Case Report of Idiopathic Pulmonary Haemosiderosis in a Child with recurrent chest infections

Renan Ibrahem[1], Anita Arasaretnam[2], Katherine Ordidge[3], John Vedelago[4], Rosemary Toy[5]

1. Nottingham University Hospitals NHS trust, Nottingham, UK

2. Maidstone and Tunbridge Wells NHS trust, Kent , UK

3. UCL Centre for Advanced Biomedical Imaging, London, UK

4. Royal Prince Alfred Hospital, Sydney, Australia

5. Breast Screening Unit, Medway Maritime Hospital, Kent, UK

* **Correspondence:** Renan Ibrahem, Nottingham University Hospitals NHS trust, City Hospital Campus, Hucknall Road, Nottingham, NG5 1PB, UK
(*Renan.ibrahem@doctors.org.uk*)

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ABSTRACT

Idiopathic pulmonary haemosiderosis (IPH) is a rare condition that usually presents as a triad of haemoptysis, iron deficiency anaemia and pulmonary infiltrates. We report a case of IPH diagnosed in a 7 year old boy who had recurrent hospital admissions with severe chest infections and haemoptysis from his first few months of life. He was found to have microcytic hypochromic anaemia, diffuse infiltrate shadowing on his chest X-ray (CXR) and ground-glass opacification on his computed tomogram (CT). Perl's Prussian blue staining of his bronchoalveolar lavage fluid revealed haemosiderin-laden macrophage infiltration. After exclusion of infective, cardiac, immunological and glomerular causes, he was diagnosed with idiopathic pulmonary haemosiderosis. He has since been treated intermittently with steroids, which have failed to control his symptoms fully.

CASE REPORT

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We present a case history of a 7 year old boy, who has had recurrent admissions with cough, shortness of breath and severe chest infections since birth. His mother had also noticed occasional episodes of blood-stained sputum production. Further investigations revealed unexplained profound microcytic hypochromic anaemia and a negative faecal occult blood test. His haemoglobin electrophoresis was normal and serology for viruses and autoimmune antibody screening was negative. Chromosomal studies revealed no abnormalities.

The patient was born prematurely at 28 weeks following a maternal history of pre-eclampsia. His respiratory condition deteriorated suddenly, immediately post delivery for which he required intubation and ventilation for 5 weeks. Following this, he required nocturnal oxygen for the first 9 months. Since then, he has only been oxygen dependent during significant chest infections. Jaundice had been noticed on day nine post-delivery and was treated with phototherapy. He also required blood transfusions and erythropoietin for anaemia of prematurity.

CXR's were obtained postnatally, which identified diffuse infiltrate shadowing (Figures 1, 2). A chest CT was subsequently performed, which revealed patchy areas of ground-glass opacification with non-specific appearances (Figures 3, 4). The patient was therefore sent for a bronchoscopy and bronchial lavage which grossly appeared entirely normal. The lavage demonstrated a slight neutrophilia and on Perl's Prussian blue staining highlighted moderate levels of haemosiderin-laden macrophages (Figure 5). His subsequent blood tests were normal apart from a raised IgG.

On the basis of the above results, a diagnosis of IPH was made at the age of 2.5 years. Repeat CT chest at 6 years of age showed widespread homogeneous ground-glass opacification throughout both lung fields, with localized areas of denser opacification peripherally (Figure 6). He is currently being treated with hydroxychloroquine and prednisolone. In addition he is on prophylactic azithromycin. Four pulses of methylprednisolone have been given with minimal impact on his haemoptysis. He failed an initial trial of azathioprine due to intolerance although it is planned to re-challenge him in the near future.

DISCUSSION

IPH is caused by diffuse alveolar haemorrhage leading to abnormal accumulation of haemosiderin in the lungs [1]. It was described first by Ceelen 1931 as a triad of haemoptysis, anaemia and pulmonary infiltrates [2]. IPH is characterized by abnormal accumulation of iron as haemosiderin in alveolar macrophages due to repeated episodes of intra-alveolar haemorrhage. The aetiology of the condition is unknown and it can be fatal if left untreated due to progressive pulmonary fibrosis [3].

