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Chylothorax and Chylopericardium Complicating Behcet's Disease: A Case Report

Hanaa Harraz[1], Afaf Amir[2*], Nahid Zaghba[1], Hanane Benjelloun[1], Khadija Chaanoun[1], Najiba Yassine[1]

[1]Respiratory Diseases Department, Ibn Rochd University Hospital, Casablanca, Morocco, 20100
[2]Faculty of Medicine and Pharmacy- Hassan II University, Casablanca, Morocco

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***Corresponding author:** Afaf Amir
Faculty of Medicine and Pharmacy- Hassan II University, Casablanca, Morocco

Abstract **Case Report**

Behçet's disease is a complex multisystem vasculitis of unknown aetiology, characterised by different clinical manifestations and the occurrence of successive attacks interspersed with remissions. Mediastino-pulmonary involvement is rare but severe, resulting mainly in thrombosis of the superior vena cava and pulmonary artery aneurysm. The association between Behçet's disease and chylothorax and/or chylopericardium is rare, with a few cases reported in the literature. We present a case of chylothorax and chylopericardium with thrombosis of the superior vena cava as clinical manifestations in a patient with Behçet's disease. Management was multidisciplinary, based on pleuropericardial drainage, immunosuppressants, anticoagulants and nutritional management.

Keywords: Behçet's Disease, Chylothorax, Chylopericardium, Thrombosis.

INTRODUCTION

Behçet's disease is a multisystemic vasculitis affecting vessels of all sizes, with a venous tropism, and is characterized by successive flare-ups interspersed with remissions. Mediastino-pulmonary involvement is rare but serious, essentially involving thrombosis of the superior vena cava and pulmonary artery aneurysm. The occurrence of chylothorax and chylopericardium during the course of the disease is rare. We present a case of chylothorax and chylopericardium with thrombosis of the superior vena cava (SVC) as clinical manifestations in a patient with Behçet's disease.

CASE REPORT

A 57-year-old man, a chronic weaned smoker. He had been diagnosed with Behçet's disease at 23 years old, based on the presence of recurrent oral aphthosis (6 episodes/year), pseudofolliculitis, and retinal vasculitis. He had been treated with corticosteroids for 10 years and then lost sight of. He had undergone surgery for a left cataract at the age of 33, and had undergone retinal laser surgery on the right eye 10 years previously.

For the past 2 months, the patient had been progressively presenting with stage II mMRc exertional dyspnoea, associated with oedema of the face and neck, evolving in a context of conservation of general condition. Dyspnoea worsened with the onset of right-sided basithoracic pain one month before admission.

On physical examination, he was apyretic, with an oxygen saturation of 93% on room air, a respiratory rate of 24 cycles per minute, a heart rate of 98 beats per minute and a blood pressure of 116/81mmHg. Somatic examination revealed a superior vena cava syndrome in the state phase, with pellucid oedema, turgidity of the jugular veins and collateral venous circulation. Examination revealed a syndrome of fluid effusion in the lower two-thirds of the right hemithorax, multiple mouth ulcers, and extensive pseudofolliculitis of the face and back.

The chest X-ray revealed a pleural opacity occupying almost the entire right hemithorax. (Figure 1), exploratory pleural puncture revealed a thin pleura, and yielded a lactescent fluid, whose chemical and cytobacteriological analysis showed aseptic lymphocytic cellularity, a triglyceride level of 6.55 g/L, with the presence of chylomicrons, and a cholesterol level of 0.63 g/L. Biological tests showed a white blood cell count of 5820/mm3, haemoglobin of 13.8 g/dl and platelet count of 434,000/mm3. Protein levels were normal at 68g/l. The serum lipid profile was unremarkable. Renal and hepatic function were normal. Thoracic angioscan revealed the right pleural effusion already described,

associated with pericardial effusion and thrombosis of the superior vena cava extending to the innominate vein and the right internal jugular vein, as well as significant mediastinal and subcutaneous collateral circulation of the anterior cervicothoracic-abdominal wall (Figure 2). Ophthalmological examination and retinal angiography revealed blindness in the left eye, areas of retinal ischaemia and iridial rubeosis. Transthoracic echocardiography confirmed the presence of a large pericardial effusion with significant variations in flow (60% in the mitral region and 50% in the aortic region) with collapse of the cardiac chambers. Puncture revealed a lactescent pericardial fluid rich in triglycerides.

