Pulmonary haemosiderosis associated with left atrial myxoma

A A Chaudhry, C M Dobson, F G Simpson

Abstract
A patient with features suggesting pulmonary haemosiderosis was found to have a myxoma. The pulmonary lesion cleared after excision of the tumour.

Idiopathic pulmonary haemosiderosis is a condition of unknown aetiology characterised by recurrent haemoptysis, iron deficiency and transient radiological pulmonary infiltrates in the absence of infection, systemic disease, disordered haemodynamics or coagulopathy.1 It has been associated, perhaps coincidentally, with various conditions. We report a case associated, probably causally, with atrial myxoma.

Case report
A 53 year old white housewife presented with a six month history of daily small haemoptysis and dyspnoea limiting her to 100 metres walking on the flat. There were no other respiratory symptoms or past medical history of note. She smoked 20 cigarettes a day and was having no regular medication.

On examination she appeared to be anaemic and had no lymphadenopathy or finger clubbing. The pulse was 72 beats/min, of normal character and volume, and blood pressure 130/70 mm Hg. The apex was of normal character and position. The first heart sound was well defined with a 2/6 pansystolic murmur radiating into the axilla and a scratchy 2/6 ejection systolic murmur in the pulmonary area followed by a loud pulmonary second sound. The clinical examination otherwise showed nothing abnormal.

The full blood count showed a hypochromic microcytic anaemia of 7·1 g/dl; results of faecal occult blood tests and urine analysis were negative. Her erythrocyte sedimentation rate was 25 mm in the first hour; clotting studies, liver, renal, and thyroid function gave normal results. The electrocardiogram was unremarkable. Her chest radiograph showed diffuse ground glass shadowing in the mid and lower zones of both lung fields (fig 1). Lung function testing showed some airways obstruction, consistent with her smoking history. FEV₁ and forced vital capacity were 1·69 and 2·24 litres (predicted 2·39 and 2·92 l). Total lung capacity (helium dilution) was reduced at 3·32 (predicted 4·69) litres, as was the transfer factor for carbon monoxide when corrected for lung volume (Kco) at 1·66 (predicted 2·02) mmol min⁻¹ kPa⁻¹ l⁻¹. Her Heaf test was grade 2 and serum angiotensin converting enzyme 24 U/l (normal). A ventilation perfusion scan was normal. An autoantibody screen, antineutrophil cytoplasmic antibodies, and avian and aspergillus precipitins were negative; immunoglobulins were normal.

Fibroptic bronchoscopy was unremarkable but bronchial lavage fluid contained huge numbers of haemosiderin laden macrophages. Histological examination of the transbronchial biopsy specimen showed numerous siderophages within the alveoli (fig 2). The alveolar walls showed minimal fibrous thickening but were otherwise unremarkable. There was no evidence of vasculitis. Connective tissue stains confirmed the absence of any vascular occlusive lesions, though in such a small biopsy specimen this did not entirely exclude veno-occlusive disease.

Upper gastrointestinal endoscopy found no gastric cause for blood loss and duodenal biopsy showed normal villi. On catheterisation of the right heart by Swan–Ganz catheter the mean right atrial pressure was 8 mm Hg, right ventricular pressure 47/0 mm Hg, and pulmonary artery pressure 44/14 mm Hg. A satisfactory wedge pressure was not obtained. Her anaemia and breathlessness rapidly improved with iron treatment.
Echocardiography unexpectedly showed a large left atrial myxoma causing obstruction at the mitral valve and occupying most of the atrium during diastole. Surgical removal was successful. Six months after surgery she is well with no dyspnoea or haemoptysis. The alveolar shadowing on her chest radiograph has largely cleared.

Discussion

Idiopathic pulmonary haemosiderosis has been reported in association with coeliac disease, rheumatoid arthritis, thyrotoxicosis, IgA gammopathy, and in one case mild mitral valve disease. All are common conditions and the association may have been coincidental. We are unaware of any reported association with atrial myxoma. Because of the rarity of both conditions and the improvement in the lung condition after surgery this association seems likely to be causal.

It is difficult to be certain of the pathogenesis of haemoptysis in this patient, but intermittent obstruction at the mitral orifice might result in a rise of pulmonary venous pressure with consequent haemorrhage into the alveoli. The intermittent nature of this may explain the difficulty in obtaining a satisfactory waveform during the attempt to measure the wedge pressure. If this is the case it is surprising that there were no more overt clinical manifestations.

Atrial myxoma should be considered as a possible cause of pulmonary haemosiderosis. As clinical signs may be atypical or absent echocardiography should be performed in all patients.

Haemoptysis is a common late manifestation of the Eisenmenger syndrome in adults, and may be a terminal event. The underlying pathological process is usually pulmonary infarction, due to spontaneous thrombosis of branch pulmonary arteries. We report an unusual case in which pulmonary infarction was complicated by life threatening intrapleural haemorrhage and pneumothorax, which required a lobectomy to arrest the haemorrhage.

Successful pulmonary resection after spontaneous haemopneumothorax in the Eisenmenger syndrome

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Abstract

In an unusual case of the Eisenmenger syndrome, in which pulmonary infarction was complicated by life threatening intrapleural haemorrhage and pneumothorax, lobectomy was carried out successfully to arrest the haemorrhage.

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