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Case Study

Idiopathic Pulmonary Hemosiderosis Presenting as Severe Iron Deficiency Anemia and Acute Respiratory Distress in A Six-Year-Old Child: A Rare Pediatric Case Report

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Idiopathic pulmonary hemosiderosis (IPH) is a rare and potentially life-threatening cause of diffuse alveolar hemorrhage in children, characterized by recurrent intra-alveolar bleeding that results in iron deficiency anemia, pulmonary infiltrates, and progressive respiratory impairment. Owing to its nonspecific clinical manifestations, the diagnosis is often delayed and confused with more common conditions such as pneumonia, pulmonary tuberculosis, asthma, or hematological disorders. We report the case of a six-year-old female who presented with cough, fever, progressive respiratory distress, generalized weakness, and severe pallor. On admission, she was hypoxemic and required oxygen supplementation with close monitoring in a pediatric high-dependency/intensive care setting. Clinical evaluation, laboratory investigations, and radiological findings revealed severe microcytic hypochromic anemia with bilateral diffuse pulmonary infiltrates. An extensive diagnostic workup was undertaken to exclude infectious, cardiac, autoimmune, hematological, and other secondary causes of diffuse alveolar hemorrhage. Based on the characteristic clinical presentation, supportive laboratory and imaging findings, and exclusion of alternative etiologies, a diagnosis of idiopathic pulmonary hemosiderosis was established. The patient was managed with systemic corticosteroid therapy, packed red blood cell transfusion, oxygen supplementation, and supportive care, resulting in marked clinical and hematological improvement during hospitalization. She was discharged on oral corticosteroids with advice for regular follow-up because of the chronic relapsing nature of the disease. This case highlights the importance of maintaining a high index of suspicion for idiopathic pulmonary hemosiderosis in children presenting with unexplained severe iron deficiency anemia and respiratory symptoms, even in the absence of hemoptysis, which may be absent in younger children. Early recognition, exclusion of secondary causes, and prompt initiation of immunosuppressive therapy are essential to reduce recurrent alveolar hemorrhage, prevent irreversible pulmonary fibrosis, and improve long-term prognosis. Reporting such rare pediatric cases contributes to increased clinical awareness and emphasizes the need for multidisciplinary management and long-term follow-up to optimize patient outcomes.

Keywords: Idiopathic pulmonary hemosiderosis, Diffuse alveolar hemorrhage, Iron deficiency anemia, Pediatric, Respiratory distress, Hemosiderin-laden macrophages, Corticosteroid therapy, Rare case report.

INTRODUCTION

Idiopathic pulmonary hemosiderosis (IPH) is a rare, potentially life-threatening disorder which is characterized by recurrent episodes of diffuse alveolar hemorrhage and accumulation of hemosiderin-laden macrophages within the alveoli. It is primarily observed in children and has an estimated annual incidence of less than one case per million and remains one of the rarest causes of chronic pulmonary disease in pediatric practice [1,2]. Due to its rarity and nonspecific clinical features, IPH is often