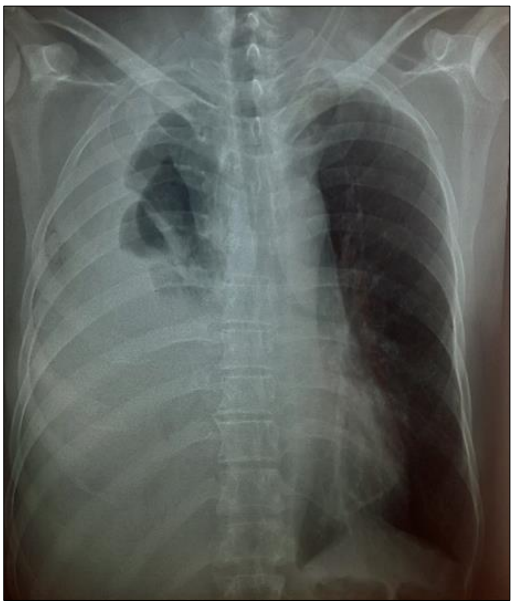

Figure 1: Chest X-ray showing a right pleural opacity

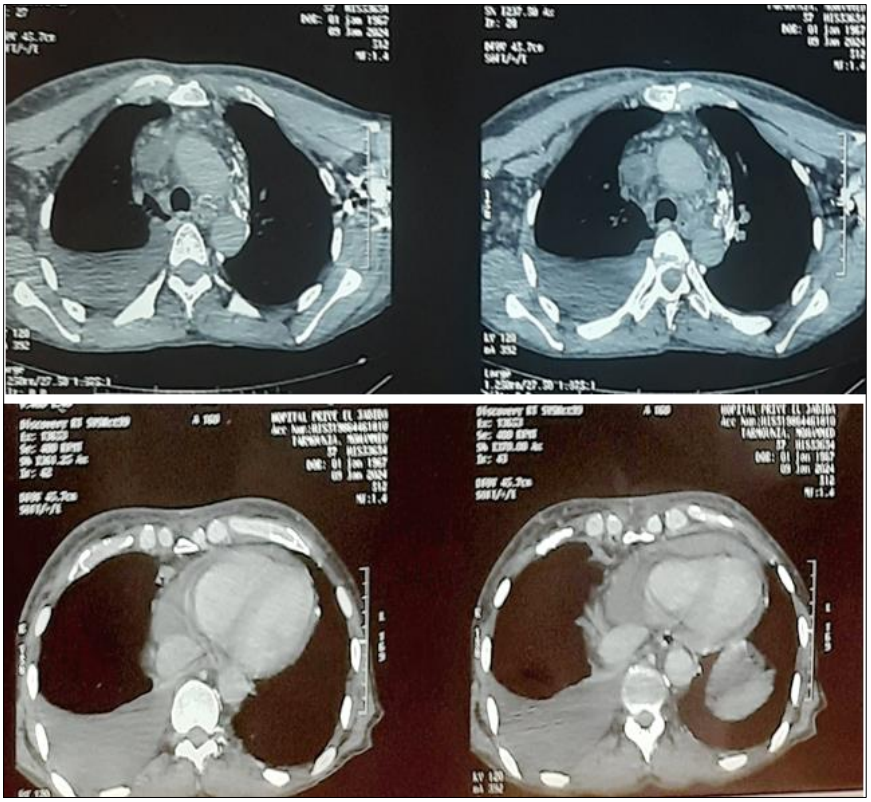

Figure 2: Thoracic angioscan showing pleural and pericardial effusion with thrombosis of the superior vena cava extending to the innominate vein and the internal jugular vein, as well as significant mediastinal and subcutaneous collateral circulation

We adopted the diagnosis of chylothorax and chylopericardium complicating superior venous thrombosis in the context of Behçet's disease. Treatment consisted of pleural and pericardial drainage combined with a low-fat diet and anticoagulation with curative dose heparin therapy. After a negative infectious work-up, the patient received boluses of methylprednisolone for 3 days, then prednisone 1mg/kg/d, combined with boluses of cyclophosphamide, followed by azathioprine. In view of the recurrence of pericardial effusion, the patient underwent a pleuropericardial window. The clinical and radiological outcome was good (Figure 3).