IPH is considered to be a rare disorder with an incidence of 0.24-1.23 cases per million in selected populations [4, 5]. 80% of cases occur in children with an equal sex distribution in childhood and a slight male predominance in adulthood. IPH can present with repeated chest infections and occasional haemoptysis, as in this case. It can also present as iron deficiency anaemia without respiratory symptoms [6, 7] or diffuse parenchymal infiltrates on chest X-ray. In young children and infants with IPH who present with iron deficiency anaemia a positive faecal blood test can often be detected due to swallowed blood-stained sputum [8]. The aetiology and pathogenesis is not yet known, although autoimmunity is thought to play a role in some cases [9]. Diagnosis is by exclusion of other systemic diseases which can cause pulmonary haemorrhage such as Wegener's granulomatosis, Systemic Lupus Erythematosus, Rheumatoid arthritis, Goodpasture's Syndrome, coagulopathies, platelet defects, pulmonary infections, pulmonary neoplasms, pulmonary veno-occlusive disease, pulmonary capillary haemangiomatosis and toxins such as cocaine, pesticides or insecticides. The distinctions between these differentials are based on both clinical presentation and radiological features [Table 1]. The radiological features are non specific and vary between interstitial changes, consolidation, pleural effusion, pulmonary nodules with or without cavitations, lung fibrosis and ground glass opacities. It has been reported that systemic vasculitis has developed in a patient with IPH, eight years from diagnosis [10].

Although rare, IPH should be part of the differential diagnosis of a child presenting with haemoptysis or other respiratory symptoms when the CXR shows patchy, diffuse alveolar infiltrates mainly in the mid and lower zones. Diagnosis of IPH is confirmed by the presence of haemosiderin-laden macrophages in the bronchoalveolar lavage fluid. Lung biopsy has been used to establish the diagnosis in the past [11].

The majority of patients with IPH respond favorably to oral corticosteroids with quick recovery during acute flares, a decreased number of IPH exacerbations and an improved overall prognosis [3, 12]. Immunosuppressant agents including azathioprine, hydroxychloroquine, cyclophosphamide and methotrexate have been used with variable results [5]. Historically, the prognosis of IPH was considered to be poor with an average survival of 2.5 years from the time of diagnosis. Conversely, Muhammad et al. [13] have reported a five year survival of 86% in their 17 study patients.

TEACHING POINT

Idiopathic pulmonary haemosiderosis is a rare but important cause of bilateral patchy air-space opacification on a CXR in a child. It should be considered in the differential diagnosis of children presenting with shortness of breath, haemoptysis and radiological evidence of pulmonary infiltrates.

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FIGURES

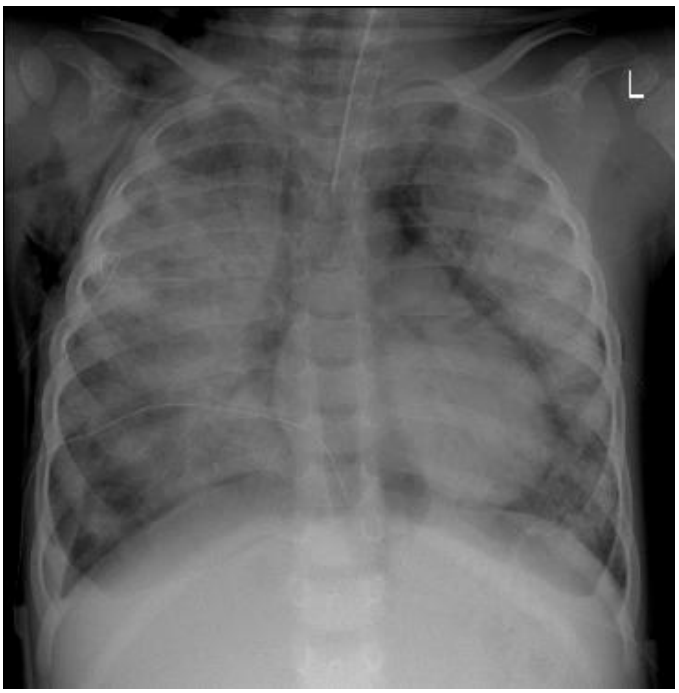

Figure 1: CXR of a 33 month old boy with idiopathic pulmonary haemosiderosis (IPH). It shows endotracheal and nasogastric tubes, a right internal jugular line and bilateral chest drains in-situ. There is diffuse opacification seen throughout both lung fields. There is a major lucency seen around the heart border in keeping with a pneumomediastinum. The lungs remain expanded.

