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RESEARCH ARTICLE

INTRA-CARDIAC THROMBUS IN BEHÇET'S DISEASE: TWO CASE REPORTS

A. Maliki Alaoui[1], B. Mesmoudi[1], Y. Amchich[2], N. Loudeyi[1], A. Zaimi[1], I. Asfalou[1] and A. Benyass[1]

1. Cardiology Department of the Military Training Hospital of Mohamed V, Rabat, Morocco.
2. Pulmonology Department of the Military Training Hospital of Mohamed V, Rabat, Morocco.

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Abstract

Behçet's disease (BD) is a multi-systemic inflammatory disorder characterized by a recurrent oral and genital aphthosis associated with multiple organ disorders: ophthalmic, neurological, vascular, and digestive.Cardiac involvement in BD especially Intra-cardiac thrombus (ICT) is extremely rare; only 50 cases have been registered worldwide [1-5]. We report a rare case of two patients aged respectively 18-year-old and 50-year-old who are diagnosed with Behçet's disease complicated by right-sided heart thrombi extended to the pulmonary artery.

Introduction:-

Described first by the Turkish dermatologist Hulusi Behçet in 1937, Behcet's disease (BD) also known as oculo-orogenital syndrome or malignant aphthosis, is a rare systemic vasculitis involving arteries and veins of all sizes and types.

Cardiac involvement is rare but serious and life-threatening. Its prevalence varies from 1 to 6% in clinical series [5,6]. It can affect all three tunics: endocardium, myocardium, and pericardium. Endomyocardial involvement remains exceptional, manifested by intracavitary thrombus and endomyocardial fibrosis [7].

The seriousness of cardiac damage in BD implies its systematic search during the follow-up of patients as well as its suspicion in case of atypical cardiac manifestations in a young man without risk factors [8].

CasePresentation:

Case1:

A 50-year-old man, having ascardiovascular risk factor a chronic smoking cessation, presented with a long-term fever associated with significant weight loss and tiredness for one year. The evolution was marked by the appearance, 6 months later, of mouth and genital ulcers which disappear and reappear episodically.

The patient developed, at the same time, Abdominal pain and Intermittent arthritis of the knees and ankles. There is no family history of Behçet's syndrome or other inflammatory diseases.

The bioassay showed hyperleukocytosis at 11.000/mm³, thrombocytopenia at 131.000/mm³, an acceleration of the sedimentation rate. The CRP value was normal. Our patient meets the International Criteria for Behçet's Disease. He was put on prednisolone and colchicine with good evolution.

Corresponding Author:- Amine Maliki Alaoui
Address:- Department of Cardiology, Mohamed V Training Military Hospital, University of Mohammed.V.Rabat, Morocco.

The patient was admitted to our hospital for 1-Week exertional dyspnea. On admission the patient was asymptomatic. An electrocardiogram showed sinus rhythm with a complete right bundle branch block and secondary repolarization changes.

An echocardiogram revealed a voluminous thrombus of the right ventricle extended to the infundibulum and the trunk of the pulmonary artery (Figure 1), a dilated right ventricle and atrium, a dysfunction of the right ventricle, a distended inferior vena cava with diminished inspiratory collapsibility, ejection fraction:55%, mild pulmonary hypertension, a normal filling pressure of the left ventricle and a low abundance pericardial effusion.

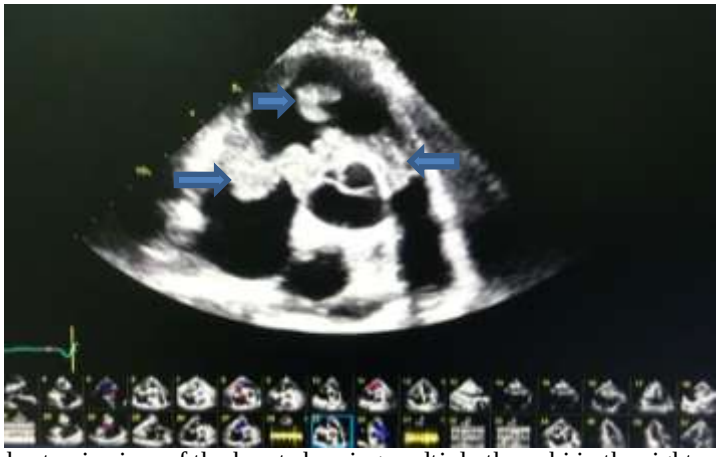


Figure 1:- Parasternal short axis view of the heart showing multiple thrombi in the right ventricle and the trunk of the pulmonary artery.

These findings have been confirmed by trans-esophageal echocardiography (figure 2). Anticoagulant treatment with low Molecular Weight Heparin and Vitamin K antagonists was started. In later echocardiographic control, the thrombus size has decreased. The patient did well without any adverse events.

