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Behçet's disease with pulmonary involvement, superior vena cava syndrome, chyloptysis and chylous ascites

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Behçet's disease is a chronic multisystem vasculitis of unknown aetiology. This case report describes a patient who applied to the hospital because of dyspnoea, ascites, oedema of lower extremities and recurrent episodes of haemoptysis. For the last 12 yr, he had superior vena cava syndrome (SVCS) and cardiac and pulmonary involvement of Behçet's disease, and biochemical examination of ascite fluid yielded a chylous effusion containing triglyceride 421 mg dl^{-1} and cholesterol 49 mg dl^{-1}. Chyloptysis was also detected by Sudan III stain. The patient died from cardiac tamponade in spite of cardiac fenestration. To the authors' knowledge, this is the first reported case of Behçet's disease with chylous ascites and chyloptysis in the English literature.

Introduction

Behçet's disease is a multisystem vasculitis which was first described in 1937 by a Turkish dermatologist named Hulusi Behçet. In the distribution of the old Silk Route from Asia across the Mediterranean into Europe (1), Behçet's disease is most often observed in Turkey. This case report describes a patient with Behçet's disease who had pulmonary and cardiac involvement, and superior vena cava syndrome (SVCS) for 12 yr. In addition, he had chylous ascites and chyloptysis.

Case Report

A 40-year-old man with a history of Behçet's disease since 1982 was admitted to the Department of Chest Diseases of Dokuz Eylül University Hospital, Turkey, because of chest pain, dyspnoea, cough, abdominal distention and lower extremity oedema.

He was first diagnosed as having Behçet's disease because of recurrent oral and genital ulcerations, eye involvement and positive Pathergy test. Colchicin 1·5 g daily had been given until cardiac (pericarditis) and vascular involvement [superior vena cava syndrome (SVCS)] developed and then oral prednisolone 8 mg daily was added to the treatment. In 1989, right hydropneumothorax developed and chest tube thoracostomy was applied. Since then, he had been doing well on colchicine 1 g day^{-1} and prednisolone 8 mg day^{-1}. He was admitted to Dokuz Eylül University Hospital in 1994 with chest pain, dyspnoea and abdominal distention. On physical examination, the patient was frail and appeared chronically ill. There was marked oedema over the neck, shoulders and lower extremities. The abdomen was distended and non-tender; a fluid wave and shifting dullness were present. The liver was palpable 6 cm below the costal margin, the spleen was not felt and no masses were palpated. The urine was normal.

The haematocrit was 43%, the white cell count was 12 900, and the platelet count was 304 000. Erythrocyte sedimentation rate was 13 mm h^{-1}. The prothrombin time was 13·4 s and the activated partial thromboplastin time was 42 s. The blood urea nitrogen was 29 mg ml^{-1}, and creatinine was 1·0 mg dl^{-1}. The potassium was 4·8 mmol l^{-1}, AST was 89 IU l^{-1} and ALT was 59 IU l^{-1}. Serum total protein was 5·4 g dl^{-1} and serum albumin was 2·9 g dl^{-1}. All other biochemical tests were within normal limits. An electrocardiogram disclosed a normal rhythm at a rate of 74 bpm and low voltage in all leads was noted. An X-ray film of the chest showed bilateral pleural effusion, right pleural thickening and cardiomegaly. A computed tomographic (CT) scan of the thorax showed mediastinal shift to the right, bilateral pleural thickening, and pericardial

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effusion. In addition to massive calcification at the proximal part of the main pulmonary artery, many abnormalities in the venous vascular structures were noted. The filling defect seen in the superior vena cava was thought to be due to a thrombus. Azygos and hemiazygos veins were dilated. Due to venous stasis, contrast material in the right thoracic wall and axillary veins were not blushed away. No evidence of lymphadenopathy was found in this region. Abdomen CT showed massive ascites, and distended and dilated inferior vena cava. The liver, spleen and kidneys were within normal limits. Magnetic resonance imaging (MRI) of the thorax revealed bilateral pleural effusion and segmental atelectasis on the lower lobe of the right lung, and increased signal intensity in the azygos and hemiazygos veins due to decreased flow. Magnetic resonance imaging examination of the abdomen showed massive ascites. Inferior vena cava was dilated but no evidence of stasis was noted. Portal and splenic veins were patent, no evidence of an intraluminal mass or thrombus was observed. The liver, spleen and kidney had normal architecture and signal intensity. Doppler ultrasound revealed normal flow and calibration of vena porta, vena splenica and vena hepatica, but revealed calcified thrombus in right and left jugular vena and total flow occlusion in the left subclavian vena.

