Original Research Article

Dilated cardiomyopathy with biventricular thrombi : a severe manifestation of behcet disease

Abstract:

Behcet's disease is a systemic vasculitis of the vessels for all calibers, touching arterial and venous territories. The causes of disease are unknow . BD reaches young age subjects from 10 to 45 years and affects both men and women. BD is ubiquitous but more frequent in patients from Mediterranean basin, the middle East and Asia. The diagnosis of BD is essentially clinical. The diagnostic criteria make it possible to carry the diagnosis with good sensitivity and specifity. BD evolves by recurrent inflammatory attack. BD can affect all of the organs; cardiacs manifestations are dominated by intracardiac thrombosis, the damage of three tunics, coronaryarteritis with or without myocardial infarction, coronaries aneurysms and endomyocardial fibrosis. The vascular manifestations are dominated by arterial or venous thrombosis. The presence of dilated cardiomyopathy with reduced left ventricular ejection fraction is rare. It can be explained by ischemic or inflammatory origin by cytokines. We report a case of young woman aged of 33 years to the history of 3 episodes of bipolar aphtae which presented dilated cardiomyopathy with reduced left ventricular function, biventricular thrombosis, bilateral distal pulmonary embolism with pulmonary infarction.

Keys word: Dilated cardiomyopathy, Behcet disease, Thrombus, left ventricular dysfonction

Introduction:

The BD is a rare systemic vasculitis of unkwon cause which is characterized by recurrent inflammatory attacks affecting many organs [1]. Pathophysiology hallmark is an occlusive vasculitis local and systemic. BD is observed worldwide but more frequentin countries located on the old silk road especially in the middle East, Japan, Turkey. In Africa the cases are rare essentially found in the Maghreb [1-3]. BD often touch young patients aged between 30 and 40 years is exceptional before puberty or after age of 50 year. It usually affects young subjects between 30 and 40 years old, is rare before puberty and after 50 years [1-4]. It is associated with significant morbidity and mortality particularly in men when it occurs at a

very young age [1-4]. It is mainly manifested by damage to mucocutaneous, ocular, joint, neurological, vascular (venous and arterial) as well as cardiac damage [1, 3, 4]. Cardiac involvement is dominated by intracardiac thrombosis, involvement of the tunics, arteritis and coronary aneurysms, endomyocardial fibrosis [1, 3, 4]. The diagnosis is clinical. Treatment is symptomatic using anti-inflammatories and/or immunosuppressants [1,5].

Patient and observation:

This is a 33-year-old patient, with no particular cardiovascular risk factors apart from gestational diabetes put on a diet, with no notion of known heart disease, having 3 episodes of bipolar aphtosis per year, on estrogen-progestogen pill for 1 month. , without notion of miscarriage (last child aged 17 months). For 8 months, she has had NYHA stage II dyspnea, aggravated 15 days before her admission to the emergency service with dyspnea at rest with productive cough, moderate hemoptysis in a context of fever with poor general condition. She was hemodynamically stable with arterial pressure at 110/70 mmHg, polypnea at 32 cycles/min, tachycardia at 110 bpm. Examination found bilateral crackles, signs of right heart failure, edema of the lower limbs, turgor jugular veins). The electrocardiogram shows sinus tachycardia, left anterior hemiblock. Previously, the patient had presented to the emergency room and the diagnosis of a bilateral distal pulmonary embolism with an area of infarction had been made after performing a chest CT angiography (Fig 1) with diffuse ground glass images; the diagnosis of a Covid-19 infection was ruled out after a negative PCR as well as serologies. Biologically, there is a biological inflammatory syndrome with a CRP at 88 mg / l, leukocytes at 9.8 giga / l, platelets at 289 giga / l, moderate impairment of renal function with clearance at 42 ml / min.

Immunological assessment and Serological test were negative.

The BK sputum to eliminate atypical tuberculosis came back negative.

Transthoracic ultrasound revealed an aspect of global hypokinetic heart disease at the dilated stage (DTD/DTS: 63/55 mm), with severe systolic dysfunction at 25% (SBP), low cardiac output at 1.7 l/min. We noted the presence of an adherent thrombus at the left apical level measuring 21 x 40 mm, and several right apical thrombi, the largest of which measured 12 x 23 mm. There were no significant mitroortic valvulopathies. The mitral profile was restrictive, indicating an increase in the filling pressures of the left ventricle. The atria were dilated, echo-free; the right ventricle was undilated with moderate longitudinal systolic dysfunction. There was a moderate pulmonary hypertension at 60 mmHg with a dilated inferior vena cava (fig 2).

