An Uncommon Cause of Hemoptysis Presenting Like Hemolytic Anemia

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Abstract

Idiopathic pulmonary hemosiderosis (IPH) usually presents with hemoptysis and iron deficiency anemia. In this report, an unusual presentation of IPH is described. 11-year-old boy presented with clinical features suggestive of hemolytic anemia. However, on detailed evaluation, he was diagnosed to have IPH and confirmed by the presence of hemosiderin-laden macrophages in bronchoalveolar lavage fluid (BALF). The child was managed with oral corticosteroids, and he was doing well on follow-up.

Keywords: Hemolytic anemia; Hemoptysis; Idiopathic pulmonary hemosiderosis; Bronchoalveolar lavage fluid; Hemosiderin-laden macrophages

Introduction

IPH is an unusual condition that is responsible for chronic incidents of diffuse alveolar hemorrhage in children. The clinical manifestation is characterized by a triad of iron deficiency anemia, hemoptysis, and pulmonary infiltrates on chest imaging. Children with IPH appear to experience a rapid course and have a worse prognosis than adults more commonly. Diagnosis is confirmed by demonstrating the presence of hemosiderin-laden macrophages in bronchoalveolar lavage fluid (BALF) or lung biopsy specimens[1].

In this report, we describe an unusual presentation of IPH in which a child who initially presented with a clinical picture of hemolytic anemia, was finally diagnosed as IPH based on the detailed evaluation.

Case presentation

11-year-old boy was admitted for evaluation of severe anemia. Anemia had been noted three months earlier in an
outside hospital, and since then he had received three packed cell transfusions. There was also a history of hemoptysis off and on for the last six months, although no history of bleeding from any other site. There was no history of fever, weight loss, chest pain, or other constitutional symptoms. There was no significant history or history of contact with tuberculosis (TB).

**Clinical findings**

On examination, there was severe pallor and icterus. Height and weight were below the third centiles, and a BCG scar was present. There was no cyanosis, clubbing, or lymphadenopathy. System examination revealed hepatosplenomegaly. The diagnosis of hemolytic anemia was considered based on the clinical findings of hepatosplenomegaly with anemia and icterus. A final clinical diagnosis of hemolytic anemia with hemoptysis was made.

**Investigations**

Investigations revealed low hemoglobin percent (3.2 gm%), normal total leukocyte count (9600 cells/mm³), elevated platelet count (6,50,000 cells/mm³), low mean corpuscular volume (MCV) (66.1 fL), elevated reticulocyte count (5%), hyperbilirubinemia (total 3.94 mg/dL, direct 0.42 mg/dL), elevated lactate dehydrogenase (LDH) (345 U/L). Peripheral smear, however, showed microcytic, hypochromic RBCs, normal WBCs, thrombocytosis, and there was no evidence of hemolysis. The corrected reticulocyte count (0.43%) was within the normal range. The levels of serum iron (35 mcg/dl) and serum ferritin (5 µg/l) were low.

Chest X-ray showed diffuse alveolar infiltrates. Based on these clinical and lab findings, pulmonary hemosiderosis was suspected. BALF examination confirmed the presence of hemosiderin-laden macrophage (Figure 1). CT thorax revealed features of chronic pulmonary hemorrhage.

![Figure 1: Bronchoalveolar lavage showing hemosiderin- laden macrophages](image-url)
Differential diagnosis

Further workup for gluten enteropathy and other causes of pulmonary hemorrhage like TB, vasculitis, cardiovascular disease, and connective tissue disorders were negative.

Therapeutic interventions

A final diagnosis of IPH was made. The child was started on oral steroids and hematinic.

Outcome and follow-up

Hemoptysis resolved within one week of starting steroids, hemoglobin was steadily improving, and the child was doing well on follow-up.

Discussion

IPH is a rare disease found primarily in children that causes recurrent episodes of diffuse alveolar hemorrhage. The disease is characterized by the triad of hemoptysis, iron deficiency anemia and alveolar infiltrates\(^2\). However, this child presented with features of hemolytic anemia. Such a presentation has also been previously described\(^3,4\). Following an acute bleeding episode, a high number of reticulocytes may be present in the blood. Absorption of hemoglobin from the lungs induces a rise in plasma bilirubin. Therefore, in many aspects, IPH can mimic hemolytic anemia\(^5\). Once corticosteroids control the bleeding episodes, the blood picture returns to normal, just as in our case. Prognosis in cases of IPH is variable. Most of the patients with IPH seem to respond favorably to chronic oral corticosteroids\(^6\). Other immunosuppressant agents, including azathioprine, hydroxychloroquine, cyclophosphamide, and methotrexate, have been tried with variable results.

Conclusion

IPH can present with clinical features of hemolytic anemia. Diagnosis can be confirmed by the presence of hemosiderin-laden macrophages in BAL or lung specimens.

Patient consent: N/A
Conflict of Interest: Nil
Financial Disclosure: None
References