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Acute Myocardial Infarction in Behcet's Disease: A Case Report

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Abstract **Case Report**

Background: Behcet's disease is an autoimmune disease characterized by diffuse vasculitis and oral and genital ulcers. Coronary vasculitis is a rare finding in Behcet's disease and only reported in few cases. ***Case summary:*** A 47-year-old male patient was recently diagnosed as Behcet's disease and admitted as inferior ST elevation myocardial infarction. Coronary angiogram revealed coronary aneurysm in posterior descending artery with TIMI III flow. The patient was managed medically with immunosuppressive therapy and he discharged after 5 days in a stable condition. Patient was seen in the clinic after one month and he was asymptomatic. ***Conclusion:*** Coronary involvement in patients with Behcet's disease is rare; however, it can be associated with myocardial infarction. Medical management with steroid and immunosuppressive therapy is the mainstay of therapy. Coronary intervention might be required in some cases with ongoing ischemia.

Keywords: Behcet's disease, Myocardial infarction, Coronary aneurysm, coronary angiography, Immunosuppression.

INTRODUCTION

Behcet's disease is an autoimmune disease characterized by mucocutaneous, articular, vascular, ocular, gastrointestinal, and neurologic manifestations. Vascular involvement in Behcet's disease is reported to be between 8%-39% in different series and can involve both arteries and veins of any diameter. Vasculitis in Behcet's disease affects venous system more than arterial system. The frequently involved major arteries are the abdominal and thoracic aorta, pulmonary, iliac, and femoral arteries [1]. However, coronary vasculitis is reported rarely in Behcet's disease and can lead to coronary stenosis, aneurysm or thrombosis [2]. No definite treatment is available for patients with coronary vasculitis but the main treatment is anti-inflammatory and immunosuppressive therapy. Interventions in the form of percutaneous coronary intervention with cover stents and coronary artery bypass grafting are done sporadically based on the patient's presentation [3].

CASE PRESENTATION

A 47-year-old male patient was recently diagnosed as Behcet's disease after a 6-month history of oral and genital ulcers associated with low grade fever and weight loss. He was started on colchicine 0.5 mg twice daily and clinically improved. Then he presented to emergency department of our institute with a 12h history of typical chest pain. He was vitally stable and clinical examination was unremarkable. Electrocardiogram (ECG) was done and revealed ST segment elevation in inferior leads (Figure 1) then subsequent ECG showed Q wave with mild residual ST segment elevation in inferior leads (Figure 2). The patient was shifted immediately to Cath lab and coronary angiogram done and revealed aneurysm in the posterior descending artery a branch of the right coronary artery (Figure 3). The flow was TIMI III and thus, no intervention done and decision was to treat him conservatively. Other coronaries were normal (Figure 4). The patient shifted to coronary care unit and rheumatology consultation done and they recommended to start him on pulse steroid and then shifted to oral steroid along with antiplatelet and statin therapy. Echocardiogram was done and revealed a low normal ejection fraction (EF) (EF was 52% by modified Simpson Method) with mild basal inferior wall hypokinesia. Speckle tracking echocardiogram confirmed the regional wall motion abnormality; however, his global longitudinal strain was normal at -22.1% (Figure 5). Patient condition improved and he was discharged from hospital 5 days later in stable condition. After one month, patient came and reviewed

in the clinic and he was totally asymptomatic and on oral prednisolone.

