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Octopus-Like Thrombus: An Unusual Complication with Behcet's Disease

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Abstract

Behcet's disease (BD) is a chronic inflammatory multisystemic disorder with an unknown cause. Although BD may involve many organs or systems in the body, cardiac pathologies related to BD have seldomly been reported. Intracardiac thrombus formation is a rare but very serious complication of BD. In the following text, we'll share a patient with BD who has massive thrombus formation in right heart chambers and large veins.

Keywords:

Behcet's Disease, thrombus, right ventricle, right atrium

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Manuscript

A 30-year-old male patient with complaint of fatigue and dispnea for two months was referred to our cardiology department with a diagnosis of intracardiac mass. He was diagnosed as Behcet's Disease five years ago and has given up his medical treatment for two years without the counselling of his doctor. His physical examination was normal except for sinus tachycardia and subfebril fever at the administration of our clinic. The laboratory tests resulted in a normal leukocyte count, mild anemia, marked elevation of eritrocyte sedimentation rate (ESR) and serum C-reactive protein (CRP) level. The protein C, protein S and anticardiolipin antibody levels were within normal limits. Electrocardiogram, chest X-ray and deep-vein Doppler ultrasound analyses were evaluated normal. On transthoracic echocardiography there were heterogeneous hyperechoic mobile masses seen in the inferior vena cava (IVC), in the basal portion of the right atrium, and on the septal leaflet of tricuspid valve. Also a fixed, huge mass plastered on the whole surface of the right ventricular wall filling more than one half of the right ventricular cavity was seen. All of these mass images were compatible with thrombus (*Figure 1A and 1B, asterisk*). In addition to these findings mobile thrombus images along superior vena cava (SVC) and IVC were shown with transesophageal echocardiography (TEE) (*Figure 1C, asteriks*). Cardiac MRI revealed mass images in the right atrium, right ventricle and SVC at thrombus density (*Figure 1D, 1E and 1F, arrows*). We offered surgical treatment to the patient, but he refused the operation. Then we administered prednisolon, colchicine and cyclophosphamide treatment and anticoagulated the patient with warfarin.

Behcet's disease (BD), first described by Dr. Hulusi Behçet in 1937, is a chronic, multisystem, inflammatory disorder characterized with recurrent oral and genital ulcers and relapsing uveitis.

Venous or arterial thrombosis is reported in 7% to 38% of patients in different series (1). Venous thrombosis is seen more commonly than arterial thrombosis, with relative frequencies of 90% and 10%, respectively (2). Although vascular complications may be seen commonly as a component of the disease, cardiac involvement is very rare. Intracardiac thrombosis is an unusual and severe condition during the course of the disease with a tendency to recur and has a mortality rate of approximately 30%. It usually occurs among young male patients and located mostly in the right heart chambers, especially the right ventricle (3). Transthoracic and transesophageal echocardiographical evaluation is usually enough for the diagnosis. Images on echocardiography may resemble other conditions such as tumour or vegetation, so we must consider the clinical findings for the correct diagnosis (4). Although there is no consensus on the treatment of intracardiac thrombi, the aim should be to control the underlying disease and resolve the thrombus. Anticoagulant and antiplatelet drugs are the first choice for this purpose (3). Immunosuppressive treatment with cyclosporine and glucocorticoids (prednisolone) is commonly used in order to take control of Behcet's disease (5).

In conclusion, we suggest that Behçet's syndrome should be kept in mind in the differential diagnosis of intracardiac thrombi.

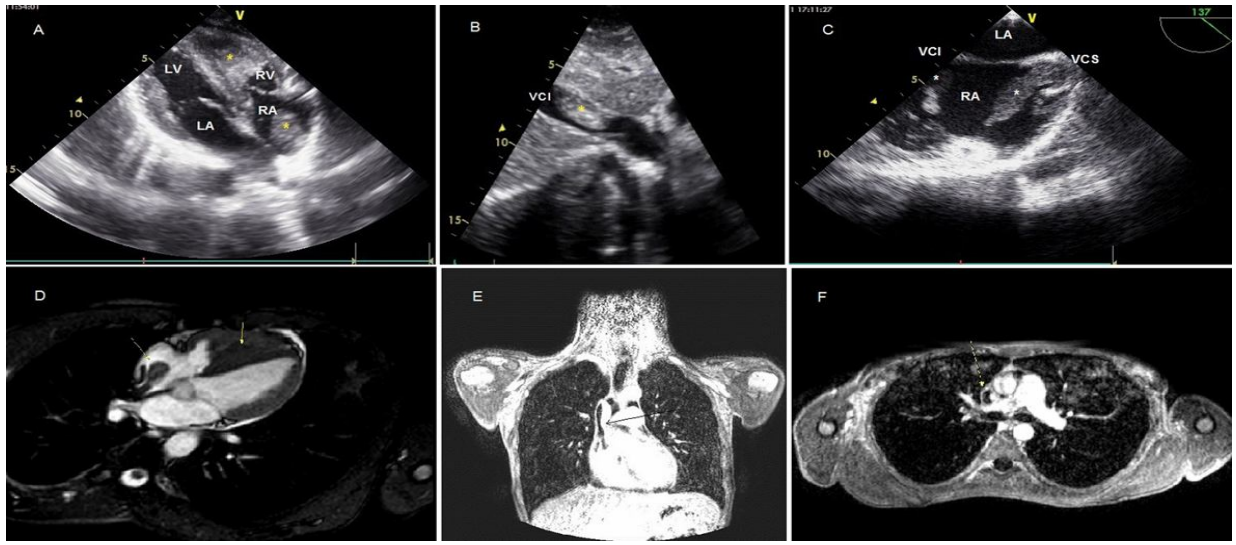

Figure 1: Transthoracic echocardiography shows mobile thrombus images in RA and RV in apical 4-chamber view (1-A), and in the IVC in subcostal view (1-B) (asterisk), and also mobile thrombi seen from IVC and SVC extending into the RA in bicaval view on transesophageal echocardiography (1-C) (arrows). MRI reveals mobile thrombus images in the SVC (1-E,1-F) and in the base of RA and plastered thrombus on RV walls, filling almost one half of the RV cavity(1-D) (arrows).
RA: right atrium, RV: right ventricle, IVC: inferior vena cava, SVC: superior vena cava

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