# Idiopathic pulmonary haemosiderosis: diagnosis by gastric lavage

Y Khonglah,V Raphael, A Bhuyan, P Gogoi, and R Dass

## Introduction

Idiopathic pulmonary haemosiderosis (IPH) is a rare disorder of unknown etiology characterised by iron deficiency anaemia, recurrent or chronic pulmonary symptoms, such as cough and haemoptysis, and diffuse pulmonary infiltrates.[1] The incidence is reported from 0.24 to 1.23 cases per million selected populations.[2] It affects paediatric patients in approximately 80% of cases with equal gender incidence.[3] The presence of iron or haemosiderin in macrophages obtained in gastric or bronchoalveolar lavage is considered crucial in the diagnosis of the cinical syndrome of haemosiderosis.[4]

## Case report

A 3.5-year-old boy presented to the paediactric outpatients department with complaints of fever and cough for 1 month. He was initially admitted to a local hospital and transfused two units of blood for severe anaemia.

On examination he was pale, febrile, with a respiratory rate of 60 beats a minute and with intercostal retractions. Lung fields were clear on auscultation and he had a hemic murmur. There was no hepatosplenomegaly. His weight and height were normal for his age. There was no history of contact with tuberculosis (TB).

A working diagnosis of pneumonia with nutritional anaemia with differential diagnosis of pulmonary haemosiderosis and pulmonary Koch was made. Investigations showed haemoglibin of 7.7 gm/dl; haematocrit of 23.1%; erythrocyte sedimentation rate (ESR) of 14 mm at the end of 1 hour; total leukocytes of 7400 $mm^3$ with neutrophils 62%; lymphocytes 34%; and plateletes 5.8 $mm^3$. Peripheral blood smear revealed microcytic hypochromic anaemia with reactive thrombocytosis. C-reative protein, rheumatoid factor, antinuclear antibody (ELISA), antiphospholipid antibody were all negative. Renal function tests, liver function tests, and serum electrolytes were within normal limits.

A computed tomography (CT) scan of the chest showed bilateral, diffuse ground-glass haziness with few patchy areas of sparing.

*Dr Yookarin Khonglah, Dr Vandana Raphael, Dr Ashim Bhuyan, and Dr Priyanka Gogoi, Department of Pathology; and Dr Rashna Dass, Department of Pediatrics, all from the North Eastern Indira Ghandhi Research Institute of Health and Medical Sciences, Mawdiangdiang, Shillong-793018, Meghalaya, India. Correspondence to: Dr Yookarin Khonglah. Email:*

## Cytological findings

Gastric lavage revealed a good number (75%) of haemosiderin-laden macrophages (HLM) confirmed by Perl's stain for hemosiderin, suggesting the possibility of intra-alveolar haemorrhage (see Figure 1). The presence of bilateral diffuse ground-glass haziness and iron deficiency anaemia (IDA), along with HLM in gastric lavage confirmed the diagnosis of pulmonary haemosiderosis.

## Management and follow-up

At admission the patient was administered methyl prednisolone 30 mg/kg/day by injection once daily for 5 days; then oral prednisolone 20 mg, gradually tapered and maintained at 5 mg once daily. He was also given azathioprine and hydroxychloroquine (disease-modifying agents). Later prophylactic co-trimoxazole was introduced for the prevention of *Pneumocystis carinii* pneumonia.

The patient was on regular follow-up and was admitted to hospital three times for respiratory distress and anaemia. He finally succumbed to the disease 3 years later due to progressive respiratory failure leading to cardiac arrest.

## Histopathological findings

A postmortem lung biopsy was performed and histopathology examination revealed expanded alveoli filled with haemosiderin pigment and haemosiderin-laden macrophages (see Figure 2). The alveolar wall was thickened and showed intense fibrosis with deposits of haemosiderin pigment. The bronchiole also showed deposits of hemosiderin. There was no evidence of vasculitis. A diagnosis of chronic idiopathic pulmonary haemosiderosis (IPH) was tendered.

