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Case Report

Steroid-responsive idiopathic pulmonary hemosiderosis in adulthood: A case report.

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Abstract

Background

Idiopathic pulmonary hemosiderosis (IPH) is a rare cause of diffuse alveolar hemorrhage (DAH), most commonly reported in children but occasionally seen in adults.

Case Presentation

We describe a 30-year-old woman with recurrent hemoptysis, anemia, and dyspnea for two years. Investigations revealed iron deficiency anemia, diffuse ground-glass opacities on HRCT, and hemosiderin-laden macrophages on bronchoalveolar lavage. Transbronchial biopsy confirmed pulmonary hemosiderosis. She was treated with oral corticosteroids (0.75 mg/kg/day), resulting in complete clinical and radiological resolution within one month.

Conclusion

This case highlights the importance of considering IPH in adults presenting with unexplained DAH and anemia, particularly in tuberculosis-endemic regions. Early recognition and corticosteroid therapy can lead to favorable outcomes.

Keywords: *Idiopathic pulmonary hemosiderosis, diffuse alveolar hemorrhage, hemoptysis, corticosteroids, case report*
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Introduction

Idiopathic pulmonary hemosiderosis (IPH) is an uncommon and poorly understood cause of diffuse alveolar hemorrhage (DAH). Although predominantly described in children, adult presentations have been documented. The incidence is extremely low, estimated at 0.2–1.2 per million. The classical clinical triad includes recurrent hemoptysis, anemia, and dyspnea, but the disease often presents with variable severity. Because IPH lacks a specific diagnostic marker, it is considered a diagnosis of exclusion after other causes of DAH have been ruled out.

The rarity of IPH frequently leads to delayed recognition, which can cause significant distress for patients and families. Pathogenesis remains unclear, and evidence for optimal treatment is limited, with few randomized clinical trials available. Hemoptysis is the most common symptom, occurring either intermittently or persistently, and the amount of bleeding varies considerably. Clinicians should maintain a high index of suspicion and include IPH in the differential diagnosis of DAH and chronic anemia, even in atypical cases (Butt et al., 2020). The wide spectrum of clinical manifestations—from mild recurrent bleeding to severe respiratory failure—underscores the need for a comprehensive diagnostic approach and individualized therapeutic strategies (Saha & Milman, 2022).

Case Report

A 30-year-old woman presented with a 2-year history of chest pain, recurrent hemoptysis, and exertional breathlessness. She denied fever, abdominal pain, skin rash, joint pain, mucosal ulcers, ocular redness, hematuria, or

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anorexia. There was no history of prior medications, comorbidities, or addictions.

Examination

Pallor was noted, with no clubbing, cyanosis, icterus, or lymphadenopathy. Vital signs: pulse 86/min, blood pressure 110/70 mmHg, respiratory rate 20/min. Chest auscultation revealed normal vesicular breath sounds bilaterally; systemic examination was otherwise unremarkable.

Investigations

- **Blood tests:** Iron deficiency anemia (serum iron 35 µg/dl, saturation 5%). Peripheral smear showed microcytic hypochromic RBCs with macrocytes, teardrop cells, pencil cells, and target cells. WBC count 6700/µL, platelet count 2.67 × 10^5/µL, reticulocyte count 4.62%.
- **Other labs:** Coagulation profile, liver and renal function tests, and urine microscopy were normal. Autoimmune markers (ANA, RF, anti-dsDNA, ANCA, anti-GBM, antiphospholipid antibodies) were negative. Sputum CBNAAT was negative.
- **Bone marrow biopsy:** Cellular marrow with trilineage hematopoiesis.
- **Pulmonary function:** Spirometry showed moderate restriction with increased DLCO.
- **Imaging:** Chest X-ray revealed bilateral patchy ground glass opacities and increased bronchovascular marking in mid and lower zones (Fig.1). HRCT thorax showed bilateral extensive ground-glass opacities with right lower lobe bronchiectasis (Fig.2).
- **Bronchoscopy:** BAL demonstrated hemosiderin-laden macrophages. Transbronchial lung biopsy report consistent with pulmonary hemosiderosis(Fig.3).
- **Other:** Upper GI endoscopy showed fissuring of the D2 fold; biopsy was normal.

Management

Oral glucocorticoids (0.75 mg/kg/day) were initiated. Within one month, the patient became asymptomatic, and chest imaging showed complete resolution(Fig.4).

Figure 1: chest X-ray PA view bilateral patchy ground glass opacities and increased bronchovascular marking

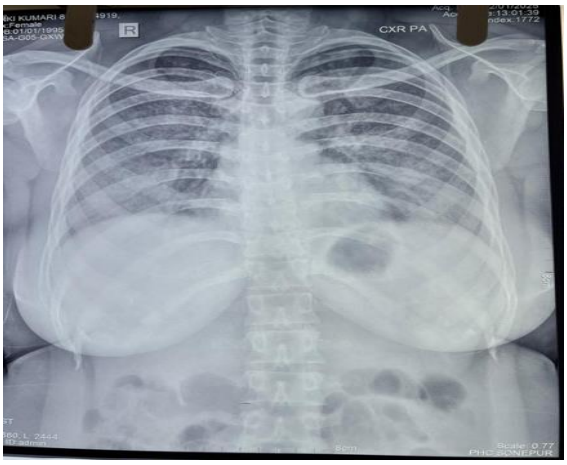

Figure 2: HRCT thorax showed bilateral extensive ground-glass opacities with right lower lobe bronchiectasis

