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RESEARCH ARTICLE

HEMOPTYSIS REVEALING INTRACARDIAC THROMBOSIS: A CASE REPORT OF BEHÇET'S DISEASE

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Abstract

Behcet's disease is a chronic multisystem disorder characterized by relapsing inflammation for which the underlying histopathology is an occlusive vasculitis. Intracardiac thrombosis is a rare complication of Behçet's disease. The management of this involvement is difficult due to the risk of recurrence. We prresent the case of a young man admitted to our hospital for hemoptysis. We discovered a right ventricle thrombus on CT angiogram and we confirmed the diagnosis on echocardiography.

Introduction:-

Behçet's disease (BD) is a systemic inflammatory disease having a chronic and prolonged course, it is characterized by oral and genital ulcerations, ocular inflammatory involvement, skin lesions, vascular involvement and numerous systemic manifestations that may affect mortality. The pathogenesis of the disease still remains poorly characterized, although infectious, genetic (HLA-B51 antigen polymorphism), and environmental factors have been implicated [1].

Vascular involvement and thrombotic tendency is a potentially lifethreatening condition in patients with Behçet´s disease. Deep vein thrombosis and pulmonary embolism are the most common manifestation but other locations may also occur. A risk factor of thrombosis was observed in many patients[2].

The over all prevalence of vascular involvement across cohorts of patients with BD ranges between 14 and 39 %, with venous lesions and thromboses being more frequent than arterial ones [3-4]. Intracardiac thrombus formation (ICT) is a rare but serious manifestation of BD. Untill 2014, there were just 89 cases previously published in 76 reports in which BD was associated with intracardiac thrombus[5].

Here, we report an another case of right ventricular thrombus in a patient with BD discovered during a hemoptysis.

Case report :

A 22-year-old male patient, without any cardiovascular or systemic illness and was not taking any medication, admitted to pneumology department of our hospital for hemoptysis of small abundance for 03 weeks, without dyspnea or chest pain. In addition, the patient reports the notion of recurrent oral aphthosis with two attacks per month with inflammatory polyarthralgias..

On physical examination, the patient was respiratory and hemodynamically stable, his blood pressure was 12/06 and his respiratory rate was 20 cycles per minute. An ulcer scars on oral mucosa and pseudofolliculitis were found.

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During the cardiac evaluation a diastolic murmur at the pulmonary area was heard.Fundus examination did not show uveitis.

Hematological investigations revealed white blood count 10.30 103 cells/µl with normal differential; hemoglobin level was 13.8 g/dL; hematocrit, 41.2% and platelets were 205 103 cells/ml. C-reactive protein was 60 mg/L. The liver function tests, urea and electrolytes were normal.

Chest X-ray showed accentuated convexity of the right inferior arch of the cardiac silhouette and a sub diaphragmatic projection of the tip of the heart [Figure1].The thoracic angioscanner revealed a right intraventricular thrombus adherent to the wall, measuring 3.1 cm in long axis with good opacification of the pulmonary artery up to the segmental branches [Figure2].

The echocardiography showed a right intraventricular thrombus, consisting of two parts, the first measuring 1.5 cm long axis, adherent to the posterior wall, the second measuring 1.9 cm long axis, mobile and protruding into the trunk of the pulmonary artery during ventricular systole [Figure3]. A pulmonary arterial hypertension (56 mmHG) was detected.

The patient was treated by anticoagulant (sodium heparin in curative dose with an autopulsed syringe), oral cyclophosphamide (100 mg/day), and prednisone (1 mg/kg/day). The glucocorticoid dose was gradually reduced.

Discussion:-

BD is a vasculitis which can involve all arteries and veins irrespective of diameter. Cardiac involvement is relatively uncommon [6]. The heart and great vessels are not primary targets of BD, but although not well recognized, arterial or cardiac involvement is life-threatening with associated strong prognostic implications in BD.

Cardiac involvement includesmyocardial infarction, endocarditis, pericarditis, endomyocardial fibrosis, aneurysms of the coronary arteries of sinus of Valsalva. Their prevalence changes between 7% and 29% according to the different ethnic group [7].Intracardiac Thrombosis is a rare and serious complication. It is reported mostly in case reports and case series, with <100 cases reported in the medical literature [8].

The pathogenic mechanism underlying thrombotic propensity in patients with BD is not completely understood.. It is however reported that that endothelial cell ischemia or disruption leads to enhancement of platelet aggregation [9].Prothrombin gene mutation was identified in some BD patients. Increased plasma homocysteine levels are also a risk factor for thrombosis in BD[10]. Another conceivable pathogenic mechanism is the presence of antiphospholipid antibodies, which have been reported in 18% of cases in a study [5].

