Case Report

Idiopathic Pulmonary Hemosiderosis

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ABSTRACT

Idiopathic pulmonary hemosiderosis (IPH) is a rare disorder that is usually characterized by the triad of bilateral pulmonary infiltrates, hemoptysis and iron deficiency anemia. The disease is well known to affect the pediatric age group with conflicting treatment trials. We report a case of a 19 year-old girl with IPH and respiratory failure who had excellent initial response to systemic steroids. To the best of our knowledge this is the first reported case in this age group in the region.

KEY WORDS: anemia, hemoptysis, immunosuppressive agents, steroid

INTRODUCTION

Idiopathic pulmonary hemosiderosis (IPH) is a rare disorder that is usually characterized by the triad of bilateral pulmonary infiltrates, hemoptysis and iron deficiency anemia. The disease is well known to affect the pediatric age group with conflicting treatment trials. We report a case of a 19 year-old girl with IPH and respiratory failure who had excellent initial response to systemic steroids. To the best of our knowledge this is the first reported case in this age group in the region.

Her anemia was severe enough to require blood transfusion twice. She denied any skin rash, joint pain or swelling, mouth or genital ulcers.

On admission to the referring hospital, she was febrile, tired and short of breath. Her initial chest X-ray showed bilateral alveolar infiltrates (Fig. 1). She was initially managed as community acquired pneumonia. Soon after admission, her condition deteriorated with marked hypoxemia requiring 70% inspired oxygen. CT chest revealed bilateral diffuse infiltrates, mixed alveolar opacities and interstitial infiltrates.

She was then transferred to our facility for further workup and management. On arrival she was significantly hypoxic, but hemodynamically stable. On a FIO₂ of 0.7 her O₂ saturation was only 91%. She was admitted to the intensive care unit and started on broad spectrum antibiotics (maxipime and intravenous erythromycin). Bronchoscopy was performed and transbronchial biopsies were taken. Her bronchoalveolar lavage done prior to the biopsy was consistently bloody. Full immunological investigations were sent to exclude any pulmonary vasculitis including: ANA, C and P ANCA, Anti-GBM, complements and rheumatoid factor. These were all negative except for a non-specific weak positive C ANCA and ANA. The transbronchial biopsy showed extensive hemosiderin in the tissue as well as hemosiderin laden macrophages (Fig. 2). There was also marked interstitial fibrosis with no evidence of vasculitis or capillaritis.

CASE REPORT

Ms. F is a 19-year-old girl, who was transferred to Mubarak hospital from another institution with respiratory failure. Her illness started five days prior to her presentation with fever, cough and shortness of breath. She described episodes of hemoptysis during her illness with few blood clots occasionally. On further questioning, she described similar attack three years ago, where she had been coughing streaks of blood and was febrile. This was followed by a protracted course of mild hemoptysis for almost one year followed by spontaneous resolution. Since then she continued to feel short of breath on exertion, but attributed it to the iron deficiency anemia that she suffered from.

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During her admission she had a 2D echo which was normal. On reviewing her old chest X-Ray done in 2003 (Fig. 3), it did show a large air space lesion on the right lower lung zone (which in retrospect could represent alveolar hemorrhage). She was started on systemic steroid at a dose of prednisolone 40 mg daily with complete resolution from a radiological as well as clinical aspect. Her maintenance treatment consisted of steroids with chloroquine.

When her steroids were tapered down to 10 mg over four months, she started to experience multiple episodes of minor hemoptysis, none of which was clinically significant. Currently, azathioprine has been added as steroid sparing agent.

DISCUSSION

IPH is a rare disease with an estimated incidence of 0.24 -1.23 cases per million in the pediatrics population[4]. It is usually diagnosed after combining clinical and radiological parameters with exclusion of more common disorders that leads to pulmonary hemorrhage. Clinically IPH manifest with a triad of pulmonary infiltrates, iron deficiency anemia, and hemoptysis. An absolute requirement for the diagnosis is identification of hemosiderin laden macrophages in sputum, bronchoalveolar lavage, lung biopsy specimens and gastric washings[5]. Multiple blood transfusion for severe anemia in IPH patients has been reported in the literatures[6]. Our patient was labeled as iron deficiency anemia and required blood transfusion twice. The exact etiology of IPH is unknown, although most therapeutic attempts used immunosuppressive agents for treatment[7].

Treatment is based on case series, but in the initial phase of presentation, steroids in high dose (e.g., prednisone 2 - 5 mg/kg/d or equivalent) are considered the treatment of choice[4,8]. This has proven to control the acute bleeding episode and decrease the frequency of pulmonary hemorrhage. However, the long term benefit is still controversial. A retrospective review of 23 children diagnosed with IPH in whom steroids in low-dose have been tried after initial high-dose on presentation, showed prevention of crises and milder disease course[6]. However, there are some patients who fail to respond to steroids alone and in whom another form of immunosuppression has been tried.

Such therapies include azathioprine, chloroquine, or cyclophosphamide. There are several case reports and retrospective studies that showed beneficial effect of azathioprine in long
term control of symptoms and as a steroid sparing agent[5,9,10]. However, mortality benefits are difficult to prove in such types of publications. The usual starting dose varies from 1 to 2 mg/kg daily or on alternative days. Chloroquine has long been the favored immunosuppressive therapy of choice due to acceptable side effects profile. There are several case reports of the effectiveness of chloroquine, where it has been used for long term control of pulmonary hemorrhage[11,12]. The reported dosage ranged from 200 to 400 mg daily, but retinal changes were detected in one patient on 400 mg of chloroquine, which resolved after discontinuation of the drug. Cyclophosphamide, on the other hand, is the least used immunosuppressive therapy reported[7]. The range of dosage in these reports was from 1.5 to 2.5 mg/kg/d.

Given all these data, the main concern in choosing between the different regimens of immunosuppression would be the side effect profile of the drug and patient tolerance. The five years survival of IPH has been recently reported to be 86%, in contrast to previous reports of 2.5 years average survival[10]. There are different reports of favorable survival factors but with conflicting results.

Lastly, the presence of c-ANCA in low titre in the serum of our patient, although not associated with systemic or pathological manifestation of vasculitis, has been reported in the literature[13]. It has been linked to classifying IPH according to severity, and was thought to represent a prognostic value. This, though has not been consistent through the literature.

REFERENCES