A 23-year-old man presented with progressive swelling in the left supraclavicular area and worsening dyspnea of 7 days duration. Histories of fever, chills, night sweats, cough or weight loss was denied. Medical and family histories were unremarkable. Physical examination revealed usage of accessory muscles of respiration, facial plethora, acne, multiple dilated veins on chest wall, firm non-tender bilateral supraclavicular lymph nodes and decreased air entry and dullness in both lung bases. Chest X-ray revealed bilateral pleural effusions (Figure 1A). Computed tomography (CT) scan of the neck and chest demonstrated thrombotic occlusion of the superior vena cava, right subclavian, innominate and internal jugular veins, mediastinal fibrosis, subcentimeter mediastinal adenopathy and a mass on the left cardiac atrioventricular wall (Figure 1B and C). Pleural fluid revealed a triglyceride concentration of 799 mg/dl and lactate dehydrogenase (LDH) of 947 U/l with negative cultures and cytology. Lymph node and pleural biopsies were benign. Cardiac magnetic resonance imaging revealed a cystic 3.1 × 3.1 cm mass, which was delineated to be a thrombosed pseudoaneurysm of the left circumflex coronary artery by cardiac catheterization. During hospitalization, the patient developed oral aphthous ulcers and folliculo-pustular rash on chest. Biopsy revealed superficial and perivascular inflammation. A positive pathergy test followed, clinching the elusive diagnosis of Behcet's disease. The patient was started on anticoagulation along with cyclophosphamide and prednisone. For the persistent chylothorax, triglyceride restriction, chest tubes with attempts at pleurodesis proved futile. After an attempt at thoracic duct embolization, ligation and partial decortication of the right lung were performed. Six months into follow-up, the patient still has a loculated right-sided pleural effusion, with the disease in remission.

Discussion

Behcet's disease is a chronic multisystem inflammatory disease characterized by recurrent oral and genital ulcers and ocular lesions. Diagnosis of Behcet's is based on the criteria proposed by the International Study Group for Behcet's disease in 1990. These include oral ulcers and at least any two of the following: recurrent genital ulceration; uveitis, skin lesions including erythema nodosum, pseudofolliculitis or papulopustular lesions and positive pathergy test. The disease is more common in the middle-eastern population and in the far east, with more severe manifestation in young males. Though the pathogenesis is obscure, the main pathologic lesion is the systemic perivasculitis with early neutrophil infiltration and endothelial swelling. Perivasculitic process involving the vasa vasorum of the large vessels can ultimately lead to thromboembolic phenomenon, occlusion and in 5% of cases, arterial aneurysm formation. Though both arterial and venous involvement is known, predilection is more for the venous. In rare instances when the thrombotic complications of the large vessels become the primary manifestation of the disease, without temporal association of the
well-known clinical triad, it becomes a diagnostic challenge with unavoidable delays.

Pulmonary involvement is noted in only 5% of cases with the more commonly reported ones being pulmonary infarctions, pulmonary artery thrombus or aneurysm and sometimes pleural and pericardial effusions. Chylothorax, defined as triglyceride concentration in the pleural fluid >110 mg/dl, is a very rare presenting manifestation, with only 16 reported cases so far in the medical literature. It is primarily a consequence of the vasculitic process affecting the large vessels of the thorax. Thrombosis of the left subclavian and the innominate vein obstructs the drainage of the thoracic duct, and subsequent increase in the intraluminal pressure forces out chyle from the pleural lymphatics. Simultaneous thrombosis of the superior vena cava lends a larger volume to the pleural effusion.4

Once spontaneous chylothorax is encountered and various other neoplastic, infectious and prothrombotic disorders are ruled out, the diagnosis of Behcet’s disease can be instituted clinically. The key to diagnosis remains a high index of suspicion and a thorough physical examination. Chylothorax resulting from Behcet’s disease can be very recalcitrant to treatment. Aside from anticoagulation and immunosuppression, conservative measures like dietary restriction to medium chain fatty acids are of little benefit. Complete drainage of the chylous effusion via chest tubes along with octreotide therapy and pleurodesis is an acceptable approach, however for extreme cases, aggressive surgical measures like ligation of the thoracic duct and lung decortication are mandated.5

To conclude, our case reiterate the often formidable task of diagnosing Behcet’s disease and the hurdles in management of chylothorax, a rare pulmonary complication of the same.

Conflict of interest: None declared.
References


