

Case Report

Takayasu's Arteritis: A Case Report With Global Arterial Involvement

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ABSTRACT

A patient with global arterial involvement in Takayashu Arteritis is described with emphasis on early diagnosis. (Rawal Med J 2005;30:43-45).

Key Words: Takauasu disease, arteritis, aortitis, hypertension

INTRODUCTION

Takayasu's arteritis is a rare inflammatory stenotic disease which effects large to medium sized arteries typified by a strong predilection for the aortic arch and its main branches. Although the etiology remains elusive, an autoimmune pathogenesis has been suggested, as several associations have been made with human leukocyte antigen (HLA)-B51, B52, DRB1 1502 etc. in Japan, Korea¹ and India.² Clinical manifestations may range from non-specific symptoms to catastrophic sequelae due to carotid or other major arterial involvement. We report the case of a 55-yr old Pakistani male with global involvement with Takayasu's arteritis.

KEY WORDS: Takayasu arteritis, pulseless disease, aortography

CASE REPORT

A 55 year old Pakistani male presented to the cardiology outpatient department with left arm pain of several months duration. His past medical history was significant for type II diabetes mellitus and hypertension. Six years earlier, he had developed progressive pain and weakness of his entire right arm. The findings of diminished pulses lead to an angiogram which revealed aneurysmal dilatation and critical stenosis of right subclavian artery. He underwent surgical revascularization with an end-to-end gortex graft. However, one month latter, the graft occluded with moderate residual dysfunction of his right arm. Biopsy of the arterial segment, at the time of the surgery, showed non-specific inflammation and thrombosis. He subsequently developed left arm claudication and presented to this hospital.

A moderate size, pulsating mass was obvious in the left clavicular region. Distal pulses in the left arm were 2 + with warm skin. No peripheral pulses were palpable in the right arm. Cardiac auscultation was unremarkable. Blood chemistries including renal function were normal. A duplex ultrasound scan done at another hospital revealed a very tortuous and aneurysmal left subclavian artery. He underwent aortography, left subclavian and coronary angiography. Arch aortography revealed dilated aortic arch with complete occlusion of the right subclavian artery (fig-1). Selective left subclavian angiography revealed severe segmental aneurysmal dilatation of the entire vessel extending into the proximal part of the axillary artery. The largest aneurysm diameter was 1.7 cm.

There were multiple areas of severe stenosis interposed between the aneurysmal segments (fig-2). Coronary angiography showed aneurysmal dilatation of the left main and proximal left anterior descending artery (LAD) with moderate (50%) mid LAD narrowing. Abdominal aortogram revealed aneurysmal dilatation of the entire aorta. Both renal arteries had severe proximal segment dilatation with significant subsequent narrowing. The aneurysmal involvement of the aorta extended into the proximal part of both common iliac arteries as well.

Patient was placed on high dose prednisone (40 mg twice daily) for two weeks which was tapered to 10 mg daily over the ensuing two weeks. There was substantial improvement of his claudication. An erythrocyte sedimentation rate (ESR) was 45 mm/h with negative C-reactive protein. He was maintained on 10 mg/day with instructions for monthly ESR checks. For hypertension, he was placed on beta-blockers and dihydropyridine calcium channel blockers with good results.

DISCUSSION

Takayasu's arteritis is a chronic granulomatous vasculitis that typically involves the aorta and its major branches with potential for catastrophic outcome. This rather rare disease, also known in the literature as "pulseless disease", "Martorell syndrome" and "occlusive thromboaropathy" is characterized by progressive inflammation in vessel wall leading to thickening, dilatation, stenosis and thrombosis ¹. Named after professor Takayasu, an ophthalmologist at Kanazawa University Japan in 1905, this disease was initially confined to females from Eastern Asia. Though its incidence has rapidly involved both sexes in most parts of the globe, the female to male predominance remains.

Patient can initially present with obscure systemic symptoms, such as fever, malaise and night sweats. The age at presentation is highly variable, though most cases present between the 2nd and 3rd decade of life . The classic symptoms of takayasu's arteritis depict end-organ damage i.e. stroke from carotid involvement, hypertension from renal artery stenosis or aortic regurgitation from aortitis. Bruits and diminished pulses are the most reliable signs. Most patients have bilateral involvement with stenosis and aneurysmal segments coexisting in the same vessel.³□ Though there is a clear predilection for aortic arch, renal arteries are involved in 28-75%,³ coronary arteries in less than 10 % □ and pulmonary arteries in 14-100 % of patients.❶ Based on region of involvement, takayasu's arteritis is classified as Type I (aortic arch), Type II (aortic arch and descending thoracic aorta), Type III (descending thoracic aorta and abdominal aorta) Type IV (abdominal aorta only) and Type V (aortic arch, descending thoracic aorta and abdominal aorta).□

The differential diagnosis includes syphilitic or tuberculous aortitis, lupus and other collagen vascular disorders, giant cell arteritis and Kawasaki's disease. There are no diagnostic pathologic signs and the diagnosis rests on combination of clinical features complemented by imaging studies. While angiography is the gold standard, inflammation and dilatation in the vessel wall can be visualized by Doppler studies and magnetic resonance imaging. Though association with certain HLA types favors an autoimmune pathogenesis, this hypothesis remains unproven.¹

The course of the disease is variable and spontaneous remissions may occur. Five year survival has been reported to be 83%.□ Steroids are the mainstay of therapy with 50% of response. This response rate can be increased by using methotrexate or other

immunosuppressive agents, in addition to providing steroid sparing benefit.¹ Surgery or percutaneous intervention is recommended for non-responders to medical therapy or critical ischemia, with variable results. While most studies have used acute phase reactants (ESR) to monitor therapy, the validity of this approach has been challenged and perhaps the best way is to complement acute phase reactants with non-invasive imaging modalities.□

Our patient with Type V disease responded very well clinically to steroid therapy and the goal is to keep his ESR less than 20 mm/h. The benefit of doing complete aortogram is underscored by the fact that we detected unsuspected bilateral renal artery disease which precluded ACE-I therapy for hypertension.

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LEGENDS:

Figure-1: Arch aortogram showing dilated arch, totally occluded right subclavian (arrow)

Figure-2: Selective left subclavian angiogram showing severe aneurysmal dilatations and stenosis. Arrow indicates the largest aneurysm.

