

Eight Years Follow-up of a Case with Idiopathic Pulmonary Hemosiderosis After Corticosteroid Therapy

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Abstract

Idiopathic pulmonary hemosiderosis is characterized by recurrent episodes of alveolar hemorrhage, hemoptysis, and secondary iron deficiency anemia with unknown etiology. It generally emerges in childhood and adolescence periods but rarely found in adulthood. Definite diagnosis is established by appearing the hemosiderin-laden macrophages at sputum, bronchoalveolar lavage, or open lung biopsy. We reported a male patient who was born in 1975, expectorated blood since 1995, and was diagnosed in 1998. He received many blood replacements. He admitted to our clinic in 2003 with complaints of coughing up blood, shortness of breath, and tiredness. We gave the corticosteroid therapy to patient for 6-month period. After treatment, the patient did not have any complaints. Clinicians should keep in mind that idiopathic pulmonary hemosiderosis may differ in localization on chest X-ray and corticosteroid treatment should be started when diagnosis is established.

Keywords: Alveolar hemorrhage, Corticosteroid therapy, Pulmonary hemosiderosis

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Introduction

Idiopathic pulmonary haemosiderosis (IPH) is a rare cause of diffuse alveolar hemorrhage (DAH) with unknown etiology.^[1] DAH is characterized by haemoptysis, dyspnea, alveolar infiltrates on chest X-ray, and various degrees of anemia. In IPH, intra-alveolar bleeding is persistent or recurrent, and often marked.^[2,3] It is classic disease of childhood, but approximately 20% of patients are presented during adulthood. Radiographical manifestations of IPH include patchy areas of opacities that usually involve both lungs and opacities similar to pulmonary edema. The costophrenic angles and periphery of the lungs are usually spared. Absolute diagnosis is based on the presence of hemosiderin-laden macrophages (HLM) without any evidence of pulmonary vasculitis, nonspecific granulomatous inflammation or

deposition of immunoglobulins in respiratory secretion or biopsy materials.^[1] We reported the young adult man case because of 16-year period of IPH disease; responding to corticosteroid treatment very well and 8-year follow up after treatment.

Case Report

A 29-year old man was admitted to our clinic with complaints of fatigue, pallor, cough, bloody sputum, and shortness of breath in 2003. In history, his complaints began in 1995 and HRCT was taken in 1998. It showed a decrease in aeration and patchy areas of ground glass pattern in both lungs, especially in the lower lobes. Open lung biopsies were taken by video-assisted thorascopic technique. In pathology, a large number of HLM, interstitial edema and alveolar fibrosis, and foci of fresh hemorrhage were determined and he was diagnosed as idiopathic pulmonary hemosiderosis [Figure 1]. Patient's hemoptysis continued for years. In 2003, he was hospitalized. After blood transfusions, peripheral white cell count (WBC) and platelet (PLT) count were normal, hemoglobin (Hb) was 5.3 g/dl. Serum biochemistry, ANA, p-ANCA, C-ANCA, and RF were normal. Erythrocyte sedimentation rate (ERS) was 28 mm/h, serum iron was 18 mg/dl, iron binding capacity was 300

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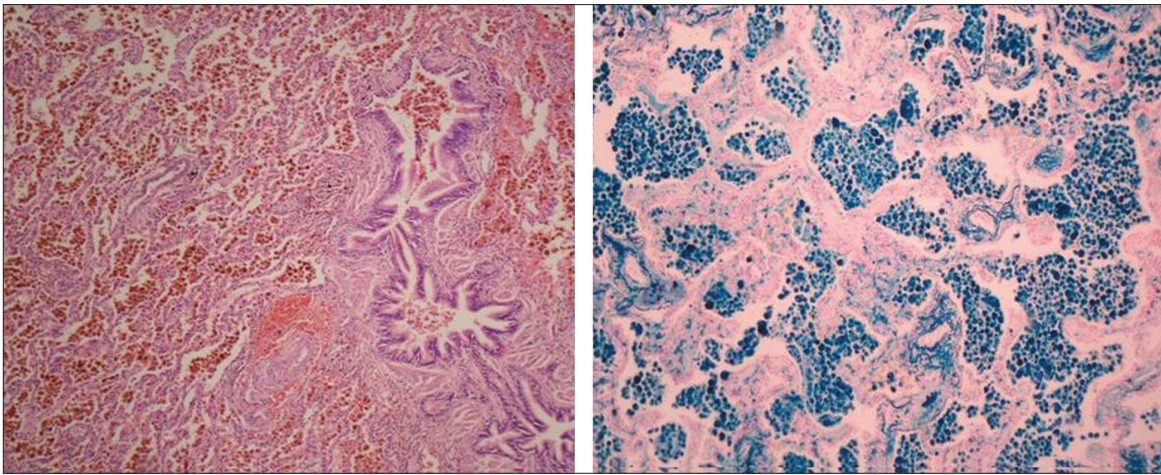


Figure 1: A large number of hemosiderin-laden macrophages and foci of fresh hemorrhage were determined. H and E, $\times 100$ and Prussian blue, $\times 100$

