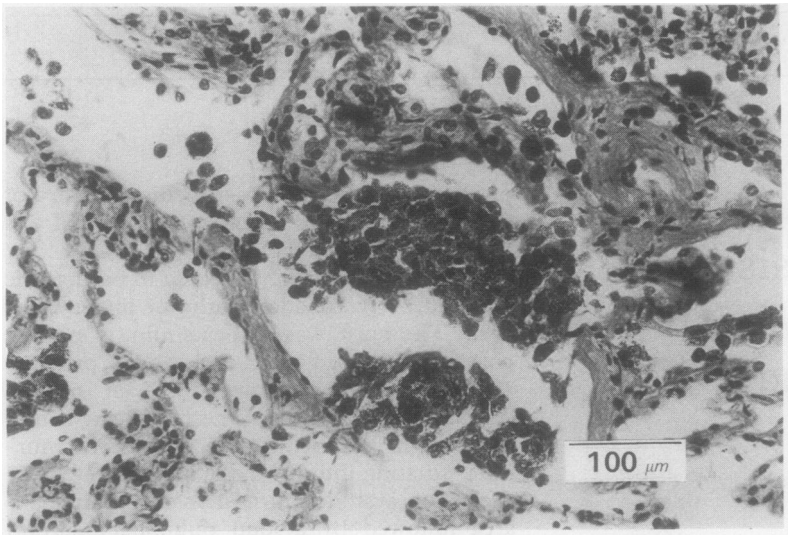

Figure 2 Transbronchial lung biopsy specimen: dense intra-alveolar aggregates of macrophages packed with refractile granules of haemosiderin.

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,[2] rheumatoid arthritis,[3] thyrotoxicosis,[4] IgA gammopathy,[5] and in one case mild mitral valve disease.[6] 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 wave form 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.

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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.

Haemoptysis is a common late manifestation of the Eisenmenger syndrome in adults, and may be a terminal event.[1,2] 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.

Case report

A 24 year old man presented to the accident and emergency department with a three day history of malaise, left pleuritic chest pain, and haemoptysis. He was known to have the Eisenmenger syndrome as a result of persistent truncus arteriosus, a ventricular septal defect, and patent ductus arteriosus. On examination he was cyanosed and shocked with a blood pressure of 75/50 mm Hg. His trachea was deviated to the right and air entry

was diminished over the left mid and lower zones. Chest radiography showed a moderate left effusion with an associated basal pneumothorax. Arterial blood gas values while he was breathing air were: carbon dioxide tension ($Paco_2$) 4·6 kPa, oxygen tension (Pao_2) 5·1 kPa, and H^+ 55 nmol/l.

An intercostal drain was inserted and 1500 ml of blood was drained immediately. A further litre was lost over the next hour as the patient was resuscitated and prepared for surgery. A coagulation screen showed a prothrombin time ratio of 1·8, a partial thromboplastin time of 49 (control 36) seconds, a platelet count of 63×10^9/l, and a normal thrombin time and fibrinogen concentration. Fresh frozen plasma and platelets were transfused.

At surgery 1 litre of blood was found in the pleural cavity, and the left lower lobe was noted to be haemorrhagic and infarcted. There was profuse active bleeding, and the visceral pleura had been stripped off from the underlying lung by a haemotoma. Telangiectatic vessels were present over the surface of the entire lung, and very large collateral vessels were present in the pulmonary ligament and in the peribronchial tissues. Left lower lobectomy was performed, with considerable difficulty in achieving haemostasis: profuse haemorrhage occurred from all cut surfaces and the operative blood loss was 2400 ml. Perioperative arterial blood gas analysis showed: Pao_2 3·4–4·1 kPa, $Paco_2$ 9·8–20·9 kPa, and H^+ 86·5–130·8 nmol/l.

Postoperatively he received assisted ventilation for 18 hours with a high frequency jet ventilator, and was then extubated. Arterial blood gas analysis at all times showed profound hypoxaemia with a mean Pao_2 of 3·7 (range 2·7–4·6) kPa and normocapnia with 4–6 l oxygen/min. Despite this cerebral and renal function remained entirely normal. He made good progress and was discharged home 10 days after surgery. He has since been referred for heart-lung transplantion. Pathological examination of the resected specimen confirmed pulmonary infarction with massive pulmonary haemorrhage and advanced pulmonary hypertensive vascular disease.

Discussion

This is an unusual case of massive haemorrhage and pneumothorax complicating pulmonary infarction in a patient with the Eisenmenger syndrome. Haemothorax complicating pulmonary infarction is rare but has been reported in relation to anticoagulant treatment.[3,4] Pneumothorax complicating infarction is equally uncommon.[5] In this case pulmonary hypertension and "autoanticoagulation" contributed to the severe pulmonary haemorrhage, which disrupted the parietal pleura and caused the pneumothorax. Coagulopathy is well documented in cyanotic congenital heart disease,[6,7] though the mechanism is unclear.

Our patient tolerated a remarkable degree of chronic hypoxaemia (mean resting Pao_2 3·7 kPa) and perioperative acidosis without evidence of organ dysfunction. An acute fall in Pao_2 to 4 kPa or below is usually associated with loss of consciousness,[8] and acclimatised residents nearly 5000 metres have a Pao_2 of around 6 kPa.[9]

From our experience of this case we suggest that patients with the Eisenmenger syndrome who present with haemoptysis should be observed carefully for intrapulmonary haemorrhage and any coagulopathy should be corrected aggressively with blood product transfusion. Despite the very abnormal arterial blood gas tensions, surgical removal of a bleeding lobe is feasible and may be required as a life saving measure.

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