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Case Report

Idiopathic Pulmonary Hemorrhage in Saudi Girl Mimicking Iron Deficiency Anemia

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Abstract

We report an extremely rare case of a 7-years-old girl diagnosed with IPH at the age of 18 months successfully treated with azathioprine which, to the best of our knowledge, is the first case in Saudi Arabia.

The girl was found to have hemolytic anemia with diffuse fluffy bilateral shadows in chest x-ray in addition, hemosiderin-laden macrophages in the bronchial wash were positive. Workup for vasculitis, immune diseases, and connective tissue disorders were negative. The patient was started on Methylprednisolone and azathioprine which showed a notable effect in reducing the frequency and severity of the pulmonary hemorrhage in comparison to prednisolone alone.

This case was reported to highlight the importance of attention to detailed history in suspected cases and to demonstrate the need for IPH management in the pediatric age group as there is no standard management

Keywords: Idiopathic Pulmonary ; Hemorrhage ; Iron Deficiency Anemia

Introduction

Diffuse alveolar hemorrhage (DAH) is rare, a life-threatening condition that constitutes dyspnea, hemoptysis, and alveolar infiltration in imaging studies. The causes for DAH are variable from primary and secondary causes. Idiopathic pulmonary hemosiderosis (IDH) is a rare cause of diffuse alveolar hemorrhage which usually presents with repeated episodes of hemoptysis, diffused pulmonary infiltration, and iron deficiency anemia. The first clinical presentation is usually iron deficiency anemia since it is a common presentation on children, which renders the early diagnosis of IDH quite difficult then proceeded by other symptoms [1]. The exact etiology of IDH is still unknown, but few proposed theories exist. Some studies suggest genetic background since they found a higher incidence of IDH with consanguineous parents (2). In our case whole exome sequencing was negative. In addition, studies suggested allergic causes because some reported cases of IDH have been linked with celiac disease and the starting of a gluten-free diet led to complete remission in these cases [2]. The estimated patients of IDH reported were 0.24 per million per year in a study published in Sweden and 1.24 per million per year in a retrospective study in Japan [2]. IDH is more common in children; thus, only a few reported cases in adults. This study observes the

diagnosis and treatment of a 2-year-old girl who presented with respiratory distress in the emergency department.

Case Presentation

A 2 years-old Saudi girl presented to our emergency department with a cough, shortness of breath, and posttussive bright red color sputum. Her parents reported a history of chronic cough for the last one year, which was not relieved by medication. In addition, the patient was admitted to our hospital one month back as a case of hemolytic anemia, in which she received a blood transfusion. Regarding her past medical history, the patient had a severe pneumonia at the age of one month. Parents denied any history of seizures, rash, joint swelling, epistaxis, hematuria, oliguria, ear discharge, diarrhea, or skin infection.

Upon admission to the emergency department, the patient received a blood transfusion as her blood hemoglobin (Hb) level was 5.5, then she developed three episodes of frothy sputum with bright color blood followed by increased respiratory distress. She was shifted to the pediatric intensive care unit (PICU), where she was intubated. During intubation, blood was seen coming through the endotracheal tube. The physician started conventional ventilation, but the patient situation deteriorated; thus, high-frequency oscillatory ventilation (HFOV) was started for two days. The patient improved and was shifted back to the conventional ventilation.

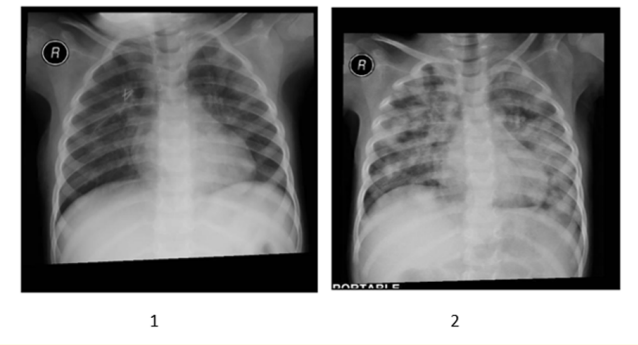

Figure 1 and 2: Posterioranterior chest radiography revealing acute phase of pulmonary infiltrations of idiopathic pulmonary Hemorrhage.

