

Left main coronary artery thrombosis revealing angio-Behçet syndrome

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Although acute myocardial infarction commonly results from coronary atherothrombosis, there are several other etiologies that should be taken initially into account, especially in young adults without significant atherosclerotic risk factors. Thrombophilia and coronary arteritis are, in this context, examples of etiologies that should be looked after. Through this article, we present a case of Behçet's disease with arterial involvement diagnosed after myocardial infarction resulting from thrombosis of the left main coronary artery in a 38-year-old young man without any particular past medical history.

Keywords:

behçet's disease, left main coronary artery, ST-elevation myocardial infarction, thrombolytic therapy, thrombosis

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Introduction

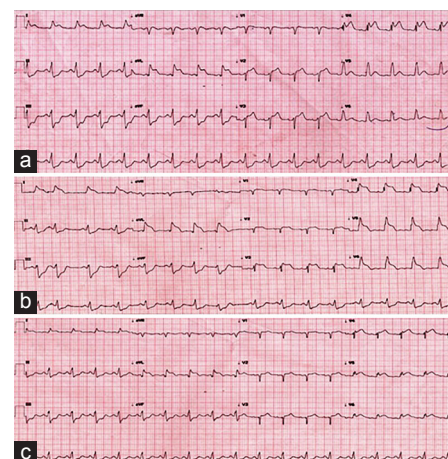
Behçet's disease (BD) represents a frequent systemic vasculitis in Japan and Mediterranean basin. It commonly associates buccal and genital ulcers with cutaneous, ocular, neurological, vascular, and gastrointestinal lesions. Although vasculitis represents the anatomic substratum of BD, its presence is not necessary to make diagnosis. In such disease, the incidence of vasculitis varied from 7 to 29% [1], with marked predominance of venous lesions [2]. Coronary involvement was rarely reported [3–5]. Literature data in this regard are scarce; hence, real incidence of coronary occlusion in BD remains unknown. Our article illustrates the management of angio-Behçet, diagnosis of which was made following ST-elevation myocardial infarction (STEMI) secondary to left main thrombosis.

Case report

We report the case of a male patient aged 38 years, without any particular past medical history and having active smoking (10 pack-years) as the only atherosclerosis risk factor. This patient presented in August 2011 with acute and constrictive chest pain evolving since 3 h and highly evocative of infarction scene. Physical examination noted initially stable hemodynamic status without signs of heart failure. On ECG (Fig. 1), ST-segment elevation of about 5 mm in anterior leads and mirror images in inferior leads were noted. This presentation was consistent with the diagnosis of a noncomplicated anterior STEMI seen at the third hour. Given the fact that primary angioplasty was not available at

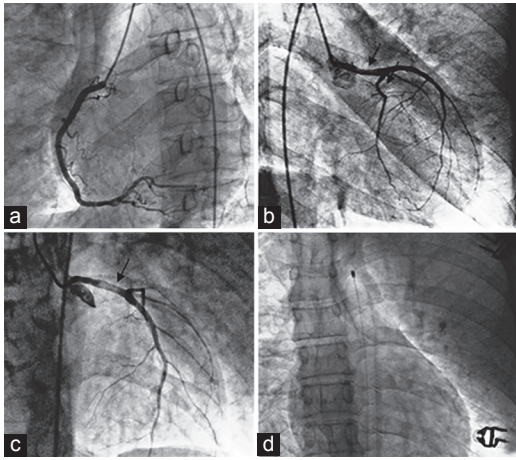
that hour, we immediately opted for pharmacologic revascularization by streptokinase. After fibrinolysis, clinical and electric success criteria were obtained. The course was marked by setting of cardiogenic shock and nonsustained ventricular tachycardia. On transthoracic echocardiography performed in extreme urgency, there were not any mechanical complications. Transthoracic echocardiography showed, however, extended akinesis of the anterior and lateral walls and of the apex without intraventricular thrombus. Left ventricle ejection fraction was estimated at 25%. At the fourth hour and 30th minute, the patient underwent emergency coronary angiography and intra-aortic counterpulsion balloon. The former showed a fresh thrombus floating in the distal part of the left main with TIMI3 flow

Figure 1



ECG at the acute phase of myocardial infarction. (a) Elevation of the ST segment in anterior leads (beginning of fibrinolysis). (b) Forty minutes after the beginning of fibrinolysis. (c) Fifty minutes after the termination of fibrinolysis: electric criterion of success.