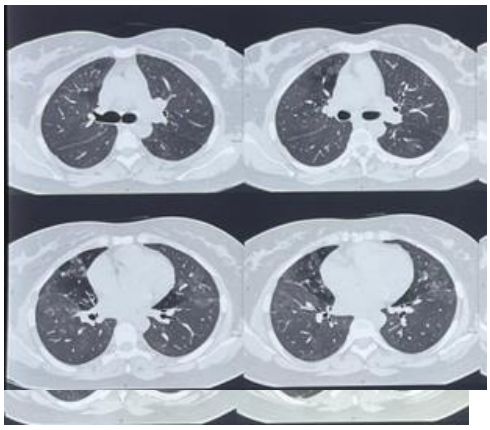

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Figure 3: Trans bronchial lung biopsy showing alveolar tissue**,** septa are thick due to fibrosis and infiltration of **hemosiderin laden macrophages**

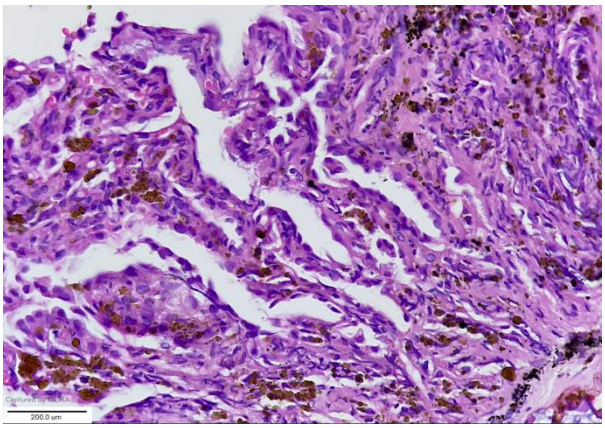
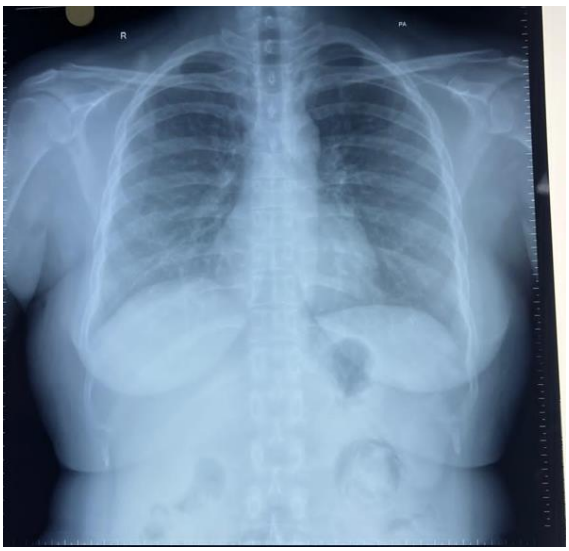


Figure 4: Chest X-ray PA view after 1 month of treatment showed complete resolution



Discussion

IPH is a rare disorder, with most cases occurring in children under 10 years. Adult presentations, such as in this patient, are unusual. Hemoptysis with anemia is a common presentation, but in tuberculosis-endemic regions, IPH is often misdiagnosed as pulmonary tuberculosis, delaying recognition and treatment.

The pathogenesis remains unclear. Proposed mechanisms include oxidative stress, immune-mediated capillary injury, and endothelial damage, all contributing to recurrent hemorrhage, hemosiderin deposition, inflammation, and fibrosis (Saha & Milman, 2022). Structural defects in pulmonary capillary endothelium or basement membrane have also been suggested.

Clinical manifestations vary widely. Some patients remain asymptomatic, while others develop recurrent dyspnea, cough, hemoptysis, or even respiratory failure. Chronic disease may present with pallor, growth retardation, or features of fibrosis such as bilateral crackles and clubbing. In this case, recurrent hemoptysis and exertional dyspnea were the predominant symptoms.

Diagnosis requires exclusion of other causes of DAH, including infections, Goodpasture syndrome, ANCA-associated vasculitis, and systemic diseases. Investigations such as chest imaging, spirometry, bronchoscopy, and lung biopsy are essential. BAL has higher diagnostic yield than sputum analysis, while histology showing hemosiderin-laden macrophages remains the gold standard.

Corticosteroids are the cornerstone of therapy. Immunosuppressants (e.g., azathioprine, mycophenolate mofetil) may be considered in steroid-resistant cases. Our patient responded well to corticosteroids, with complete clinical and radiological resolution.

Conclusion

Idiopathic pulmonary hemosiderosis is a rare but important cause of DAH and chronic anemia. Adult presentations are uncommon and often misdiagnosed. Early recognition and prompt corticosteroid therapy can lead to favorable outcomes. This case emphasizes the need for heightened clinical suspicion in patients with unexplained recurrent hemoptysis and anemia.

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Conflict of Interest

The authors declare that they have no conflict of interest related to this case report.

Data Availability

The data supporting the findings of this case report are contained within the manuscript. Additional patient information is not publicly available due to patient confidentiality and privacy considerations. Further details may be made available from the corresponding author upon reasonable request, subject to appropriate ethical and confidentiality requirements.

Author Contributions

Dr. Krishnan Namboothiri E. contributed to the conception and preparation of the case report, clinical evaluation and management of the patient, interpretation of the investigations, literature review, and drafting of the manuscript. Dr. Deependra Kumar Rai, Dr. Somesh Thakur, Dr. Javeeria Shabbir, Dr. Jisha G. Panicker, and Dr. Thaodem Collin contributed to the clinical evaluation and management of the patient, interpretation and discussion of the clinical findings, critical review of the manuscript, and approval of the final version. All authors reviewed and approved the final manuscript and agree to be accountable for the content of the work.

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Informed Consent

Written informed consent was obtained from the patient for publication of this case report.

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