Takayasu arteritis: An unusual cause of stroke in a young person

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Abstract

Takayasu Arteritis (TA) is a rare granulomatous vasculitis that occurs primarily in females. It affects large and medium vessels, primarily the aorta and its large branches, and proximal portions of pulmonary, coronary, and renal arteries. There is a paucity of epidemiological data on the disease. Numerous cases have been reported worldwide, though it is more prevalent in Asian countries such as Japan. It is thought to be rare in the African continent. If diagnosed or treated late, Takayasu's can have grave complications. Early diagnosis is vital, though it can be challenging to many physicians due to its non-specific presentation, such as fever, fatigue, arthralgia, myalgia, and weight loss, which can often be missed. About 84-96% of these patients present as diminished or absent pulses associated with limb claudication and blood pressure discrepancies between the two arms. Here we report a case of a 26-year-old lady who presented with an ischemic stroke secondary to Takayasu.

Key words: Takayasu arteritis, Stroke, Young patient, Kenya

Introduction

Takayasu's arteritis is a rare and challenging entity characterized by chronic inflammation of the large vessels, primarily the aorta and its major branches[1]. However, the multifaceted nature of this rare autoimmune vasculitis occasionally presents clinicians with atypical manifestations, challenging the traditional understanding of the disease. This case report unravels a unique presentation of Takayasu's arteritis, diverging from its more conventional manifestations, as it manifests itself unexpectedly and uncommonly – mimicking the clinical features of a stroke[1,2]. This case report aims to shed light on an unusual presentation of Takayasu's arteritis, wherein a young patient initially presented with symptoms mimicking an acute stroke. One should consider vasculitis as a potential aetiology in patients presenting with stroke-like symptoms. This case underscores the need for heightened awareness among healthcare professionals to ensure timely and accurate diagnosis, thereby facilitating appropriate management strategies for this uncommon but potentially debilitating condition, especially in regions like Africa, where the disease is thought to be rare.

Case report

A 23-year-old procurement officer with no known history of comorbidity presented to casualty with a one-day history of left-sided weakness. According to the patient's mother, she had been in her usual state of health until two days ago when she developed a headache, dizziness, and chest pains followed by an altered level of consciousness. She collapsed, and after regaining consciousness, she was found to have left-sided weakness and difficulties swallowing food. There was no history of fever, vomiting, convulsions, visual symptoms, urine or faecal incontinence. She had, however, complained of fatigue, malaise, palpitations, bilateral upper limb numbness, and intermittent unilateral headaches, for which she had been treated for malaria and typhoid fever severally over the past three years. She is a para 0+1, which resulted from an ectopic pregnancy. She had no history of use of oral contraceptives or tobacco but socially takes alcohol. The remainder of the past medical, surgical, and family history was non-contributory.

On arrival, she was anxious, had a blood pressure of 140/90 in the right arm but non-recordable in the left arm, pulse was 70 beats/minute (only recordable in the right arm), and temperature was 36.9°C and respiratory rate 26 breaths/minute. Examination of the central nervous system revealed she was aphasic and had a cranial nerve palsy of the third nerve. Motor examination revealed reduced power in both the left upper and lower limbs. Her cardiovascular examination showed muffled heart sounds and no murmurs. She had bilateral carotid

bruit more pronounced on the left side. Examination of the respiratory and abdominal examinations were unremarkable. We ordered pertinent laboratory and other specific tests based on the history and examination. She had a normal haemogram, urea, and electrolytes with elevated C reactive protein of 90mg/L (normal is 0-10mg/L). A brain CT scan showed an infarct on the right middle cerebral artery territory. Echocardiogram and ECG were normal. Due to the audible carotid bruit bilaterally, we also performed carotid artery color Doppler imaging evaluating the Common Carotid Artery (CCA), Internal Carotid Artery (ICA), External Carotid Artery (ECA), and Vertebral Artery (VA). Carotid Doppler revealed bilateral carotid artery atheromatous plaques thrombus with partial lumen occlusion.

Following the above results and her young age, a decision was made to order a CT carotid, angiogram which revealed a complete occlusion of the left common carotid artery from the point 1cm from its point of origin through to the internal and external carotid arteries—approximately 90% luminal stenosis of the proximal third right common carotid artery thus confirming the clinical diagnosis of Takayasu. She was started on 1 gram of solumedrol for three days and then converted to oral steroid at 1gm/kg taper and addition of azathioprine as a disease modifying agent. Our cardiothoracic team advised continuation of medical management and not stenting at this stage. Subsequent reviews as an outpatient showed marked improvement in peripheral circulation with restoration of the previously absent L pulse and the blood pressure is now measurable in both arms. The paresis is still present, for which she undergoes regular physiotherapy and can now communicate as her speech has recovered. We have been unable to repeat the CT angiogram due to financial limitations.

