Isolated Coronary Ostial Stenosis Associated With Coronary Vasospasm

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A 50-year-old woman was brought to the emergency room in a preshock condition. An emergency coronary angiogram revealed 90% ostial stenosis of the left coronary artery with delayed distal filling. After intracoronary nitrate, the degree of stenosis was reduced to 75%; no other coronary lesions were evident. The patient was found to have hyperthyroidism and she became euthyroid after a 2-month regimen of methimazole. A follow-up coronary angiogram showed that the left coronary artery had 50% ostial stenosis without delayed distal filling. At the same time, an aortogram showed complete occlusion of the right subclavian artery in its proximal site, a slight dilatation of the truncus brachiocephalic artery, and a diffuse wall irregularity of the abdominal aorta, suggestive of Takayasu's arteritis. (*Jpn Circ J* 2000; 64: 985–987)

Key Words: Coronary ostial stenosis; Coronary vasospasm; Hyperthyroidism; Takayasu's arteritis

Isolated coronary ostial stenosis in which disease is not found in other coronary arteries is a rare condition. Takayasu's arteritis is a chronic inflammatory disease of unknown etiology involving the aorta and its major branches, and the ostia of the coronary arteries can be affected, resulting in angina or myocardial infarction! Coronary vasospasm appears to be one of the causes of myocardial ischemia and we describe a case of isolated coronary ostial stenosis caused by Takayasu's arteritis associated with coronary vasospasm.

Case Report

A 50-year-old woman was brought to the emergency room complaining of chest pain and dyspnea. She was conscious, but cyanotic and in a preshock condition. The patient had manifested slight hypertension since the age of 45, but had not received treatment. She was pre-menopausal and had no risk factors for cardiovascular disease except for hypertension. She had lost 5 kg in weight within the past 6 months and had a history of anginal attacks upon effort with attacks of unstable angina during the month prior to her admission.

On admission, blood pressure in her left arm was 80/50 mmHg, with a regular pulse rate of 110 beats/min. Blood pressure in the right arm could not be measured because a pulse was absent. A bilateral cervical bruit was audible and the thyroid gland was slightly enlarged. Heart sounds were decreased in intensity and moist rales were present throughout the entire lung field. Although abdominal vascular

bruits were audible, palpation of the peripheral vessels of the lower extremities revealed that pulsations of the dorsalis pedis and tibialis posterior arteries were equal bilaterally. An electrocardiogram revealed ST segment elevation in leads V₁ and V₂, ST segment depression in I, II, III, aV_F and V₄₋₆, and T wave inversion in aV_L (Fig 1). The chest radi-

Fig 1. An electrocardiogram reveals ST segment elevation in leads V₁ and V₂, ST segment depression in I, II, III, aV_F and V₄₋₆, and T wave inversion in aV_L.

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ograph showed slight cardiomegaly (cardiothoracic ratio (CTR) 51.4%), with slight congestion in the bilateral lung fields (Fig 2). Echocardiography showed a diffusely severe hypokinesis in the left ventricular wall (ejection fraction 30%) without hypertrophy and aortic regurgitation. Laboratory findings on admission were slightly elevated: leukocytes, $8,300/\text{mm}^3$ with normal distribution; serum creatine kinase, 108 IU/L (normal, 9–93); serum aspartate aminotransferase, 74 IU/L (normal, 5–37); and serum alanine aminotransferase, 54 IU/L (normal, 6–43). Although the serum total cholesterol level of 92 mg/dl (normal, 150–219) was markedly depressed, the values for alkaline phosphatase (214 IU/L), lactic dehydrogenase (334 IU/L) and C-reactive protein (0.3 mg/dl), and the erythrocyte sedimentation rate (7 mm/h) were within normal limits. Thyroid function tests (enzyme immunoassays) revealed marked elevations of free thyroxine of 4.30 ng/dl (normal, 0.78–2.10) and free triiodothyronine of 16.0 pg/ml (normal, 2.9–6.0), an undetectable level of serum thyroid-stimulating hormone (normal, 0.20–

$3.20 \mu\text{U}/\text{ml}$) and a high titer for antithyroid-receptor antibody.

We performed an emergency cardiac catheterization on admission, which showed an elevated left ventricular end-diastolic pressure of 25 mmHg. The coronary angiogram revealed 90% ostial stenosis of the left coronary artery with delayed distal filling, and the arterial blood pressure tracing was damped, but collateral blood flow could not be seen (Fig 3A). After intracoronary nitrate, the degree of the stenosis was reduced to 75%, and other coronary lesions were not present (Fig 3B). The right coronary artery was free of any obstructive lesions or coronary calcification and there was not any collateral blood flow. The findings indicated isolated left coronary ostial stenosis associated with coronary vasospasm.

