Right Atrium Thrombus and Pulmonary Artery Aneurysm in a Man with Behçet’s Disease

Mohamed Elqatni, Youssef Sekkach, Ali Abouzahir and Driss Ghafir

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A 30-year-old man, with no prior pathologic antecedent was admitted to the internal medicine department for a 3 month history of cough and bloody sputum. These symptoms occurred in the context of apyrexia, night sweating and 6 kilogram weight loss. On physical examination the patient appeared pale, BMI was 17 kg/m², crackles were heard in the left lung. The skin examination noted painful genital and oral ulcers with pustular lesions in the back.

Chest X-ray showed left hilar opacity. Angio CT scan revealed an aneurysm of the left inferior pulmonary artery (Picture 1) with parietal thrombus, right basal pulmonary infarction, and right intra-atrial thrombus (Picture 2). Echocardiography noted the mobile thrombus in the right atrium (Picture 3) and another thrombus in the vena cava junction. Laboratory tests found inflammatory anemia. The infectious analysis (VDRL-TPHA, Mycobacteria) was negative. There were definite findings for thrombophilia or connectivitis, however HLA-B51 was positive. The diagnosis of Behçet’s disease was made. The patient was treated with cyclophosphamide and methylprednisolone pulses relayed by oral prednisone 1 mg/kg/j and anticoagulation. After 5 months there was disappearance of intracardiac thrombi and marked regression of the pulmonary artery aneurysm.

The location is rare in Behçet’s disease: from 1 to 5% in clinical series, 16.5% on a register of autopsies in Japan and...
15% in an echocardiographic series (1). Intracardiac thrombosis is rare. It may be isolated or associated with aneurysms of the pulmonary artery and accompanied by an increased risk of mortality.

A similar case was reported by Marc et al (2) in a 60-year-old man. The aneurysm of the pulmonary artery and intracardiac thrombosis occurred two years after the diagnosis of Behçet’s disease and the thrombus was located in the right ventricle which is a relatively frequent site.

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References