

Acute Myocardial Infarction as the Initial Presentation of Behcet's Syndrome

Abstract

Behcet's disease (BD) is clinically characterized by recurrent oral and genital ulcers and ocular symptoms. However, systemic manifestations involving the lungs, gastrointestinal tract, kidneys, and heart are well known. Acute myocardial infarction (MI) is a rare manifestation of Behcet's syndrome and usually results from coronary arteries vasculitis, leading to aneurysms. More uncommon is the occurrence of acute MI as the initial presenting symptom. Hereby, we report a case of a young male who presented with acute anterior wall MI sans any other conventional risk factors with a normal angiogram. And 3 years later, he was diagnosed with BD.

Keywords: *Angiography, anterior wall infarction, corticosteroids, mucosal lesions*

Introduction

Behcet's disease (BD) is a widespread vasculitis, first described by a Turkish physician Hulusi Behcet in 1937.^[1] The pathology of BD is an inflammatory response in the arteries and veins. Some patients of this disease manifest only skin and mucosal lesions, while others may manifest life-threatening central nervous system and gastrointestinal involvement and pulmonary artery aneurysms.^[2] Cardiac involvement is uncommon and can be seen in the form of intracardiac thrombus, endocarditis, myocarditis, pericarditis, endomyocardial fibrosis, coronary arteritis, myocardial infarction (MI), and valvular disease.^[3] The prognosis of cardiac disease is unfavorable than that of other organs involved in BD. Acute MI as the primary presentation of BD is very uncommon, as found in our case.

Case Report

A 20-year-old young male presented to the emergency department with a history of acute-onset severe retrosternal chest pain for the past 6 h. On first examination in the emergency department, his blood pressure was 100/70 mmHg and heart rate was 90 beats/min. The patient looked pale and apathetic. Systemic examination did not reveal any abnormality in cardiovascular as well as respiratory system.

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His 12-lead electrocardiogram revealed sinus rhythm with ST-segment elevation in leads V1–V6, I, and aVL and isoelectric ST-segment in leads II, III, and aVF [Figure 1]. Two-dimensional echocardiography demonstrated regional wall motion abnormality in the apical, anterior wall; distal interventricular septum; and lateral wall with an ejection fraction of 37%. A diagnosis of anterior wall MI was entertained, and immediate thrombolytic therapy was initiated. Streptokinase was administered as an intravenous infusion (1.5 million IU) over 45 min. The patient's clinical symptoms improved 1 h after thrombolysis, and his electrocardiogram showed ST-segment resolution. The patient's biochemical parameters were within normal limits. On further history taking, the patient denied any history of cigarette smoking, tobacco chewing, family history of coronary artery disease and neither was there any suggestion of any recurrent oral ulcer, genital ulcers, uveitis or skin abnormality, or any systemic illness in the past. On the following day, the patient was taken up for routine coronary angiography as a part of pharmaco-invasive therapy. Coronary angiogram done by transradial route revealed the absence of significant plaque in any of the coronary arteries, and a possibility of recanalization of the left anterior descending artery was entertained [Figure 2]. The patient's in-hospital course was uneventful, and he

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was discharged on dual-antiplatelet therapy (aspirin and clopidogrel), statins, beta-blockers, angiotensin-converting enzyme inhibitors, mineralocorticoid receptor antagonist, and loop diuretics. The patient was asymptomatic and was on guideline-directed medical therapy during routine follow-up.

Three years later, he presented to the outpatient department with a history of oral ulcers, genital ulcers, blurring of vision, and marked papulo-pustular lesion all over the torso for the past 10 days [Figures 3 and 4]. On taking a detailed history, he confirmed that previously he had three episodes of similar oral ulcers in the last 3 months for which he did not take any treatment as the ulcers had healed spontaneously. We referred the patient for rheumatological as well as ophthalmological examination. Slit-lamp examination of the anterior chamber of the eye confirmed anterior uveitis. A battery of investigations for vasculitis workup revealed normal anti-nuclear antibody, extractable nuclear antigen, and complement and other markers. The pathergy test was affirmative, and based on the International Study Group for BD Diagnostic Criteria,

a diagnosis of Behcet's syndrome was made.^[2] Owing to ocular involvement, the patient was put on high-dose oral corticosteroid therapy to which he responded very well. Currently, the patient is symptom free with no new episode of acute coronary syndrome.

Discussion

Behcet's is a multisystem vasculitis with the triad of oral ulcer, genital ulcer, and uveitis. Vascular involvement is seen in 7%–38% of cases with a high mortality in severe affliction.^[4,5] Interestingly, there is a greater predilection of the venous system compared to arterial involvement. Cardiac involvement can be life-threatening and has been reported variably anywhere between 1% and 16%. However, disease manifesting first time with cardiac manifestation is rare.

Although BD can involve all types of vessels, coronary involvement is extremely uncommon (0.5%–1.0% in autopsy series), with literature of acute MI in BD primarily

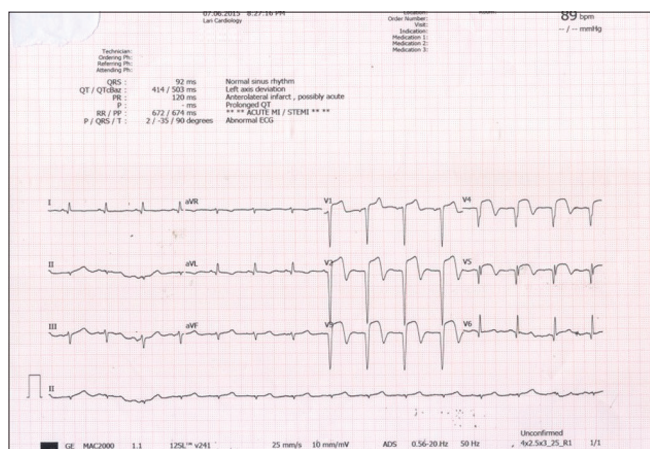


Figure 1: A 12-lead electrocardiogram showing, normal sinus rhythm, ST-segment elevation on V1–V5, and isoelectric ST-segment in leads II, III, and aVF

