

Right heart thrombus dissolved after medical management in a patient with Behçet's Disease

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ÖZET

Behçet hastalığı, etyolojisi bilinmeyen, kronik, rekürren, multisistemik bir hastalıktır. Hastalığın bulguları; rekürren aftöz stomatit, genital ülserasyonlar, diğer mukokutanöz lezyonlar, oküler, artiküler, nörolojik, ürogenital, kardiyovasküler, gastrointestinal ve pulmoner bulgulardır. Vasküler komplikasyonlar hastalığın yaygın görülen bir bulgusudur. Fakat kardiyak tutulum çok nadir bir bulgu olup, kötü prognoz işaretidir. 48 yaşında erkek hasta nefes darlığı, atipik göğüs ağrısı ve hemoptizi şikayeti ile başvurdu. Özgeçmişinde Behçet hastalığı dışında özellik yoktu. Yapılan ileri tetkiklerde sağ atrial trombus ve pulmoner tromboemboli saptandı. Hastaya antikoagulan ve immünsupresif ilaçlar içerikli medikal tedavi başlandı. 3 aylık takipte, trombusun tamamıyla gerilediği ve klinik durumun dramatik olarak iyileştiği saptandı.

Anahtar Kelimeler: Sağ Kalp Trombusü; Behçet Hastalığı; Medikal Tedavi

SUMMARY

Behçet's disease is a chronic relapsing multisystem disease of unknown etiology. The manifestations of the disease are: recurrent aphthous stomatitis, genital ulcerations, other mucocutaneous lesions, ocular, articular, neurological, urogenital, cardiovascular, gastrointestinal and pulmonary manifestations. Vascular complications are a common component of this disease but cardiac involvement is a very rare and the sign of poor prognosis. A 48-year-old male patient presented with dyspnea, atypical chest pain, and haemoptysis. In his past history; he was unremarkable except for Behçet's disease. Further investigations revealed a free right atrial thrombus and pulmonary thromboembolism. We preferred medical management which consisted of immunosuppressive drugs and anticoagulation. On a follow-up period of three months we observed complete dissolution of the thrombus and dramatic improvement of clinical status.

Key words: Right Heart Thrombus; Behçet's Disease; Medical Management

Introduction

Behçet's disease is a chronic relapsing multisystem disease of unknown etiology. The manifestations of the disease are: recurrent aphthous stomatitis, genital ulcerations, ocular disease, as well as mucocutaneous, articular, neurological, urogenital, cardiovascular, gastrointestinal and pulmonary manifestations (1). Cardiac involvement is a very rare and the sign of poor prognosis. The management of this complication is difficult because of the risk of thrombosis recurrence even after surgical resection of the thrombus (2). We are reporting a case of a 48-year-old male patient with Behçet's disease who presented with right heart thrombus dissolved after medical management.

Case Report

A 48-year-old male patient presented with dyspnea, pleuritic chest pain, and haemoptysis. Physical examination was unremarkable except for oral ulceration. The patient had been suffering from oral and genital ulceration for almost three years. With a positive pathergy test and HLA B5 genotype the diagnosis of Behçet's disease was established. A complete blood count revealed leukocytosis, and the erythrocyte sedimentation rate was 38 mm/h. D-dimer levels are elevated in plasma. Arterial blood gas analysis revealed mild hypoxia and hypocarbia. ECG showed sinus tachycardia. Although chest X-ray was normal, tomographic examination of the chest showed infiltrates at the anterior segment of the right upper lobe, anterior segments of the left lower lobe. Ventilation-perfusion scintigraphy (V/Q scan) revealed multiple perfusion defects. His abdominal and deep-vein Doppler ultrasounds were normal. Transthoracic echocardiographic (TTE) examination revealed a right atrial thrombus. The function of the left and right ventricles were normal. Two and three-dimensional transoesophageal echocardiography (2D and 3D TEE) were performed for visualizing the thrombus and evaluating its relationship with cardiac structures better. 2D and 3D TEE revealed a pedunculated, large and highly mobile thrombus which was attaching to tricuspid valve (Figure 1A and B, arrow). It occasionally protruded into the tricuspid valve orifice. Screening tests for hypercoagulability included normal levels of protein C, protein S, homocystine and positive antiphospholipid antibodies. Genetic analysis for prothrombin gene mutation and Factor V Leiden were negative. The patient was started oral prednisone (1 mg/kg per day) and colchicine (50 mg) three times daily. An immunosuppressive therapy with intravenous cyclophosphamide (1 gram monthly) was added to the treatment, yielding immediate improvement in clinical and laboratory parameters. Furthermore, warfarin was administered as an anticoagulant. He did well on follow-up, and a TTE examination at three months showed that the thrombus had been completely disappeared, and the patient was free of symptoms. To better definition, we performed TEE and we confirmed this situation.