misdiagnosed as pneumonia, pulmonary tuberculosis, bronchial asthma and hematological disorders, leading to delayed diagnosis and treatment [1,3]. The classic clinical triad of IPH is hemoptysis, iron deficiency anemia, and diffuse pulmonary infiltrates. However, this triad is often incomplete in young children, as hemoptysis may be absent or unnoticed due to swallowing of blood. Severe iron deficiency anemia might be the dominant presenting feature, often with recurrent hospitalizations before the diagnosis is made [1,4]. The exact cause of IPH is unknown, but immune-mediated injury of the alveolar capillary basement membrane is thought to be an important factor in its pathogenesis. Several studies supporting the immune dysregulation hypothesis have reported associations with autoimmune diseases, especially celiac disease (Lane–Hamilton syndrome). Hence, screening for autoimmune disorders and celiac disease is recommended in all the newly diagnosed patients with IPH [1,5]. Diffuse alveolar hemorrhage is the main pathological process in IPH and should be considered in children with recurrent respiratory symptoms, unexplained anemia, or bilateral pulmonary infiltrates. Chest radiography and high-resolution computed tomography usually show diffuse ground-glass opacities or bilateral alveolar infiltrates. Bronchoalveolar lavage revealing hemosiderin-laden macrophages is considered the diagnostic investigation of choice. Lung biopsy is the last resort for diagnostically difficult cases after exclusion of secondary causes of pulmonary hemorrhage [1,6,7]. Systemic corticosteroids are the mainstay of IPH therapy and decrease recurrent alveolar hemorrhage and improve survival and remain the mainstay of therapy. In patients with relapsing disease or steroid-dependence, steroid-sparing immunosuppressive agents such as azathioprine, hydroxychloroquine, mycophenolate mofetil, cyclophosphamide, and rituximab have demonstrated promising results for long-term disease control [1,8,9]. Although treatment has improved, delayed diagnosis may lead to recurrent pulmonary hemorrhage, pulmonary fibrosis, chronic respiratory insufficiency and increased mortality, highlighting the necessity of early recognition and multidisciplinary management [1,10]. We report a case of a 6-year-old girl with idiopathic pulmonary hemosiderosis presenting with severe iron deficiency anemia and acute respiratory distress requiring intensive pediatric care. The case illustrates the diagnostic difficulty in this rare disorder and highlights the necessity of considering IPH in children with unexplained anemia and diffuse pulmonary infiltrates for early diagnosis, timely initiation of immunosuppressive therapy, and improved clinical outcomes.

MATERIALS & METHODS

Study Design: Single-patient retrospective case report

Study Setting: Department of Pediatrics, SSIMS Hospital, Davangere, Karnataka, India

Diagnostic Tools Used: Clinical examination, complete blood count (CBC), arterial blood gas (ABG) analysis, chest radiography, high-resolution computed tomography (HRCT) of the chest (if performed), and relevant laboratory investigations to exclude secondary causes of diffuse alveolar hemorrhage

Data Sources: Patient medical records, clinical presentation, physical examination, laboratory investigations, radiological findings, treatment records, and follow-up

Ethical Considerations: Written informed consent was obtained from the patient's parent/legal guardian for publication. Patient identity has been kept confidential in accordance with ethical guidelines

CASE PRESENTATION

A 6-year-old female was brought to the pediatric emergency department with complaints of fever, cough, progressive breathing difficulty, generalized weakness, and reduced activity for several days. The child had developed worsening respiratory distress associated with marked pallor, which prompted hospitalization. There was no documented history of foreign body aspiration, congenital heart disease, bleeding disorders, or known chronic respiratory illness. The past medical and family histories were otherwise non-contributory. On admission, the child appeared acutely ill with severe pallor and respiratory distress. She was tachypneic, hypoxemic, and required supplemental oxygen with continuous

