Case report

Acute myocardial infarction due to sinus of Valsalva aneurysm in a patient with Behçet's disease

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ABSTRACT

Cardiovascular manifestations have been reported in 7-38% of patients with Behçet's disease (BD), and mortality occurs in up to 20% of those with marked vascular involvement. Sporadic cases of endocarditis, myocarditis, pericarditis, acute myocardial infarction, aortic aneurysm, ventricular thrombosis, congestive cardiomyopathy, and valvular dysfunction have been reported. Here we report a case of acute myocardial infarction that resulted from the compression of coronary arteries by a sinus of Valsalva aneurysm in a patient with BD.

Introduction

The prevalence of vascular involvement in Behçet's disease (BD) varies from 7-43% (1-3). Cardiovascular involvement in BD is extremely rare, but it sometimes leads to a poor prognosis, with mortality occurring in up to 20% of those with marked vascular involvement (4). Here we report a patient with a 3-year history of BD who presented with acute myocardial infarction resulting from compression of the coronary arteries by a sinus of Valsalva aneurysm.

Case report

A 45-year-old Korean woman with BD who had been experiencing intermittent severe chest pain for one month was admitted. Three years earlier she presented with recurrent oral ulcers, genital ulcers, erythema nodosum and acne-like skin lesions. BD was diagnosed based on the criteria of the International Study Group for BD (5) and the patient was placed on treatment with colchicine. She had no known risk factors for acute coronary syndrome (such as smoking, diabetes mellitus, hypertension, or dyslipidemia) and no previous history of congenital heart disease or Kawasaki disease.

On admission she had oral ulcers, but abnormal skin lesions and genital ulcers were not found. Laboratory tests showed leukocytosis and an elevated erythrocyte sedimentation rate (53 mm/hour). However, cardiac enzyme levels and serum lipid levels were within normal ranges, and anti-nuclear antibodies, anti-neutrophil cytoplasmic antibodies, lupus anticoagulant, and anti-cardiolipin antibodies were negative. HLA typing was not performed and the results of a pathergy test were negative. Plain chest radiography revealed mild cardiomegaly and electrocardiography showed no abnormalities.

Transthoracic echocardiography showed a normal left ventricular ejection fraction (76%), grade II aortic regurgitation due to incomplete coaptation of the aortic valve, no regional wall motion abnormalities of the left ventricle, and no evidence of congenital heart defects such as ventricular septal defect, bicuspid aortic valve and so on, which have been reported to be associated with the formation of aneurysms of the sinus of Valsalva (6, 7). Conventional coronary angiography revealed 80% stenosis of the proximal to middle segment of the left anterior descending artery (LAD) (Fig. 1). However, there were no significant luminal changes in response to nitrate stimulation and no atherosclerotic plaques at the stenotic lesion on intravascular ultrasonography.

A 3-dimensional computed tomography (3D-CT) cardiac scan showed an aneurysm originating in the left sinus of Valsalva with a maximal diameter of 35 mm that was externally compressing the proximal to middle segment of the LAD (Fig. 2A). After determining the cause of this sinus of Valsava aneurysm to be coronary arteritis with

