



CASE REPORT

IDIOPATHIC PULMONARY HEMOSIDEROSIS- A CASE REPORT

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ABSTRACT

We report a case of idiopathic pulmonary hemosiderosis (IPH) in a 12 year old boy who presented with anemia without any pulmonary symptoms. He had been investigated in a number of hospitals and had received multiple blood transfusions for iron deficiency anemia. On his first admission he was misdiagnosed as having gastrointestinal bleeding because of hematemesis and occult blood in the feces. Six months later, a history of coughing blood guided us to the probability of pulmonary hemosiderosis. The diagnosis of IPH was made after a lung biopsy showed a number of hemosiderin-filled macrophages in the alveoli.

Keywords: Hemosiderosis, Iron, Hemoptysis, Pediatrics

İDYOPATİK PULMONER HEMOSİDEROSİS TANILI OLGU SUNUMU

ÖZET

Bu yazıda; hiçbir pulmoner semptomu olmadan anemiyle başvuran ve idyopatik pulmoner hemosiderosis tanısı alan 12 yaşındaki erkek olgu sunulmuştur. Hastaya daha önce çok sayıda hastanede çeşitli tetkikler yapılmış ve demir eksikliği anemisi için çok sayıda kan transfüzyonu uygulanmıştı. Hasta ilk başvurusunda hematemezi ve gaitada gizli kan varlığı ile gastrointestinal kanama tanısı aldı; ancak 6 ay sonra tariflediği hemoptizi anemnezi bizi pulmoner hemosideroz olasılığına yönlendirdi. Akciğer biopsisi sonucunda alveollerde hemosiderin yüklü makrofajların görülmesiyle idyopatik pulmoner hemosiderosis tanısı koyuldu.

Anahtar Kelimeler: Hemosiderosis, Demir, Hemoptizi, Çocukluk çağı

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 infiltrates¹. The incidence is reported from 0.24 to 1.23 cases per million in selected populations². 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 reported a case with no pulmonary signs and with a misleading presentation of iron deficiency anemia, due to hematemesis and occult blood loss in the feces.

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CASE REPORT

A 12 year old boy was admitted to our hospital suffering from fatigue, pallor, anorexia and blood in his vomit. There was no history of hemoptysis or dyspnea. He had been investigated at a number of other hospitals and diagnosed as having iron deficiency anemia. The boy had received multiple blood transfusions and iron preparations.

Physical examination revealed a child with extreme pallor, normal respiratory rate, a normal precordium with a grade 2/6 systolic ejection murmur, normal pulmonary auscultation. The liver and spleen were not enlarged. His weight and height were normal for his age. Laboratory findings showed a severe microcytic anemia with hemoglobin (Hb) 4.7 gr/dl, hematocrit (Hct)

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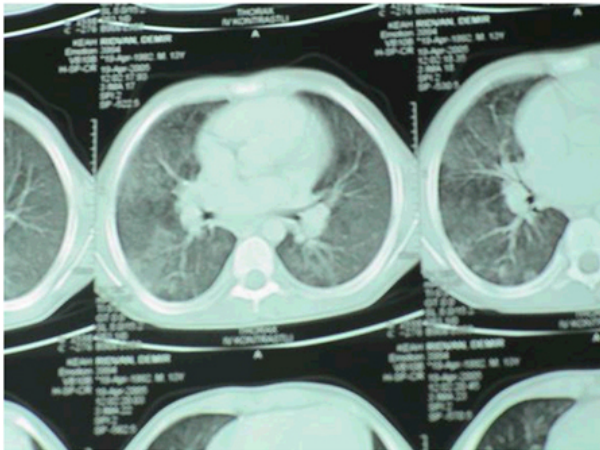


Fig. 1-a: A.C.T. scan of the thorax showing diffuse alveolar interstitial infiltration

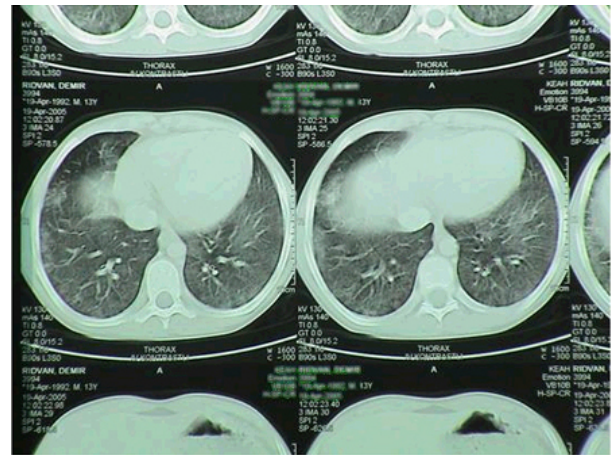


Fig. 1-b: A.C.T. scan of the thorax showing diffuse alveolar interstitial infiltration

17.8%, mean corpuscular volume(MCV) 66 fl, leukocyte count $6.8 \times 10^9/L$, platelet count $398 \times 10^9/L$, serum iron 10mg /dl, serum iron binding capacity 361 mg /dl, and ferritin 47.4 ng/ml. 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 and the history of hematemesis, a gastroduodenoscopy was performed and antral gastritis and erosive duodenitis was found. Helicobacter pylori was cultured and shown on biopsy. This led to a conclusion of iron deficiency most likely due to blood loss. The patient received blood transfusion and medical treatment for the helicobacter pylori.

Six months later we saw him 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. At this time the boy told us that he had seen some blood after coughing. We considered the probability of pulmonary hemosiderosis. A computerized tomography (CT) scan of the thorax showed diffuse alveolar interstitial infiltration (Figs. 1a- 1b). Gastric aspiration and BAL for macrophages containing hemosiderin remained negative, so biopsy was performed and pulmonary hemosiderosis was diagnosed. For differential diagnosis of idiopathic pulmonary hemosiderosis antinuclear antibody (ANA), antineutrophil cytoplasm antibodies (ANCA), RAST for cow's milk were checked and were found negative. The levels of complement C3 and C4 were normal.

Therapy with prednisolone (2 mg/kg/day) was initiated. The patient's clinical course improved after starting treatment and he was followed up.

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 presents as a microcytic anemia. Individuals of any age can be affected and it occurs with equal frequency in males and females^{3,4}.

Although its etiology remains unknown, IPH is considered to be an immune mediated disease¹. The presence of ANCA is thought to be a sign of poor prognosis 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^{2,5}. Diagnosis can be delayed in patients without pulmonary symptoms^{6,7,8}. We had a six month delay before diagnosis and treatment because our patient had no respiratory findings on his first admission. The initial presentation with iron deficiency anemia, hematemesis, occult blood loss and erosive gastritis was misleading. Kipper et al³ 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 hemosiderin-laden macrophages in the gastric aspirates or BAL. Biopsy is essential for



diagnosis if BAL results are cleared within two weeks.^{9,10}

In our patient IPH was confirmed by the finding of hemosiderin filled macrophages in lung biopsy material as in other studies^{2,7}.

Studies indicate that patients show varied response to immunosuppressants. Early active and extended courses of immunosuppressive therapy (prednisone, hydrochloroquine, azothioprine, cyclophosphamide) may improve the prognosis. Saeed et al² found that 5 year survival of patients with IPH was 86% because of long term therapy with immunosuppressants. 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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