Coronary Arterial Involvement in Takayasu's Disease

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SUMMARY

This report describes the findings in a young woman with Takayasu's disease, resulting in angina pectoris due to severe narrowing of the left coronary ostium. The patient underwent aortocoronary bypass surgery. The disappearance of angina, with reversal of positive findings on the treadmill exercise test, was noted postoperatively. Selective coronary angiography should be performed in patients with Takayasu's disease with a positive exercise test and symptoms of chest pain.

Additional Indexing Words:

Angina pectoris Aortocoronary bypass Treadmill exercise test

TAKAYASU'S disease is an arterial inflammatory disease of unknown etiology, which commonly affects the aorta, its large branches, and the pulmonary artery.¹⁾⁻³⁾ The disease has a worldwide distribution.^{4),5)} It predominantly affects young women and most often is first detected during the second or third decades of life. Symptoms referable to cardiac involvement range from congestive heart failure to angina pectoris.^{3),6),7)} Postmortem studies have revealed coronary occlusion and myocardial infarction in 9-10% of cases^{3),6)} of Takayasu's disease with angina. There is also documentation of 14 patients in whom stenotic lesions of the coronary arteries were angiographically demonstrated.⁸⁾⁻¹⁹⁾ We recently treated a 23-year-old woman with Takayasu's disease with an associated angina pectoris. Coronary angiography revealed a left coronary ostial stenosis.

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CASE REPORT

A 23-year-old Japanese woman was hospitalized with angina pectoris of 5 months duration. Attacks were readily precipitated by minimal physical exertion, such as walking up a slope. She reported a right cervical pain of few months duration 3 years earlier which was associated with leucocytosis, increased erythrocyte sedimentation rate (ESR), and positive C-reactive protein (CRP). There was no family history of coronary artery disease. Physical examination on admission revealed a regular heart rate of 82/min. The blood pressure was 124/70 mmHg, and there was no laterality or difference between the upper and lower extremities. Optic fundi were normal. The temporal arteries were easily palpable and not tender. There was no cardiac enlargement. The heart sounds were normal and there was a grade II systolic bruit at the right side of the neck. All peripheral pulses were palpable.

Laboratory studies revealed normal urinalysis, normal blood cell counts, blood urea nitrogen, serum creatinine, and serum lipid levels. Serum protein electrophoresis and thyroid function tests were normal. ESR was 17 mm in 1 hour. CRP was one plus positive. A serological test for syphilis and a test for rheumatoid factor, antinuclear antibody and lupus erythematosus prep-

Fig. 1. A: electrocardiogram in the resting state is within normal limits.
B: positive treadmill exercise electrocardiogram with 0.4 mV ST segment depression in lead II, III, aV_F, and V₃₋₆.

arations were all negative. The resting electrocardiogram was within normal limits (Fig. 1A). A treadmill exercise test showed ST segment depression of 0.4 mV in II, III, aV_F, and V₃₋₆ after exercising to the Bruce stage 2 (Fig. 1B), when she developed a typical chest pain of effort angina. At the end point

Fig. 2. Selective coronary angiogram. The left main coronary artery is almost completely occluded. LTMCA=left main coronary artery; LCX=left circumflex coronary artery; LAD=left anterior descending coronary artery.

Fig. 3. Thoracic aortogram shows an irregularity and narrowing at the right carotid artery (arrows).

Fig. 4. Histological section of the ascending thoracic aorta. Mildly thickened adventitia (A) and outer side of the media (M), and infiltration of mononuclear cells. (Hematoxylin-eosin, $\times 120$)

of the exercise, the heart rate and blood pressure increased to 146/min and 156/90 mmHg respectively, without complications. The ST segment depression persisted for 9 min. Cardiac catheterization showed normal pressure and oxygen values. A selective coronary arteriogram revealed a greater than 90% occlusion at the ostium of left main coronary artery (Fig. 2). There was no stenosis or wall irregularity in the other coronary arteries. The left ventricular angiogram showed an ejection fraction of 70% with no abnormal wall motion. Thoracic aortography demonstrated stenotic lesions at the right carotid artery (Fig. 3). The remainder of the aorta and its branches were normal. A pulmonary perfusion scintigraphy, using I^{131} labelled macroaggregates of albumin, revealed no reduction of the blood flow in the lung. The patient underwent aortocoronary bypass surgery. The aortic wall was found to be mildly thickened; the lumen of the coronary arteries appeared normal on inspection through arteriotomy. There was no obvious inflammation surrounding the great vessels of the heart. The wall of the ascending aorta was biopsied at operation for the microscopic examination. This specimen demonstrated a mildly thickened adventitia and outer side of the media. There were infiltrations of mononuclear cells, particularly lymphocytes, in the adventitia, the outer part of media and the vasa vasorum (Fig. 4).