The patient was depleted by injectable furosemide with a rapid oral relay, the treatment of heart failure was introduced gradually after passing the acute stage. It was decided to start SGLT2 inhibitors but the high cost was a hindrance for the patient. Oral anticoagulation with vitamin K antagonists was initiated with a target INR of 2-3. After discussion with the internists, the patient received a bolus of corticosteroids with an oral relay. The evolution 3 months later found a patient with few symptoms with NYHA stage II dyspnoea, without clinical signs of heart failure; cardiac echography noted a slight improvement with an ejection fraction of 35%, disappearance of the left apical thrombus, regression of the thrombotic magma at the right ventricular level (Fig 3). The global longitudinal strain was altered at -4.2% (fig 3). We titrated the treatment of heart failure gradually in view of an optimal dose.

Result & Discusssion:

Behcet's disease is a systemic inflammatory disease of unknown etiology. Its pathophysiology is based on local and systemic occlusive vasculopathy [1,2,6-8]. Both sexes are concerned and the predominance varies according to the regions and the series. It most often occurs in young people between the ages of 30 and 40. It occurs most often in Mediterranean regions, the countries of the Middle and Far East with an estimated prevalence of 80-420 cases per 100,000 inhabitants in Turkey, on the other hand it is around 20 cases per 100,000 inhabitants; Its prevalence is lower in Europe and America, in sub-Saharan Africa, it predominates in North Africa but there are no national registers, there are only very few cases described in sub-Saharan Africa [1,2]. The role of genetic factors is incriminated, in particular the presence of the type I histocompatibility complex (HLA-B51) as well as environmental factors that are still poorly understood to this day [1, 5, 6]. It is a complex nosological entity manifested by mucocutaneous, ocular, joint, neurological, vascular, venous and arterial as well as cardiac damage [2, 6, 7, 9]. Vascular involvement is one of the most frequently encountered elements in BD and is not seen in all patients; all vessels, of any caliber and at all sites can be affected: this is why BD is classified in the variable vasculitis group by the 2012 Chapel Hill International Consensus Conference [2]. Cardiac manifestations are very rare, and represent only 0.6% in an Iranian series [2], and 6% in a French series [8]. Cardiac manifestations include myocarditis, pericarditis, endocarditis with sometimes severe valvular involvement, intracardiac thrombus, myocardial infarction, endomyocardial fibrosis and cardiac aneurysm [2, 5, 7, 8].

Some cases of cardiomyopathy have also been described [2, 6, 7, 10]. The diagnosis of BD is essentially based on clinical criteria because there is no reliable biological test and it associates the presence of bipolar aphtosis with skin lesions (erythema nodosum, pseudo-folliculitis, pustules, acneiform nodules) with ocular lesions (Anterior uveitis, posterior uveitis, cellular infiltrate in the vitreous body, retinal vasculitis) to a positive pathergic test within 24-48 h [1, 11]. Recently, new international classification criteria assigning points to each of the manifestations were published in 2013 and include: oral aphthosis (2 points), genital aphthosis (2), ophthalmological involvement (2), cutaneous (1), central neurological (1), vascular (1), pathergic test (1). A patient with a score of at least 4 is classified as having Behcet's disease with a sensitivity of 94.8% and a specificity of 90.5% [11]. Our patient had a clinical score of 7 points. She also presented biventricular large thrombi located on ventricular apex. She had bilateral distal embolism due to presence of multiple thrombus on apical site of right ventricular complicated with pulmonary infarction which infected. Our patient had dilated cardiomyopathy with reduced systolic function. The presence of dilated cardiomyopathy in MB is very rare, it is either related to an ischemic or inflammatory component; inflammatory damage is explained by the action of pro-inflammatory cytokines on left ventricular function. Clinically, it is manifested by systolic or diastolic heart failure, sometimes it is asymptomatic systolic or diastolic dysfunction [2, 4, 6, 7]. The treatment is based on the administration of colchicine, immunosuppressor; corticosteroid therapy is the cornerstone of treatment for Behcet's disease flares [1, 2, 8]; in the event of thrombosis, treatment is based on the initiation of anticoagulant treatment [1, 2, 8, 10, 11]. Our patient was put on oral anticoagulant, heart failure treatment indicated by European society of cardiology. The corticoherapy was initiated. The prognosis depends on the presence or absence of complications, it is often favorable under treatment and seems to be more severe in young subjects [1, 10, 12].

Conclusion:

Behcet's disease is a multisystemic vasculitis of unknown etiology that occurs in countries along the ancient Silk Road. It affects the young subject between 30 and 40 years old, with a male predominance. Cardiovascular damage is rare but makes the prognosis pejorative. The diagnosis is clinical and is based on a clinical score. The treatment is based on anti-inflammatories and immunosuppressants, corticosteroid therapy is the cornerstone of the treatment of flare-ups, anticoagulation is required in the event of venous or arterial

thrombosis, in the event of heart failure the treatment must follow the recommendations of the learned societies.