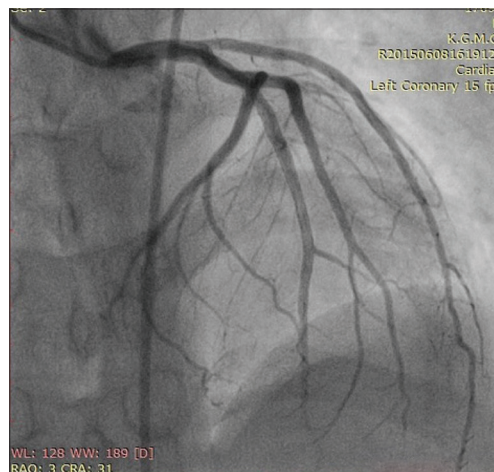


Figure 2: Coronary angiogram in anteroposterior view with cranial angulation demonstrating the absence of any significant atherosclerotic plaque in the left anterior descending artery

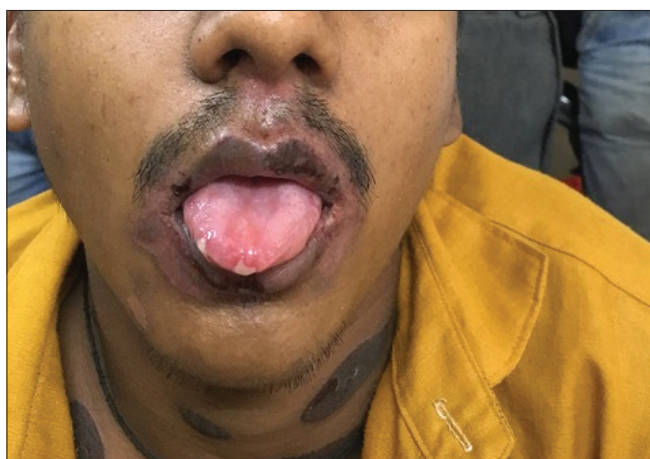


Figure 3: Two mucosal ulcers at the margins of the tongue

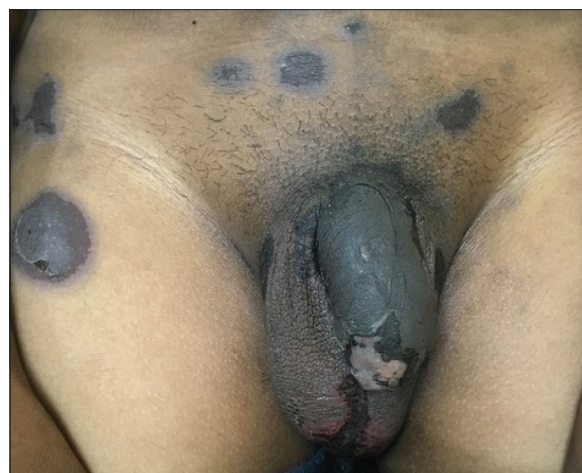


Figure 4: Multiple papulo-pustular lesions are visible over the torso. Also seen are ulcers over the glans penis and scrotum

restricted to case reports.^[6] Vasculitis of the coronary artery can lead to the formation of aneurysm or stenosis and the former is more common.^[7,8] The aneurysms can predispose to thrombosis and even rupture, leading to pseudoaneurysm. The thrombosis associated with BD may be due to vascular endothelial cellular dysfunction and/or a reduction of systemic fibrinolytic activity and a rise in fibrinogen and factor VIII, but the precise mechanism of thrombosis is not known.^[4,5] Factor V Leiden mutation, protein C deficiency, protein S deficiency, and antiphospholipid antibodies are also postulated as additional factors for thrombosis. High-dose corticosteroids used in the management of the disease can also accelerate atherosclerosis.

Interestingly, majority of the published cases have (at least 30) reported acute MI in a patient previously diagnosed with Behcet's disease or rarely, the diagnosis was made during an acute episode. However, in our case, acute MI was the only and initial presentation. The systemic manifestations appeared only years later. Second, the angiogram was remarkable in the absence of aneurysm or stenosis unlike other cases. Coronary microvascular dysfunction and intimal thickening with plaque erosion could be the pathologies responsible in such cases.

In BD, although vasculitis rarely involves the coronary arteries, coronary involvement is especially important because it affects young individuals, and it often presents as acute coronary syndromes. Other reported cardiac manifestations include occlusion of the subclavian artery, aortic arch syndromes, pericarditis, myocarditis, conduction abnormality, ventricular aneurysm, endomyocardial fibrosis, mitral valve prolapse, mitral and aortic valve insufficiency, and pulmonary arterial hypertension. The overall prognosis of BD with cardiac involvement is poor, with 1-year mortality ranging from 2% to 4%. Ocular involvement is the most common cause of morbidity with vascular involvement being the most common cause of mortality.^[5]

Our case report sheds light on an important aspect in a young patient with acute MI who lacks conventional risk

factors for atherosclerotic coronary artery disease. A search for the underlying immunological disorder should be made and these patients should be followed up closely as was done in our case (delayed manifestation of underlying immunological disorder).

Consent

Appropriate informed consent was obtained for publishing audio-visual data. All possible attempts have been made to conceal the identity of the subject/patient.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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