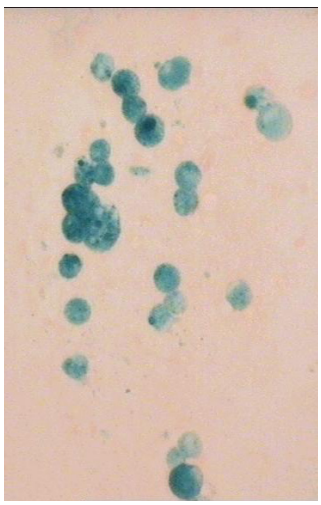

*Figure 1 Haemosiderin-laden macrophages in gastric lavage (Perl's stain, 40x)*

## Discussion

IPH is a disorder of unknown etiology that is characterised by recurrent or chronic haemorrhage and accumu-

lation of haemosiderin in the lungs.[4] The symptoms of IPH usually occur early in childhood before the age of 10 years.[5] There have also been reports of IPH with seasonal occurrence.[6] Clasically, IPH clincially manifests as a triad of haemoptysis, diffuse parenchymal infiltrates on chest radiograph, and iron deficiency anaemia. Haemoptysis is unusual as children swallow blood-stained sputum and alveolar bleeding does not readily gain access to the central airways.[7] Swallowed blood may lead to positive occult blood in stools, a misleading clue pointing to gastrointestinal blood loss as a cause for anaemia.[7] It is difficult to diagnose IPH because nutritional iron deficiency anaemia is common in children, as is TB, which may present with lung infiltrates not responding to routinely used antibiotics.[8]

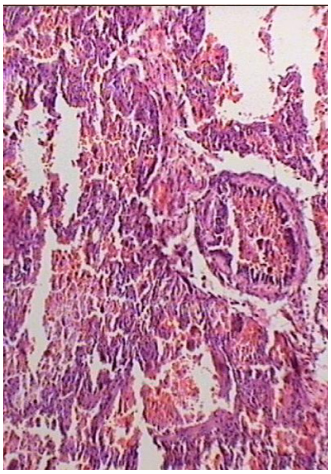

*Figure 2 Alveoli filled with haemosiderin-laden macrophages and haemorrhage (haematoxylin and eosing x 10))*

The pathogenesis of IPH is still not well established. Numerous theories have been proposed mostly suggesting an autoimmune basis, because alveolar haemorrhage in IPH is similar to that in systemic lupus erythematosus (SLE), an established autoimmune disorder. This autoimmune theory is strengthened by the fact that immunosuppressants are effective in improving the clinical status.[3,6] A study by Kabra et al on the clinical profile and follow-up of 26 children concluded that the outcome of IPH can be improved with inhaled corticosteroids and hydroxychloroquin.[9]

IPH is a diagnosis of exclusion and there should be a high degree of suspicion. Laboratory and radiological findings that have been found helpful in diagnosing the disease are anaemia, chest X-ray showing diffuse parenchymal infiltates, pulmonary function tests showing interstitial lung disease, and increase in single breath carbon monoxide (CO) uptake. A diagnosis by lung biopsy is considered gold standard by some authors, but many have accepted the presence of HLM in gastric or bronchoalveolar lavage fluid as diagnostic if typical clinical featrues are present and are not accompanied by evidence of extrapulmonary disease.[4] The use of gastric or bronchalveolar lavage fluid is indicated for investigating the aetiology in case of a pulmonary haemorrhage, since it is simple and only a mildly aggressive procedure and yields significant sensitivity and specificity.[3] Salih et al reviewed data from bronchoalveolar lavage studies done in children to correlate the presence of HLMs with pneumonia and haemosiderosis and to determine what proportion of HLMs has the optimal sensitivity and specificity for the diagnosis of haemosiderosis. They concluded that a HLM index of 36% from a percentage of 200 cells gave the highest sensitivity and specificity of 1 and 0.96 respectively.[4]

## Conclusion

IPH is a rare disorder and the diagnosis may be delayed considering the fact that children in developing countries are affected by more common diseases such as TB and pneumonia. If the classical triad of hemoptysis, diffuse parenchymal infiltrate, and IPD are present, IPH should be suspected and every effort made to rule it out. Obtaining sputum samples in children can be difficult and in centres where bronchoscopy is not available, the present of HLM in gastric lavage fluid is equally diagnositic and also the simplest, reliable diagnostic test in infants and young children.[4]

**References**


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