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Surgical Treatment of a Recurrent Intracardiac Thrombosis Complicating Behcet's Disease

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Abstract: ***Cardiovascular involvements are frequent in Behçet's disease (BD), cardiac involvement is rare, affecting 1 to 6% of patients with BD. We report a case of a 19-year-old man, with a recent diagnosis of a BD, who presented with hemoptysis. An intra-right atrial thrombosis was diagnosed, and the patient was treated by medical treatment with a good initial result. 5 months later, he presented a recurrence of the intracdiac mass witch increase on volume and became very mobile. The CT-scan revealed a pulmonary embolism. He was operated under cardiopulmonary bypass and the mass was excised successfully. The postoperative course was favorable without any recurrence during 3 months follow-up.***

Keywords: Behçet disease, intracardiac thrombosis, surgery

1. Introduction

Originally described by a Turkish dermatologist; Hulusi Behçet in 1973, Behçet's disease (MB) was defined by the triad: mouth ulcers, genital ulcers and ocular involvement (1).

It is a multisystem vasculitis of unknown origin responsible for chronic inflammatory disease. Apart from the classic triad, it can manifest as venous thrombosis, arterial aneurysms, gastrointestinal involvement and skin lesions. Cardiac manifestations are rare, found in 1-6% in clinical series and 16.5% in an autopsy series (2). It may be pericarditis, myocardial involvement, valve involvement, and coronary or conduction tissue damage. Intracardiac thromboses are exceptional and serious complications.

They preferentially affect the right cavities and they are hard to manage (3).

We report the case of a young patient presented a recurrent right atrium thrombosis after medical treatment, complicated by pulmonary embolism and requiring surgical excision.

2. Case Report

We report the case of a 19-year-old man with a recent diagnosis of Behçet's disease. He developed low-abundance hemoptysis associated with a stage II NYHA classification dyspnea. Exploration revealed a mass in the right ventricle (RV), poorly mobile and adherent to the wall, complicated by pulmonary embolism. The patient received medical treatment combining corticosteroid, colchicine and immunosuppressants. The one-month check follow up have noted the resolution of the inflammatory syndrome and reduction in the size of the mass. The patient was kept under corticoid therapy.

Five months after, he presented a recurrence of hemoptysis. The physical examination found a blood pressure at 110/51mmHg, a heart rate at 71cpm, the cardiac auscultation was normal, as well as the pleural-pulmonary auscultation, and the rest of the physical examination was unremarkable.

The electrocardiogram shows a regular sinusal rhythm, a deviated QRS axis to the right and an incomplete right bundle branch block. The chest X-ray found cardiomegaly with a cardio-thoracic index at 0.6 with a normal pleuro-parenchymal transparency.

Biological examinations showed an inflammatory syndrome with a C-Reactive-Protein at 43mg/l, white blood cells count at 13300 elements/mm3.

The thoracic CT angiography showed a right ventricular mass with proximal pulmonary embolism. The pulmonary arteries were normal (Figure 1).

The transthoracic echocardiography (TTE) revealed a mass at the outlet portion of the RV witch increased in volume measuring 30x15mm and becoming very mobile and engaging through the pulmonary valve at each systole (Figure 2).

Given the increasing volume of the mass despite a well-conducted medical treatment and the threatening aspect of the mass witch prolapsing in the pulmonary artery, the surgical indication was retained.

The patient was operated on by vertical sternotomy under conventional cardiopulmonary bypass. The tumor mass was approached by a right longitudinal atriotomy. It was a voluminous mass, adherent to the anterior wall of the RV and to the ventricular face of the anterior tricuspid valve. It was also extending towards the outlet chamber of the RV until the pulmonary orifice. The entire resection required the disinsertion of the anterior tricuspid valve (Figure 3). The

pulmonary artery trunk and proximal branches were free from thrombosis. The anterior tricuspid valve was reinserted using an autologous pericardial patch (Figure 4).

The histopathological examination was in favor of a constituted fibrino-cruoric thrombus. In the postoperative and discharge TTE, the right ventricle was free of thrombosis. The patient discharged from the hospital after 10 days under corticoid therapy. During 3 months follow-up any recurrence was registered.