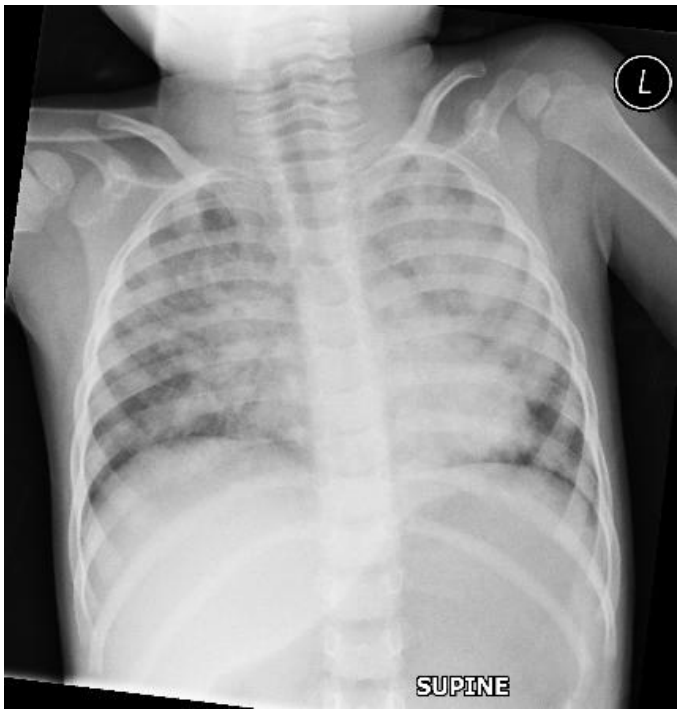

Figure 2: CXR of a 4 year and 9 month old boy with idiopathic pulmonary haemosiderosis showing bilateral diffuse perihilar peribronchovascular consolidations.

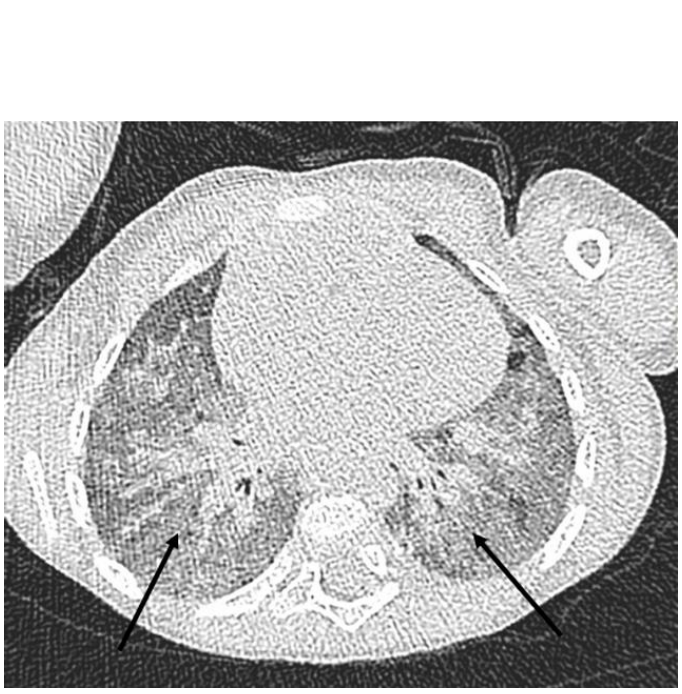

Figure 3: Non-contrast CT scan of a 3 year old boy with idiopathic pulmonary haemosiderosis (IPH). Dose length productivity (DLP) = 4, CTDI vol = 0.29, mAs= 28-30 ms, kVp=100 tube energy and slice thickness= 10mm. The transverse plane shows patchy areas of ground-glass opacity, despite the confounding factor of being taken in the expiratory phase. The ground-glass opacity has a non-specific appearance.

