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RESEARCH ARTICLE

A CASE SERIES OF INTRACARDIAC THROMBOSIS REVEALING BEHCET'S DISEASE IN MOROCCAN PATIENTS

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Abstract

Aim of the work: The clinical presentation of Behcet's disease (BD) is varied, While cardiovascular effects such as pericarditis, acute myocardial infarction, conduction disorders, and valvular disease have been reported, they are infrequent. The formation of intracardiac thrombus, as seen in our patients, is an exceptional occurrence. Patients and methods: We present a retrospective study of 4 cases of intracardiac thromboses in a series of 50 patients followed for BD in our department (over a period of four years), The diagnosis is made on the basis of the criteria proposed by the International Study Group (ISG).

Results: All were male, the mean age at the time of the ICT diagnosis was 29.5 years. The main presenting symptoms were hemoptysis, chest pain, and dyspnea. It was associated with pulmonary artery aneurysm in two cases, and pulmonary embolism in all cases. In three cases, echocardiography was sufficient to reach the diagnosis; but, in one, the difficulty of differential diagnosis with a cardiac tumor has been raised. The diagnosis has been confirmed by examining the operative specimen histologically in the case of this deceased patient, who experienced postoperative complications. Three patients were treated with immunosuppressant agents, corticosteroids, and anti-TNF alpha, one of whom showed a good response to treatment.

Conclusion: A potential diagnosis of BD should be considered if a patient presents with an intracardiac mass, this is especially relevant in young male patients.

Introduction:-

Behcet's disease (BD) is a systemic vasculitis is characterised by recurrent bipolar aphthosis and joint involvement. Additionally, it may involve varying levels of severity of ocular, neurological, and vascular manifestations [1]. Intracardiac thrombus (ICT) is a rare and serious complication of BD, which can have a life-threatening outcome. We present four cases of ICT from a group of 50 patients diagnosed with BD in our department over a period of four years, as well as their evolution in response to corticosteroids, cyclophosphamide and anti-TNF alpha therapy.

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Cases reports:
Four Moroccan patients, with Behcet's disease, revealed by ICT, Characteristics of the four patient series including clinical features are summarized in Table 1.

Case 1:
A 18-year-old male patient, who was recently diagnosed with Behcet's disease, revealed by cerebral thrombophlebitis and inaugurated by bipolar aphthosis. The patient complained of moderate hemoptysis and mild chest pain in the emergency department. A chest angioscan was performed and showed a pulmonary embolism (PE). Considering that BD is not very emboligenic, a transthoracic ultrasound echocardiogram was performed, which showed the presence of an ICT. It was decided to start the patient on monthly boluses of methylprednisolone combined to cyclophosphamide for 6 months, based on severity of vascular involvement, followed by azathioprine. The evolution was noted a clinical improvement, radiological control will be scheduled at the end of his six boluses.

Case 2:
A 17-year-old male patient was admitted to our hospital with an unknown origin cough, dyspnea and mild hemoptysis. He had been experiencing recurrent oral ulcers for one year. During the physical examination, we observed the patient had tachypnea and signs of healing scars from genital ulcers. Additionally, the pathergy test yielded a positive result. Further, a chest angioscan revealed multiple partially thrombosed pulmonary arterial aneurysms (PAA) with PE. Cardiac echocardiography confirmed the presence of a thrombus adhering to the side of the right ventricle. The patient fulfilled the ISG criteria for the diagnosis of BD [2]. The patient received a first bolus of methylprednisolone and cyclophosphamide, followed by anti-TNF-alpha. The clinical and radiological evolution was favorable.

Case 3:
A 44-year-old patient with no significant medical history presented to the emergency department with severe hemoptysis and chest pain. They had a history of recurrent oral and genital ulcers for the past two years. Physical examination revealed tachycardia, papulopustular lesions on the upper back, large ulcers in the oral cavity and on the penis. Chest X-ray and electrocardiogram showed no abnormalities, but laboratory tests indicated high acute phase reactants and elevated D-dimers levels. An angioscan revealed two small embolisms in the right lung's arteries. The patient was diagnosed with Behcet's disease with vascular involvement based on the ISG criteria for BD. A cardiac ultrasound examination indicated the presence of an echogenic mass in the right atrium. Cardiac MRI confirmed the presence of an intracardiac tumor. While the cardiology team initially suspected a right atrial myxoma, the internal medicine team agreed with the diagnosis of ICT as a complication of BD, with the pulmonary embolism being a secondary outcome. The patient underwent open-heart surgery, but the histology of the surgical material revealed an ICT. Unfortunately, the patient passed away due to post-operative complications.