Figure 2

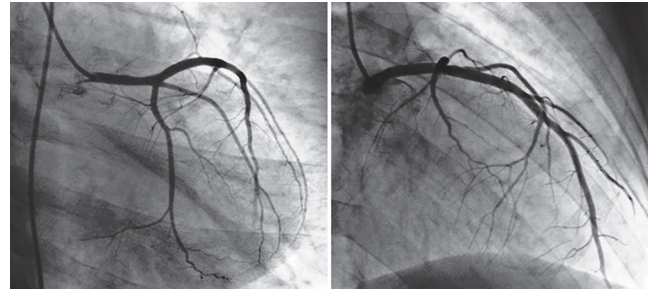


Coronary angiography at the acute phase of myocardial infarction. (a) Angiographically 'healthy' right coronary artery. (b, c) Distal left main coronary artery thrombosis with TIMI3 flow. (d) Intra-aortic counterpulsation balloon.

(Fig. 2) and angiographically healthy coronary arteries. The patient was put on anti-glycoprotein IIb–IIIa (anti-GpIIb–IIIa) (tirofiban) during 48 h. At the fifth hour following fibrinolysis, peak creatine phosphokinase and troponine Ic amounted, respectively, for 8353 IU/l and 322 ng/l. C-reactive protein and erythrocyte sedimentation rate amounted, respectively, for 82 mg/l (first day of STEMI) and 94 mm/h (second day postmyocardial infarction). Dobutamine and intra-aortic counterpulsation balloon were withdrawn on the fourth day postmyocardial infarction.

On the seventh day post-STEMI, control coronary angiography showed angiographically healthy and patent coronary arteries (Fig. 3). Etiological investigation did not reveal other atherothrombosis risk factors such as diabetes mellitus or dyslipidemia. Similarly, screening of hyperhomocysteinemia and of anticardiolipin antibodies, anti- β 2 microglobulin-1 antibodies, and protein S, C, and antithrombin deficiencies was negative. Careful questioning and physical examination revealed bipolar ulcers (buccal with a scrotal scar consistent with a sequel of scrotal ulcer), which were highly evocative of BD; hence, it was associated with a positive pathergic test and inflammatory arthralgias. Thoracoabdominopelvic multislice computed tomography scan did not draw evidence of other arterial or venous involvement such as aneurysms or occlusions. There were no signs of uveitis on eye fundus. Given this body of evidence, the diagnosis of angio-Beçet was retained. As a consequence, the patient received, in addition to anti-ischemic medications, colchicine 1 mg/day and boli of solumedrol for 3 consecutive days. After that, he was put on oral corticotherapy (prednisone) with monthly boli of cyclophosphamide. The patient was, moreover, under double antiplatelet therapy (acetyl salicylate

Figure 3



Control coronary angiography: patent and angiographically 'healthy' coronary arteries.

and clopidogrel), bisoprolol, furosemide, enalapril, and atorvastatin. Clinical course was good; the patient did not develop signs of left heart failure, and repeated echocardiographic controls showed improved and stable left ventricle ejection fraction at 40%.

Discussion

The incidence of vascular involvement in BD varies from 7 to 29% [1]. Cardiac involvement is present in 0.14% of patients [1]. Intraventricular thrombosis, regurgitant valvular heart diseases, coronary arteries aneurysms, and myocardial infarction are some of the reported manifestations of cardiac involvement in such disease [6]. Sporadic cases of granulomatous endocarditis, myocarditis, pericarditis, aortic aneurysms, and conductive tissue disorders were also reported during BD [5].

