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Case Report

Intra-Left Ventricule Thrombus in Behcet's Disease About 3 Cases

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Abstract: ***Introduction:*** Behçet's disease (BD) is a systemic vasculitis with a triad of oral and genital aphthosis and hypopyon uveitis, potentially accompanied by various systemic manifestations. While venous thrombosis is common, cardiac involvement, including intracardiac thrombi (ICT), is rare. The occurrence of left ventricular thrombi (LVT) in BD presents significant diagnostic and therapeutic challenges. ***Observations and Patients:*** We present three cases of BD with LV thrombus diagnosed at the Cardiology Department of University Hospital Ibn Rochd, Casablanca. The patients, aged 28 to 54 years, included two men and one woman. All had a history of BD symptoms before cardiac issues were identified. Clinical examinations revealed oral and genital aphthae, skin lesions, and positive Pathergy tests in one patient. Cardiac imaging confirmed left ventricular thrombi in all patients, with varying degrees of left ventricular ejection fraction (LVEF) impairment. Treatment included corticosteroids, immunosuppressants, anticoagulants, and heart failure management. Two patients showed improvement, while one patient with severe multi-organ involvement unfortunately died. ***Discussion:*** Cardiac involvement in BD is rare, and ICTs are exceptional. These thrombi, typically adherent to the wall and less emboligenic are discovered primarily through echocardiography. The treatment is not standardized but generally involves anticoagulants, corticosteroids, and immunosuppressants. Early detection and treatment are crucial, as the prognosis is significantly influenced by cardiovascular involvement. ***Conclusion:*** ICT in BD, while rare, is a serious complication that requires careful diagnostic and therapeutic management. Echocardiography should be considered in the evaluation of BD, even in the absence of cardiac symptoms, due to the potential severity of cardiovascular involvement.

Keywords: Behçet's Disease, Left Ventricular Thrombus, Anticoagulants, Corticosteroids, Immunosuppressants.

INTRODUCTION

Behçet's disease (BD) is a systemic vasculitis, the etiology of which remains unclear. It consists of a symptomatic triad described by Behçet in 1937 [1] associating buccogenital aphthosis and hypopyon uveitis, which may be associated with various systemic manifestations: cutaneous, vascular and articular, with the possibility of neurological manifestations.

Vascular involvement includes venous thrombosis, which is common in Behçet's disease, while arterial thrombosis and/or aneurysms are rarer [2].

Cardiac involvement is rare in Behçet's disease, ranging from 1% to 6%. Intracardiac thrombi (ICT) are an exceptional complication [3], especially left ventricular thrombi.

Their occurrence during the course of this disease poses a diagnostic and therapeutic problem, and worsens the prognosis. In half of all cases, ICTs are discovered prior to the diagnosis of the disease, and are exceptionally revelatory.

OBSERVATIONS AND PATIENTS

We report a series of 3 cases of MB with left ventricular thrombus diagnosed at the Cardiology Department of University Hospital Ibn Rochd, Casablanca.

In our series, age ranged from 28 to 54 years, with 2 men and 1 woman. As for Cardiovascular risk factor, only one patient was diabetic.

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The age of diagnosis of MB in our patients fell within each of the age ranges [>45 years], [35-45 years] and [25-34 years] for each of them, with an age of onset of symptoms of less than 25 years for all our patients.

On examination, one of our patients reported long-term fever, weight loss and NYHA stage IV dyspnoea, while the other patients were asymptomatic.

Clinical examination revealed oral aphthae in all patients, one case of aphthous ulcer of the upper lip, two cases of scrotal aphthous ulcer, and one case of genital aphthous ulcer of the upper lip. We also observed pseudofolliculitis lesions on the thighs and one case of erythema nodosum. The Pathergy test was positive in 1 patient. In cardiopulmonary terms, one patient presented with a left-sided cardiac decompensation. The rest of the examination in all our patients was unremarkable.

Ophthalmological findings included papilledema, uveitis, retian vasculitis and maculopathy.La radiographie thoracique de face montrait une cardiomégalie avec un indice cardio-vasculaire augmenté chez 2 malades de la série.

The electrocardiogram showed sinus rhythm in 2 patients and one case of atrial fibrillation, with no conduction disturbance, anterior necrotic Q waves in one patient with previous MI and inferior negative T waves in another.

Echocardiography showed intra-left ventricular thrombi and impaired LVEF in all our patients [Figure 1], with extremes ranging from 25% to 30%. Among the thrombi, 2 were adherent to the cardiac apex, one of which was associated with a cardiac apex aneurysm and a very thinned wall, while one was intracavitary and multiple. The LV was dilated in 2 patients, with elevated Left Ventricular Wedge Pressure in 1. LV systolic function was normal in all our patients, with absence of Pulmonary Hypertension.

Doppler ultrasonography of the lower limbs revealed Deep Vein Thrombosis in only one patient with thrombophlebitis of the left lower limb extending to the primitive iliac vein, whereas it was normal in the others.

Vascular signs such as splenic infarction and sequelae of Ischemic Stroke were found in one patient.

Biologically, only one patient showed an inflammatory syndrome, with an accelerated sedimentation rate of 60mm in the first hour. The rest of the patients' work-ups were normal.

The immunological work-up was also normal, including protein C, protein S and antithrombin III levels, antiphospholipid antibodies, anti-CCP, ANCA, AAN, rheumatoid factor, cryoglobulin and HLA B51. Hepatitis, HIV and syphilis viral serologies were negative.

The diagnosis of MB in its severe form was based on the association of clinical criteria: oral aphthosis and cutaneous signs, as well as the presence of cardiac involvement in the form of intracardiac thrombi.