Discussion

Takayasu arteritis (pulseless disease) is an autoimmune, inflammatory disorder affecting large and medium-vessels vasculitis that predominantly affects women of childbearing age[3]. It is rare and considered a disease of young females (9:1), predominantly below the age of 40 years, with a worldwide incidence rate of 1 to 2 per million[3,4]. The disease has a worldwide distribution, mainly seen in Asia[4]. Our patient was female and aged below 40 years in keeping with known literature. A review on vasculitis in Africa found it more in females affecting mainly the aortic arch and its branches. The most common presentations were intermittent claudication and hypertension[4]. Our female patient had claudication symptoms; the site affected was part of the aortic arch. Diagnosis is established using The American College of Rheumatology criteria (3 of 6 are necessary). The presence of any three or more criteria yields a sensitivity of 90.5% and a specificity of 97.8%[5]. Our patient met 4 out of the 6 criteria. The criteria are as follows:

- Age of 40 years or younger at disease onset
- Claudication of the extremities
- Decreased pulsation of one or both brachial arteries
- Difference of at least 10mm/Hg in systolic blood pressure between arms
- Bruit over one or both subclavian arteries or the abdominal aorta.
- Arteriographic narrowing or occlusion of the entire aorta, its primary branches, or large arteries in the upper or lower extremities that is not due to arteriosclerosis, fibromuscular dysplasia, or other cause

Early identification of Takayasu can be a challenge more especially in a tropical environment where infectious diseases commonly cause fever and malaise contributing to the risk of a delayed diagnosis. During the asymptomatic phase only the incidental finding of a vascular abnormality will alert the clinician (10%). About 15-50% of patients initially experience non-specific symptoms such as headache (50-70%), malaise (35-65%), and arthralgia (28-75%)[6-8]. Other symptoms include weight loss, fever, dizziness, syncopal attacks, and numbness of the limbs[6-8]. The clinical progression of the disease is divided into an early active inflammatory and late chronic phase[6-8]. The active phase usually follows a remitting and relapsing course and may last a few months to years. Our patient had prodromal symptoms (variously attributed to malaria and typhoid) for about three years before the ischaemic event. The only possible route to a Takayasu diagnosis at this stage is to detect a vascular abnormality via absent pulses, carotid bruits or a significant upper limb differential BP reading but note that neither of the latter two is a routine in clinical practice.

However, all of these signs were present when our patient presented with her stroke and raised inflammatory markers. We were able to delineate with imaging the precise vascular gross pathology and brain damage. This are expensive tests and inevitably there will be restricted access in a resource poor setting. Nevertheless within the context of a sound clinical diagnosis treatment with high corticosteroids should commence immediately.

The specific symptoms are dependent on the region of the aorta predominately affected. With the aortic branches, malaise, headache, and dizziness are common early symptoms progressing to visual and orientation disturbances, syncopal attacks, and decreased or absent pulses in upper extremities[9-13]. Our patient had most of these symptoms due to carotid vessel involvement.

Involvement of the aortic arch may cause congestive heart failure and arterial hypertension. Angina and myocardial infarction are complications of coronary arteritis. Abdominal aortitis can present with pain, vomiting, and nausea, while unexplained arterial hypertension and chronic renal disease result from renal artery involvement[9-13].

Our patient presented with a stroke in a young person (below the age of 45 years). Strokes in young people contribute about 5% of the total strokes. About half of the patients with Takayasu's present with neurological deficits[14]. Ten percent of these patients have strokes presenting as hemiparesis and hemi hypesthesia. The most commonly affected sites are the basal ganglia and watershed zone[14]. Our patient had an infarct in the middle cerebral artery territory, keeping with what is known in the literature.

Treatment of Takayasu arteritis is mainly by immunosuppression decrease to eliminate inflammatory activity. The therapy of choice is high-dose steroids but with a high relapse up to 50%. Methotrexate, azathioprine, and cyclophosphamide are often added to the steroid regimen. There is a role for IL-6 and TNF inhibitors in management especially for inadequate response to other immunosuppressive therapies[15]. These treatment options can improve the 5-year survival rate to 94%[7,8]. Surgery has a role, especially in the presence of symptomatic stenotic lesions, for example, uncontrolled hypertension secondary to renal artery stenosis and symptomatic carotid artery disease[7,8,11,15]. The procedures include endovascular revascularization procedures like bypass grafts, patch angioplasty, endarterectomy, percutaneous transluminal angioplasty, or stent placement[7,8,11,15].

Conclusion

In a tropical area like Kenya, where infection would be high on the list, Takayasu's should be considered in a young female who presents with stroke, especially in the background of recurrent fever, malaise, and high inflammatory markers. Examination of the patient can give additional clues, from the blood pressure and pulse differentials on the limbs to the presence of bruits over blood vessels, especially the aorta and its main branches. Early diagnosis helps improve the outcomes and prevent morbidity from the disease, as in our patient who had been unwell for three years before the diagnosis was made following an ischemic stroke. Diagnosis of the disease has its challenges, from the low index of suspicion to the high costs of investigating and managing these patients. More studies on Takayasu's needs to be done in Kenya to raise awareness about the disease and to have a better picture of the disease presentation and prognosis in this part of the world.

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