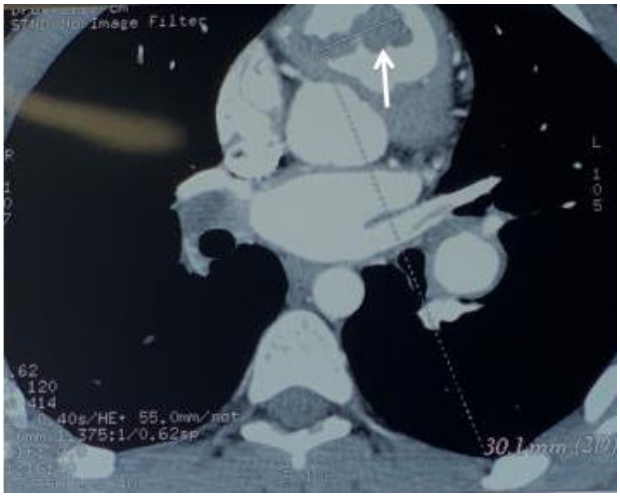

Figure 1: CT-scan shows intra-right ventricular mass

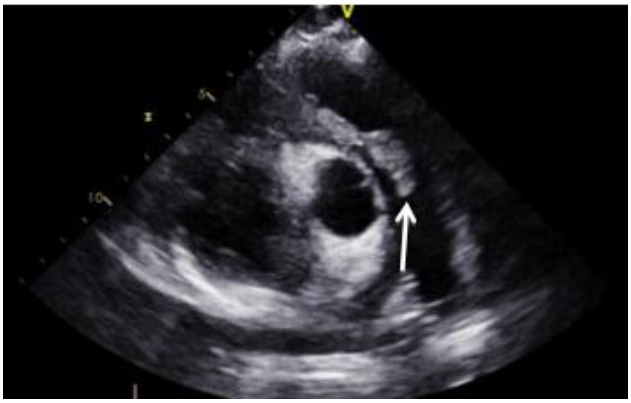

Figure 2: Echocardiographic image shows right ventricular outlet mass that engaging through the pulmonary valve.

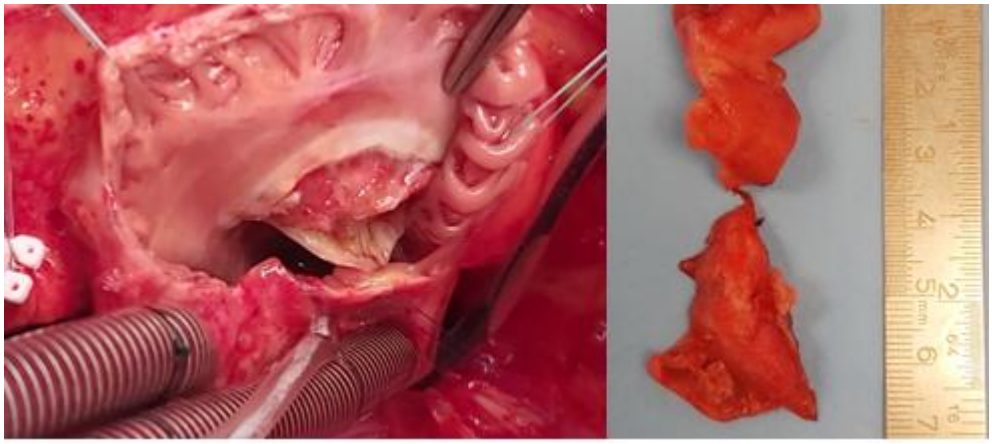

Figure 3: Operative view: desinsertion of tricuspid anterior leaflet (image on the left) ant total removal of the entire mass (image on the right)

