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Life threatening chylous pleural and pericardial effusion in a patient with Behçet's syndrome

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Abstract

Chylothorax and chylopericardium secondary to thrombosis of the superior vena cava and the innominate and subclavian veins were diagnosed in a patient with Behçet's syndrome. Immunosuppressive treatment, diet, and underwater seal drainage led to a diminished volume of pleural fluid and pericardial fluid and to a diminished concentration of triglyceride in them; pleurodesis with tetracycline was then performed.

Pulmonary complications of Behçet's syndrome include thromboangiitis and pulmonary artery aneurysm, both of which may cause life threatening haemoptysis.[1 2] A case of chylothorax in Behçet's syndrome has been reported.[3] We report the first case of life threatening chylous pleural and pericardial effusion secondary to thrombosis of the superior vena cava and the innominate and subclavian veins.

Case report

A 25 year old man was referred with a recalcitrant left sided pleural effusion. Behçet's syndrome had been diagnosed seven years previously on the basis of orogenital ulcers and uveitis. He had been given immunosuppressive agents irregularly. Frequent thoracocentesis over 11 months had led to the removal of 43 litres of pleural fluid for symptomatic relief. On examination he was acutely breathless and distressed and had signs of a massive left sided pleural effusion. Several maculoerythematous lesions 2–5 mm in diameter were present on his arms and trunk and several distended veins on the chest wall. The left eye was enucleated as a result of phthisis bulbae. The right cornea was cloudy, and pupillary reflexes were absent. There were four ulcers on the scrotum and oedema and erythema nodosum on both legs.

Laboratory tests showed that the erythrocyte sedimentation rate was 70 mm in one hour, haemoglobin 12·8 g/dl, white blood cells 11 × 10^9/l, and platelets 351 × 10^9/l. Coagulation studies showed a raised fibrinogen concentration (480 mg/dl). There were no anticardiolipin antibodies and the antithrombin III concentration was normal. The concentration of total serum protein was 5·2 g/dl and of albumin 2·9 g/dl. His chest radiograph showed a massive left sided pleural effusion. An electrocardiogram showed sinus tachycardia (120 beats/min) and low voltage. Echocardiography showed a large pericardial effusion (above 1000 cm^3) with right atrial and ventricular diastolic collapse. A pleural tap yielded a chylous effusion containing triglyceride 177 mg/dl, cholesterol 64 mg/dl, total protein 32 g/l, and white blood cells 3·4 × 10^9/l (77% lymphocytes and 6% neutrophils). Lipoprotein electrophoresis of the pleural fluid showed a chylomicron band; the adenosine deaminase activity was 30 (normal 0–30) IU/ml. Gram staining revealed no organisms and cytological examination showed no abnormality. Thoracoscopy showed numerous collateral veins over the diaphragmatic and costal pleura. The histological appearances of several pleural biopsy specimens were non-specific. An intercostal tube drain connected to a water seal was inserted, and drained about 300 ml daily. Despite this the patient's tachycardia and respiratory distress increased. As the large pericardial effusion was unchanged a pericardial tube was inserted. Laboratory analysis of the fluid confirmed a chylous effusion. There was no bacterial growth and no malignant cells were seen on cytological examination. Pericardial biopsy showed only fibrosis, with no malignant cells or granulomatous changes. A pericardial tube was left in place for three days and drained 1000 cm^3 of fluid. A perfusion lung scan showed a subsegmental perfusion defect in the right lower zone. Upper extremity venography with digital substraction angiography showed total obstruction of the subclavian and innominate veins on the left and obstruction of the superior vena cava above the azygos vein. Contrast material filled the intercostal, lateral thoracic, and azygos veins on the right (fig 1). Bilaterial pedal

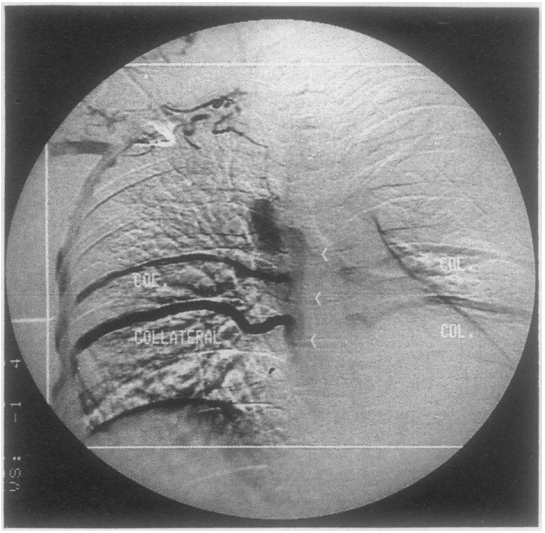

Figure 1 Upper extremity venography with digital subtraction angiography showing obstruction of the superior vena cava; the contrast material fills the intercostal, lateral thoracic, and azygos veins on the right.

