

Case Report

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Myocardial Infarction and Ventricular Tachycardia in a Patient with Behçet's Disease

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Abstract

Behçet's disease is a chronic vasculitis with heterogeneous manifestation. Cardiovascular involvement, although rare, is described and of extreme severity. We report the observation of a 33-year-old man diagnosed with Behçet's disease complicated with a thrombosed aneurysm of the right coronary responsible of a myocardial infarction and an aneurysm of the lower LV wall admitted for management of a life threatening ventricular tachycardia with an indication of an implantable cardioverter-defibrillator (ICD) implantation.

Keywords: Behçet's disease; Arrhythmia; Myocardial infarction; Ventricular tachycardia

Introduction

Behçet's disease is a chronic, recurrent and multisystemic vasculitis characterized by muco-cutaneous lesions, articular, ocular and nervous manifestations. The cardiovascular involvement, although rare, is described and requires special attention given the heterogeneity of the presentations from pericarditis to rhythm disorders and the high risk of morbidity and mortality [1].

Case Report

A 33-year-old patient, previously diagnosed of Behçet's disease, presented to the ER with palpitation due to a sustained, poorly tolerated ventricular tachycardia.

The patient had a history of recurrent thrombophlebitis in 2001 and 2007 and has had Behçet's disease since 2007. The diagnosis was made based on the association of recurrent oral and genital aphthoses and the presence of pseudofolliculitis. Few months after the diagnosis, the patient presented with chest pain and he was treated for myocardial infarction based on electrocardiographic and enzymatic data. Coronary angiography revealed a thrombosed aneurysm of the right coronary second segment, measuring 3 centimeters in diameter (Figure 1). The rest of the coronary arteries were normal. An echocardiographic study demonstrated a reduced left ventricular systolic function, mainly related to inferior wall hypokinesis and a large aneurysm at the inferior wall of the left Ventricle. A coronary angiotomography done in 2009 had objectified a thrombosed aneurysm at the level of the right coronary and a small thrombus of the right ventricle's free wall. Control coronary angiography in 2010 showed a decrease in the diameter of the right coronary aneurysm to 2 centimeters.

From 2013 until October 19th 2016, the medical staff was unaware of the patient's vital status. On that day, he presented to the emergency room for palpitations associated with profuse sweating and lipothymia. Clinical examination showed a low Blood Pressure of 80/60 mmHg and a rapid pulse. Pulmonary auscultation was without abnormalities and there were no signs of heart failure. At the ECG, there was a wide shaped regular tachycardia with left axis deviation (Figure 2). The diagnosis of a life threatening ventricular tachycardia was made. Sinus rhythm was restored by external electric shock of 200 Joules. Post-cardioversion ECG showed a regular sinus rhythm with inferior Q waves of necrosis and T inversions in the same leads (Figure 3).

He was put on antiarrhythmic (amiodarone) and was monitored by scope. Transthoracic echocardiography showed a dilated spherical left ventricle with a diastolic diameter of 60 mm, an estimated ejection



Figure 1: Coronary angiography revealing a thrombosed aneurysm of the right coronary second segment, measuring 3 centimetres in diameter.

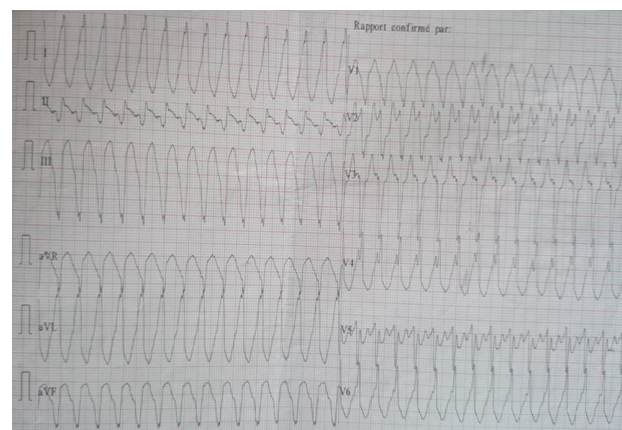


Figure 2: Left-bundle-branch-block-shaped-regular tachycardia with upper axis deviation.