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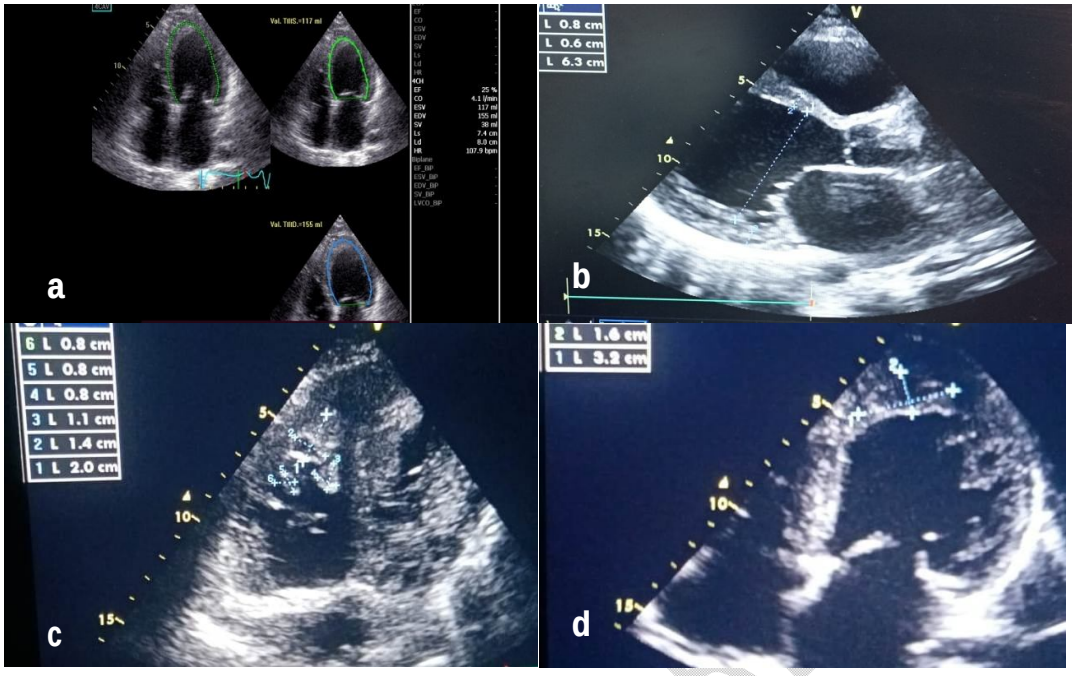

Figure 1 : Transthoracic echography 2D initial evaluation : a) : Severe left ventricle dysfunction by automatical systolic ejection fraction ;b) :large dilatation of left ventricle ; c):Magma of thrombus in right ventricule apex; d): adherent thrombus in left ventricle apex

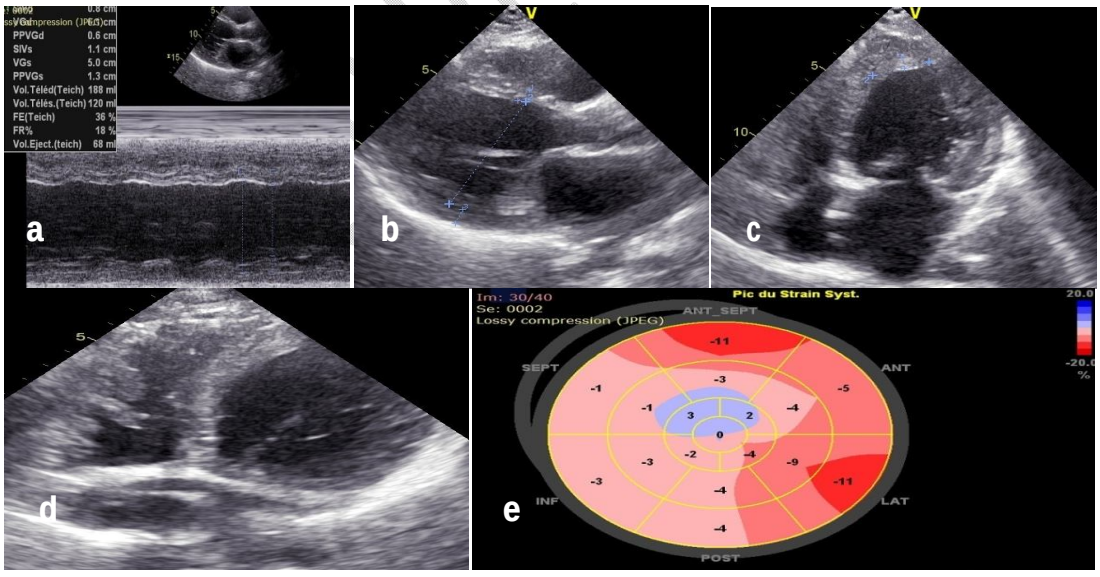

Figure 2: Control 2D TTE after 3 months: a and b: Persistence of left ventricle dilatation; c) and d): large regression of ventricular thrombus; e): severe alteration of global longitudinal strain