

# Idiopathic pulmonary hemosiderosis in an adult female patient

A Uysal, M Boktor, M Tapolyai

## Citation

A Uysal, M Boktor, M Tapolyai. *Idiopathic pulmonary hemosiderosis in an adult female patient*. The Internet Journal of Pulmonary Medicine. 2006 Volume 8 Number 1.

## Abstract

Idiopathic pulmonary hemosiderosis (IPH) is a rare condition that is rarely seen in the adult population<sup>1</sup>. It is more common in younger patient between the ages of 1 to 171. It can present with onset of anemia, hemoptysis, dyspnoea and presence of opacifications on cat scan (CT) of the chest.

The patient is a 40 year old female who presented to the hospital with gradual onset of dyspnoea and cough as well as hemoptysis that developed over the course of several weeks. Her vital signs were stable on admission to the hospital. She had intensive work up of her condition which led to a bronchoscopy and an open lung biopsy. These findings were consistent with diagnosis of IPH.

The patient once diagnosed was started on corticosteroid therapy. She was sent home with clinic appointment. Pathogenesis, presentation and diagnosis of IPH are reviewed for this rare presentation in an adult patient.

## INTRODUCTION

We report a case of a 40 yr old female patient with history of asthma, hypothyroidism, obstructive sleep apnoea and avascular hip necrosis who presented with gradual onset of dyspnoea over 4 months and cough as well as hemoptysis.

She also had evidence of bilateral parenchymal changes on cat scan (CT) of the chest.

Her bronchoscopic alveolar lavage and open lung biopsy were consistent with diagnosis of idiopathic pulmonary hemosiderosis (IPH).

We will review the current literature on IPH.

## CASE REPORT

The patient presented to the hospital with gradual onset of dyspnoea over 4 months and cough as well as hemoptysis that developed over the course of several weeks. Patient's dyspnoea became progressively worse 10 days before hospitalization. She also reported 13.3 pound weight gain over the last 7 days. She previously had a cardiac catheterisation that was unremarkable.

Her social history was remarkable for 20 pack-year history of smoking.

On admission her vital signs were normal and had low grade fever. Relevant findings on physical exam were presence of presence of bilateral rales on chest auscultation. There was evidence of moderate leg edema on both sides. She also had mildly distended abdomen.

Relevant laboratory findings include white blood cell count of 7.7 k/cmm and hemoglobin of 11.3 g/L with mean corpuscular volume of 88.6 fl. Patient had an elevated sedimentation rate of 59 mm/hr.

Her angiotension converting enzyme (ACE) levels were slightly elevated at 72 u/l. Her p-ANCA and c-ANCA (antineutrophil cytoplasmic antibodies) as well as antigliadin antibody levels, cardiolipin antibodies, rheumatoid levels were all within the normal range. She also had a normal brain natriuretic peptide level (BNP) at 92 pg/ml.

She also had a normal ejection fraction on echocardiographic exam. There was also no evidence of pulmonary hypertension nor atrial and ventricular enlargement.

The patient's ct scan of her chest revealed bilateral hilar lymphadenopathy and scattered bilateral glass ground opacifications (Figure 1). There was no evidence of pulmonary embolism.

**Figure 1**

Figure 1: CT scan of the chest showing bilateral opacities.



The spirometry revealed a decrease in FEV1 (forced expiratory volume) and FVC (forced vital capacity) with a normal FEV1/FVC ratio that was suggestive of a mild restrictive ventilatory impairment.

She was evaluated by pulmonology team and subsequently underwent bronchoscopy. Bronchoalveolar lavage (BAL) findings showed evidence of hemosiderin laden alveoli. Patient's cytology results did not reveal any evidence of malignancy. There was also no evidence of fungal or mycobacterial and bacterial infection.

She later went on to have an open lung biopsy. Multiple sections from both lung specimens showed multiple patchy areas, 0.1 to 0.5 cm. of interstitial and alveolar fibrosis. These areas were associated with recent and old intra-alveolar hemorrhages with hemosiderosis. There were numerous hemosiderin-laden macrophages in alveolar

spaces. The alveolar walls were thickened due to fibrosis. Intra-alveolar fibroblastic proliferation was also seen. There was no evidence of vascular lesion nor thrombus. These findings were consistent with IPH which led to steroid therapy.

**DISCUSSION**

Idiopathic pulmonary hemosiderosis (IPH) is an uncommon disorder and is particularly rare over the age of 17<sup>1</sup>. It most commonly affects individuals between the ages of 1 to 17<sup>1</sup>. Aetiology of the disease is unknown<sup>1</sup>. Patients can present with anemia and development of pulmonary symptoms such as cough, shortness of breath, hemoptysis and presence of parenchyma lesions on chest X-ray (CXR)<sup>1</sup>.

Celeen syndrome comprises hemoptysis, onset of anemia and presence of opacities on CXR<sup>2</sup>.

It is a diagnosis of exclusion of other conditions which can also present with diffuse alveolar hemorrhage<sup>2</sup>. It presents with recurrent hemorrhage and accumulation of hemosiderin in lung parenchyma<sup>3</sup>. It can be a recurrent condition that can result in fibrosis<sup>2</sup>. Open lung biopsy is often warranted for diagnosis<sup>1</sup>. Treatment consists of high dose corticosteroids and sometimes immunosuppressive drugs<sup>2</sup>. Hoca et al described a case of IPH that was associated with celiac disease<sup>4</sup>. It is a rare disease in the adult population so disease progression could be unpredictable.

**References**

1. Turay UY, Ergun P, Erdogan Y, Biber C, Demirag F, Keyf IA: Idiopathic pulmonary haemosiderosis. *Tuberk Toraks* 2004; 52:382-385.
2. Willms H, Gutjahr K, Juergens UR, et al: Diagnostics and therapy of idiopathic pulmonary hemosiderosis. *Med Klin (Munich)* 2007; 102:445-450.
3. Deniz O, Onguru O, Ors F, et al: Idiopathic pulmonary hemosiderosis in an adult patient responded well to corticosteroid therapy. *Tuberk Toraks* 2007; 55:77-82.
4. Hoca NT, Dayioglu D, Ogretensoy M: Pulmonary hemosiderosis in association with celiac disease. *Lung* 2006; 184:297-300.

**Author Information**

**Askin Uysal, M.D.**

Department of Internal Medicine, Veterans Hospital Overton Brooks Medical Center

**Moheb Boktor, M.D.**

Department of Internal Medicine, Veterans Hospital Overton Brooks Medical Center

**Mihaly Tapolyai, M.D.**

Department of Internal Medicine, Veterans Hospital Overton Brooks Medical Center