Recurrence of Variant Angina Pectoris due to Behçet’s Syndrome

Zhuang Tian¹, Shuyang Zhang¹ and Qingjun Wu²

Abstract

We report a case of recurrent variant angina pectoris which occurred even while receiving antispastic treatment. Coronary angiography revealed no obvious lesion. Behçet’s syndrome was diagnosed based on recurrent oral aphthous and genital ulcer as well as skin manifestation. After treatment with immunosuppressant, neither angina nor oral ulcer relapsed. We strongly believe that the variant angina is due to coronary vasospasm induced by Behçet’s syndrome in this patient.

Key words: angina pectoris, Variant, Behçet’s syndrome, Coronary vasospasm

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Introduction

Behçet’s syndrome is a systemic inflammatory disorder which can involve the cardiovascular system. Several cases of acute coronary syndrome (ACS) due to Behçet’s syndrome have been reported (1-7). Here we report a case of recurrent variant angina pectoris caused by Behçet’s syndrome.

Case Report

A 64-year-old man was referred to our Emergency Department with the complaint of retrosternal chest pain and loss of consciousness in November 2007. He had felt severe chest pain and collapsed during defecation 2 hours earlier. ECG showed nodal rhythm with ST segment elevations on inferior leads and reciprocal depression on all precordial leads (Fig. 1a). Thirty minutes later he regained consciousness and his pain was eased. Repeat ECG showed normal sinus rhythm with ST segment back to the equipotential line (Fig. 1b).

Dating back from 2001, he had had six similar attacks. Each time the symptoms were relieved 30 minutes later spontaneously or after intravenous nitroglycerine. ECG showed a similar abnormality this time. Coronary angiography had been performed 3 times and revealed an almost normal coronary artery. The latest angiography was done in July 2006, which found only 40% stenosis on the first diagonal branch with a normal right coronary artery (Fig. 2).

He had been diagnosed as variant angina pectoris and was given aspirin, nitrates and calcium channel blocker (CCB) after the first attack. Angina still relapsed. He had no history of hypertension, diabetes or familial history of premature coronary heart disease. He had been a smoker for 30 years (1 pack/day) but quit in 2001.

Physical examination showed blood pressure of 105/64 mm Hg, and heart rate 88 beat per minute. No abnormality was found on heart, lungs or other organ systems. Ventricular wall motion was normal on echocardiogram. The labs results were: serum total cholesterol 4.51 mmol/L, low density lipoprotein 2.78 mmol/L, and triglyceride 1.41 mmol/L. Biochemical analyses displayed normal creatine kinase (CK), CKMB and troponin-I activities, normal hemogram, kidney function and D-dimer level. Erythrocyte sedimentation rate (ESR) was 28 mm in the first hour and C-reactive protein (CRP) was 72.8 mg/L (normal range: 0-8 mg/L). In a thorough interview, the patient described he had a history of oral aphthous ulcer which occurred at least 3 times per year for 12 years, and had recurrent genital ulcer for 2...
years. We found acne-like papules and pustules on his face and trunk. Test results for anti-nuclear antibody, antineutrophil cytoplasmic antibody, HLA-B5 and pathergy were negative. After discussion with our rheumatology department, we made the diagnosis of Behçet’s syndrome and thought variant angina was caused by right coronary vasospasm due to Behçet’s syndrome. Therefore, we did not attempt to conduct coronary angiography this time. The patient was discharged on a treatment protocol of aspirin, CCB, β-blocker, statins, prednisone and thalidomide. Three months later, his levels of ESR and CRP had decreased to normal levels. He persisted in taking all the medicines for almost 3 years and neither angina nor oral ulcer occurred again. Coronary tomography angiography done recently showed a 50% stenosis on the same part of first diagonal branch, and no obvious obstruction on right coronary artery.

Discussion

Behçet’s syndrome is designated as a multisystem disorder, the etiology of which is unclear. Vascular involvement is the main characteristic of Behçet’s syndrome. Arterial involvement is generally expressed by thrombosis, stenosis and/ or aneurysms, as shown in early studies (3-6). Biopsy and autopsy samples from the involved arteries usually exhibit histological evidence of vasculitis and thrombosis. Coronary involvement can present as silent ischemia, stable angina pectoris, or a myocardial infarction (MI) (8). MI can result from inflammatory coronary disease induced by Behçet’s syndrome such as coronary aneurysm or stenosis; however, it also can occur in a Behçet’s patient with normal coronary arteries. A thrombus (4), coronary arteritis (1), prolonged vasoconstriction, or vasoconstriction combined with a coronary lesion (2, 7) can lead to a MI in Behçet’s patients.

The present patient presented with recurrent variant angina. He had no risk factors for coronary artery disease other than a smoking history and aging. Coronary angiograms revealed no obvious atherosclerotic lesions or thrombosis. Because the angiographies were performed after remission of the symptoms each time, no coronary spasm was detected. However, the transient nature of the illness and quick resolution of ECG supported a diagnosis of coronary artery vasospasm as the etiology of acute coronary syndrome in our patient. The patient was diagnosed as incomplete Behçet’s syndrome which could involve the coronary artery. There was no unequivocal proof supporting coronary arteritis in this patient, however, there was no thrombotic tendency found and coronary angiogram excluded aneurysm, thrombus or stenosis which could lead to acute coronary syndrome in Behçet’s patients. Coronary spastic angina is a frequent complication in patients with connective tissue disease and the inflammatory condition is associated with coronary spastic angina (9). Some previous reports (1, 2) also showed that myocardial infarction in Behçet’s syndrome may possibly be due to vasculitis of coronary arteries. So we thought variant angina was induced by coronary vaso-
spasm due to arterial vasculitis in this patient. After we treated him with corticosteroid and thalidomide, the variant angina and oral ulcer did not relapse along with decreased levels of ESR and CRP, while nitrate and CCB did not have a beneficial effect. This suggested that the treatment suppressed the coronary vasospasm via suppression of inflammation or vasculitis. The mechanism of coronary vasospasms in Behçet’s syndrome is not clear. Some researchers have suggested vascular endothelial dysfunction or cytokines released by inflammatory cells are the reason for coronary spasm in patients with vasculitis and apparently normal coronary arteries (10, 11). As we considered there was coronary arteritis in the current patient, the vasospasm may have been due to the reasons mentioned above.

Such a diagnosis may be difficult and offers a diagnostic and therapeutic challenge, due to the diversity of clinical manifestations and the lack of specific laboratory tests for this disease. In the present case, the recurrent oral ulcerations and genital ulcers were unspecific and failed to get the patient’s and doctor’s attention. In addition to this, the low incidence of Behçet’s syndrome and the rare complication of cardiovascular system were also the reason for misdiagnosis. Without the immunosuppressant, nitrates and CCB, which are commonly used antispastic agents, cannot prevent the angina relapsing when used alone. To date, the most appropriate approach in a patient with acute coronary syndrome and Behçet’s syndrome is not clear. Treatment may consist of supportive medication, thrombolytic agents (1), percutaneous interventional therapy (12), as well as parenteral corticosteroids and immunosuppressant (7). We used steroid, thalidomide, as well as the antispastic agents on this patient, which have improved his prognosis significantly.

We have described a rare case of recurrent variant angina in Behçet’s syndrome. In patients presenting ACS, Behçet’s syndrome should not be overlooked in the differential diagnosis, particularly when there are no risk factors for coronary atherosclerosis.

The authors state that they have no Conflict of Interest (COI).

References