Coronary Ostial Stenosis in Takayasu’s Arteritis

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A patient with Takayasu’s arteritis with left coronary ostial narrowing is presented. The dramatic clinical and pathologic findings are discussed in detail. Emphasis is placed on making the diagnosis as soon as possible, in order to expedite bypass surgery to prolong life.

Coronary arterial disease in Takayasu’s arteritis is rare. Its presentation as coronary ostial narrowing is a potentially lethal but correctable problem. This case report illustrates how dramatic the clinical and pathologic features can be and the importance of considering this surgically remediable entity in the differential diagnosis of chest pain, especially in women.

Case Report

The patient was a 45-year-old woman who in December 1976 noted discoloration of her left hand, with increased sensation to touch and cold. She had a positive lupus erythematosus preparation and an antinuclear antibody titer of 1:40; however, her rheumatoid arthritis factor, level of complement, hemolytic complement, VDRL test, and double-stranded DNA binding were normal.

An arteriogram demonstrated thrombosis of the left subclavian artery, with retrograde thrombosis into the left vertebral branch. The patient underwent surgery to implant a Dacron bypass graft from the left carotid artery to the left subclavian artery. The subclavian artery was resected and revealed minimal intimal fibrosis without adventitial thickening. Therapy with sodium warfarin (Coumadin) was begun, with relief of the patient’s symptoms.

The patient did well until May 1978, when she noted tingling sensations in her wrists bilaterally, radiating up both arms and crossing her chest, accompanied by sweating and mild shortness of breath. The sensations occurred with rest and with exertion and lasted 30 minutes. With administration of nitroglycerin, they were less intense and averaged six per day. With isosorbide dinitrate and propranolol, the episodes occurred twice daily. During these episodes the electrocardiogram manifested marked T-wave inversions over the lateral precordial leads (Fig 1). The patient denied any history of syncope, palpitations, orthopnea, paroxysmal nocturnal dyspnea, seizures, rheumatic fever or arthralgias. She was premenopausal.

There was no history of hypertension. The patient had smoked five cigarettes per day over the last 30 years. She had received birth control pills for eight months, but discontinued them eight years ago. Her level of cholesterol was 184 mg/100 ml and the triglyceride level was 193 mg/100 ml. The results of physical examination were unremarkable. The chest x-ray film and echocardiogram were normal. A treadmill test on Bruce’s protocol reproduced the patient’s chest pain with 2 mm of lateral ST-segment depression after four minutes of exercise at a heart rate of 100 beats per minute; four minutes after the study, she had marked T-wave inversions, which resolved.

Prior to and during the coronary injections, the patient developed her typical chest pain with marked ST-segment depression on electrocardiographic leads, necessitating therapy with nitroglycerin. She also had “ventricularization” of

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the aortic pressure curve during injections of the left coronary arteries.

The left ventricular angiogram demonstrated normal contrac-
tion. There was a lengthy 95 percent obstruction of the left coronary ostium, with thread-like filling prior to the takeoffs of the left anterior descending and circumflex arteries (Fig 2). No other disease was noted in the left or right coronary arteries. The shot of the aortic root showed a 100 percent obstruction of the left subclavian takeoff. The remainder of the aortic arch appeared normal.

The patient was scheduled for surgery; however, during preoperative weighing, she developed chest pain and demonstrated seizure activity. Cardiopulmonary resuscitation was performed without success.

At autopsy, prominent smooth, white, firm intimal plaques were seen in the ascending arch and descending segment of the aorta. Some of these plaques encircled the ostia of the vessels of the aortic arch. The brachiocephalic ostium was 30 percent occluded, and the ostium and proximal 1 cm of the left subclavian artery were totally occluded. All other vessels arising from the thoracic and abdominal aorta were similarly affected. A plaque measuring 1.5 × 2.0 cm located in the left sinus of Valsalva reduced the ostium of the left coronary artery to a narrow slit, and the proximal 0.5 cm of the left main coronary artery was virtually occluded (Fig 3).

The aorta and the proximal segments of its affected branches showed marked intimal fibrous proliferation, disruption of the elastic fibers of the media, and focal and medial adventitial thickening and injury. Lymphocytes and plasma cells were seen in association with a few vasa vasorum; other inflammatory cells were absent from the arterial walls. The myocardium did not show evidence of a remote or recent infarct.

**DISCUSSION**

Takayasu’s arteritis of the coronary arteries has been reported; however, symptoms of coronary involvement are rarely the initial presentation. More commonly, systemic complaints such as headaches, fever, and heart failure precede angina. Angina pectoris was the present-
ing symptom in only four of 16 patients described in the literature. Two of the four patients underwent successful bypass surgery. Our patient’s ostial lesion was amenable to grafting, but prior to surgery, she died, probably from an arrhythmia. The success of bypass surgery emphasizes the importance of recognizing coro-
nary arterial disease in the setting of Takayasu’s arteritis prior to myocardial infarction, heart failure, and sudden death. When a woman has ischemia of the arms and angina pectoris, positive findings on collagen vascular workup, abnormal ECGs with pain, and markedly posi-
tive findings on an exercise test, the possibility of coro-
nary ostial lesions in the setting of Takayasu’s arteritis prompts an expeditious but cautious coronary and aortic root angiographic study.

Several features of Takayasu’s arteritis, especially with coronary ostial narrowing, are exemplified in this case. First, the response to complete ostial occlusion with the tip of the catheter was manifested (ie, hypotension, left ventricular pressure-like tracing, and reproducible pain in the chest). Careful monitoring and prompt withdrawal of the catheter are imperative. Secondly, left ostial stenosis connotes the same or a worse prognosis than that of a lesion of the left main coronary artery. Furthermore, sudden death is common. Thus, surgery should be performed as soon as possible. Thirdly, there were histologic findings of Takayasu’s arteritis. Adventi-
tial thickening was present and localized to the aortic arch. Intimal and adventitial proliferation with fibrotic thickening accounted for the narrowed left coronary ostium.

Symptomatic nonaortic coronary ostial stenosis is rare. Even rarer is coronary ostial stenosis secondary to Takayasu’s arteritis. The dramatic clinical and pathologic features of this patient are those of an aortic arch syndrome. Syphilis, atherosclerosis, giant cell arteritis, and rheumatic arteritis are unlikely causes in this case. The features are consistent with those previously described for Takayasu’s arteritis or sclerosing aortitis. Urgency in making the diagnosis cannot be overempha-
sized, in light of the problem being a surgically cor-
rectable lesion.

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Figure 3. Plaque significantly narrowing orifice of left main coronary artery, viewed from left sinus of Valsalva.