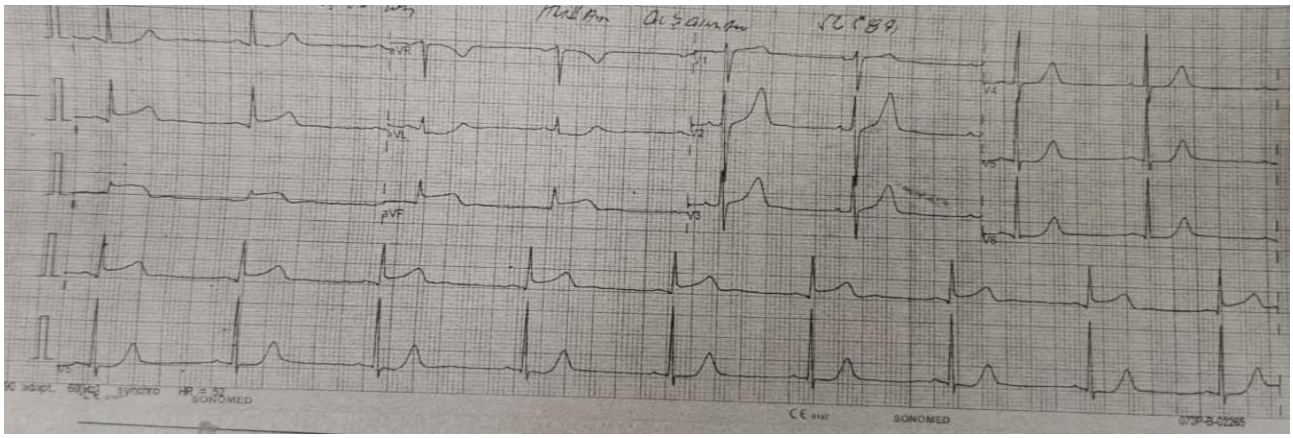

Figure 1: 12 lead surface ECG showing ST elevation in II, III, aVF.

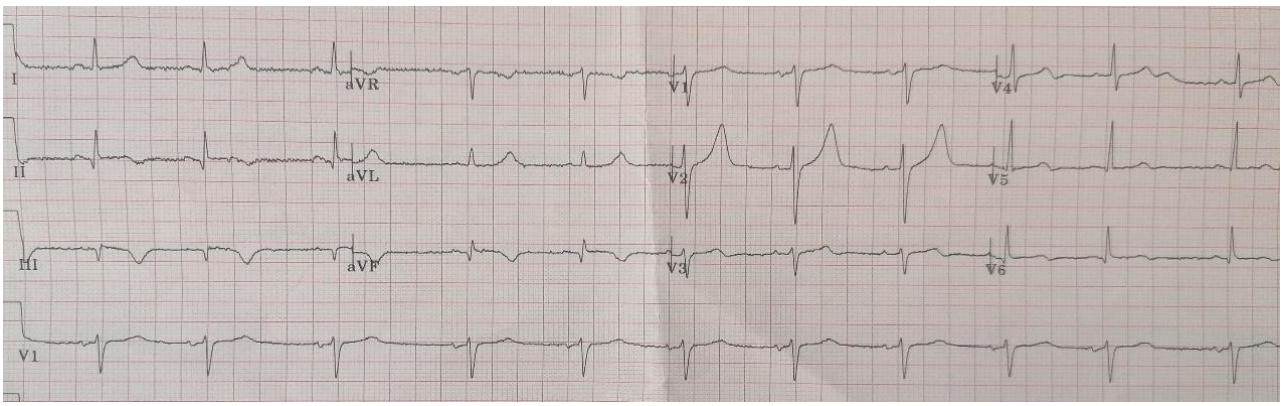

Figure 2: 12 lead surface ECG showing Q wave and residual ST elevation in II, III, aVF.

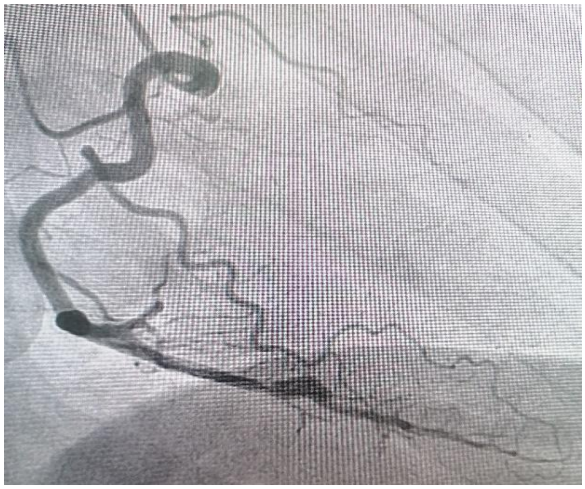

Figure 3: PDA of RCA with mid aneurysm with TIMI III flow.

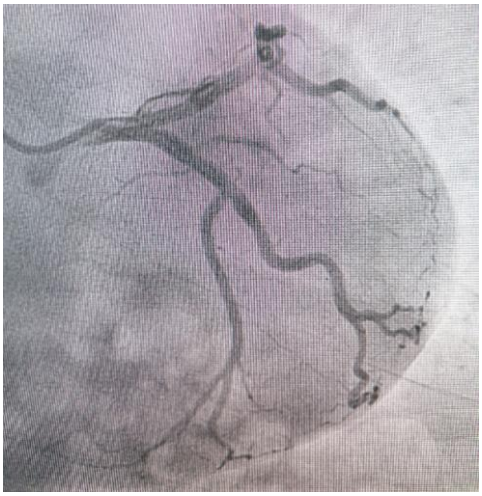

Figure 4: LAD and LCX are normal without significant lesions or aneurysm.