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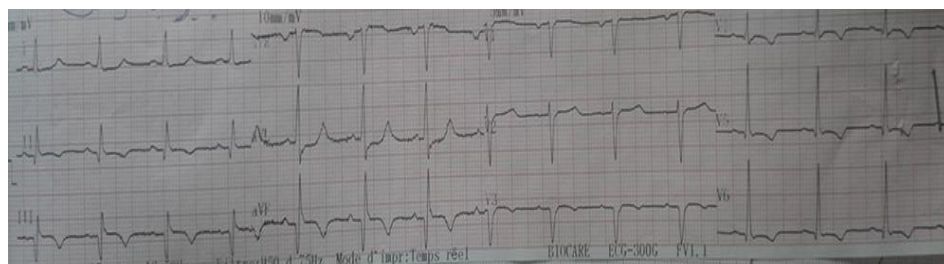


Figure 3: Post-cardioversion ECG showing a regular sinus rhythm with inferior Q waves of necrosis and some slight T inversions in the same leads.

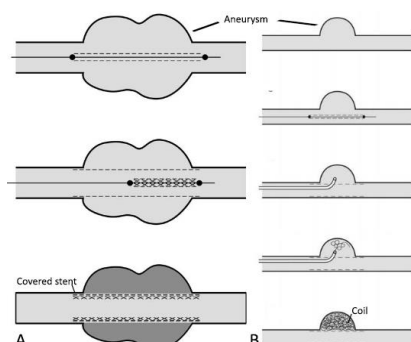


Figure 4: Coronary aneurysms' treatment using covered stents (A) or coils (B).

fraction of 40, a large and previously known inferolateral akinesia and a moderate mitral regurgitation without pulmonary hypertension. The patient was deemed candidate for an ICD implantation and transferred to a specialized center.

Discussion

Behçet's disease was described for the first time in 1937 as a triad associating repeated oral and genital aphthoses and uveitis [2]. Other manifestations can affect the cutaneous, ocular, respiratory, urogenital, articular system and less frequently but nonetheless with alarming severity, the neurological and cardiovascular systems [3].

Cardiac involvement in Behçet's disease may take the form of endocarditis, pericarditis, myocarditis, intracardiac thrombi or valvulopathy [4,5]. In the vascular aspect, the involvement is both venous and arterial, being the latter either aneurysmal or occlusive. Coronary arteries are rarely affected [6]. Apart from these manifestations, an increase in the incidence of arrhythmias has been described [7,8].

In 1958, Stucchi et al described for the first time a case of atrial fibrillation in a patient with Behçet's disease [9]. A few years later, the arrhythmias observed in this type of patients were associated with acute coronary syndromes (ACS) or myocarditis [3,10] in fact infiltration of the myocardium by granulocytes and histiocytes as is the case in chronic myocarditis has been described in Behçet's disease [11]. In 1976 a study found diastolic dysfunction in more than a third of a series of 24 patients followed for Behçet, none of them had signs pointing towards a cardiac pathology. The authors concluded that cardiomyopathy may exist in Behçet's disease, with no obvious clinical signs [12].

In our patient's case ventricular rhythm disorder was explained by the sequelae of MI. This coronary event was included in the symptoms of Behçet's syndrome for the following reasons: physical finding of atherosclerosis on examination were absent, our patient did not have

cardiovascular risk factors, the cardiac event occurred at a young age and the coronary angiography revealed only a thrombosed aneurysm in the right coronary artery. The other angiographic images were devoid of visible atherosclerosis.

There are two main different techniques to treat coronary artery aneurysms: The first involves the use of covered stents where their structure is encircled by a tissue that excludes the aneurysmal wall (Figures 4A and 4B) thus attenuating the repetitive impact of the blood pressure. The alternative technique relies simply on coil embolization of the aneurysm (Figure 4) [13]. No strategy is superior over the other.

The literature reports a little more than 25 cases of myocardial infarction in patients followed for Behçet's disease [6,14], they are all young, generally male, without significant cardiovascular risk factors. Indeed, vasculitis in Behçet's disease may be the only etiology underlying thrombosis, stenosis and false aneurysms of the coronary artery [15]. Its yet poorly understood pathophysiology, encompasses multiple pathways, such as a reduction in endothelial diameter decreased systemic fibrinolytic activity or increase in fibrinogen and factor VIII [16].

Conclusion

Cardiac involvement in Behçet's disease has a high risk of morbidity and mortality especially in young patients. Therefore, careful attention in diagnosis and treatment of this cardiovascular complication, even in the absence of known risk factors is imperative.

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