Intracardiac thrombosis is located usually in the right side of the heart, the right ventricle being the most common location [11], this might partly attributed to extending of thrombi in vena cava and lower pressure of the right heart.Involvement of the left heart has been rarely reported [12]. It can be associated with a venous thrombosis of the superior and/ or inferior vena cava (45.3%), pulmonary embolism (41.5%), and aneurysms of the pulmonary arteries (38.3%) [11].

Transesophageal and transthoracic echocardiography are generally sufficient to establish thediagnosis of cardiac thrombosis, it appears usually as aheterogeneous and echogenic masson intracavity. Other Imaging tests such as CT chest and MRI could be helpful in the assessment of associated thoracic manifestations of BD [13].

Treatment of intracardiac thrombus is unclear, because there is no predefined consensus. The aim is to clean the cardiac thrombus and to prevent the recurrence of this complication.In previous reports, initiation of combination therapy for Behçet’s disease with prednisone and an immunosuppressant was associated with the resolution [14]. Anticoagulants in the treatment of thrombosis in Behçet's disease have been widely discussed, and their results are controversial. The European League Against Rheumatism (EULAR) does not recommend the use of anticoagulant alone in the management of thrombosis [15]. However, anticoagulation treatment together with immunosuppressive therapy is an advised treatment option in Behçet’s cases with intracardiac thrombosis.

Surgery, performed as a first line treatment incardiac thrombosis inaugurating a Behçet disease, has a high rate of complications and death [16].It might be limited to the cases with massive thrombosis, recurrence of the complication despite an optimal medical treatment, and when there is a cardiac congestion [17].

Conclusion:-

In conclusion, Intracardiac thrombus formation is a rare but serious manifestation of BD.Hemoptysis is an alarming sign and should lead to thoracic angioscanner and transthoracic echocardiography. The combination of methylprednisolone, cyclophosphamide, and sodium heparin is a good option to treat such a complication.

The authors state that they have no Conflict of Interest (COI

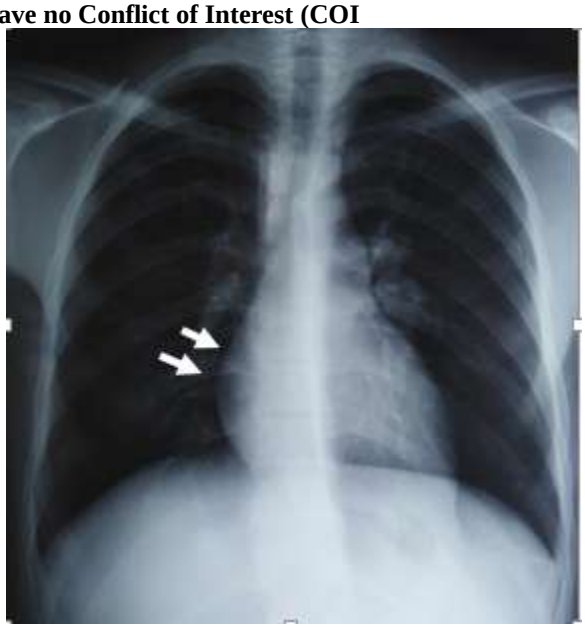

Figure 1:- Chest X-ray:Accentuated convexity of the right inferior arch of the cardiac silhouette and a sub diaphragmatic projection of the tip of the heart.

