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Tricuspid Aseptic Endocarditis Revealing Right Endomyocardial Fibrosis During an Unrecognized Behçet's Disease: Two Cases Report

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BENHALLA ET AL: Tricuspid Aseptic Endocarditis Revealing Right Endomyocardial Fibrosis During an Unrecognized Behçet's Disease: Two Cases Report. We report two cases of endomyocardial fibrosis, revealed by verrucous tricuspid valvulitis extending to the right ventricular endomyocardium and complicated by a right heart failure, initially misdiagnosed and treated as infective endocarditis, during an unrecognized Behçet's disease. (J HK Coll Cardiol 2015;23:75-78)

Behçet's disease, Cardiac MRI in Behçet's disease, Endomyocardial fibrosis

Introduction

Endomyocardial fibrosis is rare in Behçet's disease (BD), we report 2 cases suffering from BD complicated by ventricular pseudo-tumor formation shown in the echocardiography. This deceptive appearance evoked the initial diagnosis of infective endocarditis with thrombosis.

Case 1

Patient of 20-year-old with no particular medical history, admitted for an initial diagnosis of infective endocarditis in the right heart. Physical examination disclosed murmur of tricuspid regurgitation, signs of right sided cardiac failure, and bilateral papillary oedema. Brain resonance magnetic imaging showed superior sagittal and right lateral sinus thrombosis. Electrocardiography showed right atrial hypertrophy and incomplete right bundle branch block.

Echocardiography disclosed an enlarged right atrium, severe narrowing of the inflow tract and the middle part of the right ventricle. Bright echoes were seen within the inferior and the middle parts of the right ventricular endocardium (Figure 1).

Nine blood cultures were negatives contrasting with disturbed inflammatory analysis and a fever, initially the patient was treated by intravenous antibiotics without any improvement, and it is with the onset of genital and oral ulceration, pseudo folliculitis in her skin that BD was established as a final diagnosis after an extensive work-up to exclude de infective endocarditis.

Cardiac magnetic imaging showed a mass of intermediate signal intensity on T1 weighted images with right ventricular dilation complicated by a cavitary...
thrombosis, late gadolinium enhancement was observed, with endocardial fibrous tissue present only in the subendocardium, appearing as a continuous area, commonly extending from the subvalvar region to the apex of the ventricle (Figure 2).

The endomyocardial biopsy was not possible because of the cardiac thrombi the patient was treated with cyclophosphamide boluses, prednisone (10 mg/d), colchicine (1 mg/d), and anticoagulants with a good evolution and partial regression of the masses in the right ventricle after 7 weeks.

**Case 2**

A young patient of 19-year-old, admitted in the cardiac emergencies for right heart failure with a fever lasting for three months, treated initially as an infective right endocarditis with 6 negatives blood cultures, without any improvement.

The patient reported having recurrent oral and genital aphthae, a posterior uveitis 2 months ago without healing; erythema nodosum in the abdomen and limbs. We completed by a pathergy test that was positive.

The echocardiography showed a dilated right ventricle with pseudotumoral formations lining the entire wall of the right ventricle and the septum inter atrial unlike the first case, with images of thrombi in the right ventricle, tricuspid valve was not affected by this process neither the others valves (Figure 3). Cardiac magnetic imaging for this patient was not available nearby.

The diagnosis of endomyocardial fibrosis in BD was established and a first dose of cyclophosphamide

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**Figure 1.** Parasternal small-axis showing the exuberant formations lining the entire wall of the right ventricle.

**Figure 2.** Cardiac Cine MRI, long-axis cine sequence, late gadolinium enhancement viewing the subendocardial thickening appearing as a continuous area in the right ventricle associated with the presence of several thrombi.
combined with oral corticosteroid therapy were given to the patient, with a clinical improvement within 15 days.

**Discussion**

BD is an inflammatory vasculitis, characterized by its frequency, in general with benign mucocutaneous, articular manifestations, but sometimes the severity of ocular, neurological, cardiac and vascular complications remains crucial.¹

This disease mainly affects men (twice the woman) between 20 and 40 years. It is common in the Far East and the Mediterranean. The diagnosis is clinical and based on international criteria.² It is a disease that progresses in spurts sometimes spontaneously regressive and which treatment is largely symptomatic, of the fact many unknowns about its etiology,¹ but, as described in pathophysiology, the interplay between infectious-agent exposure and genetic factors may have a role. An environmentally triggered hyperactive primed state of autoimmunity ensues, resulting in two types of vascular damage. The first is vasculitic lesions that may be widespread. Sequelae depend on the various organ systems affected.

Some of the pathologic changes are due to thrombosis and / or clot formation caused by the development of a hypercoagulable state. The mechanism is still undetermined; however, studies have demonstrated excessive thrombin formation and the potential role of impaired fibrinolytic kinetics in the generation of the hypercoagulable/prothrombotic state. Pathologic activation of the procoagulant cascade via endothelial injury has also been demonstrated in patients with Behçet disease.³

The frequency of cardiac involvement varies from less than 1% to 6% in clinical series and 16.5% in an autopsy series.³ The three cardiac layers may be affected with pericarditis, myocardial injury, valvular and coronary tissue conduction. Intracardiac thrombosis is very rare, endomyocardial fibrosis still a part of the differential diagnosis of restrictive heart disease, and which can be presented as an intracardiac tumor also.⁴

Transthoracic echocardiography is the first-line examination and allows accurate systolic and diastolic functional assessment. It is however limited for tissue characterization and differential diagnosis of restrictive heart disease. Cardiac magnetic imaging has a key role in the diagnosis and prognosis of this disease, although few data have been reported in the literature.⁵⁻⁶ It allows precise morphological evaluation of the endomyocardial fibrosis most often characterized by a diffuse thickening under endocardial right ventricle with the presence of several associated thrombus. Auricular areas are often increased in size due to severe diastolic dysfunction with restrictive disorder.²

The sequences of delayed enhancement affirm the diagnosis by showing a typical late enhancement, limited to subendocardium, and extended from the valve to the apex regions under the two ventricles where it usually dominates. Key element, a raise is not distributed in a vascular territory and is not accompanied by myocardial thinning in most cases. The presence of a thrombus is common at the apex of the LV and / or RV and. The prognostic role of MRI has also been recently suggested as the treatment of choice is the surgical resection of the endomyocardial fibrosis areas, in patients with Stage 3 or 4 NYHA.⁷⁻⁸ MRI may help in the future planning of surgery and monitor its effectiveness. For our 2 patients, surgical treatment was deferred with the improvement under medical treatment with corticosteroids and anticoagulants.

**Figure 3.** Four chamber bidimensional end diastolic echocardiogram: bright echoes in the right ventricular endocardium.
Conclusion

The discovery of endocardial masses in a patient suspected having an infective endocarditis, negative blood cultures with criteria of Behçet diseases, should arouse suspicion of the diagnosis of endomyocardial fibrosis. The cardiac MRI allows precise characterization of particular tissue fibrosis with the delayed enhancement sequences, and could help prognostic stratification and planning for therapeutic intervention in those patients.

Disclosures

The authors declare that there is no conflict of interest.

References