron deficiency most likely due to blood loss. The patient received blood transfusion. Six months later we saw her again with anemia, fatigue and pallor. Severe microcytic anemia was again present. The laboratory findings were as follows; Hb 3.6 gr/dl, Hct %12.9, MCV 66.5 fl, RDW:24.3, serum iron 12 mg /dl, serum iron binding capacity 344 mg /dl, and ferritin 133 mg /ml. We considered the probability of pulmonary hemosiderosis. A computerized tomography (CT) scan of the thorax showed diffuse alveolar interstitial infiltration . Gastric aspiration remained negative, so we went for bronchoalveolar lavage which was positive for macrophage containing haemosiderin and pulmonary hemosiderosis was diagnosed.

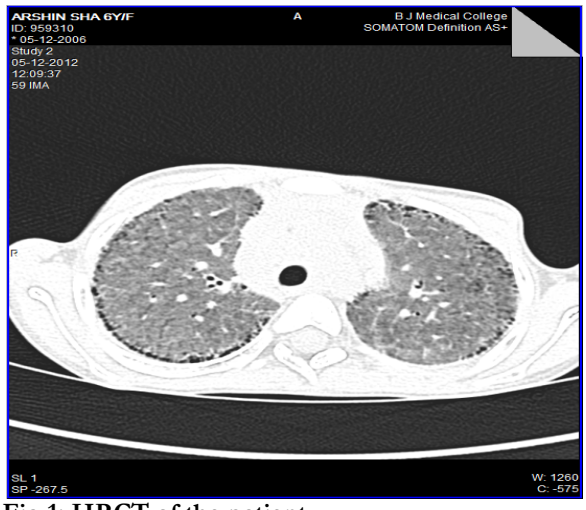

Fig 1: HRCT of the patient

DISCUSSION

IPH is a rare disorder characterized by recurrent or chronic diffuse alveolar hemorrhage and accumulation of hemosiderin in the lungs. The anemia caused by blood loss is enforced by the iron deficiency and pre-

sents as a microcytic anemia. Individuals of any age can be affected

and it occurs with equal frequency in males and females. Although its etiology remains unknown, IPH is considered to be an immune mediated disease. The presence of ANCA is thought to be a sign of poor diagnosis for pulmonary progression. The presence of other antibodies seems to be the predictor for development of an immune response. Several cases were reported with iron deficiency anemia, pulmonary symptoms including cough, dyspnea and hemoptysis, diffuse paranchymal infiltrates on chest radiographs. The initial presentation with iron deficiency anemia, occult blood loss and erosive gastritis was misleading. Kipper et al

noted there was a long delay (4 months- 10 years) between the starting of symptoms and the time of correct diagnosis. The diagnosis can be confirmed by demonstration of hemosiderosin-laden macrophages in the gastric aspirates or BAL. Biopsy is essential for diagnosis if BAL results are cleared within two weeks. Studies indicate that patients show varied response to immunosuppressants. Early active and extended courses of immunosuppressive therapy (prednisone, hydrocloroquine, azothıoprıne, cyclophosphamide) may improve the prognosis. Saeed et al found that 5 year survival of patients with IPH was 86% because of long term therapy with immune supressants. Treatment is continued according to patient's needs. We concluded that IPH should be considered when investigating a possible cause of iron deficiency anemia especially in patients who require multiple blood transfusions. A more rapid diagnosis could have prevented unnecessary laboratory analyses and blood transfusions.

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