

Case report

Right ventricular thrombus and tricuspid valve dysfunction in a patient with Behçet's syndrome

C. Arslan¹, B. Arapi¹, E. Seyahi², C. Tel¹, K.H. Tüzün¹

¹Department of Cardiovascular Surgery, and
²Department of Rheumatology, Cerrahpasa
Medical Faculty, Istanbul University,
Istanbul, Turkey.

Caner Arslan, MD
Berk Arapi, MD
Emire Seyahi, MD
Cigdem Tel, MD
Kamil Hasan Tüzün, MD

Please address correspondence to:

Caner Arslan, MD,
Cerrahpasa Medical Faculty,
Department of Cardiovascular Surgery,
Fatih, Istanbul,
38400 Turkey.

E-mail: canerkvc@yahoo.com

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ABSTRACT

Behçet's syndrome (BS) is a systemic inflammatory disease generally presented with triad of uveitis, oral and genital ulcers. However, it may present with gastrointestinal, central nervous system, skin, vascular disease manifestations. Cardiac involvement like intracardiac thrombus and valvular involvement in BS are rarely seen entities. Here we present the management of a 23-year old male BS patient who had a right ventricular thrombus and tricuspid valve dysfunction which was resistant to immunosuppressive treatment. He has been doing well for 4 years after intraventricular thrombus resection and tricuspid valve replacement with bioprosthesis.

Introduction

Behçet's syndrome (BS) is a multi-systemic disorder that can generally present with orogenital ulcers, uveitis and skin lesions and rarely arthritis, gastrointestinal, central nervous, and cardiovascular system manifestations, including arterial aneurysms, venous thrombosis and intracardiac lesions (1). Endomyocardial fibrosis and intracavitary thrombus are the main reasons for the rarely seen intracardiac masses in patients with BS (2, 3). Here we present the successful treatment of a right ventricular mass and tricuspid valve (TV) dysfunction in a young male patient with BS suffering from dyspnea.

Case report

The patient, a 23-year-old male, was diagnosed with BS at the age of 16 because of recurrent oral ulcers, genital ulcers and skin lesions. He was HLA-B51 (+) and was positive for pathergy test. Two years before his current admission, he was hospitalised in a university hospital because of fever,

weight loss, chest symptoms, and high acute phase response. He was found to have right ventricular thrombus and pulmonary artery aneurysm (PAA) in the right descending branch. He was started monthly cyclophosphamide pulses (750 mg/m²) along with prednisolone (1 mg/kg for the first 1 month then tapered). After 4 months of this treatment, fever, acute phase response and chest symptoms except dyspnea had subsided and PAA aneurysm had disappeared. However, the right ventricular mass remained unchanged. He was switched to a combination of oral 100 mg cyclophosphamide with 10 mg prednisolone daily, which he received for 15 months. Because he continued to have severe dyspnea and right ventricular mass despite this treatment, he was eventually referred to our institution.

At the current admission, his physical examination was normal except a 3/6 systolic murmur and a 2–3/6 diastolic tricuspid murmur on the lower left side of the sternum. His C-reactive protein value was slightly higher than normal 5.2 mg/dl (n=0–5 mg/dl). Erythrocyte sedimentation rate was also higher than normal (60 mm/hr). Leukocyte count was high, 28.6 x 10³ /mm³ (n=4.1–10.3). ALT, LDH and haptoglobin levels were also higher than normal (98 U/L; n=0–45, 403 U/L; n=125–243, 337 mg/dl; n=34–200, respectively). FANA and ANCA tests were negative. Right bundle branch block was present on the electrocardiogram. In the transthoracic echocardiography (TTE) performed to define cardiac pathology, it was demonstrated that TV was stenotic having maximum gradient of 8 mmHg, mean 5 mmHg, the valve area was near 1.4 cm² and valve leaflets had calcifications on some parts. Systolic pulmonary artery pressure was estimated as 40 mmHg. Two separate masses were revealed;

Competing interests: none declared.

one originating from the lateral right ventricular (RV) wall, mobile 1.0x0.8 cm in dimension, the other one filled the middle part of the ventricular septum on the right side, measuring 2.3x1.8 cm in . There was also moderate tricuspid regurgitation. Transesophageal echocardiography (TEE) was carried out to verify these findings.

Dynamic contrast enhanced magnetic resonance imaging revealed that right atrium was severely dilated (7.8x6.3 cm) and inside the right ventricle a mobile thrombus (1.1x0.7 cm) originating from the right ventricle free wall extending to the TV preventing its closure during systole. Also, there was another thrombotic material immobile, 2.4x 1.2 cm in size, occupying the mid to apical regions of the RV (Fig. 1).

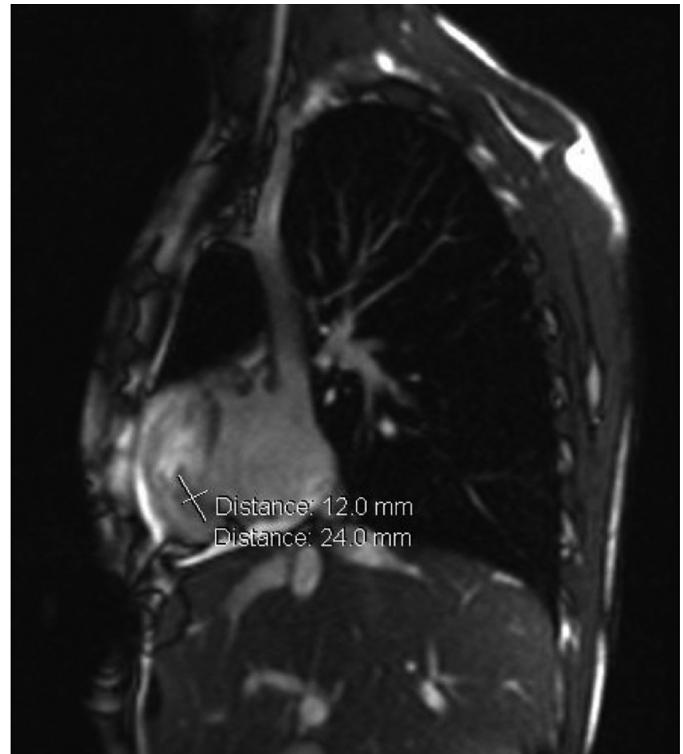
Also, ultrasonographic examinations revealed no thrombosis in portal veins, vena cavae and lower extremity veins.

