

Delayed Diagnosis of Takayasu's Arteritis: Total Abdominal Aorta Occlusion Treated with Axillo-Bifemoral Bypass

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ABSTRACT. Takayasu's arteritis is an inflammatory vasculitis that primarily affects the aorta and its major branches. Delayed diagnosis is common, and is largely attributed to the variable and nonspecific presentation of the initial symptoms. Involvement of the abdominal aorta causes severe claudication of the lower extremities, and bypass surgery is required in some cases. We describe a case in which diagnosis was delayed. Total occlusion of the abdominal aorta was successfully treated with axillo-bifemoral bypass. (J Rheumatol 2004;31:393-5)

Key Indexing Terms:

DELAYED DIAGNOSIS TAKAYASU'S ARTERITIS AXILLO-BIFEMORAL BYPASS

Takayasu's arteritis (TA) is a chronic inflammatory vasculitis that primarily affects the aorta and its major branches¹. In some asymptomatic cases, physical examination reveals different blood pressure levels in the left and right arms, or pulselessness in an extremity. Most cases of delayed diagnosis are due to variable and nonspecific initial symptoms²⁻⁶. The lesions tend to appear as long, smooth, segmental narrowing of vessels, with little evidence of plaque formation. Occlusion is common, and some patients also develop aneurysms⁷⁻⁹. TA is known to affect a variety of vessels, but the subclavian artery, axillary artery, carotid arteries, renal arteries, and infraabdominal aorta are most frequently affected^{1,2,7-11}. The majority of patients are treated medically with corticosteroids and immunosuppressive agents such as methotrexate, azathioprine, and cyclophosphamide (CYC)^{12,13}. Angioplasty, stent placement, and bypass surgery may be necessary when ischemic symptoms do not improve with medical therapy^{14,15}.

We describe a case of Takayasu's disease with total occlusion of the abdominal aorta in which diagnosis was delayed because of nonspecific initial symptoms. The patient was successfully treated by axillo-bifemoral bypass.

CASE REPORT

In February 2002, a 44-year-old woman presented to our clinic with leg pain, weakness, and fatigue. She described claudication of the lower extremities, and reported that she had had this problem for 10 years. Initially, claudication had developed only after long walks, but at the time of presentation she had pain after walking even short distances. Her history included diagnosis of hypertension in 1985, and she had been taking anti-hypertensive medication since that time. In addition, 8 years before presentation, she had been hospitalized for chest pain. Coronary angiography at that time revealed nothing abnormal. She had had headache problems for the past 7 years, and had attributed this to hypertension. The patient also exhibited emotional lability during this period, and was diagnosed with depression. She reported intermittent myalgia and arthralgia as well, but had no arthritis issues.

In 1994, she had undergone initial abdominal and renal ultrasonography studies during the examination for hypertension. At this stage, all sonography findings had been normal. Another ultrasonography examination performed in 1999 revealed mild stenosis of the abdominal aorta, with a 4 cm lesion below the origin of the renal arteries. Two years later, in 2001, this lesion was 9 cm long, was causing marked stenosis (> 50% occlusion), and showed rim thrombus formation. These changes were attributed to atherosclerotic disease, and the patient was given coumadin. She was not a cigarette smoker, and there was no coronary artery disease or sudden death in her family history. She reported that her lipid profile had always been normal or shown mild elevations. There were no records to document previous erythrocyte sedimentation rate (ESR) or C-reactive protein (CRP) concentration.

On initial examination at our clinic, her blood pressure measurement in the right arm was 165/120 mm Hg, and in the left arm 160/120 mm Hg. Livedo reticularis was noted in both lower extremities. There were no palpable pulses from the bilateral tibialis posterior, dorsalis pedis, popliteal, or femoral arteries in either leg.

The laboratory findings were as follows: hemoglobin 14.5 g/dl, white blood cell count 12,600/mm³, creatinine 0.78 mg/dl, ESR 98 mm/h, CRP 21 mg/l, total cholesterol 237 mg/dl, high density lipoprotein 53 mg/dl, low density lipoprotein 139 mg/dl, and triglyceride 190 mg/dl. Urinalysis was normal, and there were no signs or symptoms of infectious disease. Serological tests for antibodies to hepatitis C virus and hepatitis B virus surface antigen were negative. Testing for antinuclear antibodies, anticardiolipin antibodies immunoglobulin G and immunoglobulin M, lupus anticoagulants, cytoplasmic and perinuclear types of antineutrophil cytoplasmic antibodies, and complement 3 and 4 revealed no abnormal findings.

