Acute coronary syndrome revealing a Behcet's Syndrome in a young man

Syndrome coronarien aigu révélant un syndrome de Behçet chez un jeune hommeà propos d'un cas

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Summary

Behcet'syndrome is a chronic and rare multisystemic vasculitis with uncommon coronary involvement. This is a case of a 37 years old male patient, without past medical history or any atherothrombosis risk factors, admitted for an anterior STEMI. Coronarography has showed an aneurysmal LAD coronary artery with a fresh floating thrombus. Physical examination showed bipolar ulcers (Oral and anal), eye and oral dryness, and inflammatory arthralgias with a positive pathergy test. Thus, an angioBehçet was diagnosed.

Our case shows an unusual aetiology of coronaropathy in young patients

Keywords

Acute coronary syndrome; Behçet's syndrome; Young patient

Résumé

Le syndrome de Behçet est une vascularite multisystémique chronique avec atteinte coronaire exceptionnelle. Il s'agit d'un patient de 37 ans, sans ATCDS ni facteurs de risque cardiaques, admis pour un IDM antérieur . La coronarographie a révélé un anévrisme thrombotique de l'IVA. L'examen physique a révélé des ulcères bipolaires (buccaux et anaux), une sécheresse oculaire et buccale, ainsi que des arthralgies inflammatoires, avec un test de pathergie positif. Un angio-Behçet a été donc diagnostiqué.

Notre cas illustre une étiologie inhabituelle de coronaropathie chez les patients jeunes.

Mots-clés

Syndrome coronarien aigu; Syndrome de Behçet; Sujet jeune

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INTRODUCTION

Behcet's Syndrome (BS) is a chronic and rare multisystemic vasculitis characterized by recurrent oral and genital ulcers, ocular inflammation, skin lesions and frequent articular involvement (1). Vascular involvement in BS is reported between 8-39% in different series and can affect both arteries and veins of any diameter (2, 3). In fact, arterial involvement in BS is far less common than venous involvement. Vasculitis may manifest as an aneurysm, thrombosis or occlusion. The frequently involved major arteries are the abdominal and thoracic aorta and pulmonary, iliac, and femoral arteries. However, coronary arteries are rarely affected and are only reported as case presentations in the literature (4, 5). Here, we present a case of myocardial infarction due to BS

PATIENT AND OBSERVATION

Herein, we report a case of a 37 years old male patient, without any particular past medical history, who was admitted to our cardiology department for acute and constrictive chest pain evolving for 4 hours. Electrocardiography (ECG) showed an ST-segment elevation of about 5 mm in anterior leads and mirror images in inferior leads. The diagnosis retained was a noncomplicated anterior ST-elevation myocardial infarction (STEMI). Primary angioplasty was performed. Coronary angiography showed an aneurysmal anterior interventricular artery with a fresh thrombus floating in the proximal part with a TIMI3 flow (figure 1).

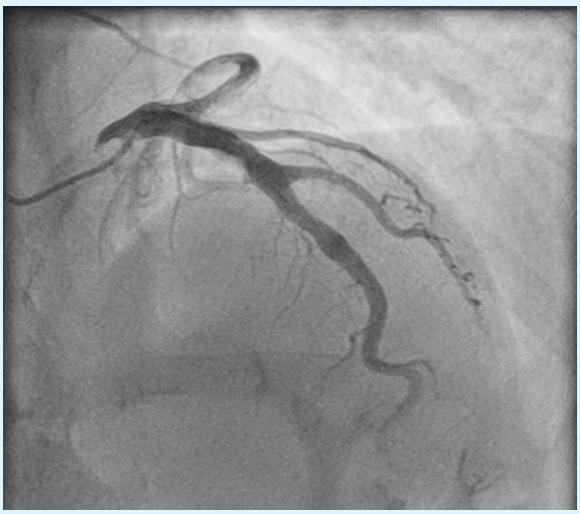

Figure 1 . Coronary angiography performed at admission: Aneurysmal anterior interventricular artery with a fresh thrombus floating in the proximal part.