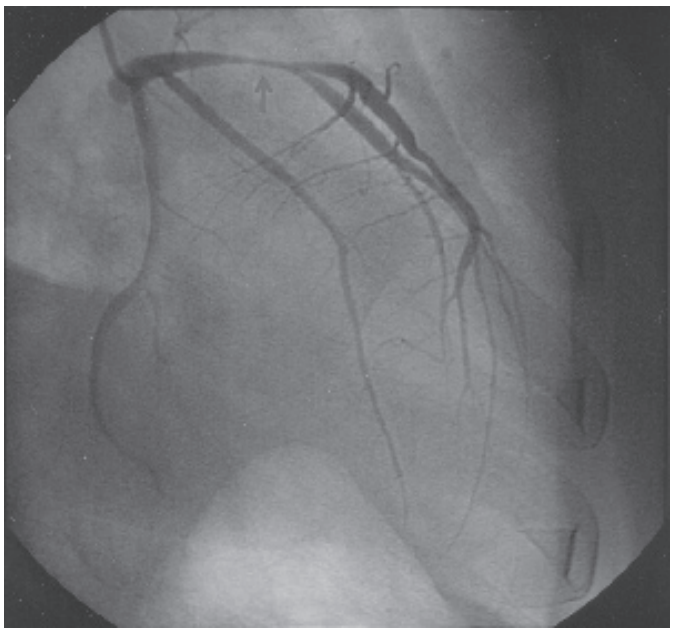
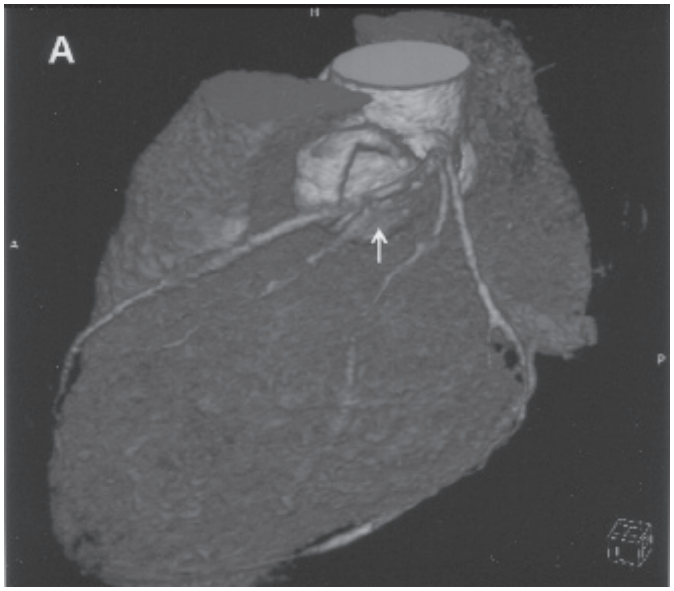


Fig. 1. Conventional coronary angiographic findings. Coronary angiography revealed 80% flattened stenosis of the proximal to middle segment of the left anterior descending artery without evidence of an atherosclerotic plaque.



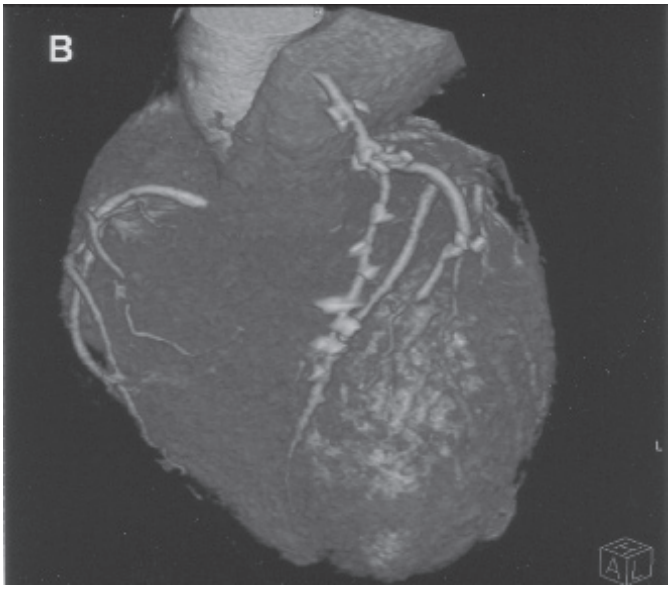


Fig. 2. Cardiac 3-demensional CT scan findings before and after surgery. **A**. Initial CT scan, revealing a huge aneurysm originating from the sinus of Valsarva compressing the left anterior descending artery. **B**. A follow-up CT scan showed a patent LAD pathway with a good flow graft pathway and thrombus formation in the previous aneurysm..

resultant external compression of the LAD, patch closure of the aneurysm and a coronary artery bypass graft (CABG) were scheduled and oral prednisolone was prescribed at an initial dose of 60 mg/day to prevent any postoperative complications.

On the 8th day of high-dose glucocorticoid treatment, the patient complained of typical acute chest pain and ST segment elevation was seen at the anterior pre-cordial leads of the electrocardiograph. Compared to levels before the development of chest pain, serum creatinine phosphokinase isoenzyme MB and cardiac troponin-T gradually rose from 0.9 to 5.0 ng/ml (reference range: 0~5.8) and from <0.01 to 0.05 ng/ml (reference range: 0~0.1), respectively. A diagnosis of acute myocardial infarction was made, and an emergency operation was performed. The diagnosis was confirmed with the development of new pathologic Q waves on electrocardiography after surgery (8).

