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Massive unilateral chylous pleural effusion: a rare initial presentation of Behcet's disease

Husain S J, Sadiq F, Zubairi A B, Khan J A

ABSTRACT

Pulmonary manifestations of Behcet's disease are not very common and usually include pulmonary artery aneurysms, central venous thrombosis, pneumonia and pleurisy. Chylothorax secondary to superior vena caval obstruction is a rare complication and has been reported in only a few cases. We report a case of a 24-year-old man presenting with massive chylothorax as the initial presentation of Behcet's disease that was successfully treated conservatively.

Keywords: Behcet's disease, chylothorax, pleural effusion, superior vena cava thrombosis

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INTRODUCTION

Behcet's disease is a multisystem chronic inflammatory vasculitis of young adults characterised by recurrent attacks^(1,2). Pulmonary complications due to Behcet's disease are uncommon. The usual pulmonary involvement includes thromboangiitis, pulmonary artery aneurysms, recurrent pneumonias, and pleurisy⁽¹⁻³⁾. We present a case where massive chylous pleural effusion was the presenting feature of Behcet's disease.

CASE REPORT

A 24-year-old man presented to the emergency department with complaints of progressively-worsening dyspnoea for three months. He also complained of a chronic nonproductive cough and low grade fever for the last two and a half months. His appetite had been poor for a few weeks and he had lost five kg. His past history was significant for recurrent painful oral and genital ulcers with serous discharge. On examination, he was tachypnoeic and tachycardic. He had distended superficial veins over the right anterior chest wall, neck and right upper arm. His right upper extremity and face were swollen. He also had dullness to percussion and diminished breath sounds over the right hemithorax. There was evidence of tender raised nodules on both his legs. The rest of the systemic examination including the eye examination

was normal. There were no visible oro-genital ulcers.

The chest radiograph showed complete opacification of the right hemithorax (Fig. 1). The serum studies were unremarkable except for a raised sedimentation rate of 41 mm in the first hour. A diagnostic and therapeutic pleural tap was done, and 1 L of milky fluid was aspirated (Fig. 2) The fluid contained total protein



Fig. 1 Chest radiograph shows complete opacification of the right hemithorax.

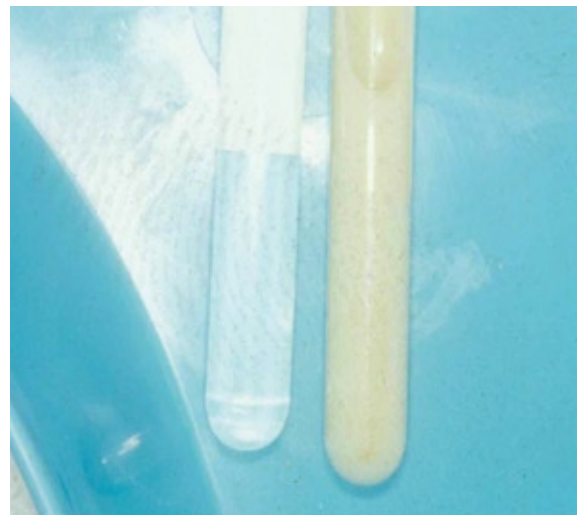


Fig. 2 Photograph shows milky fluid aspirated, compared with water.

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of 4,300 mg/dL, triglyceride of 1,320 mg/dL and cholesterol of 32 mg/dL. The cell count of the fluid showed a white blood cell count of 7,600/L with a predominance of lymphocytes (95%). The microbiological and cytopathological studies were all normal. Doppler ultrasonography of the neck showed thrombosis of right internal jugular and right subclavian veins. Spiral computed tomography (CT) of the chest showed thrombosis of the innominate vein and the superior vena cava, without any distal embolus.

