Nigerian Journal of Clinical Practice
Dec. 2010 Vol. 13(4):467-469

Case Report

TAKAYASU'S DISEASE IN A YOUNG BLACK BOY

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ABSTRACT

Background: Takayusu's disease is a rare disease affecting women predominantly during the child- bearing age. It is a primary vasculitis condition of large-vessels that responds well to steroid therapy. Immunosuppressives and vascular reconstruction may be needed as necessary.

Procedure: Reference was made to the case note of this young boy who was being co-managed by cardiology and vascular clinics. The diagnosis of Takayasu's disease was confirmed by the rheumatology unit and appropriate literature search was done.

Result: Takayusu's disease responds well to steroid therapy as exemplified by this patient. There was no relapse of the active inflammation after six months of steroid therapy.

Conclusion: A high index of suspicion must be exercise in diagnosing Takayasu's disease. It could be difficult to have a clue early in the disease process because of non-specific presentations. Appropriate referral should however be made to Rheumatologist when the diagnosis is suspected. This will go a long way in delaying the morbidity that is associated with this rare disease.

Key Words: Takayasu, vasculitis

INTRODUCTION

Takayasu's disease was described by a Japanese ophthalmologist in 1908. It is a primary vasculitis affecting aortic arch and the major branches of the aorta. The average age at diagnosis is in the mid twenties, but the disease may begin as early as age seven or as late as seventy years. It affects women eight times more frequently than men.

Reports have linked Takayasu's disease with infectious agents, including spirochetes, bacterial, mycobacterial and viruses. There is no convincing evidence that any of these play a pathogenetic role in Takayasu's disease[1]. Genetic studies remain controvertial. Earlier studies had shown a relationship with HLA-BW52 in Takayasu's disease in Japanese patients[2]. Other studies however did not confirm the HLA-BW52 association[3].

The earliest change seems to be a granulomatous inflammation in the adventitia and outer layers of the affected arteries with gradual progression to a panarteritis[4]. Histology shows an infiltration of lymphocytes, plasma cells, histiocytes, multinucleated giant cells and occasional polymorphonuclear cells with resultant disruption of the normal architecture of the vessels[4].

Clinical features are of two groups: those due to vascular damage and those due to systemic inflammation. Presentations include hypertension, pain over the carotid artery, dizziness and visual abnormalities. Others include chest wall pain, myalgia and joint pain[5]. Constitutional symptoms such as fever, malaise, weight loss and night sweats precede vascular complications by weeks or months[5]. Bruits over the subclavian artery, carotid and abdomen may be present. Absent or reduced pulse volume in one or more large arteries is detected and blood pressure discrepancy of greater than 30 mmHg between arms[5]. With progression, cardiac symptoms are prominent and massive haemoptysis indicates pulmonary artery involvement[6]. Neurological symptoms are present in 80% of patients with Takayasu's disease that involves the brachiocephalic arteries[6].

Four late-phases of Takayasu's disease have been described on the basis of the sites of vessel involvement[7]: they are type 1- classic pulseless type that involves the brachiocephalic trunk, carotid arteries, and the subclavian arteries. Type 2 which is a combination of type 1 and 3. Type 3- atypical coarctation, type that involves the thoracic and abdominal aortas distal to the arch and its major branches. Type 4-dilated type that involves extensive dilatation of the length of the aorta and its major branches.

The most common type is the type 3, which is found in as many as 65% of patients. The most commonly involved vessels include the left subclavian artery (50%), the left common carotid artery (20%), the brachiocephalic trunk, the renal arteries, the celiac trunk, the superior mesenteric artery, and the pulmonary artery (50%). Infrequently, the axillary,

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brachial, vertebral, coronary and the iliac arteries are involved[7].

Laboratory findings are non-specific. Anaemia, mild leucocytosis, elevated ESR (erythrocyte sedimentation rate) and hypoalbuminaemia may be detected in active inflammation[8]. The diagnosis is confirmed by arteriography. Other useful investigations include CT scan, MRI, and gallium scintigraphy[9].

Management is with steroid (prednisolone), disease modifying anti-rheumatic drugs (DMARDS) and vascular bypass surgery.

