P 208: Right Ventricular Thrombus with Behçet's disease: Successful Treatment with anticoagulant therapy and Immunosuppressive Agents

**Introduction:**
Behçet's syndrome is a chronic multisystem disease that presents with recurrent oral and genital ulceration and recurrent uveitis. Cardiac involvement is an extremely rare manifestation of this disorder (less than 50 cases had been reported).

**Case presentation:**
A 22-year-old Tunisian man with a two-month history of fever of unknown origin, weight loss and inspiratory thoracic pain was admitted. He had suffered from both genital and oral ulcers over five months. The initial physical examination revealed a temperature of 38°C, multiple pseudofolliculitis, oral and scrotal ulcerations. Laboratory tests on admission revealed: haemoglobin of 14 g/dl, erythrocyte sedimentation of 105 mm/hr and C reactive protein concentration of 134 mg/l. The tests looking for antiphospholipid antibodies, protein C, protein S and antithrombine III deficiencies were negative. HLAB5 and pathergy tests were positive. Electrocardiogram showed sinus rhythm tachycardia, and chest X-ray was normal. Transoesophageal echocardiography revealed a cardiac mass in the right atrium of 20/23 mm size attached into atrial septum protruding through the tricuspid valve into the right ventricle. These findings were confirmed by CT scan that also showed a partial obstruction of the terminal portion of the inferior vena cava and thrombosis of the left lobar pulmonary artery with multiple pulmonary infarcts. We started treatment with intravenous heparin then oral anticoagulant, 1 gr of methyl prednisolone per day for 3 days, 1 mg/kg/day of oral, which was tapered gradually and 1 g pulse of cyclophosphamide monthly associated with colchicine 1 mg/day. Consequently, the thrombus in the right atrium has substantially decreased in size. At four months follow-up, a complete resolution of the thrombi in the right atrium, vena cava and pulmonary artery tree was observed.
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Discussion

Epidemiology. Thrombosis is the most frequent lesion. Usually, arterial thrombosis occurs later than does venous thrombosis. Thrombosis is very often associated with (and predictive of) ocular involvement in Behçet syndrome.

Despite the high incidence of vascular damage in this disease (more than one third of patients experience it), cardiac involvement is uncommon. It varies from 1% to 5% in clinical series and was 16.5% in a register of Behçet autopsy cases in Japan. The appearance of such thrombi often precedes other manifestations of Behçet syndrome, and it is rarely inaugural.

Diagnosis. Transesophageal and transthoracic echocardiography are generally sufficient to enable a diagnosis of cardiac thrombosis. The mass is usually heterogeneous and echogenic, involving the ventricles rather than the atria and the right heart more often than the left. In any event, the resolution of the mass under anticoagulant or antiplatelet therapy establishes a retrospective diagnosis, because it distinguishes thrombus from tumor.

Therapy. There is no consensus regarding the treatment of cardiac thrombosis in association with Behçet syndrome. The aim of treatment is to control the underlying disease and to resolve the cardiac thrombus. Anticoagulant and antithrombotic agents are the 1st line of therapy. However, an associated aneurysm of the pulmonary artery—especially the presence of large or bilateral aneurysms—makes the use of these therapies difficult because of the risk of a devastating hemoptysis.

Surgery might become necessary in cases of cardiac thrombosis that is massive, is recurrent after medical treatment, or is associated with cardiac congestion.

In our case, we find that Behçet syndrome was controlled by immunosuppressive drugs, corticosteroids and by means of 3 to 6 months of anticoagulant treatment with vitamin K antagonists (target INR range, 2–3).

Conclusion.

Intracardiac thrombus formation should be considered when a young Behçet's disease patient presents with an intracardiac mass. Medical treatment including corticosteroids, immunosuppressive drugs and anticoagulants should be considered as the first line treatment and surgery should be considered when there is no resolution of the thrombus or when it becomes massive and extensive. In certain cases, thrombolytic treatment becomes an interesting alternative to surgery.