An abdominal paracentesis yielded milky white fluid that contained 7 cells mm^{-3} of which 100% were lymphocytes. The triglyceride content of the fluid was 421 mg dl^{-1}, cholesterol 26 mg dl^{-1}, glucose 133 mg dl^{-1}, protein 0·5 g dl^{-1}, amylase 35 U l^{-1} and LDH was 48 U l^{-1}. Microscopical examination and culture of the fluid showed no fungi, acid-fast bacilli, or other micro-organisms. Cytologic examination of the fluid revealed a predominance of mesothelial cells. Sudan III stain revealed fat globules in fluid.

The patient was treated with 40 mg prednisolone and 80 mg furosemide daily. His complaints decreased and physical examination revealed improvement. The patient was discharged and called for the follow-up period. One month later, he presented with pericardial tamponade because of massive pericardial effusion, bilateral pleural effusions and massive ascites. Sputum and ascites fluid had a milky appearance. Chyloptosis was confirmed by Sudan III stain. Massive pericardial and pleural fluids had sero-haemorrhagic biochemical analysis. Pericardial fenestration and pleural tube thoracostomy were made because of hypotension. Anuria and shock progressed despite medical support and the patient died. Pericardial mononuclear cell infiltration and hepatic periportal fibrosis were detected by post-mortem needle biopsies. His family did not give permission for autopsy.

Discussion

Pulmonary involvement in Behçet's disease is not usual. The prevalence is reported to be around 5% by Raz *et al.* In Turkey, studies have reported 92, 34, 32, 20, 15 and nine cases with pulmonary involvement (2–4). To the authors' knowledge, this is the first reported case of Behçet's disease with chylous ascites and chyloptysis in the English literature.

Chylothorax is much more common than chylous ascites, although both are rare. In 50% of cases, chylothorax is related to a tumour (5). Normally, chylomicrons are absorbed from the gastrointestinal tract, enter the intestinal lacteal vessels, empty into the cisterna chyli which overlies the body of the second lumbar vertebra, and then drain into the thoracic duct across the oesophageal hiatus into the thoracic vein cavity on the right side in an extrapleural location until the level of T4 or T6, where it crosses the mediastinum and then empties into the left subclavian vein.

Chylous ascites is rarer. In 30% of cases, it is due to a neoplasm; in 35% of cases, it is due to inflammation; in 11% of cases, it is due to trauma; in 1% of cases, it is due to congenital causes; and in 23% of cases it is idiopathic. In a recent report, neoplastic disease was the cause of chylous ascites in 87% of cases. Lymphomas constitute about half of these; the other half is caused by a variety of primary and metastatic neoplasms. The inflammatory causes of chylous ascites are quite varied, including tuberculosis, pancreatitis, portal vein thrombosis, mesenteric adenitis, adhesions with intestinal obstruction, radiation and pulmonary fibrosis. With thoracic duct obstruction, chylous ascites may also be seen rarely in uncomplicated cirrhosis (6). In the present case, CT and MRI did not show any intra-abdominal or intrathoracic masses which compressed or infiltrated the duct. Chylous ascites in this patient may have developed due to a mechanism similar to that of other inflammatory causes. In this patient, SVCS may have caused retrograde stasis in the ductus thoracicus and may have contributed to development of chylous ascites, although, in the authors' opinion, this alone should not be wholly responsible, because the retrograde pressure transmitted by the SVCS may increase the pressure in the ductus thoracicus, mainly immediately before it empties into the venous system. If any rupture was present at this level, chylothorax would have accompanied the chylous ascites.

This patient had multiple organ involvement of Behçet's disease such as vascular [SVCS and inferior vena cava syndrome (IVCS)], pulmonary (bilateral pleural effusions, history of pneumothorax), cardiac (pericarditis) and eye (decolman of the vitreus) involvements and chylous ascites and chyloptysis. The authors suggest that Behçet's disease should be considered in the differential diagnosis of chylous ascites.

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