CASE REPORT

Idiopathic pulmonary haemosiderosis: an unusual case of anaemia with pulmonary involvement

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Abstract

Idiopathic pulmonary haemosiderosis is a rare disorder, with recurrent life-threatening alveolar haemorrhages and chronic lung parenchymal changes. It is associated with a triad of haemoptysis, iron deficiency anaemia, and diffuse pulmonary infiltrates. Although most cases are idiopathic, secondary haemosiderosis linked to known diseases has also been observed. Most of the cases remain undiagnosed because the disease is very low on the list of differentials.

There is no specified age for the disease. The present study reports on an adolescent female patient who presented with microcytic anaemia and bilateral lung infiltrates to the National Institute of Child Health (NICH), Karachi, a tertiary care hospital. She was diagnosed with Idiopathic pulmonary haemosiderosis after ruling out other possibilities.

Keywords: Pulmonary haemosiderosis, Alveolar haemorrhages, Lung disease, Alveolar hypoventilation syndrome.

DOI: 10.47391/JPMA.7323

Submission completion date: 15-11-2022
Acceptance date: 06-04-2023

Introduction

Pulmonary haemosiderosis resulting from diffuse alveolar haemorrhage is a life-threatening condition resulting from damage to pulmonary microvasculature and blood seepage into alveolar spaces. In children, it is mostly idiopathic, but it is also associated with many systemic autoimmune diseases like Wegener's granulomatosis, Systemic lupus erythematosus, and Goodpasture disease resulting in pulmonary capillaritis. Other conditions like cardiac and non-cardiac conditions can also lead to diffuse alveolar haemorrhage without capillaritis.[1]

Diagnosis of Idiopathic pulmonary haemosiderosis is a diagnosis of exclusion. Clinically, it can present acutely such as haemoptysis, dyspnoea, and worsening anaemia or it can present with chronic cough, dyspnoea, and repetitive episodes of haemoptysis leading to fatigue, anaemia, and failure to thrive. Radiological findings are mostly non-specific; however, bronchoalveolar lavage usually helps in the diagnosis of diffuse alveolar haemorrhage, though lung biopsy is recommended.[2]

Approximately 80% of the cases are reported in paediatric age group[3] and the exact prevalence of Idiopathic pulmonary haemosiderosis is generally unknown due to the rare nature of the disease. A study from Sweden estimated a prevalence of 0.24 million children per year.[4]

We present this case with the intention of adding to the existing literature and highlighting the importance of considering Idiopathic pulmonary haemosiderosis in the differential diagnosis if the patient presents with refractory iron deficiency anemia whether there are any pulmonary symptoms or not

Case Report

An adolescent female patient aged 6 years presented to National Institute of Child Health (NICH) Karachi, a tertiary care hospital, on March 7, 2020, with a history of repeated chest infections since the past six months. For the past eight months, she had fever off and on with a non-productive cough, and iron deficiency anaemia. She had been transfused fresh red blood cells six times along with multiple courses of oral antibiotics. Before reaching our hospital her workup for haemoglobinopathies, malaria, coeliac disease, and immunodeficiency had been done, and all were normal. On investigation, her serum ferritin level was elevated. She was referred to the NICH for further workup but before her appointment scheduled on March 9, 2020, she again developed fever which was high grade along with cough and severe respiratory distress, due to which she presented in the emergency department and was shifted to the ICU. Here she was electively intubated and put on a ventilator support.

On examination, she weighed 14 kg, her height was 108 cm, and had a heart rate of 128 beats/min, RR of 40 breaths/min, and oxygen saturation of 89% on pulse oximeter in room air without any oxygen support. Her

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cardiorespiratory examination showed an apex beat at the left 5th intercostal space near the midclavicular line accompanied by mild bilateral basal creptitations on both sides. Neurological and abdominal examinations were unremarkable.

On the first day, chest X-ray was done which showed bilateral pulmonary infiltrates with hyperinflation. Laboratory tests were performed and revealed microcytic and hypochromic anaemia (Hb:8.9 g/dL) with mildly elevated total leukocyte count showing increased eosinophils specifically and normal platelet count. A blood sample was obtained and sent for blood culture and sensitivity. The coagulation studies, hepatic, and renal function tests along with ultrasound and echocardiogram all were normal. The next day, CT of the chest was performed which showed bilateral pulmonary infiltrates along with ground glass haze at peripheral and infrahilar regions suggesting infective aetiology. PCR for Covid-19 was advised which was also negative. Her autoimmune workup was positive for ANA and negative for anti-dsDNA. Viral serology for HBV, HCV, and HIV was also negative. Bronchoscopy was performed to obtain bronchoalveolar lavage and sent for cytology which showed haemosiderin-laden macrophages. This finding supported the diagnosis of diffuse alveolar haemorrhage.

She was initially given intravenous Ceftazidime for six days along with IV Magnesium sulfate and Dexamethasone (0.2 mg/kg/QID). She was nebulized regularly, and packed cells were transfused. The blood C/S report showed Pseudomonas species resistant to Ceftazidime but sensitive to Vancomycin and Imipenem, therefore she was shifted to IV Meropenem and Vancomycin.

Her condition deteriorated day by day with off and on bleeding from the endotracheal tube for which IV vitamin K, transaminase, fresh frozen plasma, and activated Factor VII-a were administered. On day 12 of her admission, due to recurrent bleeding, tracheostomy was performed but she kept on bleeding and expired the next day due to cardiopulmonary arrest.

Discussion

Idiopathic pulmonary haemosiderosis is an unusual condition of recurring alveolar haemorrhage resulting in deposition of hemosiderin in lungs without a known cause and is a diagnosis of exclusion after ruling out primary and secondary causes of pulmonary haemosiderosis.[5] Clinically, it can present as haemoptysis, dyspnoea, and severe anaemia acutely to chronic cough and dyspnoea with recurrent haemoptysis leading to anaemia and failure to thrive. Some cases present with asymptomatic anaemia only.[6] A complete blood picture can show varying degrees of anaemia without any qualitative or quantitative platelet defect, kidney or liver injury, or any coagulopathies. Plasma ferritin levels can also be elevated.[7, 8] Only after all the possible sources of pulmonary haemorrhage have been ruled out, the diagnosis of idiopathic pulmonary haemosiderosis is made. Diagnosis of Idiopathic pulmonary haemorrhage is made with the help of lung biopsy, and it can also be made with a bronchoalveolar lavage if it reveals haemosiderin-laden macrophages. Bronchoalveolar lavage is recommended as it is less invasive with significant sensitivity and specificity.[8] Chest X-ray can be normal at the beginning but later it may reveal reticular and nodular opacities in perihilar and basal regions.[9]

Treatment depends on clinical presentation. Various trials support the role of high systemic steroids and inhaled steroids. Corticosteroid-resistant patients with pulmonary function deterioration can be treated with immunosuppressants like Azathioprine, Cyclophosphamide and Hydroxychloroquine therapy. Blood products are transfused to reverse anaemia. After diagnosis, a mean survival age of 2.5 to 5 years has been recorded, suggesting a guarded prognosis.[10, 11]

Conclusion

Idiopathic pulmonary haemosiderosis is an unusual condition that mostly affects children and has an unpredictable prognosis. It is, therefore, suggested that in cases where the patient suffers from iron deficiency anaemia and does not respond to usual treatments, Idiopathic pulmonary haemosiderosis should be considered even if no pulmonary symptoms are observed.

Consent: Written consent was obtained from the patient's father for publishing the case.

Disclaimer: None.

Conflict of Interest: None.

Source of Funding: None.

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