CASE PRESENTATION

A 17 year old South African boy was referred from vascular surgery clinic to the rheumatology clinic of the Inkosi Albert Luthuli Central Hospital, Durban, South Africa in October 2005 with a working diagnosis of Takayasu's disease. He was being co-managed by the cardiology clinic for heart failure and vascular clinic for valvular heart disease for two years before referral. His initial echocardiography revealed significant aortic regurgitation and mild mitral regurgitation.

He presented with generalized body weakness, left anterior chest wall pain, fever, and joint pain involving the knees and the elbows. He was still on anti-failure drugs at the time of presentation. There were no symptoms referable to the central nervous system.

When examined, he was found to be small for age, with temperature of 37.6^0c, he had swollen and tender left knee. The left radial artery was barely palpable and a blood pressure difference of 30mmHg between the two arms. Bruit was picked over the left axillary region. A diagnosis of Takayasu's disease was made by clinical and radiological assessment.

Unenhanced coronal magnetic resonant angiogram revealed focal stenosis of the right external iliac artery while the post contrast dynamic magnetic angiogram revealed non-visualisation of the proximal 3.5cm of the left subclavian artery from the aortic arch. There was flow noted distally from the level of the vertebral artery suggesting a retrograde flow via the posterior circulation of the brain into the left vertebral artery-subclavian steal. The right vertebral artery was prominent. The left profunda brachial artery was not visualized. All other arteries were patent. The overall angiogram finding was compatible with Takayasu's disease. Erythrocyte sedimentation rate was 106mm/hr, PCV was 26% and he had a leucocytosis of 14,300cells/mm^3.

He was placed initially on high dose prednisolone which was tapered after two months when symptoms had subsided. He is presently on 5mg prednisolone daily. He however continued his visit to the cardiology unit for the management of the heart failure. He is presently doing well on low dose prednisolone.

DISCUSSION

Takayasu's disease was named after Japanese ophthalmologist who first described the ocular manifestation in 1908. It is also variously known by a number of synonyms. These include pulseless disease, aortic arch arteritis, non-specific aortoarteritis and Takayasu's arteritis[10].

Takayasu's disease was said to be rare in the black race. Underdiagnosis of the disease might have accounted for this rarity. Ogunbiyi and Falase had reported four cases, all women in the South West of Nigeria[11]. In 2003, Okeahialam et al also reported a case in the Northern part of Nigeria[12].

Diagnosis of Takayasu's disease requires a high index of suspicion. The American College of Rheumatology has however proposed criteria useful in the diagnosis of Takayasu's disease. The presence of three out of six criteria is needed. This is said to be 91% sensitive and 98% specific for Takayasu's disease[11].

The criteria met by this patient include age of onset before 40 years, decreased brachial artery pulse, unequal arm blood pressure, subclavian bruit and angiographic evidence of occlusion of subclavian artery on the left. The last criterion which was not present in this patient is limb claudication.

This patient presented with minimal joint complaint. Joint pain may however be severe but actual synovitis is uncommon and where present is usually mild[12].

There was no ocular complaint in this patient, and this is in agreement with the recent presentation of Takayasu's disease. The ischaemic retinopathy that was originally reported by Takayasu is very rare today[13].

The symptoms of this patient were well controlled with prednisolone without additional drugs. Some patients will however in addition to Prednisolone need immunosuppressive agents. The commonly used immunosuppressives in Takayasu's disease are methotrexate and mycophenolate mofetil[14].

This patient was in remission two months after the commencement of high dose prednisolone. Remission in Takayasu's disease is defined as resolution of signs, symptoms, and laboratory markers of inflammation as well as lack of progression of angiographic abnormalities[15].

It is too early to determine the prognosis in this patient. The fact however is that almost all patients will experience morbidity from Takayasu's disease[16]. Because of the chronic relapsing and remitting nature of this disease, a careful monitoring and adjustment of therapy in this patient is necessary. Mortality is caused by renal failure, stroke, cardiac failure, or infection complicating the use of

immunosuppressive agents[16]. This write-up is to heighten the awareness of medical personnel of the presence of this rare disease also in the black race.

ACKNOWLEDGEMENT
I wish to acknowledge the contribution of Prof O.O. Adelowo towards the production of this manuscript.

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