Treatment with isosorbide mononitrate (20 mg bid), aspirin (81 mg daily) and diltiazem (30 mg qid) was started and the patient became asymptomatic immediately. Subsequent improvement in cardiac dysfunction was satisfactory. A radioactive iodine thyroid scan showed a slightly enlarged gland with increased uptake, compatible with Basedow's disease. We prescribed 30 mg of methimazole daily and after 2 months on this regimen as an inpatient, the patient's free thyroxine value was 1.67 ng/dl, the free triiodothyronine level was 6.7 pg/ml, and the serum thyroid-stimulating hormone level was $0.10 \mu\text{U}/\text{ml}$. The day after the euthyroid state was confirmed, cardiac catheterization was performed. The coronary angiogram showed 50% ostial stenosis of the left coronary artery without delayed distal filling, and the left ventriculogram showed almost normal contraction with an ejection fraction of 74%. At the same time, an aortogram showed complete occlusion of the right subclavian artery in its proximal site, slight dilatation of the truncus brachiocephalic artery, and a diffuse irregularity of the wall of the abdominal aorta (Fig 3C,D). The patient was diagnosed with isolated coronary ostial stenosis caused by Takayasu's arteritis in association with coronary vasospasm. She underwent a left intermammary artery bypass graft to the left anterior descending coronary artery and a saphenous vein bypass graft to the left circumflex coronary artery. One year after surgery, she was asymptomatic and euthyroid,

Fig 2. The chest radiograph shows slight cardiomegaly, with slight congestion in the lung fields.

Fig 3. (A) Frontal view of the left coronary artery shows 90% ostial stenosis with delayed distal filling. (B) After intracoronary nitrate, the degree of stenosis is reduced to 75% without delayed distal filling. (C,D) Aortogram shows complete occlusion of the right subclavian artery at its proximal site, slight dilatation of the truncus brachiocephalic artery, and a diffuse irregularity of the wall of the abdominal aorta.

with maintenance treatment consisting of 10mg of methimazole daily. A coronary artery repeat study showed that the left coronary artery had a 75% ostial stenosis without delayed distal filling and that the bypass grafts were patent with excellent flow.

Discussion

A review of the international literature revealed that the incidence of coronary ostial stenosis varies between 0.13% and 2.7%.^{2,3} Isolated coronary ostial stenosis in which disease is not found in other coronary arteries is rare. Takayasu's arteritis, syphilis, congenital anomaly, complications of coronary angiography or after coronary perfusion at the time of cardiac surgery, and atherosclerosis, particularly early atheroma, have been considered as causes. There are very few reports of isolated coronary ostial stenosis in the absence of these causes, and in such cases the occurrence has usually been in premenopausal women.⁴

Takayasu's arteritis is a panarteritis of unknown etiology, commonly affecting the aorta, its major branches, and pulmonary arteries.⁵ Its diagnosis is based on both clinical and angiographic data, but can be delayed for months to years, because many patients manifest nonspecific symptoms and signs such as fever, myalgia, arthralgia, weight loss, and anemia. Our patient was in the late phase of Takayasu's arteritis, so the results of serological tests, such as for C-reactive protein, were negative and the erythrocyte sedimentation rate was within normal range. However, the aortogram showed complete occlusion of the right subclavian artery, demonstrating the proximal involvement of the aortic root that is suggestive of Takayasu's arteritis. Atherosclerosis was unlikely in this patient because of her age, lack of risk factors for coronary artery disease, and absence of angiographically evident coronary disease beyond the coronary ostium.

Hyperthyroidism is known to be associated with atrial fibrillation, heart failure, angina pectoris, and myocardial infarction.⁶ Coronary vasospasm is among the possible causes of myocardial ischemia in thyrotoxic patients with normal coronary arteries.⁷ However, vasospasm occurs less often in the left main trunk. Our morphological study of the coronary ostium showed that it is particularly well-defined and forms a funnel-shaped structure characterized by a longitudinal smooth muscle arrangement, which spreads out in the inner layer, and a circular smooth muscle arrangement, which surrounds the outer layer.⁸ The coronary ostium has a specific structure that differs from that of other coronary arteries, which we suggest is the reason why that vasospasm occurs less often in the left main trunk.

We could not perform provocative testing, using ergonovine or acetylcholine, for example, because of the critical lesion in the left coronary ostium. However, after intracoronary nitrate, the degree of the stenosis was reduced. After this patient became euthyroid, the coronary angi-

ogram showed less spastic activity than had been observed originally. From these findings, we suspect that coronary vasospasm may have been induced by thyrotoxicosis. Both the absence of defined collateral circulation and the abrupt onset of major symptoms in the present patient suggest the possibility of rapid development of myocardial ischemia induced by coronary vasospasm.

The pathology of Takayasu's arteritis during the early stage consists of granulomatous changes in the media and adventitia of the aorta. This disease process is variable, with intimal hyperplasia, medial degeneration, and adventitial fibrosis of the aorta. It may be fulminant, progress gradually, or may stabilize. Over time, the aorta and coronary ostium develop segmental narrowing, which leads to the clinical manifestation of ischemia. Critical vascular lesions, such as coronary artery stenosis leading to myocardial ischemia, should be treated by angioplasty or surgical revascularization. Steroid therapy alleviates symptoms, and the combination of steroid therapy and a surgical and/or angioplastic approach to stenosed vessels has markedly improved survival and decreased morbidity by improving ischemia. In the present case, after the euthyroid state was confirmed, the coronary angiogram showed 50% ostial stenosis of the left coronary artery, so then coronary artery bypass grafting was performed. However, steroid therapy was not indicated, because the Takayasu's arteritis was stabilized. One year after surgery, the patient's Takayasu's arteritis was as stable as before, and there was no evidence on angiography of progression or new lesions.

This case is, to our knowledge, the first reported case of isolated coronary ostial stenosis caused by Takayasu's arteritis associated with coronary vasospasm resulting in a preshock condition.

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