Idiopathic Pulmonary Hemosiderosis: a Rare Cause of Hemoptysis in Adults

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Idiopathic Pulmonary Hemosiderosis: a Rare Cause of Hemoptysis in Adults

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INTRODUCTION: Idiopathic Pulmonary Hemosiderosis (IPH) is an extremely rare lung disease characterized by diffuse alveolar hemorrhage (DAH) and accumulation of hemosiderin in the lungs. IPH commonly affects children. Although the exact incidence and prevalence are unknown, estimated yearly incidence in Swedish children from 1960-1979 was 0.24 per one million children.[1] The disease clinically manifests as a triad of hemoptysis, diffuse pulmonary infiltrates, and iron deficiency anemia. Recurrent alveolar bleeding may eventually produce pulmonary hemosiderosis and fibrosis. The etiology of IPH is unknown. However, the response to immunosuppressive therapy suggests that immune processes may be involved. It appears that a structural defect in the alveolar capillaries, either in the alveolar basement membrane or in the alveolar endothelial cell, may predispose to the condition.[1-5] The accumulation of neutrophils in the alveoli also may play a role.[7] Here we report a case where the patient was found to have IPH, that presented with DAH accompanied by clinical signs and symptoms of iron deficiency anemia and diffuse pulmonary infiltrates on imaging.

Case Report

History of Present Illness: A 21-year-old female with past medical history of asthma, vitamin B12 deficiency anemia with iron deficiency anemia, hyperthyroidism and tobacco abuse presented to an Outside Hospital System with shortness of breath, fatigue, and recurrent hemoptysis worsening over the past six months. The patient was transferred from current incarceration. Six months previous, the patient was initially admitted to hospital for shortness of breath and fatigue. The patient was found to have profound anemia with hemoglobin of 4.4 g/dL and responded to four units of packed red blood cells. Initial CT Scan of the Chest showed patchy bilateral alveolar infiltrates involving left upper lobe, left lower lobe, and right lower lobe (Figures 1 and 2). The patient was discharged and did not follow-up as an outpatient with Pulmonology as her hemoptysis improved, but did receive intravenous iron and Vitamin B12 injections as outpatient with Hematology. She re-presented to hospital two months later with recurrent non-massive hemoptysis, again with hemoglobin level of 4.4 g/dL. She responded again to four units of packed red blood cells. Pulmonology was consulted and patient elected to pursue follow-up as an outpatient, but was lost to follow-up secondary to incarceration. During incarceration, patient followed with physician to county jail, but no follow-up occurred. She presented to outside hospital system from incarceration with symptoms of severe lightheadedness and non-massive recurrent hemoptysis. She was given 1 unit of packed red blood cells and underwent fiberoptic bronchoscopy, which showed alveolar hemorrhage. She was subsequently transferred to Lehigh Valley Hospital-Cedar Crest campus for further management. Her hemoptysis was continuing to occur up to five spoonfuls on a daily basis and described as dark-red, purple-brown in color. The patient reported mild wheezing preceding hemoptysis. She denied fevers, chills, chest pain, epistaxis, menorrhagia, abdominal pain, nausea, vomiting, hematemesis, melena, and hematochezia.

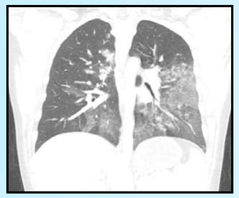

Figure 1. Coronal view of initial chest CT showing pulmonary bilateral infiltrates.

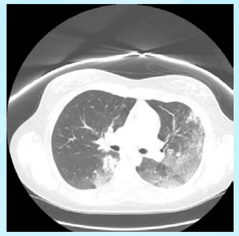

Figure 2. Axial view of initial chest CT showing bilateral pulmonary infiltrates.

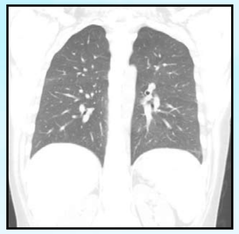

Figure 3. Follow up at 8 months. Coronal view of chest CT scan showing resolution of infiltrates after corticosteroid treatment.