A huge aneurysm from the sinus of Valsalva with a 8×15 mm opening that was filled with thrombi was discovered during surgery. Patch closure of the aneurysm with double velour and autologous pericardium and a CABG anatomizing the left internal mammary artery (LIMA) to the middle LAD were performed. Intravenous hydrocortisone 400 mg was given on the day of surgery (100 mg before and 300 mg after surgery). The dose was tapered by 50% per day for 2 days following surgery and oral prednisolone 60 mg per day was given afterward (9). Azathioprine was added for its additional immunosuppressive effect 4 days after surgery, and a coronary 3D-CT angiography performed 7 days after the operation showed an intact LIMA to middle LAD pathway and good flow from the LIMA to the LAD via the graft pathway (Fig. 2B). We gradually decreased the dose of prednisolone every 2 or 3 weeks to maintain normal ESR and CRP levels, and follow-up coronary 3D CT angiography performed 10 months after surgery showed a patent graft and thrombus formation in the previous aneurysm. The patient was placed on maintenance therapy with 5 mg/day of prednisolone and 100 mg/day of azathioprine, and

has remained asymptomatic with normal ESR and CRP levels (5 mm/hour and 1.5 mg/dL, respectively).

Discussion

BD can affect blood vessels of all types and sizes, but there is a higher risk of arterial lesions. Vascular involvement, which has been identified in 7.7% to 38% of BD patients, mainly affects the venous system and arterial involvement is unusual (1, 10). Data on the prevalence of arterial aneurysm in patients with BD is lacking, but a previous report showed the prevalence of arterial aneurysms and arterial occlusions to be similar, *i.e.,* 1-2% (11).

Here we present the unique case of a BD patient diagnosed with acute myocardial infarction. The patient had no risk factors for coronary artery diseases other than BD. Coronary angiography showed flattened stenosis of the LAD coronary artery, but there was no evidence suggesting the presence of an atherosclerotic plaque. On further evaluation, we found a huge aneurysm extending from the sinus of Valsalva that was externally compressing the LAD coronary artery and was thought to be the result of arterial inflammation.

Surgical intervention in the active inflammatory phase may lead to major complications, including suture line dehiscence, aorto-enteric fistulas, graft occlusion, and thromboembolic events (12-14). The patient in this report had recurrent oral ulcers at the time of admission, and a laboratory test showed elevated acute phase reactants. Haug *et al*. earlier demonstrated an association between inflammatory aortic aneurysm and autoimmune disease (15) and Schirmer *et al*. suggested that immune-mediated mechanisms are involved in the pathogenesis of abdominal aortic aneurysms (16). We did not perform a histological evaluation in this patient, but hypothesized that BD might have been involved in the coronary lesion (especially in the formation of the aneurysm) and therefore high-dose glucocorticoid treatment was given prior to surgery.

However, during the course of medical treatment acute myocardial infarction developed, mandating urgent surgical intervention. We speculated that early surgical intervention might prevent death, but several reports have shown that patients with acute exacerbation are susceptible to arterial wall necrosis and perforation (17, 18). Thus, Ozeren *et al*. recommended that surgical intervention for aneurysm in BD only be considered in patients with a growing aneurysm, acute rupture, or severe ischemia (19). In addition, the indications for surgical treatment of a general coronary aneurysm are severe coronary stenosis, complications such as fistula formation and compression of the cardiac chambers, and a high likelihood of rupture (20).

Recent studies have shown an immunological mechanism in the cardiovascular complications of BD and it is likely that immunosuppressive treatment could help in the management of such complications (16, 21-23). Alpagut *et al*. emphasize that the medical treatment for BD should control inflammation and regulate the immune system (22). In keeping with this is our report of a patient with BD who underwent vascular surgery and immunosuppressive treatment and did not experience surgical complications during 10 months of follow-up after surgery.

To our knowledge, there have been very few published studies that address the problem of acute myocardial infarction due to coronary aneurysm in patients with BD (24, 25). In this report, we have described a patient with BD who presented with acute myocardial infarction resulting from compression of a coronary artery by a sinus of Valsalva aneurysm. We performed surgery for the coronary lesion, adding glucocorticoids and immunosuppressive treatment, which was followed by a successful outcome. Neither re-stenosis nor an increased aneurysm was seen at a follow-up evaluation 10 months later.

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