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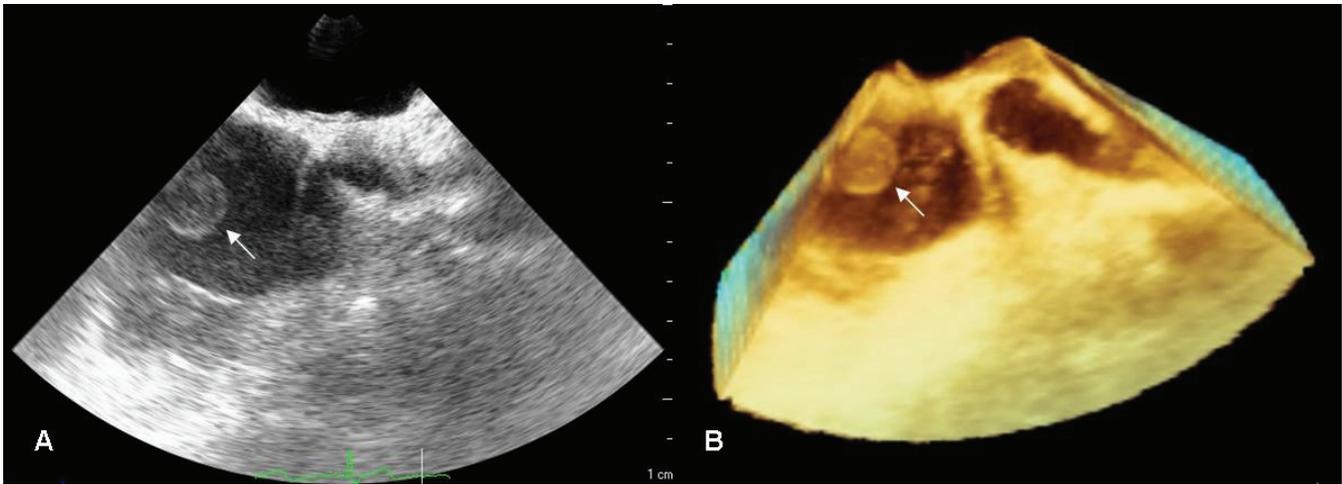


Figure 1: Two-dimensional (A) and three-dimensional (B) transoesophageal echocardiography revealed a pedunculated, large and highly mobile thrombus which was attaching to tricuspid valve.

Discussion

The prevalence of cardiovascular involvement in Behçet's disease varies from 7% to 46%, according to the different ethnic group. Venous lesions are the most common abnormality and usually consist of recurrent deep vein thromboses, most often involving the lower extremities. However cardiac involvement is a very rare and the sign of poor prognosis. Most frequently observed cardiac manifestations were dilated cardiomyopathy, coronary artery disease, mural thrombus in right ventricle and pericardial effusion, myocarditis and arrhythmia (1-5). Hypercoagulability is associated with this disorder and the presence of anti phospholipid antibodies is an independent predictive factor for increased risk of vascular thrombosis in individuals with Behçet's disease (4). Also in our patient, anti phospholipid antibodies were positive.

