Acute Myocardial Infarction in a Patient with Behçet’s Disease and Coronary Artery Aneurysm Rupture

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A 40-year-old man with a case of Behçet’s disease which lacked regular medical management was admitted under the diagnosis of acute myocardial infarction. Coronary angiography revealed multiple aneurysms along the left anterior descending and left circumflex artery. The critical lesion distal to the aneurysm of the left anterior descending artery was considered to be the culprit lesion of acute myocardial infarction. The patient underwent successful coronary bypass surgery, and subsequent surgical and pathological findings confirmed coronary artery aneurysm rupture.

Key Words: Acute myocardial infarction • Behçet’s disease • Coronary aneurysm • Interventional cardiology

INTRODUCTION

Behçet’s disease (BD) is a multisystem vasculitis that can involve vessels of all sizes, and is characterized by recurrent oral and genital ulcers with variable manifestations affecting the skin, eyes, central nervous system and musculoskeletal system. Only 3% to 6% of patients with BD have cardiac involvement, which includes conduction system abnormalities, pericarditis, myocarditis, coronary artery disease, myocardial infarction, congestive heart failure, valvular insufficiency, endomyocardial fibrosis, and intracardiac thrombus. Acute myocardial infarction (AMI) in patients with BD is rare; only about 30 cases of AMI associated with BD have been reported previously. Here, we present an unusual case of AMI in a young patient with BD and coronary artery aneurysm rupture.

CASE REPORT

This 43-year-old male patient visited our emergency department because of late night sudden onset of chest pain. The chest pain was severe, a squeezing sensation, located at the sub-sternal area, and radiating to the jaw (Canadian Cardiovascular Society, grade III). The associated symptoms included shortness of breath and palpitation. The patient had a history of BD, which had been diagnosed 10 years earlier, manifesting symptoms of recurrent aphthous ulcers, ulceration in the genital region and oral mucosa, and recurrent uveitis. His case was not routinely followed-up in our hospital due to challenges arising from patient mental retardation and poor family support.

He was afebrile, with blood pressure of 106/79 mmHg; tachycardia (100 beats per minute) with premature beats, tachypnea and diaphoresis were also noted. Additionally, patient breathing sounds revealed bilateral basal crackles, and the electrocardiogram (ECG) showed Q wave and ST elevation in leads V2 to V4 (Figure 1). The initial chest X-ray revealed mild pulmonary congestion. The laboratory data showed elevated myoglobin (417 ng/ml), creatine kinase (1438 U/L), creatine kinase-MB (192 U/L) and troponin I (12 µg/L).
Upon further review, the patient’s typical chest pain, ECG change and elevated cardiac markers confirmed the diagnosis of AMI, and he received prompt cardiac catheterization. Coronary angiography revealed multiple aneurysms along the left anterior descending (LAD) and left circumflex artery (LCX) (Figure 2A). The critical lesion distal to the aneurysm of LAD was considered to be the culprit lesion for AMI. Two other aneurysms were also noted at the middle portion of the LCX. The right coronary artery was patent without aneurysmal change. The complicated character of the coronary artery made primary coronary intervention impossible. Thus, surgical treatment was recommended due to severe coronary stenosis and the risk of aneurysm rupture.

Before surgery, echocardiographic imaging showed marked anterior, lateral and apical wall hypokinesis of the left ventricle. It was determined that there was an echogenic mass (3.1 × 5.5 cm) over the aortopulmonary window wrapped LAD. Minimal pericardial effusion in the posterior pericardial space was noted. ECG-gated computed tomography (CT) showed a mass of mixed density (4.5 × 5.5 cm) arising from the LAD, and it compressed the right ventricular outflow tract and pulmonary artery (Figure 2B). The mass was considered to be a giant LAD aneurysm with solid periphery representing thrombus formation. A penetrating ulceration with slight

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**Figure 1.** (A) The electrocardiogram in emergent department showed Q wave and ST elevation in leads V2 to V4. (B) The follow-up electrocardiogram 7 days later showed Qs pattern without ST elevation in leads V2 to V4.
Aneurysmal dilatation at the descending aorta was also noted (Figure 2C).