Hospital Course: Patient was admitted to medical/surgical floor. On physical exam, she was afebrile, heart rate was 100 per minute and regular, blood pressure was 118/84 mmHg and oxygen saturation of 94% on room air. Auscultation of the lungs revealed intermittent wheezing and crackles bilaterally. She had an initial hemoglobin of 7.5 g/dL, 5,500 WBC per mcL and platelets 246,000 per mcL. Her TSH was less than 0.022 µIU/L with normal free T4 and T3 levels. A CT scan of the chest revealed bilateral infiltrates. She was placed on intravenous azithromycin for previous elevated Mycoplasma IgM and IgG serum titers. The patient underwent a transthoracic echocardiogram showing normal left ventricular systolic function with normal regional wall motion and ejection fraction of sixty percent. Broncho-alveolar lavage cultures from bronchoscopy were negative for acid-fast bacilli. Serologies were negative for anti-nuclear antibodies, anti-neutrophil cytoplasmic antibodies, anti-glomerular basement membrane antibodies, HIV, Quantiferon-Tuberculosis Gold assay, Coombs test, and antiphospholipid antibody. Cardiothoracic surgery was consulted and the patient ultimately underwent thorascopic wedge biopsies of the right upper, middle, and lower lobes. Pathology demonstrated abundant hemosiderin deposition within the alveolar macrophages and blood vessel walls, consistent with IPH. There was no evidence of capillaritis, vasculitis, pulmonary hypertension, or granulomatous inflammation. The patient received prednisone (40 mg/day) and Bactrim (1 tab 3 times weekly). Her hemoptysis improved with prednisone therapy. The patient was transferred to Temple University Hospital for a second opinion in stable condition (Figure 3).

Discussion

This case report describes a patient with IPH, who presented with signs and symptoms of non-massive recurrent hemoptysis and infiltrates on chest imaging as well as iron deficiency anemia. In fact, histopathology demonstrated abundant hemosiderin deposition within the alveolar macrophages and blood vessel walls, consistent with anemia and a diagnosis of IPH.

The exact incidence and prevalence of IPH are largely unknown. Eighty percent of cases of IPH occur in children, generally manifesting before 10 years of age.[8,9] In adults, most cases are recognized before 30 years of age. Patients classically present with a triad of recurrent or chronic pulmonary symptoms (cough, dyspnea, wheeze, hemoptysis), pulmonary infiltrates on CXR, and iron-deficiency anemia. Our patient had all three components to suspect IPH.

A diagnosis of IPH is based on exclusion of other causes of intrapulmonary hemorrhage and systemic diseases. In the absence of systemic disease, findings of hemosiderin-laden macrophages in bronchoscopic lavage or gastric aspirate/sputum along with chronic pulmonary symptoms lead to diagnosis of IPH. Lung biopsy is the gold standard for diagnosis. It is important to exclude pulmonary capillaritis, which is a cause of DAH. Pulmonary capillaritis is a small-vessel vasculitis, which can occur as an isolated condition or in association with multiple systemic vasculitides. Isolated DAH without identifiable causation or associated disease is referred to as IPH.[10]

Daily oral corticosteroids or weekly intravenous pulse of methylprednisolone is commonly used in the induction treatment of IPH. Other immunosuppressive agents such as azathioprine, cyclophosphamide, and hydroxychloroquine have also been used alone or in combination with oral corticosteroids.[11-15] Low-dose oral corticosteroids, azathioprine, or methotrexate are used in maintenance phase. Aggressive treatment with the use of corticosteroids and immunosuppressive agents are associated with a prolonged survival and improved prognosis.[16] Long-term low-dose corticosteroid therapy was also reported to result in a milder disease course and prevent bleeding crisis.[17] Data from Saeed et al. report patients today have eighty-six percent survival beyond five years of diagnosis. However, due to the lack of large patient series and inadequate follow-up in previous studies, the prognosis of IPH remains unclear.[18]

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