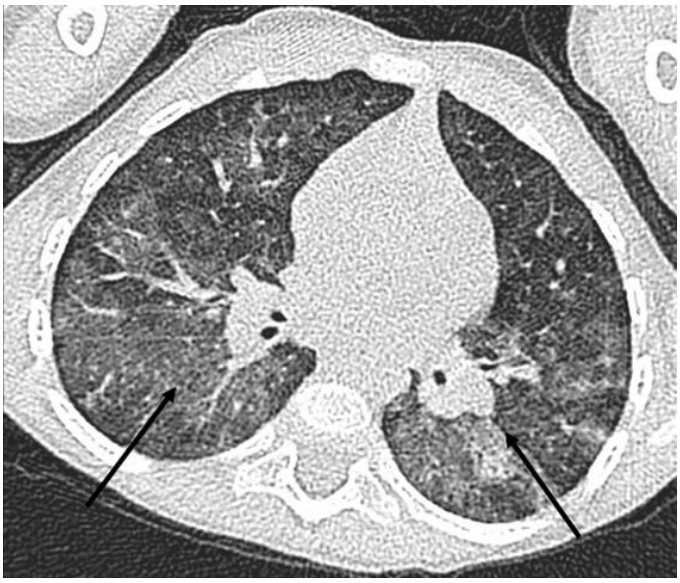

Figure 4: 3 year old boy with idiopathic pulmonary haemosiderosis (IPH). Plain CT scan repeated 4 months after Figure 3. The transverse plane shows that the patchy ground-glass opacification has increased in intensity and extent. Dose length productivity (DLP) = 7, CTDIvol = 0.49, mAs=45.51 ms, kVp= 100 tube energy and slice thickness= 10mm.

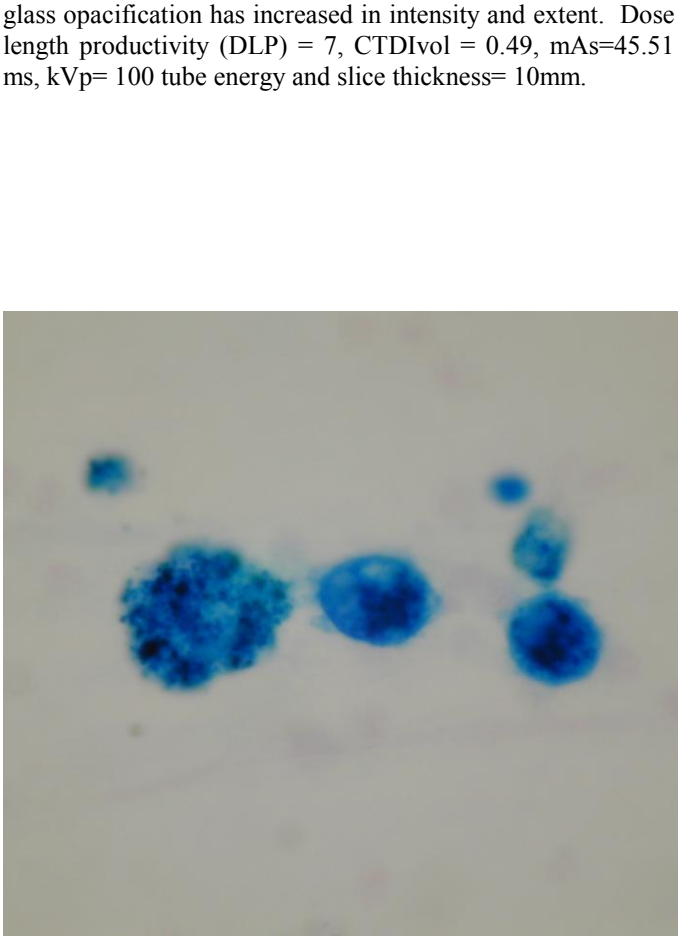

Figure 5: Perl's Prussian blue staining of the bronchoalveolar lavage fluid from a 2.5 year old boy with idiopathic pulmonary haemosiderosis (IPH). The blue stain highlights haemosiderin-laden macrophage infiltration. [Magnification x 100]

