Endomyocardial Fibrosis in Behçet's Disease: A Case Report

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Abstract

Endomyocardial fibrosis is exceptional in Behçet's disease. Less than 20 cases had already been reported. We report the case of a 28 years old man with a Behçet's disease complicated by a right endomyocardial fibrosis, associated to an intraventricular thrombus and to a pulmonary artery aneurysm confirmed by anatomopathological study. The patient underwent surgical thrombectomy, the outcome was marked by the recurrence of the intracardiac thrombus and led to the prescription of immunosuppressors with secondary favourable.

Keywords: Endomyocardial fibrosis, Behçet's disease

Introduction

Endomyocardial fibrosis (EMF) in Behçet's disease (BD) is exceptional, few than 20 cases have already been reported [1-3], we report the exceptional case of EMF involving the right ventricle associated to intraventricular thrombus and pulmonary artery aneurysm.

Case Report

A 28 years old Tunisian patient with a history of recurrent bipolar aphthosis, pseudo folliculitis and cerebral thrombophlebitis was hospitalized for dyspnea, chest pain and hemoptysis. On patient was afebrile, blood pressure was 120/80 mmHg. Cardiac auscultation noted regular tachycardia. Cutaneous mucosal examination revealed oral ulcerations, an active scrotal ulcer, several genital ulceration scars and scars of pseudo folliculitis. Ophthalmological examination revealed sequelae of bilateral venous occlusions. The rest of the somatic examination was without particularities including the neurological examination. The electrocardiogram showed in addition to sinus tachycardia, an S1Q3 pattern. The chest X-ray showed poorly limited right para-cardiac opacity measuring 6 cm long axis. The biological assessment showed an acceleration of the ESR (96 mm in the first hour) and leukocytosis (13000/mm3), there was no eosinophilia. The diagnosis of pulmonary embolism was suspected and confirmed by pulmonary perfusion scintigraphy. Transthoracic echocardiography showed dilated right cavities with a regularly contoured echogenic mass in the right ventricle. Systolic pulmonary arterial pressure was estimated at 31 mmHg. Transoesophageal echocardiography confirmed the presence of this right ventricular mass which was attached to a dyskinetic wall. Venous doppler ultrasound of the lower limbs and inferior vena cava was without abnormalities. The CT and magnetic resonance examinations (Figure 1) revealed a thrombotic aneurysm of the right lower lobar artery and a right ventricular mass that seemed thrombotic in nature (Figure 2a and 2b).

The diagnosis of a BD complicated by intracardiac thrombus and aneurysm of the pulmonary arteries was made. Treatment with heparin at a curative dose, prednisone (1 mg/kg/day) and colchicine was started, the course was marked by the persistence of the right intraventricular thrombus with recurrence of pulmonary embolism. Patient underwent surgical thrombectomy under extracorporeal circulation. The pulmonary arterial aneurysm has been respected. The perioperative exploration showed a vegetative ulcerative mass depending from endocardium. There

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been advanced, it is either a fibrous reaction secondary to an intracavitary thrombus or a primary lesion of the endocardium or myocardium, the second hypothesis seems most likely [2,6]. Macroscopic abnormalities of EMF during BD are different from those observed during EMF of hypereosinophilic syndrome and carcinoid syndrome. The fibrosis of the hypereosinophilic syndrome mainly affects the ventricular wall and rarely the valves themselves, the fibrosis of the carcinoid syndrome almost exclusively interest the valves, whereas the EMF of the BD affects both the valves and the ventricular wall. The EMF of the hypereosinophilic syndrome affects the right and left ventricles equally and causes late tricuspid involvement, whereas during BD, EMF preferentially affects the right ventricle with frequent tricuspid insufficiency [7].

The diagnosis of EMF can be challenging, as in our patient where the diagnosis was only made upon intraoperative findings and confirmed by the histopathological study. The diagnosis can be suspected on echocardiography, which can reveal a brilliant echo at the level of the endocardium, cardiac CT scan can show the presence of linear calcification surrounding the fibrosis and cardiac MRI can show apical ventricular obliteration, intracavitary thrombus and EMF mimicking a ventricular mass presenting as a mass of intermediate signal intensity in T1-weighted images, enhancing after injection of godolinium, with clear separation of the endocardium and the myocardium [1,3-4,9]. Intraoperative incidental discoveries, such as for our patient, or systematic endocardial biopsies [1] are also possible and normal echocardiographic and radiological investigations do not eliminate the diagnosis of EMF. Endomyocardial biopsy specimen can show dense fibrous tissue, inflammatory infiltrates and numerous neovessels in the endocardium [4].

Due to the high risk of thrombosis, anti-vitamin K treatment is the rule, but this treatment is difficult to handle in patients with pulmonary artery aneurysms, as in our patient, because of risk of cataclysmic haemoptysis by fissuring or rupture of the aneurysm. It is important to note that recently reported cases found that pulmonary aneurysm and cerebral sinus thrombosis are frequently associated to EMF in patients with Behçet’s disease [3], this was the case in our patient.
The medical treatment, particularly the combination of corticosteroids and immunosuppressant drugs (cyclophosphamide or azathioprine), plays an important role in reducing such accidents by reducing the inflammatory component. Several observations of disappearance of pulmonary artery aneurysms after treatment have been reported in the literature [10]. This treatment is also, in combination with the anticoagulant treatment, the first-line treatment for EMF and intracardiac thrombus [2,11]. Surgical treatment is reserved for cases where there is significant cardiovascular impairment, particularly when there is heart failure, recurrent embolic accidents or persistence of thrombus under medical treatment [12].

Conclusion

EMF is an exceptional manifestation of BD, it is often associated with intracardiac thrombi and association with pulmonary artery aneurysm is not exceptional. The first-line treatment is based on a combination of corticosteroids, immunosuppressive agents and anticoagulants. BD should be routinely sought for in patients with EMF in order to initiate this specific treatment.

References