cardiorespiratory monitoring in a pediatric high-dependency/intensive care setting. General physical examination revealed severe pallor with increased work of breathing. There was no significant cyanosis, clubbing, lymphadenopathy, edema, or skin rash. Respiratory system examination demonstrated bilateral decreased air entry with diffuse crackles, while cardiovascular, abdominal, and neurological examinations were otherwise unremarkable. Initial laboratory investigations revealed severe microcytic hypochromic anemia suggestive of iron deficiency. Additional hematological and biochemical investigations were performed to evaluate the underlying cause of anemia and respiratory compromise. Chest imaging demonstrated bilateral diffuse pulmonary infiltrates consistent with diffuse alveolar involvement. Arterial blood gas analysis showed hypoxemia requiring oxygen therapy. Microbiological investigations and further diagnostic evaluation were undertaken to exclude infectious etiologies, while cardiac and autoimmune assessments were performed to rule out secondary causes of diffuse alveolar hemorrhage. Considering the combination of severe iron deficiency anemia, diffuse bilateral pulmonary infiltrates, respiratory distress, and exclusion of infectious, cardiac, hematological, and autoimmune causes, a diagnosis of Idiopathic Pulmonary Hemosiderosis (IPH) was established. The clinical presentation was compatible with recurrent diffuse alveolar hemorrhage resulting in pulmonary hemosiderin deposition. The patient was managed with supplemental oxygen, systemic corticosteroid therapy, packed red blood cell transfusion, intravenous fluids, and supportive care according to institutional pediatric protocols. Continuous monitoring of oxygen saturation, respiratory status, and hematological parameters was performed throughout hospitalization. Following initiation of corticosteroid therapy, the child's respiratory distress gradually improved, oxygen requirement decreased, and her general clinical condition improved. Hematological parameters also showed improvement after transfusion and medical management. The patient remained hemodynamically stable during the remainder of her hospital stay and was discharged in an improved clinical condition with oral corticosteroid therapy and instructions for regular pediatric and pulmonology follow-up. The parents were counseled regarding the chronic relapsing nature of idiopathic pulmonary hemosiderosis, the importance of adherence to treatment, recognition of warning symptoms such as recurrent cough, hemoptysis, worsening pallor, or breathlessness, and the need for periodic monitoring to detect relapse and prevent long-term pulmonary complications

DISCUSSION

Idiopathic pulmonary hemosiderosis (IPH) is an uncommon but potentially life-threatening cause of diffuse alveolar hemorrhage (DAH) in children. It is characterized by recurrent bleeding into the alveolar spaces, leading to progressive accumulation of hemosiderin-laden macrophages, chronic iron deficiency anemia, and varying degrees of respiratory impairment. Although first described by Virchow in 1864 and subsequently recognized as a distinct clinical entity by Ceelen in 1931, IPH remains a diagnostic challenge because of its rarity and nonspecific clinical presentation. The estimated annual incidence in children is less than one case per million, with most cases diagnosed before 10 years of age, emphasizing the importance of maintaining a high index of suspicion in pediatric patients presenting with unexplained anemia and respiratory symptoms [1,2]. The hallmark clinical manifestations of IPH include the classical triad of hemoptysis, iron deficiency anemia, and diffuse pulmonary infiltrates. However, this triad is frequently incomplete in children because younger patients often swallow expectorated blood, making hemoptysis absent or difficult to recognize [1,3]. Consequently, severe iron deficiency anemia may precede pulmonary manifestations for several months, resulting in repeated misdiagnosis as nutritional anemia, recurrent pneumonia, pulmonary tuberculosis, or asthma. Similar to previous reports, our patient presented predominantly with severe pallor, respiratory distress, cough, hypoxemia, and diffuse pulmonary infiltrates, while the diagnosis was established only after exclusion of secondary causes of diffuse alveolar hemorrhage. This highlights the importance of considering IPH in children with persistent iron deficiency anemia associated with recurrent respiratory symptoms [1,4]. The exact pathogenesis of IPH remains uncertain. Current evidence supports an immune-mediated mechanism involving repeated injury to the alveolar capillary basement membrane,