Regarding the treatment of cardiac thrombosis associated with Behçet's disease, there is no consensus. The aim is to resolve the cardiac thrombus and to prevent the recurrence of this complication. Anticoagulants alone do not seem to be effective; systemic glucocorticoid and immunosuppressive agents are highly recommended before using anticoagulants. Surgery might be limited to the cases with massive thrombosis, recurrence of the complication despite an optimal medical treatment, and when there is a cardiac congestion (2,6).

In our patient, surgical treatment was considered because the right atrial thrombus was quite large and it protruded into the tricuspid valve orifice. However, it did not cause haemodynamic disturbances and congestive heart failure. Treatment with heparin or warfarin in addition to corticosteroids and immunosuppressive drugs (azathioprine or oral cyclophosphamide) is associated with disappearance of intracardiac thrombus in most patients (6). Considering the excessive risk of recurrence after surgery, he was managed medically with immunosuppressives and anticoagulants. The result was extremely satisfactory, with gradual resolution of the thrombus and improvement of symptoms. Patients who are haemodynamically stable and who do not show signs of congestive heart failure can usually be managed with a conservative approach. Our patient had a large and free right atrial thrombus which protruded into the tricuspid valve orifice; nevertheless, he had no haemodynamic compromise or congestive heart failure. Considering the risks of surgical treatment and possibility of recurrence, we preferred a conservative approach which proved to be successful.

Transesophageal and transthoracic echocardiography are generally sufficient to reveal this complication. In particular, transoesophageal echocardiography can be necessary in or-

der to view the ventricle or right atrium clearly (7). However, it should be emphasized that lack of contrast enhancement does not necessarily distinguish between thrombus and some forms of poorly vascularized tumors. In particular, some benign tumors, such as myxomas, lipomas, and fibromas, have sparse vascularity with subsequent partial contrast enhancement making the distinction between these tumors and avascular thrombus difficult. In these cases the differentiation can be aided by combining echocardiographic appearance and location of the mass together with clinical information (8).

We suggest that Behçet's disease be kept in mind in the differential diagnosis of intracardiac thrombus. We also suggest the combination of methylprednisolone, cyclophosphamide, and warfarin as a good option to treat cardiac thrombi in patients who have Behçet's disease.

References

1. El Menyawi MM, Raslan HM, Edrees A. Clinical features of Behçet's disease in Egypt. *Rheumatol Int.* 2009;29(6):641-6.
2. Hammami R, Abid L, Frikha F, Marzouk S, Tounsi A, Frikha Z, Hentati M, Bahloul Z, Kammoun S. Intracardiac thrombus in a young man: don't forget Behçet's disease! *Intern Med.* 2012;51(14):1865-7.
3. Cemri M. Behçet's Disease with a Large and Free Right Atrial Thrombus. *Eur J Echocardiogr.* 2002;3(3):233-5.
4. Dincer I, Dandachi R, Atmaca Y, Erol C, Caglar N, Oral D. A recurrent right heart thrombus in a patient with Behçet's disease. *Echocardiography.* 2001;18(1):15-8.
5. Kirali K, Civelek A, Dağlar B, Şişmanoğlu M, Akinci E, Berki T, et al. An uncommon complication of Behçet's disease: intracardiac thrombosis needing surgical treatment. *Thorac Cardiovasc Surg.* 1998;46(2):102-5.
6. Kaneko Y, Tanaka K, Yoshizawa A, Yasuoka H, Suwa A, Satoh T, et al. Successful treatment of recurrent intracardiac thrombus in Behçet's disease with immunosuppressive therapy. *Clin and Exp Rheumatol.* 2005;23(6):885-7.
7. Baykan M, Celik S, Erdöl C, Baykan EC, Durmus I, Bahadır S, et al. Behçet's disease with a large intracardiac thrombus: a case report. *Heart.* 2001;85(4):E7.
8. Rallidis LS, Papadopoulos CC, Kelekis NL, Kremastinos DT. The role of myocardial contrast echocardiography to assess the origin of a mass in right cardiac cavities. *Echocardiography.* 2009;26(1):88-92.