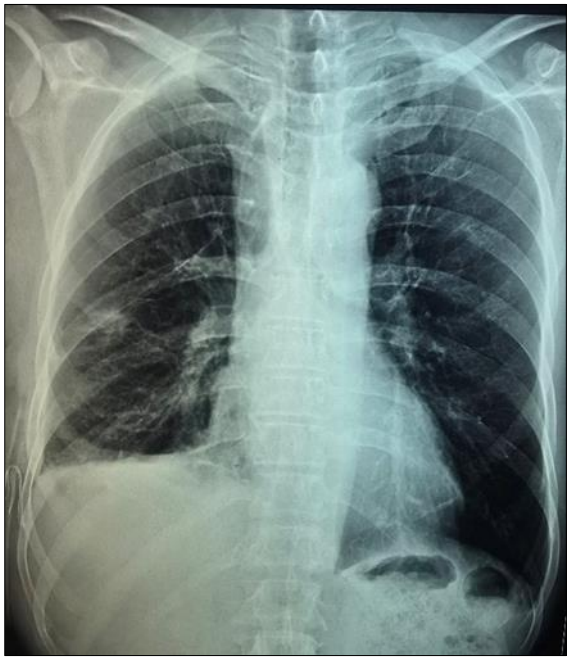

Figure 3: Follow-up chest X-ray showing regression of pleural opacity after treatement

DISCUSSION

Behçet's disease is a complex multi-systemic vasculitis of unknown aetiology characterised by different clinical manifestations, including cutaneous-mucosal, ocular, vascular, articular, neurological and gastrointestinal. It is distributed worldwide, but is most prevalent in the Mediterranean, Central Asia and the Far East [1]. Arterial or venous thrombosis due to Behçet's disease can affect any vessel in the body. Superior vena cava thrombosis is a well-recognised complication of this disease, and is a frequent cause of superior vena cava syndrome in areas where the disease is prevalent.

Our patient was diagnosed with Behçet's disease on the basis of international criteria first published in 1990 [2], and updated in 2006 [3]. He had no traumatic, neoplastic or surgical causes that could explain the presence of chylothorax.

The association of Behçet's disease and chylothorax is rare. Its prevalence is not estimated. Only a few cases have been reported in the literature. The pathogenesis of chylothorax and chylopericardium in Behçet's disease is uncertain. Superior vena cava thrombosis is the most likely mechanism of chylous effusion, and is thought to be due to increased pressure in the lymphatic system where thrombosis of the superior vena cava creates reflux through the subclavian vein, increasing pressure in the thoracic duct and compromising its drainage, a phenomenon known as 'chylous reflux'. This leads to an accumulation of chyle in the small lymphatic vessels of the thorax and in the pleural and pericardial space [4].

In cases of chylothorax reported in the context of Behçet's disease, the effusions are often bilateral or predominantly on the left. A chylous pericardial effusion may be associated in 55% of cases [5].

The initial paraclinical examination of a non-traumatic chylothorax consists of an angioscan of the thorax and abdomen to look for lymphoma, the most common aetiology [6]. Lymphatic exploration by lymphangiography or lymphoscintigraphy in search of a chyle leak or obstruction is considered when no clear aetiology emerges from the thoracic CT scan. Videothoracoscopy or pleural biopsy has no role in the investigation of chylothorax as the pleura is not involved [7].

Management of these cases based on long-term anticoagulants, corticosteroids and immunosuppressants, in addition to drainage of pleural and pericardial effusions, is the treatment of choice for thrombosis of the superior vena cava complicated by chylothorax and chylopericardium during Behçet's disease [8-10].

Anticoagulation alone is insufficient in Behçet's disease, as the ongoing inflammation may perpetuate or

create new thromboses. A search for pulmonary artery aneurysms or other arterial aneurysms should be carried out before anticoagulation, as these may lead to fatal haemorrhage [5-10].

The leakage of chyle can lead to a loss of proteins, fats, vitamins and electrolytes, resulting in undernutrition, metabolic disorders and immunosuppression linked to the loss of lymphocytes present in the chyle fluid [5]. Nutritional management is therefore essential. A low-fat diet or interruption of oral nutrition, or even entirely parenteral nutrition, can help to stop the digestive intake of triglycerides [8].

CONCLUSION

Behçet's disease complicated by chylothorax and chylopericardium remains rare, with only a handful of cases reported in the literature. Management is multidisciplinary, based on immunosuppressive therapy, and surgery may be useful, particularly in cases complicated by chylopericardium.

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