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IDIOPATHIC PULMONARY HEMOSIDEROSIS (IPH)

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ABSTRACT
Idiopathic pulmonary hemosiderosis (IPH) is an uncommon cause of diffuse alveolar hemorrhage (DAH). Patients with IPH usually present with hemoptysis, and the diagnosis is often delayed by years. However, massive hemorrhage resulting in acute respiratory failure and non-remitting hemoptysis have also been described. The classic triad includes hemoptysis, radiologic lung infiltrate, and iron deficiency anemia. Several hypotheses regarding the pathogenesis of IPH have been proposed. These risk factors include an autoimmune, allergic or genetic predisposition, and possible environmental exposure. Since IPH appears to be responsive to corticosteroids, the autoimmune hypothesis is considered to play a crucial role. A diagnosis of IPH requires exclusion of other etiologies of DAH, including infection, medications, toxic inhalation, vasculitis, and anti-glomerular basement membrane disease, among others. Corticosteroid therapy represents the primary modality of treatment. Other immunosuppressive medications have also been used with varying success, especially in the setting of steroid-refractory disease. The prognosis of IPH in adults is somewhat better compared to the pediatric population. The severity of the initial presentation does not predict future outcomes. Which risk factors and patient characteristics are associated with a poor outcome are also unknown. More research is necessary to elucidate the pathophysiology and appropriate treatment.

Keywords: *Hemoptysis, Pulmonary hemorrhage, Idiopathic pulmonary hemosiderosis*

INTRODUCTION

Idiopathic Pulmonary Hemosiderosis (IPH) is a rare cause of diffuse alveolar hemorrhage (DAH). Although more common in the pediatric population, IPH can present at any age. Due to the rarity of the condition, a definitive diagnosis is often delayed and can cause significant distress for the patient and the families.

DEFINITION

Idiopathic pulmonary hemosiderosis (IPH) is a rare disease characterized by repeated episodes of a diffuse alveolar hemorrhage, which over time, can lead to multiple respiratory complications and permanent lung damage.

EPIDEMIOLOGY

The exact incidence and prevalence of IPH are currently unknown. Based on the literature, the disease appears to be more common in children compared to adults. Children represent about 80% of reported cases. The estimated incidence ranges from 0.24 to 1.23 cases per million among children.

RISK FACTORS AND PATHOGENESIS

Despite the long-standing identification of the disease entity, the risk factors and pathogenesis of IPH remains an enigma to clinicians. It is currently unknown whether the pathogenesis of IPH is similar in adults and pediatric patients. Several risk factors have been proposed with varying degrees of supporting evidence. These include:

1. Genetic predisposition
2. Environmental exposure
3. Immunologic association
4. Allergic reaction

SYMPTOMS AND SIGNS

- Hemoptysis (intermittent episodes of blood-streaked sputum)
- Life-threatening hemorrhage with the development of acute respiratory failure
- Dyspnea and anemia.

During an episode of alveolar hemorrhage, patients become symptomatic and in between exacerbations, remain relatively symptom-free. Chronic hemoptysis without remission has been reported as well. Physical examination during the acute phase

may demonstrate mild crackles to overt signs of respiratory failure while being normal during the remission. The classical presentation of IPH includes the triad of hemoptysis, radiologic chest abnormalities and anemia; however, this constellation is absent in the majority. Other less frequent presentations include dry cough, chest pain, fever, fatigue, and clubbing.

DIAGNOSIS

In addition to the supportive clinical presentation, a diagnosis of IPH requires compatible laboratory data, chest radiology, and histopathologic findings. Since there is no specific serologic or pathologic marker for IPH, an exhaustive effort is needed to exclude competing diagnoses. Bronchoscopic evaluation is generally performed to identify DAH and rule out infection. In addition, structural airways abnormalities, including cancer, and arteriovenous malformation, which may mimic the symptoms of IPH, can be ruled out. A lung biopsy is necessary to definitively exclude capillaritis as DAH from both AAV and isolated pulmonary capillaritis can manifest with a negative serologic workup.