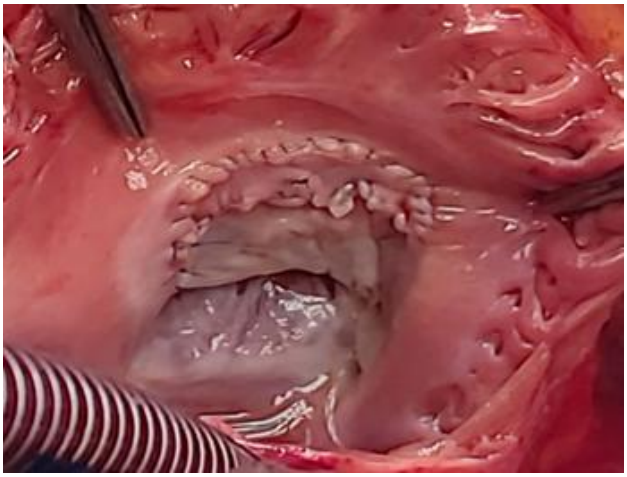

Figure 4: Enlargement of the anterior tricuspid leaflet by autologous pericardial patch

3. Discussion

Cardiovascular involvements are frequent in Behçet's disease (BD), superficial and deep venous thromboses are the most common of them. They occur in 20 to 40% of cases (4). However, cardiac involvement is rare, affecting 1 to 6% of patients with BD. This frequency was higher in an autopsy series with 16.5% of cases (5). Intracardiac thrombosis occurs preferentially in young male subjects, as is the case of our patient. It often appears in the early stages of the disease, and in 50% of cases, intracardiac thrombosis precedes the diagnosis of BD. The most frequent location is the right heart, and it is frequently associated with endomyocardial fibrosis. It can also be associated with pulmonary artery aneurysms. In our patient, intracardiac thrombosis was isolated without endomyocardial fibrosis or pulmonary aneurysms.

The exact etiopathogenesis is still not completely understood, but it seems that several mechanisms are responsible for the formation of intracardiac thrombosis: endothelial lesions, increase in pro-thrombothic factors, and the deposition of immune complexes in the vessels walls (2). Some authors have suggested that pulmonary embolisms are caused by in situ formation of thrombi rather than by embolization from the intracardiac mass (6). In our patient, the highly mobile nature of the mass and the normal appearance of the pulmonary arteries on the CT-scan suggested the embolization from the intracardiac mass. The clinical symptoms are not very specific, however the occurrence of hemoptysis is very suggestive of intracardiac thrombosis. The diagnosis is based on transthoracic echocardiography (TTE) and transesophageal echocardiography (TOE), which allow the diagnosis of intracardiac thrombosis. Cardiac CT-scan and MRI can detect pulmonary embolization and show associated pulmonary artery aneurysms or endomyocardial fibrosis.

In the absence of clear guidelines, the treatment of BD is not codified and it depends on the severity of the systemic signs and especially on the type of organ affected. In intracardiac thrombosis, medical treatment is the treatment of choice. It is based on colchicine, corticosteroids and especially immunosuppressants (2). Several studies have reported complete resolution of thrombosis with this combination, with success rates of up to 78%. Other studies have reported decreased recurrence rates with immunosuppressants (3-7). The use of anticoagulant is very debated. Some authors have reported better efficacy when using anticoagulants in combination with other medical treatments (7), others are more reticent because of the risk of bleeding, especially when pulmonary artery aneurysms are associated.

Surgical resection alone is often ineffective with early recurrence. Surgical treatment is primarily indicated for recurrences under medical treatment and for threatening thrombi with a high risk of massive embolization or valve obstruction (3,8). Surgery should be performed at a distance from the inflammatory period and medical treatment should be continued postoperatively to prevent recurrence. In our case, the surgical indication was retained because of the recurrence under medical treatment and the threatening nature of the mass.

The prognosis is favorable in the absence of recurrence. Because of the risk of recurrence (after medical treatment or surgical resection), a routine echocardiographic follow up is mandatory.

4. Conclusion

The intracardiac thrombosis is a possible complication of Behçet's disease, which can be life-threatening for the patient. Early transthoracic echocardiography is essential for the assessment of these complications. Medical treatment is initiated as a first-line treatment, but surgery is nevertheless necessary in some cases.

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