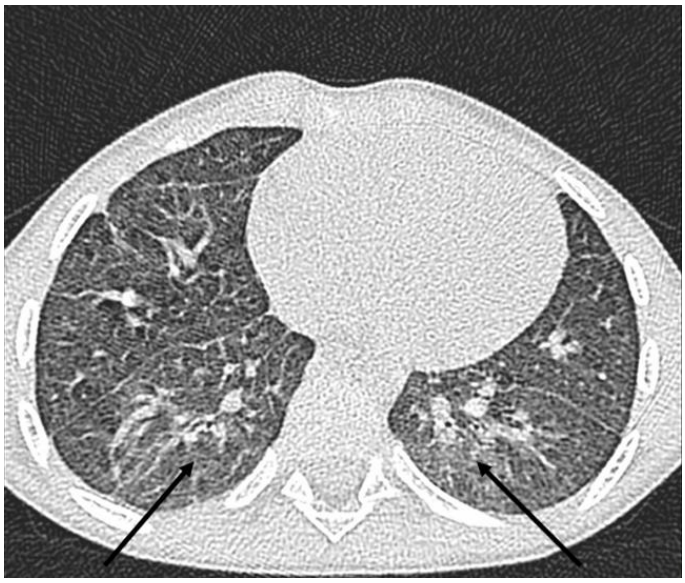

Figure 6: Non-contrast CT scan of a 6 year old boy with idiopathic pulmonary haemosiderosis (IPH). CTDI vol = 0.41, DLP = 8, mAs= 25-28 ms, kVp=120 tube energy and slice thickness=10 mm. The transverse plane shows widespread relatively homogeneous ground-glass opacification throughout both lung fields with a localized area of denser opacification peripherally in the right lower lobe.

Disease	CXR Features	Chest CT Features	Symptoms and signs
Goodpasture's syndrome	Interstitial reticular changes mainly at the bases and perihilar region similar to pulmonary oedema.	Interstitial changes, with repeated episodes of bleeding leading to interstitial reticulosis and fibrosis.	Haemoptysis, haematuria, proteinuria, fever and fatigue.
Pulmonary infections	Pulmonary opacities, consolidation, pleural effusion.	Non specific e.g. consolidation, pleural effusion.	Fever, cough, chest pain, shortness of breath.
Wegener's granulomatosis	Multiple or solitary pulmonary nodules in 40-70 % (1.5-10cm). Heterogeneous air space opacities predominant in the bases.	Pulmonary nodules with or without cavitation and air space consolidation. Interlobular septal thickening.	Non specific: rhinitis, crusting, nosebleeds, rapidly progressive glomerulonephritis, renal failure, scleritis, hearing loss, arthritis, purpura, sensory neuropathy, subglottal stenosis.
Systemic lupus erythematosus	Pleural opacity Pleural effusion.	Interstitial changes and pleural effusion.	Malar rash, vaginal and urinary tract ulcers, arthritis, anaemia, pancytopenia, pericarditis, myocarditis, endocarditis, pleuritis, pleural effusion, headache, seizure, depression, fatigue.
Rheumatoid arthritis	Pulmonary nodules, interstitial changes. Pleural effusion.	Bilateral parenchymal opacities. Bronchiectasis. Nodules.	Polyarteritis, synovitis, rheumatoid nodules, vasculitis, livedo reticulitis, lung fibrosis, fatigue, anaemia.
Diffuse pulmonary hemorrhage: Wegener's granulomatosis, Systemic Lupus Erythematosus, Antiphospholipid syndrome and drug hypersensitivities	Interstitial and air space disease.	Ground glass opacities, reticulations and nodules.	As above.

Table 1: Differential diagnosis of idiopathic pulmonary haemosiderosis (IPH).

Aetiology	Unknown
Incidence	Rare disorder, incidence of 0.24-1.23 cases per million in selected populations
Gender ratio	Balanced sex distribution in childhood Slight male predominance in adults
Age predilection	80% of cases occur in children
Risk factors	Unknown
Treatment	• Corticosteroids • Immunosuppressant agents: azathioprine, hydroxychloroquine, cyclophosphamide and methotrexate. • Supportive measures
Prognosis	Poor, average survival of 2.5 years after diagnosis
Findings on imaging	• Chest X-ray: patchy, diffuse alveolar infiltrate • Chest CT: patchy areas of ground-glass opacification with non-specific appearances

Table 2: Summary table of characteristics of idiopathic pulmonary haemosiderosis (IPH).

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ABBREVIATIONS

IPH = Idiopathic pulmonary haemosiderosis
CT = Computed tomogram/tomography
BAL = Bronchoalveolar lavage
DLP = Dose length productivity
CTDIvol = volume CT dose index, radiation dose

KEYWORDS

Child; Haemosiderosis; Emergency; Idiopathic Pulmonary Haemosiderosis

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