The patient received double antiplatelet therapy and Heparin at a curative dose. Etiological investigation did not reveal other atherothrombosis risk factors such as diabetes mellitus or dyslipidaemia. Also, the patient didn't have any smoking history. Careful questioning and physical examination revealed bipolar ulcers (oral and anal) with eye and oral dryness, and inflammatory arthralgias. Therefore, the Behcet Syndrome was suspected. Ophthalmic exam showed no signs of uveitis on eye fundus. Thoracoabdominopelvic multislice computed tomography scan did not draw evidence of other arterial or venous involvement such as aneurysms or occlusions. The pathergy test was performed, returned affirmative. HLA typing was found positive for HLA-B51.

Given this body of evidence and based on the International Study Group for BS Diagnostic Criteria, the diagnosis of angio-Behçet was retained (6).

Two weeks post-STEMI, control coronary angiography showed disappearance of the thrombus on the anterior interventricular artery (figure 2).

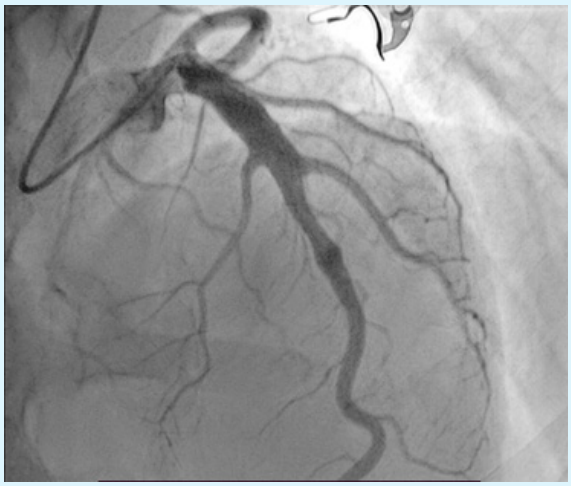

Figure 2 . Coronary angiography performed after 2 weeks: disappearance of the thrombus on the anterior interventricular artery.

The evolution was favorable and the patient was discharged on double antiplatelet therapy (acetyl salicylate and clopidogrel), Bisoprolol, Perindopril, Rosuvastatin and Colchicin.

DISCUSSION

BS is a chronic multisystem autoinflammatory condition and considered the only systemic vasculitis involving both arteries and veins in any sizes. Vascular involvement has been reported in 7.7 to 38% of BS cases, with mortality

reaching up to 20% in severe cases (7).Arterial involvement in BD is far less common than venous involvement (20% vs. 80%).

As for cardiac manifestations, it may include pericarditis, myocardial (diastolic and/or systolic) dysfunction, valve damage and intracardiac thrombus. Also, one patient may present several cardiac manifestations (8). However, vasculitis rarely involves the coronary arteries and only few case presentations are reported in the literature (9). Coronary artery involvement is very important because it affects young subjects as is the case in our patient and often presents as acute coronary syndrome. The majority of published cases have reported an acute coronary syndrome in patients previously diagnosed with BS. However, in our case, acute myocardial infraction was the initial presentation revealing the disease (10).

As for the management of acute myocardial infraction associated to BS, the definite treatment is not clear yet. Though, the use high dose of corticosteroids and immunosuppressives, in addition to thrombolytic agents and primary percutaneous coronary intervention has been reported (11, 12). However, surgical treatment using bypass grafting is a procedure with high mortality for patients with BS, especially in the acute period, as shown in a study conducted by Vural and al. (5). In fact, surgical treatment should be reserved for critical vascular lesions including aneurysms with fast progression and high risk of rupture (13).

CONCLUSION

Behcet Syndrome is a multisystem autoimmune vasculitis. Its vascular involvement is associated with a high mortality and poor prognosis, leading to the formation of an aneurysm or stenosis. However, coronary involvement is extremely uncommon, and most of published cases reported acute myocardial infraction in a patient previously diagnosed with BS. Our case report sheds light on an important aetiology of acute myocardial infraction in young patients without conventional risk factors atherosclerotic coronary artery disease.

Knowing this ethology can change the therapeutic approach an determine the prognosis of the patient.

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