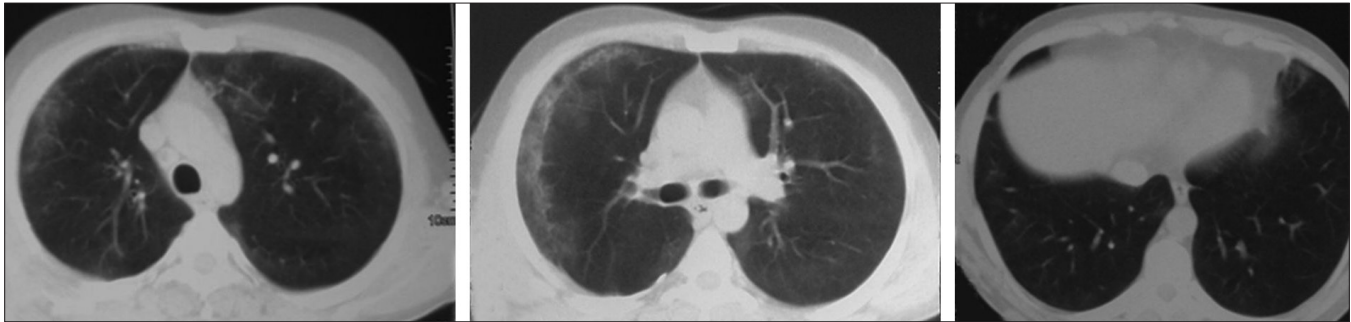


Figure 2: Peripheral ground-glass appearance at the upper and middle lobe of right lung, and at the upper lobe of left lung

mg/dl. Electrocardiography was normal, arterial blood gases were as follows: pH 7.4, PO_2 66, PCO_2 38, SaO_2 94%.

Two month later hemoptysis repeated and patient was admitted to our clinic. His and family history was not contributory. He had an 11-pack-year smoking history. Physical examination revealed pale skin and mucosa, other vital signs were normal. In respiratory system examination bilateral basal crackles were heard. Cardiovascular, gastrointestinal, and neurological examinations were normal.

A daily dose of 80 mg (1.5 mg/kg) methylprednisolone was started, tapered to 48 mg/day 1 month later, and was stopped at the end of 6th month. The patient responded to treatment very well, and after that he had no complains. We invited him to control in 2009 and HRCT was given. Peripheral ground-glass appearance at the upper and middle lobe of right lung, and at the upper lobe of left lung were determined [Figure 2]. Hb 13.2 g/dl, WBC, PLT, and ESR were normal and pulmonary function tests showed mild restriction. It has been 16 years from the beginning of hemoptysis, 13 years from the diagnosis and 8 years from the treatment. Since the beginning of treatment, there has been a very small amount of hemoptysis approximately

less than 1 cc 1-2 times per year.

Discussion

Idiopathic pulmonary hemosiderosis is a rare disorder of unknown etiology and usually appears in children.^[3] The etiology of IPH has been suspected to be autoimmune, allergic, genetic or environmental in different theories.^[4] Some reports presenting that gluten-sensitive enteropathy and sensitivity to cow's milk may accompany IPH.^[5] Our case was a young adult and such situations were not detected in our patients.

Findings of hemoptysis, iron deficiency anemia, shortness of breath, and parenchymal infiltration in chest radiography are typical as seen in many clinical articles.^[6] Our patient had typical findings too.

HRCT appearances include patchy scattered areas of ground glass opacity and consolidation that usually involve the hilary and lower regions of the lungs, but involvement may be diffused.^[7] Lymphadenopathy and pleural effusions are not common.^[8] Interstitial fibrosis was developed in our patient. But in our case peripheral ground-glass appearance was presented at upper and

middle lobe of right lung, and at the upper lobe of left lung. It is different from general articles.

For definitive diagnosis, lung biopsy is necessary, the most important features are the presence of intact erythrocytes and numerous siderophages in the alveoli and the absence of any other pathologic sign like vascular malformation, immune deposition, capillaritis, vasculitis, granuloma, malignancy.^[3] In our patient, IPH was diagnosed with the finding of HLM in lung biopsy material as in other studies.^[1,9]

The use of systemic glucocorticoids for IPH grew out of the impression of an immune pathogenesis. Although the disease is rare, information from case reports and case series suggest that systemic glucocorticoids reduce the morbidity and mortality of acute episodes of alveolar bleeding and control progression to pulmonary fibrosis.^[3,6,10] The recommended dose was <1 mg/kg/day prednisolone for two months period and then gradually reduced.^[1] In one study, methylprednisolone in dose of 1-2 mg/kg/day was suggested.^[8] We started daily dose of 80 mg (1.5 mg/kg) treatment and tapered to 48 mg/day 1 month later, and stopped at end of 6th month. The patient responded the treatment very well, as in other studies.^[6,10]

In conclusion, we think that during childhood and young adult periods IPH should be considered in patients presenting hemoptysis and anemia. For diagnosis tissue samples should be taken and as a treatment systemic corticosteroid therapy should be given.

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