Case 4:
A 39-year old male, with history of deep vein thrombosis on lower limbs had been treated 1 year before with oral anticoagulation for 12 months, and recurrent bifocal ulcers, pseudofolliculitis vein thrombosis was hospitalized with chest pain, prolonged fever, and abundant hemoptysis. Chest CT showed multiple thrombosed aneurysm of pulmonary artery, and right intraventricular thrombus Echocardiography confirmedright intraventricular thrombus. BD was retained. Methylprednisone and monthly pulses of cyclophosphamide were administered. Control angioscan realized at the sixth cycle of therapy showed an improvement of the size of the aneurysm and control echocardiography revealed a complete resolution of right ventricular thrombus. Cyclophosphamide was replaced by azathioprine. The patient is now regularly seen in our outpatient clinic and follow-up is 2 years.

Discussion:-

The most common cardiac involvement in BD is benign acute pericarditis, which occurs simultaneously with an inflammatory flare-up of the disease. Myocarditis that occurs without lesions of the large coronary trunks has also been reported. Finally, rare cases of spontaneous ventricular aneurysms or those that occur after myocardial infarction have been described. Intracardiac thrombosis is rare in BD [3]. In our BD series comprising 50 cases over four years, four cases demonstrate cardiac involvement in BD. All our patients presented ICT in BD, with a prevalence of 8%, which is markedly higher compared to other case studies [4, 5]. Our four observations are few among those described in the literature. The age of diagnosis (29, 5 years) approximates the average of 28 years as reported in the literature with a range between 12 and 51 years. TIC reveals the disease in all cases, considering that in the literature it reveals the disease in 50% of cases. The clinical presentation of ICT is unspecific, patients

presenting with dyspnea, hemoptysis, chest pain or fever. In our series, ICT was associated to PAA in two cases and PE in all four cases. It should be noted that this association of PAA with ICT carries a risk of hemoptysis and death due to rupture of the aneurysms, and the risk of hemorrhage is increased by the initiation of anticoagulant therapy for ICT. ICT is located usually in the right side of the heart, the right ventricle being the most common location [6]. Involvement of the left heart has been rarely reported [7]. In our case series, the right heart was involved in all cases with right atrial thrombosis. Echocardiography typically reveals this complication, and the primary potential differentials include cardiac tumors and infectious endocarditis, this was the situation with our third patient. Imaging examinations, such as magnetic resonance imaging (MRI), may aid in the evaluation of thoracic manifestations associated with BD, including ICT [8]. In our case, a cardiac MRI was performed, but the appearance was in favor of a tumor in the right atrium. The dossier was in the hands of the cardiology team and they were in favor of cardiac surgery as there was a strong suspicion of an intracardiac tumor. As internists, we supported the diagnosis of intracardiac thrombus in view of the fact that the patient has Behçet's disease, vascular damage and male sex, all of which are risk factors for cardiac involvement, including ICT. The patient underwent cardiac surgery, and the histological analysis of the operative specimen revealed ICT, illustrating the importance of understanding this complication. The pathogenic mechanism of ICT is incompletely understood, some authors have reported prothrombotic plasma factors in patients with cardiovascular involvement [9,10]. Others have suggested thrombogenic abnormalities specific to MB, such as the presence of antibodies directed against enolase, a target protein on the surface of endothelial cells [11]. A defect of fibrinolysis and hyperhomocysteinemia were however suggested [12, 13]. In previous studies, the administration of both prednisone and an immunosuppressant in combination therapy for Behçet's disease has been linked to disease resolution [14]. There are no real recommendations in the management of cardiac involvement due to BD. In our case study, two patients received monthly boluses of cyclophosphamide combined with methylprednisolone, for 6 months, followed by Azathioprine and one received anti-TNF alpha therapy. The three patients showed good efficacy. More studies are needed to verify these findings.

Conclusion:-

TIC is a rare yet potentially severe occurrence in BD. Our series suggests a high prevalence of this complication in our patients, which may reveal the disease. Therefore, any intracardiac mass in a young male should be regarded as a TIC.

Table 1:- Gender, age, clinical features, and management in Behçet's disease patients with intracardiac thrombus.

Case	Gender	Age	Clinical feature	ICT site	ICT diagnosis	Treatment
Case 1	Male	18	Hemoptysis/chest pain	Right atrium	Echocardiography	Methylprednisolone Cyclophosphamide
Case 2	Male	17	Dyspnea/ hemoptysis	Right atrium	Echocardiography	Anti-TNF alpha
Case 3	Male	44	Hemoptysis/ chest pain	Right atrium	Histology	-
Case 4	Male	39	Fever/ hemoptysis/ chest pain	Right atrium	Echocardiography	Methylprednisolone Cyclophosphamide

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