Our MB patients were treated with bolus corticosteroids (methylprednisolone 15mg/kg for 3 days, followed by prednisone 1mg/kg/d per os), immunosuppression and colchicine.

Anti-coagulant treatment with VKA was initiated. For their heart disease, heart failure treatment was started with beta-blockers and ACE inhibitors. In the case of decompensating ischemic heart disease, the patient was also started on aspirin and statins, combined with diuretic therapy and potassium supplementation, in view of his clinical and echocardiographic signs of congestive heart failure.

The evolution was good for 2 patients, marked by the disappearance of fever, improvement in general condition and a reduction in episodes of mucocutaneous aphthae. Respiratory status improved markedly in the patient with left-sided cardiac decompensation, under well-managed treatment.

Echocardiography Transthoracic monitoring of these patients revealed the persistence of the thrombus despite its reduction in size. Unfortunately, we noted one case of death in a patient who presented a particular picture of dyspnea, NYHA stage IV, initially due to severe MB damage of the multivisceral type.

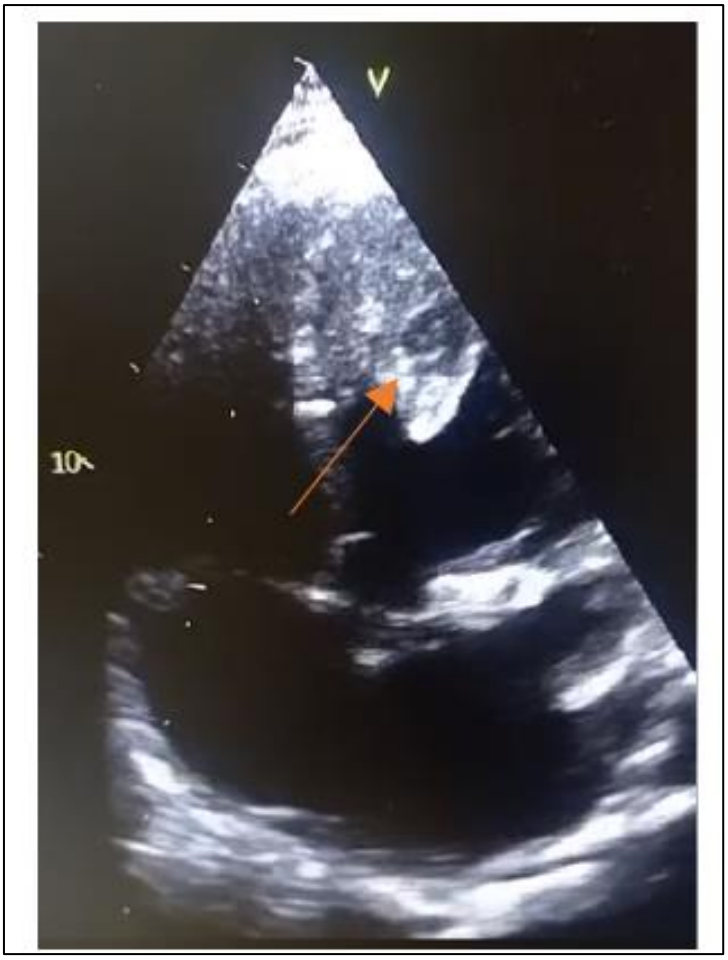

Figure 1: Images of intra-LV thrombus in BEHCET'S Disease

DISCUSSION

Behçet's disease (BD) is a systemic vasculitis of uncertain origin, characterized by the combination of oral and genital ulcers (aphthosis) along with hypopyon uveitis. It may also present systemic manifestations. Vascular involvement is common, predominantly affecting young male individuals, often in the absence of other risk factors [4]. This observation aligns with the findings from our cohort. Cardiac involvement resulting from BD is rare, reported in only 6% of patients, making it an uncommon complication of this condition.

In this instance, the intracardiac thrombus (ICT) was asymptomatic and was identified incidentally during an echocardiographic examination. The definitive diagnosis of ICT relies primarily on imaging techniques, with cardiac echocardiography being the most prominent modality. This imaging typically reveals an intracardiac mass that exhibits greater echogenicity than the surrounding blood, is heterogeneous in texture, and demonstrates adherence to the cardiac wall. Traditionally, intracardiac thrombi associated with Behçet's disease are characterized by their adherence to the wall, which accounts for their low propensity for embolism.

The relationship between intracardiac thrombus (ICT) and venous thrombosis is noted in 56% of instances [3]. One patient in our study exemplified this connection. The precise mechanism underlying ICT remains unclear, but it is likely associated with ischemia resulting from coronary artery disease and inflammatory damage to the endothelium, which may promote platelet aggregation.

Currently, there is no standardized treatment for ICT in the context of myocardial bridge (MB), although several researchers [5] have documented positive outcomes with medical management. This may involve corticosteroids, colchicine, vitamin K antagonists (VKAs) administered at therapeutic doses, potentially in combination with immunosuppressive therapy. Fibrinolytic therapy is also a viable option. Surgical intervention should be considered only in situations where medical management has proven ineffective or in cases of significant pulmonary embolism. Recurrences may respond favorably to either medical treatment or fibrinolysis.

CONCLUSION

The occurrence of ICT in myocarditis (MB) is uncommon, yet it represents a significant complication.

Echocardiography serves as the principal tool for diagnosis.

Traditional treatment approaches typically involve the administration of anticoagulants, corticosteroids, and immunosuppressive agents.

This series of observations aims to highlight the importance of incorporating echocardiography into the evaluation of MB patients, even in the absence of clinical manifestations, since the prognosis is closely linked to the degree of cardiovascular involvement.

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