resulting in recurrent intra-alveolar hemorrhage and subsequent deposition of hemosiderin within pulmonary macrophages [1,4]. Associations with autoimmune disorders, particularly celiac disease (Lane-Hamilton syndrome), rheumatoid arthritis, systemic lupus erythematosus, and antineutrophil cytoplasmic antibody (ANCA)-associated vasculitis, further support an immunological basis for the disease. Therefore, current recommendations advocate screening for autoimmune diseases and celiac disease in all newly diagnosed patients with IPH [1,4]. Diffuse alveolar hemorrhage represents the principal pathological process underlying IPH. The differential diagnosis includes pulmonary capillaritis, Goodpasture syndrome, ANCA-associated vasculitis, systemic lupus erythematosus, congenital heart disease, coagulation disorders, infections, and Heiner syndrome [3,10]. A systematic diagnostic approach is therefore essential to exclude secondary causes before confirming idiopathic disease. In our patient, extensive laboratory, microbiological, cardiac, and radiological evaluation excluded alternative etiologies, supporting the diagnosis of idiopathic pulmonary hemosiderosis. Radiological findings in IPH are generally nonspecific but provide valuable diagnostic clues. Chest radiographs typically demonstrate bilateral patchy or diffuse alveolar infiltrates during acute hemorrhagic episodes, whereas high-resolution computed tomography commonly reveals diffuse ground-glass opacities, interlobular septal thickening, and areas of consolidation. Recurrent episodes may eventually progress to pulmonary fibrosis and restrictive lung disease [6,11]. The radiological findings in our patient were consistent with diffuse pulmonary involvement, correlating with the clinical features of diffuse alveolar hemorrhage. Bronchoalveolar lavage demonstrating hemosiderin-laden macrophages remains the preferred diagnostic investigation because of its high sensitivity and relatively low invasiveness. Lung biopsy is reserved for diagnostically challenging cases or when pulmonary capillaritis cannot be excluded [1,10]. In clinical practice, diagnosis is frequently established through a combination of clinical presentation, characteristic imaging findings, laboratory evidence of iron deficiency anemia, exclusion of secondary causes, and response to corticosteroid therapy. Systemic corticosteroids remain the cornerstone of treatment for IPH and have substantially improved patient survival over the past several decades [1,12]. High-dose intravenous methylprednisolone is recommended during acute hemorrhagic episodes, followed by oral prednisolone with gradual tapering according to clinical response. Patients experiencing recurrent disease or corticosteroid dependence may benefit from steroid-sparing immunosuppressive agents such as azathioprine, hydroxychloroquine, cyclophosphamide, mycophenolate mofetil, or rituximab [8-12]. Our patient demonstrated significant clinical improvement following corticosteroid therapy, oxygen supplementation, blood transfusion, and supportive management, which is consistent with outcomes reported in previous pediatric case series. Despite improvements in diagnosis and therapy, IPH remains associated with considerable morbidity because of recurrent pulmonary hemorrhage, repeated blood transfusions, pulmonary fibrosis, chronic respiratory insufficiency, pulmonary hypertension, and impaired quality of life [1,6]. Long-term follow-up with serial clinical assessment, pulmonary function testing, chest imaging, and hematological monitoring is therefore essential to detect disease recurrence and optimize immunosuppressive therapy. Our case contributes to the limited literature on pediatric idiopathic pulmonary hemosiderosis by emphasizing the importance of recognizing unexplained severe iron deficiency anemia accompanied by respiratory symptoms as a potential manifestation of diffuse alveolar hemorrhage. Early diagnosis, exclusion of secondary causes, prompt initiation of corticosteroid therapy, and multidisciplinary follow-up are critical for reducing disease recurrence and improving long-term prognosis. Increased awareness among pediatricians, pulmonologists, intensivists, and hematologists may facilitate earlier diagnosis and prevent irreversible pulmonary complications in this rare disorder.

CONCLUSION

Idiopathic pulmonary hemosiderosis is a rare but potentially life-threatening cause of diffuse alveolar hemorrhage in children that frequently presents with nonspecific clinical features, leading to delayed diagnosis. This case highlights the importance of considering IPH in children presenting with

unexplained severe iron deficiency anemia, recurrent respiratory symptoms, and diffuse pulmonary infiltrates, even in the absence of hemoptysis. Early recognition, comprehensive exclusion of secondary causes, and prompt initiation of systemic corticosteroid therapy resulted in significant clinical improvement in our patient. Long-term follow-up remains essential because of the relapsing nature of the disease and the risk of progressive pulmonary fibrosis. Increased awareness among pediatricians, pulmonologists, intensivists, and hematologists is crucial for timely diagnosis and improved long-term outcomes.

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