He was referred to our department and underwent surgery with cardiopulmonary bypass via aortic and bicaval cannulation. The heart was arrested with antegrade cardioplegia. When the right atrium was opened, it was seen that there was a small mobile thrombus originating from the free right ventricular wall extending to the TV. Septal and posterior leaflets of the tricuspid valve were thicker than normal and fused. Below them, there was a mass, 3 cm in diameter, originating from the right ventricular side of the septum. This mass was also strongly fused to the tricuspid leaflets, so it was not possible to excise the mass without injuring the leaflets. That is why the mass and tricuspid valve were excised together and replacement of the tricuspid valve with 27 no bioprosthesis was carried out. Pathological investigation showed that the surgical material was an organised thrombus extending from the endocardium to myocardium surrounded by thick fibrotic tissue. The lesion was also shown to have mixed type T and B lymphocyte infiltrates.

Just before the operation and 2 weeks after that, the prednisolone dose was increased to 40 mg/day.

Post-op, he was followed with warfarin, azathioprine 2.5 mg/kg/day and prednisolone 10 mg/day. After the operation,

Fig. 1. Magnetic resonance imaging, 2.4x1.2 cm in dimension occupying the mid to apical regions of the RV.



he stopped having dyspnea and his pulmonary artery systolic pressure returned to normal levels. He is currently taking warfarin, prednisolone 2.5 mg / day and azathioprine 150 mg/day and has been well for nearly 4 years after the operation. He has no fever, chest symptoms or exertional dyspnea. Unfortunately, he has had hip pain for the last 6 months, due to the avascular necrosis of the hips, which most probably developed because of the high corticosteroid treatment.

Discussion

Involvement of the heart in BS is rarely seen as reported in a few case series (2-6) and several case reports (7-11). A broad range of pathology in the heart can be seen such as ventricular dysfunction, conduction system abnormalities, valvular insufficiency, myocardial infarction, pericarditis, endomyocardial fibrosis that may result from myocarditis or endocarditis. These pathologies are often associated with intracavitary thrombus. Intracardiac thromboses (ICT) generally involve the right side of the heart, probably due to the extension of the venous involvement. Similar to that seen with all vascular events in BS (12), it may cause fever, weight loss

and high acute phase response. Close association between ICT and deep vein thrombosis, vena cava thrombosis or pulmonary artery aneurysms was also noted in the related reports. So far, none of the thrombophilic factors were shown to be associated with the thrombotic tendency observed in BS (13). Detailed work-up is needed in order to differentiate ICT from cardiac tumours like myxoma covered by thrombus and also from right sided endocarditis with large tricuspid vegetations especially in patients with constitutional symptoms and fever.

A review of 25 BS patients with intracardiac thrombus revealed that:

- i. most of these patients were young males;
- ii. right heart chambers are predominantly affected;
- iii. there is strong association with venous system thrombosis involving vena cava, cranial sinuses or hepatic veins and with PAA (2).

Another important observation in this study was that the tricuspid valve could be thickened and replaced by fibrosis and could lose its functional integrity. The review also revealed that, the thrombus may contain inflammatory cell foci composed of granulocytes and/

or mononuclear cells or lymphocytes and plasma cells (2). Embolisation from these thrombi is uncommon because of their tight attachments to the endocardium or myocardium. Pulmonary lesions in these patients are supposed to be *in situ* lesions rather than due to embolisations (6).

Since there are a limited number of ICT cases in BS, there is no consensus on the antiaggregant, anticoagulant, thrombolytic, immunosuppressive or surgical treatments. It is well known that, intraventricular thrombus can recur after surgical excision despite additional anticoagulant treatment (2-3, 5). It was seen in some BS cases with ICT, only immunosuppressive treatment with or without anticoagulation was enough for thrombus resolution (2, 5, 10-11). In a current survey of 47 BS patients with pulmonary artery involvement done in our institution, ICT was observed in one third of the patients (6). Echocardiographic studies done after a median 3 years of follow-up demonstrated that ICT either had disappeared or became fibrotic small lesions with only immunosuppressive treatment (6).

Surgery may have some advantages over medical treatment. It is feasible when the diagnosis is unclear and when there is a suspicion of endocarditis or intracardiac tumour or when patients have valvular insufficiency or stenosis that

is unable to gain normal function with medical treatment.

In conclusion, similar to what was previously reported, our patient was a young male with BS. The initial and persistent symptom which led to the investigation was fever. He was found to have PAA together with thrombus in the right ventricle. Despite being treated aggressively with cyclophosphamide and corticosteroids for almost 2 years, intracardiac thrombus persisted and transformed into a dense fibrotic tissue extending to the tricuspid valve causing its insufficiency. The patient, meanwhile, continued to have severe exertional dyspnea. That is why we preferred surgical treatment. The surgical excision of the thrombotic mass and the replacement of the cardiac valve in this patient were successful in that they prevented a possible right heart failure progression. The surgery also eliminated the suspicion of tumoural growth or endocarditis. We suggest that ICT in BS should be treated with immunosuppressive treatment, and that surgery should be reserved only for complicated cases.

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