Case 2:

An 18-year-old male with a noncontributory past medical history, presented with three weeks of dry cough and intermittent hemoptysis. For several days before hospitalization, he experienced severe exertion dyspnea, high fever, and shivers. The patient acknowledged recurrent oral and genital ulcerations in the past three years. No other symptoms were documented.

The patient was conscious, oriented, and appeared weak and thin. He weighed 48 kilograms of unintentional weight loss over the preceding two months. Vital signs were as follows: temperature 39; blood pressure 125/73 mmHg; heart rate 110 bpm; respiratory rate 22 bpm and oxygen saturation of 96% on room air. He had face folliculitis otherwise; the rest of the examination was unremarkable.

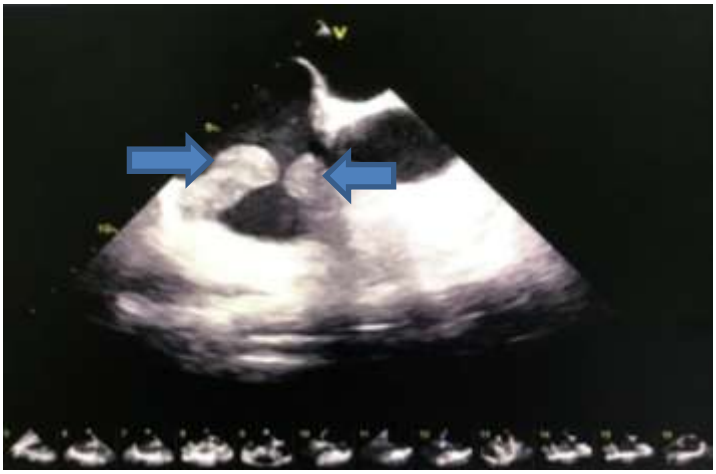


Figure 2:- trans-esophageal echocardiography view showing the thrombi of the right ventricle.

Laboratory results were obtained and noted a leukocyte count of 11.5 10^3 cells/ml; hemoglobin level of 12.5 g/dl, hematocrit of 38.5%, and platelets count of 220 100 cells/ml. C-reactive protein was 115 mg.l-¹.The liver function tests, urea, and electrolytes were normal. Chest radiography found right basal consolidation. Acid-fast bacillus testing was negative, culture was underway.

A chest CT-scan showed condensation in the apical segment of the inferior right lobe (Figure 3). The patient was put under the association of amoxicillin + clavulanic acid, without any favorable outcome.

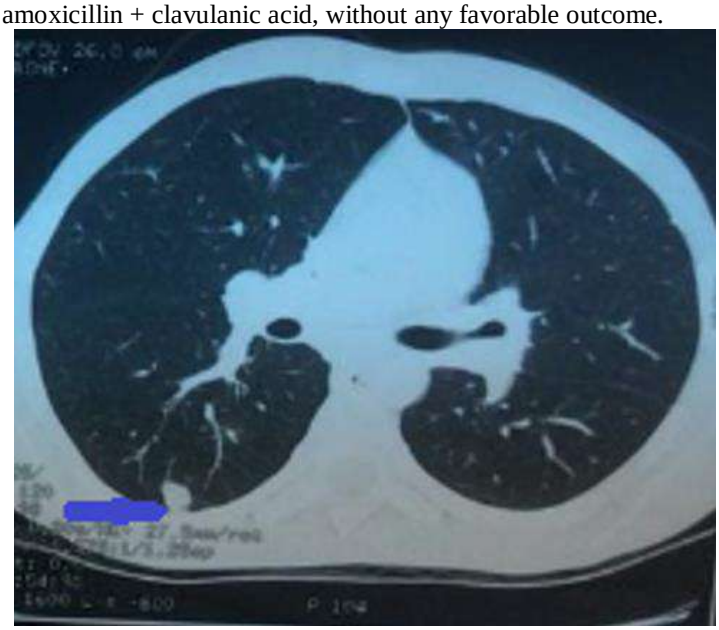
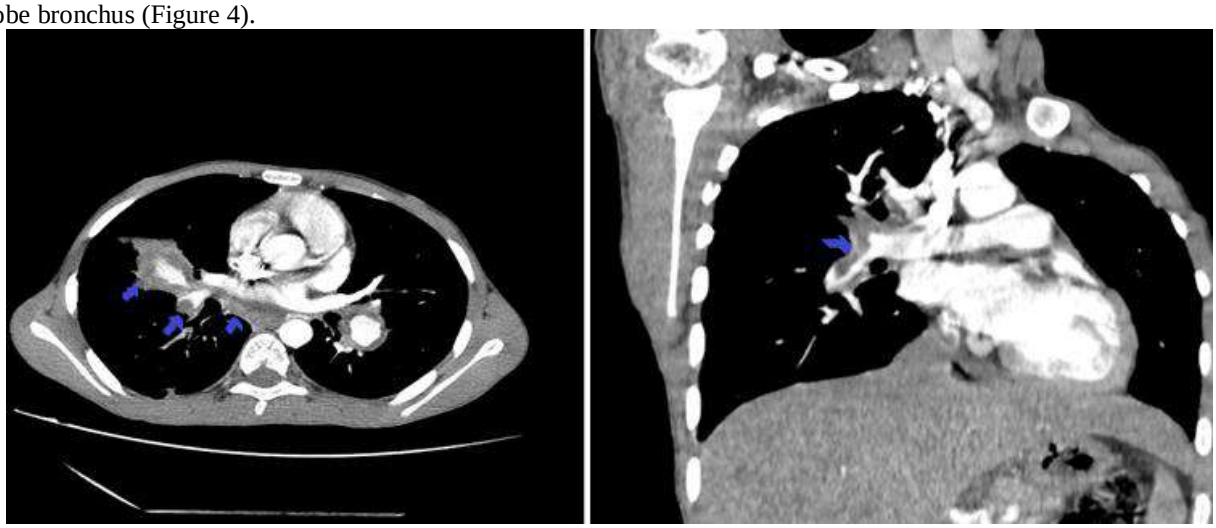