In such a disease, arterial involvement is less frequent than venous lesions. However, it is markedly more dreadful, as it was correlated to an important morbimortality. There are two types of arterial manifestations: occlusive or more commonly aneurismal lesions, which were noted in 1.5–2.2% of patients [7]. Occlusive and stenotic lesions involve not only great vessels, but also vasa vasorum. Frequently involved arteries in decreasing order are pulmonary artery, femoral, popliteal, subclavian, and carotid arteries. Coronary arteries, particularly the left main coronary artery, are rarely involved. Three forms of coronary arteries involvement during BD were reported: stenosis, thrombosis, and pseudoaneurysms. There are two mechanisms to explain thrombosis during angio-Beçet: vasculitis and hypercoagulable state. BD's vasculitis involves usually all layers of a vessel and is characterized by the presence of lymphocytic infiltrate during the acute phase. At an advanced stage, an important fibrotic and scarring reaction develops [8,9]. Hypercoagulable state observed during BD is thought to be due to inhibition of fibrinolysis on one hand and due to increased platelet aggregation on the other

hand. The latter had been explained by endothelial dysfunction leading to increased production of von Willebrand factor and decreased levels of prostacyclin (PGI₂) [9,10]. Increased production of fibrinogen and factor VIII could also represent other etiological factors explaining hypercoagulable state [11]. In our case, both hypercoagulable state and vasculitis seemed to be involved in coronary occlusion.

In a young patient with almost no significant risk factors of atherosclerosis, etiologies of acute coronary syndrome are, amongst others, coronary embolism, spasm or dissection, and also anomalous coronary arteries. Attention will be focused toward coronary arteritis when there is a particular context of systemic involvement. In our patient, thrombophilia was highly suspected, given angiographically healthy coronary arteries associated with important thrombotic burden in the left main. As biochemical assays performed to search for hyperhomocysteinemia and protein C, S, and antithrombin deficiencies were normal, the discovery of valuable signs evocative of BD allowed us to retain the diagnosis of angio-Behçet according to the reported criteria by the international study group for BD [12].

As cases are scarce, optimal therapeutic alternative in case of coronary thrombosis during BD remains unknown. Some authors suggested, however, that primary angioplasty was the most recognized revascularization option during STEMI secondary to BD's coronary thrombosis [13–15]. Nevertheless, long-term results after successful angioplasty and stent implantation remain unknown [16]. According to Ando *et al.* [17], coronary bypass grafting was not considered the best therapeutic option, as it was associated with postoperative coronary pseudoaneurysms. In our patient, in whom BD was unknown at admission, pharmacologic revascularization was undertaken at the acute phase of STEMI and was effective as demonstrated by the clinical and electric criteria on one hand and by angiographic findings (TIMI3 flow) on the other hand. Given the marked thrombotic burden in the left main, anti-GpIIb–IIIa (tirofiban) was given. Similarly, other successful cases of fibrinolysis were reported [18] in addition to efficiency of anti-GpIIb–IIIa [19], which was, in preference, indicated in case of fresh thrombus floating in coronary arteries without delayed flow or need for emergent revascularization. Moreover, according to the case reported by Ergelen *et al.* [20] illustrating left main thrombosis with stable hemodynamic status in a patient suffering from BD, successful fibrinolysis followed by the use of anti-GpIIb–IIIa had proved effective as demonstrated by further coronary angiography showing patent coronary arteries. Because of the arterial involvement, BD was considered severe in our patient justifying initial

intravenous corticotherapy followed by oral corticoids and cyclophosphamide. Nevertheless, although steroidal anti-inflammatory drugs could spare the patient from further outbreaks, such treatment could not only accelerate atherosclerosis process, but also increase the risk for cardiac decompensation. Hence, although the immediate course appears to be good, medium and long-term prognosis remain reserved.

Acknowledgements

Conflicts of interest

There are no conflicts of interest.

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