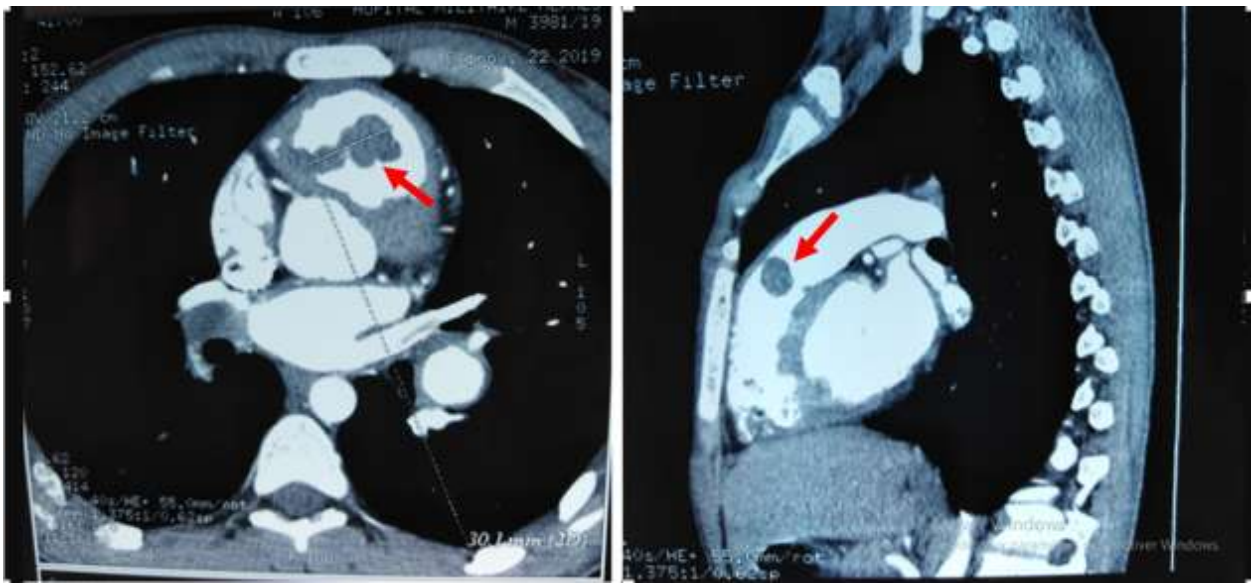

Figure 2:- Transversal and coronal of thoracic angioscanner showing right ventricular thrombus.

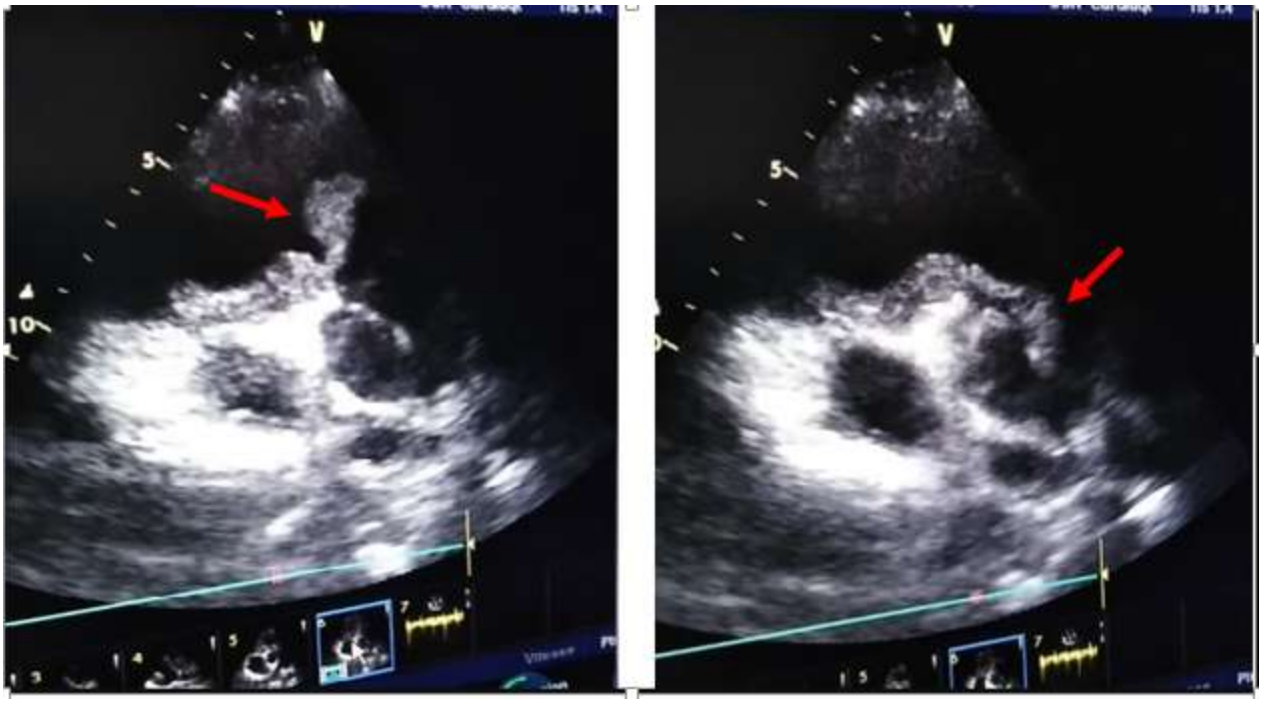

Figure 3:- The echocardiography showed a right intraventricular thrombus mobile and protruding into the trunk of the pulmonary artery during ventricular systole.

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