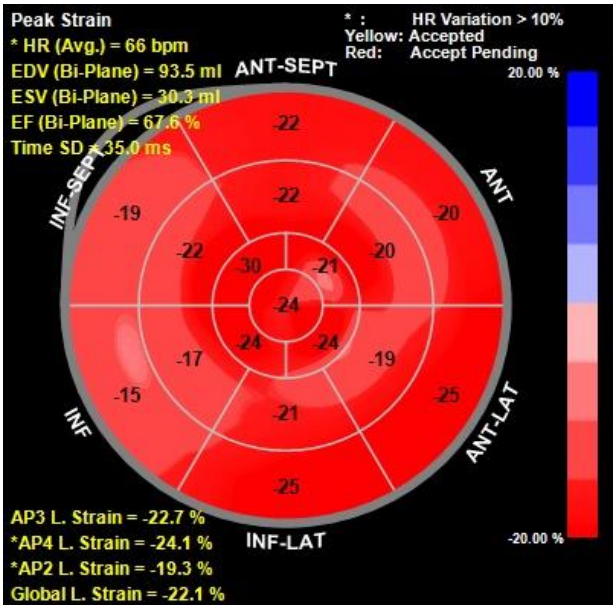

Figure 5: Bull's eye of global longitudinal strain showing lower regional strain @ basal inferior and inferoseptal walls.

DISCUSSION

Behcet's disease is an autoimmune disease characterized by multi-system involvement and vasculitis occurs in up to 40% of patients. It is reported that venous involvement is much more common than arterial disease in Behcet's disease [1]. Cardiac involvement occurs in about 6% of Behcet's patients and can affect any of cardiac structures. Pericarditis with or without effusion, myocardial injury, endocardial lesions and coronary occlusions can occur in patients with Behcet's disease [2]. Even more some patients develop cardiac manifestations before diagnosis of Behcet's disease [3, 4]. Coronary involvement is noticed more frequently in male in comparison to female as in our case. Mortality increased significantly in patients with Behcet's disease if they developed cardiac manifestations. Interestingly most of patients with cardiac involvement are relatively young with mean age: 29.7±9.9 years, therefor; it should be in the priority of differential diagnosis if young patient without cardiac risk factors presented with chest pain [2, 5]. All types of coronary syndromes ranging from chronic stable angina to non-ST segment elevation and ST segment elevation have been reported [3-7]. As seen in our patient, coronary aneurysms are more common than stenotic lesions in patients presented with picture of ischemic heart disease. Not only coronary aneurysm; ventricular and pulmonary aneurysm and aortic involvement have been seen [2, 8]. Histologically, inflammation occurs in media and adventitia leading to fibrosis and destruction of media and formation of saccular aneurysm [9]. Sometimes the risk of aneurysm formation increases in case of vascular injury that occurs during coronary intervention either percutaneous or surgical [4]. Aneurysm leads to stagnation of blood and formation of small thrombi that embolize distally and leading to myocardial ischemia with or without infarction. Diagnosis of myocardial infarction is based on troponin elevation plus evidence of myocardial ischemia (symptoms, ECG changes, Wall motion abnormality and Coronary angiogram) as shown in our case (4th universal definition of myocardial infarction) [11]. Treatment of coronary aneurysm is not unique and management is directed to restore diminished coronary blood flow and control the activity of the disease. In our case, distal coronary flow was not affected so no coronary intervention was required. However; sometimes it is necessary to do coronary intervention either percutaneous or even surgical to maintain blood flow and protect the myocardium. Immunosuppression using steroid, azathioprine and cyclophosphamide is the mainstay for stabilization of aneurysm and prevention of its progress but should be under supervision of rheumatologist. Pulse steroid is usually required as in our case to rapidly inhibit the inflammatory process and control the disease activity [3, 4, 6, 8]. We also treated the patient with anticoagulation (apixaban 5mg twice daily) as a study showed that complete remission was associated with the use of immunosuppressant, colchicine and anticoagulant therapy [2, 10]. Follow up is recommended for these patients for clinical evaluation and immunosuppressant dose adjustment.

CONCLUSION

Coronary involvement in patients with Behcet's disease is rare; however, it can be associated with myocardial infarction. Medical management with steroid and immunosuppressive therapy is the mainstay of therapy. Coronary intervention might be required in some cases with ongoing ischemia.

List of abbreviations

ECG: Electrocardiogram
TIMI: Thrombolysis in myocardial infarction

Conflict of interest

The authors declare that there is no conflict of interest regarding the publication of this article.

Consent

Written consent was obtained for submission and publication of this case report including images and associated text has been obtained from the patient in line with COPE guidance.

Ethical approval

The study was approved by our ethical committee.

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