For her anemia, blood findings showed HB 9. Normal PT, PTT, and INR. ESR 3 mm/h, normal Serum ferritin, at PICU she received blood transfusion. immunological, and connective tissue disorders workup was negative, which included: negative ANA, ds-DNA, PANCA, and CANCA antibodies, glycoprotein IgG/IgM, anticardiolipin IgG/IgM, Tb direct PCR test, celiac profile, Hepatitis B serology, and anti-GBM. HIV-1/HIV-2 immunoassay test was negative. Serum biochemistry values were within normal limits. For alveolar hemorrhage, hemosiderin-laden macrophages in the bronchial wash were positive. Chest x-rays were obtained, which showed diffuse fluffy bilateral shadows. To exclude vasculitis, capillaritis, and Systemic lupus erythematosus (SLE), a lung biopsy was ordered, which was negative. MRI brain and EEG showed no abnormality. The patient was further sent for Nasopharyngeal aspiration which appeared entirely normal. On the basis of these findings, an Idiopathic pulmonary hemorrhage (IPH) diagnosis was made.

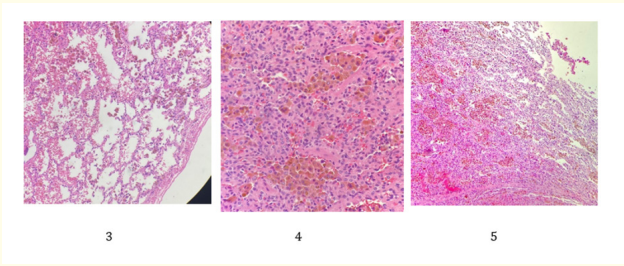

Figure 3-5: Lung biopsy specimen showing hemosiderin laden macrophages with intra-alveolar hemorrhage with no evidence of vasculitis, capillaritis, necrosis, or granuloma.

The patient was started with methylprednisolone pulse therapy for three days which showed an improvement; hence the patient was extubated and switched to low dose prednisolone. After the extubation, the patient experienced brief episodes of seizures for which a neurology team opinion was sought; MRI and EEG showed no abnormality. Moreover, the patient was followed by multidisciplinary teams including Infectious diseases, hematology-oncology, neurology, pulmonology, rheumatology, and immunology. The patient was kept in the PICU for observation, where she developed a pneumomediastinum that was resolved spontaneously. In addition, the patient received Intravenous

Ceftriaxone and Clindamycin. The patient was discharged home on a daily low dose (5 mg) of prednisolone.

Our patient now is 7 years old and doing fine with follow up in the clinic. Furthermore, all connective tissue markers were resent again without any positive result yet.

Discussion

Idiopathic pulmonary hemosiderosis (IDH) is a rare cause of diffused alveolar hemorrhage with unknown etiology up to this day. Few studies proposed different theories, including autoimmune, allergic, inherited, metabolic, and exposure to mycotoxin [2]. Idiopathic pulmonary hemosiderosis is usually characterized by a triad of iron deficiency anemia, respiratory symptoms (dyspnea, cough, hemoptysis), and chest infiltrate on chest radiograph [1,3]. Although these symptoms are reported in most of the cases of IPH, dyspnea, and iron deficiency anemia remain the most common symptoms reported [1,3]. Idiopathic pulmonary hemosiderosis is still a diagnosis of exclusion that requires high clinical suspension and knowledge about the possible differential diagnosis for diffuse alveolar hemorrhage. There are two stages for the diagnosis of IPH. The first one confirms the presence of diffused alveolar hemorrhage, which includes a multi-step investigation to prove it proceeded by the second stage to rule out the other causes of diffuse alveolar hemorrhage [2,4]. The management for IPH and response to treatment varies according to each case. The variability ranges from lack of control of the symptoms that require assisted ventilation in PICU to spontaneous remission in some cases [5]. In the acute phase of the IDH, most physicians reported using corticosteroids and other life support management when needed [5]. There is a lack of studies towards which medication should be used to treat IPH. Still, some studies showed that using systemic corticosteroids alone or with other immunosuppressive agents decreases the attacks' severity and frequency [4-6]. In our case, the patient showed a better response using methylprednisolone and azathioprine reducing the severity of the hemorrhage and its frequency. The IDH tends to be more prolonged and less severe in adults, and they respond better to corticosteroids [5]. On the other hand, children and adolescents showed more rapid disease progression and severe symptoms [5]. It is difficult to estimate the prognosis of IDH because of the lack of studies and lack of follow-up, but some studies suggested the overall survival ranges from 2.5-5 years [5,7,8].

Conclusion

The main obstacles we confront when attempting to prove the success of various treatments are the wide variety of clinical manifestations of IPH, the intermittent character of the disease, and a lack of knowledge about the mechanisms involved in its etiology. In the present study, the diagnosis of IPH was made on the basis of excluding all the possible causes of pulmonary hemorrhage. Methylprednisolone and azathioprine a notable effect in reducing the frequency and severity of the pulmonary hemorrhage in comparison to prednisolone alone, we suggest conducting further research to evaluate the efficacy of azathioprine in patients diagnosed with IPH.

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