Figure 3:- Transversal (axial) CT scan of the chest showing condensation in the apical segment of the inferior right lobe.

Considering the young age of the patient, the history of bipolar aphtosis, and the recurrent hemoptysis, a chest CT angiography was performed, showing a pulmonary embolism of the right pulmonary artery with right ventricular thrombus, the presence of a bilateral partial thrombosis ectasia of the right middle lobe bronchus and the left lower lobe bronchus (Figure 4).



The transthoracic echocardiography showed important right ventricle enlargement and a thrombus measuring 35 mmx27 mm of the right ventricular outflow tract and distal right ventricle. The pathergy test was negative. He was finally diagnosed with BD complicated by extensive venous thromboembolism including right ventricle thrombus and pulmonary embolism. Anticoagulant then immunosuppressant (methylprednisolone and cyclophosphamide) and colchicine have been introduced with marked clinical improvement.

Discussion:-

Behçet's disease (BD) is a rare systemic vasculitis characterized by a recurrent oral and genital aphthosis associated with multiple organ disorders: ophthalmic, neurological, vascular, and digestive. The etiology of BD is unknown. Viral, bacterial, genetic, environmental, toxic, and immune factors have been implicated [9]. Ethnicity is important in the incidence of the disease, with Mediterranean and Asian individuals being affected more often. Carrying the human leukocyte antigen B51 increases the risk of BD developing by 1.5 to 16 times, but it is not correlated with the severity of the disease [10].

The largest series to date demonstrated cardiac involvement in approximately 6% of individuals with BD [5]. The most common cardiac features include pericarditis, valvular insufficiency, myocardial infarction, and myocarditis. Interestingly, intraventricular thrombosis remains a relatively rare cardiac manifestation of vasculitis in patients with BD affecting the heart, reflecting the unique predilection for thrombotic complications in this disorder [11]. The male sex, arterial and venous damage, are significantly associated with heart involvement [12].

ICT autopsy was first described in 1977 by Buge et al. [1]. Since this publication, about fifty similar cases have been registered worldwide which confirm the extreme rarity of ICT [2,5]. ICT is located more frequently in the right ventricle much more than the left however left atrial location is rarely seen [6]. The association of ICT and aneurysm was often reported and contributed widely to BD diagnosis [13]. The gold standard examination for diagnosis of ICT is trans-esophageal echocardiography and the normality of trans-thoracic echography cannot exclude this complication.

Overall survival in BD patients with cardiac involvement was poorer than in those without. Complete remission of cardiac thrombi, as the case of our two patients, was associated with the use of immunosuppressants, colchicine, and anticoagulants [5].

In case of ineffectiveness of medical treatment or a massive or extensive thrombus, surgical thrombectomy can be proposed. Recurrences after surgical treatment are possible and require reinforcement of drug treatment. in certain cases; thrombolysis can constitute an interesting alternative to surgical treatment [14].

Conclusion:-

Intra-cardiac thrombus is a very rare complication of BD and it could exceptionally reveal it. It should be considered in young patients with intracardiac mass. The gold standard examination for diagnosis is trans-esophageal echocardiography and the normality of trans-thoracic echography cannot exclude this complication. Medical treatment including corticosteroids, immunosuppressive drugs, and anticoagulant is the first-line treatment, and surgery should be considered when the thrombus is not resolutive or becomes massive and extensive.

Consent:

The authors confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient.

Disclosure of Interest:

The authors declare that they have no competing interest.

Author Contributions:

All authors have contributed to the elaboration of the manuscript.

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