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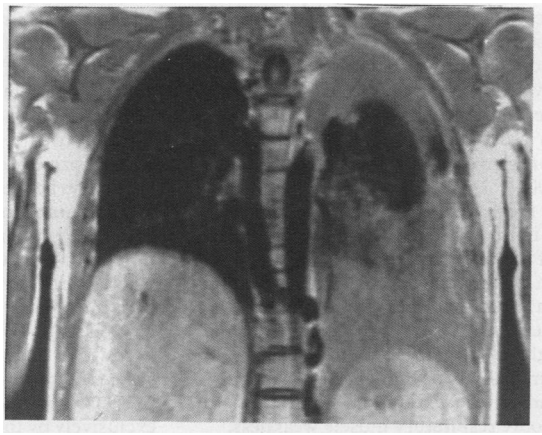

Figure 2 Magnetic resonance imaging scan of the thorax showing protein rich left pleural effusion, pronounced lumboazygous venous collaterals, and dilated azygos veins.

lymphangiography showed retrograde flow of the contrast material into the mediastinum, compatible with obstruction of the thoracic duct. Oesophageal varices were seen during oesophagoscopy. Magnetic resonance imaging of the thorax showed a left pleural effusion with a high protein content and pronounced lumboazygos venous collaterals and dilated azygos veins (fig 2).

The patient was treated conservatively with underwater seal drainage and immunosuppressive drugs (prednisolone 40 mg and azathioprine 150 mg/day) with a diet rich in carbohydrates and medium chain fatty acids. After two weeks the volume of the pleural fluid decreased to 30 ml/day and the triglyceride content to 32 mg/dl, and his symptoms and signs resolved. Pleurodesis with tetracycline 15 mg/kg via intercostal chest tube was carried out; nine weeks later his chest radiograph showed minimal obliteration of the left costophrenic angle with slightly enlarged cardiac borders. The dose of azathioprine was tapered to 100 mg and of prednisolone to 30 mg on alternate days.

Discussion

Although Behçet's syndrome was originally described as the clinical triad of oral and genital ulceration with iridocyclitis, it is a multisystem disease and its pathological hallmark is believed to be vasculitis.[4,5] The patient had all the major clinical criteria for the diagnosis of Behçet's syndrome.[5] Obstruction of the superior vena cava, innominate and subclavian veins was confirmed by imaging techniques. The lipoprotein profile of the pleural and pericardial fluid was consistent with chylous effusion.[6]

Thrombosis of the major veins is a well known feature of Behçet's syndrome. In contrast chylothorax is unusual.[3,7] A pleural transudate has been reported in Behçet's syndrome with obstruction of the superior vena cava resulting from increased high venous pressure.[7] In animals ligation of the superior vena cava above the azygos vein does not produce a pleural effusion, suggesting that obstruction of the azygos vein and vena cava is needed for the occurrence of pleural effusion.[8]

Under normal conditions lymphatic vessels in the lung do not contain chyle. Thrombosis of the subclavian vein may lead to obstruction of the orifice of the thoracic duct, which causes increased intraluminal pressure, back pressure in communicating vessels, and leakage of chyle from the pleural lymphatics into the pleural and the pericardial space.[9-11] It has also been suggested that the chylous fluid may originate through "weeping" from the lung surface into the pleural space.[12] These mechanisms may explain the development of chylothorax and chylopericardium, though obstruction of the superior vena cava may also have contributed to the development of such a large volume of pleural fluid.[8]

Chylothorax secondary to obstruction of the superior vena cava is difficult to treat. Unless obstruction in the venous system is removed, the lungs remain lymphangiectatic.[9] In this case immunosuppressive drugs, bed rest, a special diet, and underwater seal drainage led to a decrease in the volume and triglyceride concentration of the pleural fluid.

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