A diagnosis of Behcet's disease with chylothorax secondary to thrombosis of the central veins was made. The patient was started on oral prednisolone (Deltacortril, Pfizer, Karachi, Pakistan) at a dose of 60 mg a day. Anticoagulation was initiated with enoxaprin (Clexane, Aventis Pharma, Karachi, Pakistan) at 1 mg/kg twice a day initially, and later switched to oral warfarin (Marevan, Glaxo Wellcome, Karachi, Pakistan) to maintain an international normalised ratio (INR) of 2.0-3.0. He was also advised to keep to a low-fat diet. His symptoms improved dramatically, and within three months his dyspnoea and chest radiograph findings resolved (Fig. 3). Currently, eight months later, he is asymptomatic and is maintained on 5 mg of prednisolone and oral warfarin.



Fig. 3 Repeat chest radiograph taken three months later shows clearing of the effusion.

DISCUSSION

Behcet's disease is a multisystem chronic inflammatory disorder of unknown aetiology, characterised by systemic vasculitis and perivascular inflammatory infiltrates. Classically, a triad of symptoms is present, including recurrent aphthous oral ulcers, uveitis and genital ulcers. Other organs involved include skin, joints, large vessels,

central nervous system, and gastrointestinal and pulmonary involvement⁽¹⁻²⁾. Pulmonary involvement is uncommon, with studies quoting figures of 1% to 7.7%⁽³⁾. Typically, pulmonary artery aneurysms, arterial and venous thrombosis, pulmonary infarction, recurrent pneumonias, bronchiolitis obliterans with organising pneumonia, and pleurisy are the common pulmonary manifestations.

Our patient presented with a chylous effusion secondary to thrombosis of the large central veins including superior vena cava, right innominate, subclavian and the internal jugular veins. To our knowledge, chylothorax being the presenting feature of previously undiagnosed Behcet's disease, has not been previously reported. The diagnosis of Behcet's disease was made on the basis of the criteria proposed by the International Study Group for Behcet's Disease in 1990⁽³⁾. This criteria require the presence of oral ulcers and two additional findings from the following manifestations: genital ulcers, uveitis, skin lesions including erythema nodosum, pseudo-folliculitis, or papulopustular lesions and a positive reaction to a pathergy test.

Our patient was a young man with a history of recurrent oral and genital ulcers, erythema nodosum and central venous thrombosis. Thrombosis of major vessels is a well-known feature of Behcet's disease^(2,4). Propagation of peripheral venous thrombosis may lead to serious vascular complications, such as caval, portal or hepatic system occlusion. Thrombosis of superior vena cava may be life-threatening owing to complications like pulmonary emboli and chylothorax^(4,6). The analysis of the pleural fluid was consistent with an exudative lymphocyte dominant effusion, and the chemical analysis was consistent with chylothorax.

The aetiology of chylous effusion in Behcet's disease is uncertain. It has been hypothesised that thrombosis of the subclavian vein may cause obstruction of the orifice of the thoracic duct, leading to increased intraluminal pressure, back pressure in the communicating vessels and leakage of chyle from the pleural lymphatics into the pleural space^(5,6). The thrombosis of the superior vena cava also contributes to the development of the effusion^(5,6). Chylothorax due to central venous thrombosis is very difficult to treat. Previous reports^(4,6) have suggested complete drainage via a chest tube, followed by chemical pleurodesis along with anticoagulation and immune suppressive therapy.

We were able to successfully manage our patient conservatively with oral steroids, anticoagulation and a low-fat diet. Within a couple of months, our patient had a dramatic response to treatment, with

complete resolution of symptoms and clearing of the chest radiographical abnormalities. In conclusion, chylous pleural effusion is a rare presentation of Behcet's disease. However, it should be considered in the differential diagnosis of non-traumatic chylous pleural effusion. Medical management should be considered as the first option.

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Professor S. Y. Tan (author of the Stamps in Medicine series in the SMJ) will be in Singapore in November-December 2006 to conduct on-site Q/A sessions and interviews. If you are interested in an appointment, kindly send a brief resume to siang@hawaii.edu. You do not need to have completed all of your qualifying exams (USMLE parts I and 2K and 2CS) to meet up with Prof. Tan.