

# CASE REPORTS

## Takayasu's arteritis (pulseless disease) in Uganda

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### ABSTRACT

We report herein the case of a 23 year old woman who was referred to Mulago National Referral and Faculty of Medicine Makerere University Teaching Hospital because of sudden collapse, left sided weakness and headache for management. The patient underwent a battery of investigations but died five days after admission. The post mortem findings were extensive infarction of the right cerebrum extending from parietal to occipital lobes. There was thickening of the wall and complete obliteration of the right common carotid artery. The left common carotid artery was severely stenosed with marked thickening of the wall. The left subclavian artery was thickened and completely obliterated. Microscopically there was intimal thickening by fibrous connective tissue and infiltrate of chronic inflammatory cells in the walls of the three affected branches of the aorta. These gross and microscopic features were compatible with Takayasu's arteritis (TA).

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### INTRODUCTION

Takayasu's arteritis, also known as pulseless disease, is a rare inflammatory disease of large arteries especially the aorta and its larger branches. The disease most frequently affects the young with 80% being between 10 and 30 years of age and women account for 90% of the cases<sup>1</sup>. The aetiology of TA has not been elucidated but an autoimmune reaction to aortic tissue may be causative<sup>2, 3</sup>. This report describes the case of a patient with TA in an African Ugandan, which proved difficult to diagnose.

### Case Report

A 23-year-old female was referred to Mulago hospital with a history of having suddenly collapsed two days earlier and this collapse was accompanied

by left-sided weakness of the body and headache. She had had malaria treatment three weeks prior to the onset of the above symptoms. She had no significant family or past medical history. On physical examination the significant findings were left facial palsy with hypotonia in left upper and lower limbs. There was absent brachial pulse in the left arm and fundoscopy showed papilloedema. A series of investigations were done and these included a full hemogram that was within normal limits with ESR of 8mm/hour (Westgren). Doppler examination showed no flow in left common carotid artery and the left subclavian artery exhibited minimal flow. ECG and plain chest X-ray were normal. Serology for HIV type 1 was positive by ELISA.

At postmortem we found a well-nourished female (62kg and 167cm) about the stated age of 23. Significant findings were in the cranial cavity and the mediastinum. In the cranial cavity, there was flattening of gyri and widening of sulci of cerebrum and extensive infarction of left cerebrum extending from parietal to occipital region.

In the mediastinum there was thickening and obliteration of the right common carotid artery and thickening with severe narrowing of lumen of left common carotid artery (fig1).

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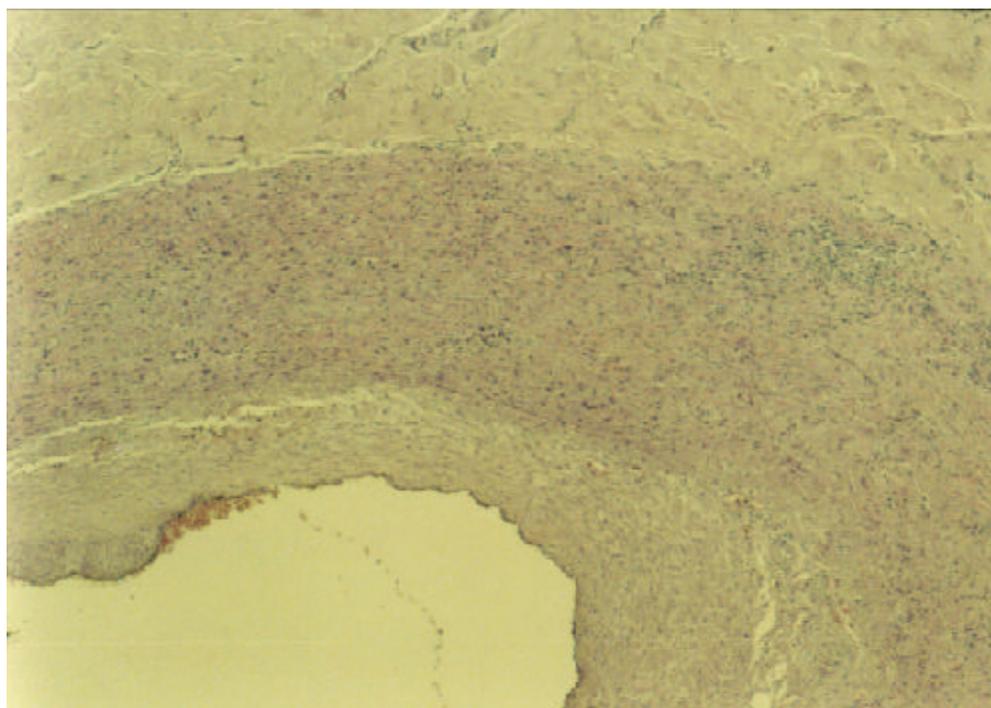
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**Fig. 1 Superior aspect of heart and aorta with thickened wall of right and left common carotid arteries x .7.**