The diagnosis was established on the basis of clinical and pathological features. The postoperative course was uneventful. One month after operation, the treadmill exercise test showed no ST segment depression and chest pain was nil. The patient is under treatment with dipyridamole (225 mg/day) and aspirin (330 mg/day).

DISCUSSION

Coronary arterial involvement in Takayasu's disease have been documented either angiographically or at necropsy since the report of Frovig and Lokens in 1951.²⁰⁾ The incidence was inferred to be 9–10% in Japan⁶⁾ or other countries.³⁾ Angiographic studies revealed lesions involving the coronary arteries in 14 cases (Table I). Included are 9 cases in Japan and 5 cases in other countries. All revealed subtotal ostial narrowing of either the right or the left coronary artery or both, and all had a history of angina pectoris. Angina pectoris in this disorder can be caused by aortic regurgitation,¹⁾ coronary ostial narrowing or the steal phenomenon with coronary-brachial arterial anastomosis.¹⁴⁾ In this case, however, aortic regurgitation was excluded by aortography and subtotal occlusion of the left coronary ostium was demon-

Table I. Clinical Features of 14 Patients with Takayasu's Disease Involving the Coronary Artery Diagnosed by Coronary Arteriography

Case	Age	Sex	Reference	Angina	Narrowed Coronary Artery	Clinical Course
1	49	F	Gotsman et al ⁹⁾	+	Ostium RCA	ACB
2	18	F	Young et al ¹⁰⁾	+	Ostium LCA 95%	
3	31	F	Ree et al ¹¹⁾	+	Ostium LCA 75% Ostium RCA 50%	
4	33	F	Huang et al ¹²⁾	+	Ostium LCA 95%	ACB
5	42	F	Shimakura et al ¹³⁾	+	Ostium LCA 100%	
6	50	F	Shibuya et al ¹⁴⁾	+	Ostium LCA	
7	38	F	Shibuya et al ¹⁴⁾	+	Ostium LCA	ACB
8	26	F	Cipriano et al ¹⁵⁾	+	Ostium LCA 80% Ostium RCA 80%	
9	37	M	Kawahata et al ¹⁶⁾	+	Ostium RCA 100%	
10	26	F	Wakiya et al ¹⁷⁾	+	Ostium LCA 99%	ACB
11	22	F	Wakiya et al ¹⁷⁾	+	Ostium LCA 99%	ACB
12	32	F	Takizawa et al ¹⁸⁾	+	Ostium LCA 90%	ACB
13	45	F	Chun et al ¹⁹⁾	+	Ostium LCA 95%	ACB
14	23	F	Makino et al	+	Ostium LCA 90%	

Abbreviations: RCA=right coronary artery; LCA=left coronary artery; ACB=aortocoronary bypass graft operation.

strated by coronary angiography as the etiology for angina pectoris. Coronary-brachial anastomosis was not observed.

The ST segment depression in women during exercise testing has been described as a less specific indicator of coronary artery disease,²¹⁾ especially in asymptomatic women.²²⁾ However, the exercise stress test in our patient brought about the typical chest pain of angina pectoris, with a maximum ST segment depression of 0.4 mV that persisted for 9 min. ST segment depression over 0.4 mV is assumed to be associated with left main or multivessel disease, especially if change occurs in Bruce stage 1 or 2.²³⁾ Moreover, the persistence of ST depression 8 min into recovery suggests the likelihood of stenosis in the left main coronary artery.²⁴⁾

There is some evidence suggesting that large doses of corticosteroids may halt or even reverse the lesions during the active inflammatory phase of Takayasu's disease.^{1),25)} However, during the chronic phase of the disease as in our patient, fibrosis and thrombus formation were responsible for the lesions and resulting symptoms and the administration of steroids would probably not have been effective. Since the left main coronary artery stenosis places a large amount of the muscle in jeopardy and may lead to myocardial infarction or even sudden death, the coronary arterial bypass graft operation is mandatory for patients with a coronary narrowing due to Takayasu's disease. The fact that the coronary arterial stenosis in Takayasu's disease tends to be confined characteristically to the ostium⁸⁾⁻¹⁹⁾ and proximal segments of coronary arteries favours consideration of the bypass graft operation.

We recommend that the treadmill exercise test be used to detect suspected coronary arterial involvement in patients with Takayasu's disease. In addition, selective coronary angiography should be performed in patients with a positive exercise test and symptoms of chest pain. If the coronary angiogram reveals a coronary ostial stenosis of a severe grade, an aortocoronary bypass graft operation should be performed as soon as possible in selected cases with no or mild inflammatory process for the treatment of this potentially lethal disorder.

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