Laboratory workup

No specific laboratory tests. Evidence of iron deficiency anemia. Normal to elevated ferritin (Reticulocytosis). Neutropenia in the immediate pre-acute phase and eosinophilia following alveolar hemorrhage. Autoimmune diseases and vasculitis need to be ruled out:

•ANCA, PANCA, c-ANCA, anti-MPO antibody, anti-PR3 antibody

•ANA, RF, anti-CCP, anti-dsDNA, anti-smith antibody, anti SSA and SSB antibody, anti-smooth muscle antibody, anticardiolipin antibody, anti-phospholipid antibody, anti-beta 2 glycoprotein antibody, C3, C4, serum cryoglobulin, anti-jo antibody, aldolase, creatine phosphokinase, IgG and IgA tissue transglutaminase antibody, anti-gliadin antibody, anti-endomysial antibody Hematologic, metabolic, and cardiac etiologies need to be ruled out:

• CBC, CMP, PT, INR, PTT, d-dimer, FDP, fibrinogen

•Transthoracic echocardiogram Anti-basement membrane antibody

Chest radiography

Chest X-ray-Diffuse alveolar opacity in the acute phase involving perihilar and lower lobes with diffuse hemorrhage, all lung zones can be affected Alveolar opacity becomes more interstitial opacity with 72 h.

High resolution computed tomography-More sensitive Ground-glass opacity in the same distribution 'Crazy paving' due to interlobular septal deposition of hemosiderin.

TREATMENT

Despite any definitive proof of autoimmune causation, IPH is often considered and treated as an autoimmune disease. Corticosteroids alone or in combination with other immunosuppressive regimens are often employed for the treatment of IPH. Based on the available literature, immunosuppressive therapy appears to be efficacious and is associated with reduced morbidity and mortality. Immunosuppressive agents such as cyclophosphamide, hydroxychloroquine, and azathioprine have been shown to help with severe disease; however, at this time, optimal dosing and duration of therapy are poorly defined. Preventative measures include maintenance doses of prednisone or prednisolone of 10 to 15 mg/kg/day. In general, if the patient does not have any recurrence in 18 to 24 months, further tapering and discontinuation of steroids can be attempted. A limited number of studies have been performed in pediatric patients, and the results are often extrapolated on the adult population. Treatment is tailored to the severity of clinical presentation.

PROGNOSIS

The prognosis is variable, with some patients showing spontaneous remission and others progressing to death. The duration of disease in the literature ranges from death within days, following a severe acute illness, to survival with corpulmonale associated with the chronic illness after twenty years. Patients with idiopathic pulmonary hemosiderosis (IPH) have a mean survival rate of 2.5 to 5 years after diagnosis. Deaths can occur acutely from massive hemorrhage or after progressive pulmonary insufficiency and right heart failure.

COMPLICATIONS

Complications and long-term effects of idiopathic pulmonary hemosiderosis (IPH) vary depending on the severity and frequency of recurrence. Iron deficiency anemia and pulmonary fibrosis are the two most common complications of IPH. In the acute phase, IPH complications vary from simple complications such as shortness of breath to death due to choking of airways because of massive bleed and shock. Chronic complications may occur from progressive pulmonary insufficiency/severe respiratory distress and right heart failure. Death may occur as a result of massive bleeding.

CONCLUSION

IPH is an extremely rare disease in adults. The risk factors and etiopathogenesis are still unclear but possibly represents an autoimmune process with a concomitant genetic contribution. The diagnosis is often delayed, sometimes by years. Exclusion of AAV, Anti-GBM antibody disease, and connective tissue diseases is crucial and often requires histopathological investigation. Patients are usually treated with systemic corticosteroid with or without other immunosuppressive medications. Although no large prospective trials exist, treatment with immunosuppressive therapy appears to be associated with improved survival. Further research is necessary to identify the association with other autoimmune diseases and determine the optimal treatment.

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