Surgery was then performed. The aneurysmal sac was opened and the thrombus was evacuated, whereafter closer examination of the wall confirmed a false aneurysm. One opening to the proximal LAD, and a second opening to the distal LAD were found. These two openings were sutured to avoid aneurysm enlargement. A sample of the sac was sent for histologic examination, and pathological report showed only thrombus without vessel wall involvement. The LAD was bypassed using the left internal thoracic artery and the LCX was bypassed using the saphenous vein graft. The size of the aorta with penetrating ulceration tapered from 3.1 cm to 2.4 cm. Thereafter, the surgeon applied 2 stent grafts (28 mm × 10 cm; 34 mm × 2.4 cm) to cover the ulceration (Figure 2D).

The patient was stable following surgery. 120 mg methylprednisolone was administered intravenously daily for BD, which was later adjusted to oral administration. Overall, the patient course of treatment after surgery was uneventful, and he was discharged later with instructions for follow-up at our outpatient department clinic.

**DISCUSSION**

BD is a systemic vasculitis of unknown cause with features of oral and genital ulcers and uveitis. This disease is prevalent in the Mediterranean region, the Middle East, and the Far East. It is well known as the “silk road disease” due to its epidemiology. Around 7% to 38% of patients have large vascular system involvement, especially the venous system. The involved vessels show aneurysmal change and may predispose to thrombosis or rupture. Coronary artery aneurysms secondary to BD are much rarer, and most cases are seen in young men without vascular risk factors. Patients may be asymptomatic or present with acute coronary syndrome. Our young patient had no cardiovascular risk factors, but he presented with a highly complicated case of BD vasculitis with coronary aneurysm rupture and ulceration of the descending aorta. The elevated C-reactive protein (8.4 mg/dl) and erythrocytes sediment rate (22 mm/hour) in our patient suggested that BD-related vasculitis might be the mechanism of the complicated vessel lesions.

The mechanisms of AMI vary in the population of BD patients. Some are caused by aneurysms which predispose to the formation of thrombus or rupture. Others may have aneurysms coexisting with atherosclerosis disease. Etiology of AMI in a patient with BD may be important with regard to treatment strategy. For example, high-dose corticosteroids may help to relieve symptoms and signs in a patient with BD, but such use accelerates atherosclerosis disease and delays the healing of injured myocardium. For our patient, the underlying mechanism of AMI could be external compression of the LAD by the pseudoaneurysm or thromboembolism of the LAD segment distal to the aneurysm, or both. However, the coronary angiography, cardiac echo, CT and surgical findings suggested the pseudoaneurysm with external compression might play a major role in the mechanism of AMI in this case. Thus, the present patient also re-
received high-dose corticosteroid to relieve BD.

Percutaneous coronary intervention (PCI) has been reported to treat a BD patient with AMI.\textsuperscript{6-8} However, it can be exceedingly difficult and quite high-risk to perform PCI across the coronary aneurysm in this type of case. Surgical treatment is reserved for critical vascular lesions including large size, rapid growing, acute or impending ruptured aneurysm.\textsuperscript{9} Besides, it is also imperative to investigate for other abnormalities of the aorta and its major branches, as we did with our patient. Our case demonstrated a successful surgical treatment in a BD case with complicated vessel lesions.

Pathologic examination of the affected vascular wall in BD patients can demonstrate an inflammatory obliterative endarteritis of the vasa vasorum causing destruction of the media and fibrosis, and thus weakening and predisposing the arterial wall to aneurysm formation that eventually ruptures.\textsuperscript{10,11} Unfortunately, we were unable to get such pathological findings in our case.

CONCLUSION

BD may cause systemic vasculitis and AMI. Appropriate imaging study is helpful for diagnosis and clarifying the extent of involvement. Properly tailored and efficacious treatment of BD patients with AMI is based on different clinical scenarios, and surgery is a reasonable option for those patients with symptomatic coronary aneurysms.

REFERENCES