The left subclavian artery was thickened and also completely obliterated. The aorta and right subclavian arteries were normal. The histological sections of the left and right common carotid and left subclavian arteries showed increase in intimal fibrosis with chronic inflammatory cells (fig 2). There was superimposed thrombosis in the right common carotid artery. The gross and microscopic features were compatible with TA.

**Fig 2. Sections of TA showing cellular accumulation of lymphoid cells x 100.**



## DISCUSSION

Takayasu's arteritis, first reported in Japan, is most frequently found in Asia while very uncommon in Europe and America. In Africa the disease is not as common as in Asia. Nair et al (2000)<sup>4</sup> while reviewing records of patients with spontaneous carotid artery aneurysm found out of 29 African patients, TA was diagnosed in two cases. Literature review has found no case of TA in Ugandan Africans. Takayasu's arteritis is now considered a widespread disease and not confined to arteries and therefore presents with protean clinical manifestations. Strachan (1964)<sup>5</sup> had classified the clinical presentation of TA into the early systemic or pre-pulseless phase and late occlusive phase also known as the pulseless phase. The pre-pulseless phase is characterized by fatigue, fevers, night sweats, anemia and a persistent ESR among many other non specific symptoms. The above symptoms were not elicited in the clinical history of our patient and this is probably because of recall bias. The late occlusive phase presents with clinical manifestation depending on the location and severity of the aortic branch lesions and may include intermittent leg claudication, intestinal infarction, renal hypertension, myocardial infarction dizziness, faint, coma and even death. There may also be a persistent rise of ESR in the occlusive phase<sup>6</sup>. This patient presented with left-sided weakness and pulseless left branchial pulses that were indicative of occlusive phase. Surprising the ESR was low in our case but it appears a high ESR is due to anemia that commonly occurs in these patients<sup>5</sup>.

Diagnosis of TA may be difficult as presented in this case because of the multi-system involvement. Currently it appears the mainstay in the diagnosis is radiological angiography especially CT scan, MRI and ECHO. Labios-Gomez et al (2002)<sup>7</sup> have stressed the role of magnetic nuclear angioresonance in the detection and follows up of this condition. A combination of difference in blood pressure and pulses between the patient's arms and angiographic findings would have led to the diagnosis of TA in our patient.

Treatment options include corticosteroids, percutaneous transluminal angioplasty or surgical bypass and certainly our patient would have benefited from the later procedure<sup>8</sup>.

Various forms of vascularitis involving small and medium-sized vessels have been described in HIV

infected persons<sup>9</sup>. Two theories have been proposed to explain the virally associated vascularitis. The virus may directly attack the vessel wall, or cellular or humoral immune mechanisms may lead to the formation of in situ or systemic circulating immune complexes that subsequently result in the development of vascularitis<sup>10</sup>. The demonstration of vascular deposits of HIV antigens, immunoglobulins and complement component suggests an immune complex deposition<sup>11</sup>. However only one case of concurrent TA and HIV has been reported in the world literature<sup>12</sup>. The possible autoimmune nature of TA and the frequent autoimmune phenomena in HIV infection could explain a possible relation between the two diseases. Clinician should be alerted to the possibility of arterial involvement in AIDS.

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