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Echocardiographic examination revealed mild akinesis in the inferior heart wall, hypokinesis of the posterior and lateral heart walls, minimal mitral regurgitation, and left ventricular hypertrophy. Thallium myocardial scintigraphy revealed a fixed perfusion defect of the inferior wall. She refused to undergo coronary arteriography.

Abdominal computed tomography revealed total occlusion of the abdominal aorta below the origin of the renal arteries. Angiographic examination of the aorta confirmed total occlusion of the abdominal aorta at the distal limit of the origin of the renal arteries, and collateral vascularization of the lower extremities (Figure 1). The aortic arch and its branches appeared normal. Electromyographic testing revealed diminished conduction velocity in the left lateral cutaneous femoral nerve.

We diagnosed TA, but felt that the initial symptoms of this disease had actually appeared in 1985. The patient was in the chronic phase, but her laboratory findings indicated she was experiencing a flare. In light of the severe and progressive disease just below the origin of the renal arteries, we administered combination therapy of oral prednisone and CYC instead of corticosteroids alone or other immunosuppressive agents. The initial prednisone dose was 1 g/kg/day for 4 weeks. After this, the daily amount was tapered by 10% every 2 weeks until a dose of 20 mg/day was reached. For CYC, a dose of 500 mg was administered intravenously, and this was repeated on Days 10, 20, 30, 50, 70, and 90 of treatment. We gave the CYC in divided doses at short intervals in order to monitor side effects more closely.

After 3 months on the prednisone and CYC therapy, her ESR and CRP had fallen to normal ranges, and she was able to undergo surgery. As determined, angiography revealed no abnormalities of the aortic arch or its branches, thus axillo-bifemoral bypass procedures were performed. There were no complications with the operation. Immediately after surgery, pulses were palpable in both lower extremities, and postoperative Doppler ultrasound revealed that both femoral arteries were patent. However, the involved segment of aorta was not examined directly; histopathologic examination of a specimen from the axillary and femoral artery at the anastomosis site revealed nothing abnormal. After surgery, she was followed up with corticosteroid that was tapered by 10% every 4 weeks until a dose of 10 mg/day was reached. CYC was stopped but azathioprine was added in dose of 2 mg/kg/day because level of ESR and CRP increased with corticosteroid alone.



Figure 1. Angiography reveals total occlusion of the abdominal aorta just below the origin of the renal arteries.

DISCUSSION

The majority of patients with Takayasu's arteritis are women, and symptoms usually arise before the third decade of life. The early systemic or "prepulseless" phase is characterized by fever, night sweats, weakness, arthralgia, hemoptysis, pleuritis, pericarditis, and chest pain. Cardiovascular manifestations may include heart failure, systemic arterial hypertension, palpitations, bruit, headaches, and syncope^{2,3}. Our patient was a 44-year-old woman, and her initial symptoms of hypertension, headache, arthralgia, myalgia, fatigue, and weakness appeared at a young age. She had started to experience claudication 10 years prior to presentation, and this problem had become worse with time. The findings when she presented to our center were unusual, with total occlusion of the abdominal aorta but no involvement of the aortic arch or its branches.

TA can be severe, and patients may exhibit 50–90% stenosis or even total occlusion of the vessel^{9,11,14,16}. Diminished or absent pulses are common during the occlusive phase^{2,9}. Some patients experience flares during the chronic phase of the disease^{2,5,6}. When we diagnosed our case with TA, we interpreted her elevated ESR and CRP as a disease flare. These measures normalized after treatment with corticosteroid and CYC, and she then underwent surgery. Angioplasty, stent placement, and bypass surgery may be necessary when ischemic symptoms are severe and do not improve with medical therapy^{12,13,14,16}.

Our patient's history, clinical findings, and laboratory results were all consistent with the diagnosis of chronic inflammatory vasculitis^{17,18}. The definitive diagnosis of TA was delayed many years due to nonspecific initial symptoms. Concerning the problems of diagnosing this illness, other authors have reported a median delay of 10 months from onset of symptoms or signs of large-vessel inflammation^{12,17}. However, the delay can be as long as 20 years^{5,19}.

Finally, this case is of interest because initial symptoms were nonspecific, diagnosis was delayed, and although she had total occlusion of aorta, the aortic arch and its major branches were normal. Axillo-bifemoral bypass surgery was performed without complication. We stress that all young patients with hypertension, and particularly those with hypertension accompanied by systemic symptoms, should be thoroughly investigated for underlying pathology. Keeping a high index of suspicion is the only way to achieve early diagnosis of underlying diseases like Takayasu's arteritis and other forms of vasculitis.

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