

# Takayasu's arteritis in an adult female from Cameroon: diagnosis via Doppler echocardiography

Leopold N Aminde<sup>1,2</sup>, Jean Jacques N Noubiap<sup>3,4</sup>, Noah Takah<sup>5</sup>, Tsi Njim<sup>6</sup> and Anastase Dzudie<sup>7,8</sup>

<sup>1</sup>Clinical Research Education, Networking and Consultancy, Douala, Cameroon

<sup>2</sup>Faculty of Medicine & Biomedical Sciences, School of Public Health, The University of Queensland, Brisbane, Australia

<sup>3</sup>Department of Medicine, Groote Schuur Hospital and University of Cape Town, Cape Town, South Africa

<sup>4</sup>Faculty of Health Sciences, University of Cape Town, Cape Town, South Africa

<sup>5</sup>School of Medicine, University of Glasgow, UK

<sup>6</sup>Regional Hospital Bamenda, Bamenda, Cameroon

<sup>7</sup>Department of Medicine, Faculty of Health Sciences, University of Cape Town, Cape Town, South Africa

<sup>8</sup>Department of Internal Medicine, Faculty of Health Sciences, University of Buea and Cardiology Unit, General Hospital Douala, Cameroon

**Corresponding author:** Leopold N Aminde. Email: amindel@gmail.com

## Lesson

Takayasu arteritis is rare in black people. Doppler echocardiography may assist in its diagnosis with good response to steroids.

## Keywords

Takayasu's disease, pulselessness, arteritis, doppler echocardiography, Cameroon

## Case presentation

A 30-year-old lady presented to our unit with a history of progressive ipsilateral left upper and lower limb weakness on exertion for over four years. This was associated with intermittent aches in affected limbs which worsened on exertion. She occasionally experienced shortness of breath but no orthopnoea, chest pain, cough nor leg swelling. She consulted at several primary care hospitals with no amelioration of symptoms.

On examination, she was stable and not in any distress, afebrile with absent pulses on left upper limb. There was no blood pressure reading on affected limb but the right arm had a blood pressure of 169/79 mmHg and pulse of 69 beats per minute. There was a right carotid bruit and very faint left carotid pulse. The rest of the peripheral pulses were normally palpable. Heart, neurological and the rest of the examination was unremarkable. Investigations included: a normal full blood count; raised erythrocyte sedimentation rate: 25 mm at first hour (normal  $\leq 10$  mm) and 59 mm at second hour (normal  $\leq 25$  mm); normal C-reactive protein: 0.29 mg/L (normal  $\leq 5$  mg/L); normal serum creatinine: 7 mg/L; negative hepatitis B and C serological tests; normal thyroid function

tests; negative anti-nuclear antibody test; and negative Venereal Disease Research Laboratory and Treponema pallidum hemagglutinin antigen tests. Chest X-ray, electrocardiography and cardiac echography were normal. Doppler vascular assessment revealed significant reduction of abdominal aortic lumen from inflammation, occlusion of left carotid, subclavian, axillary and vertebral arteries. There was also involvement of the brachial, ophthalmic and radial arteries. A reverse flow pattern was observed in the left common carotid, with perfusion of left upper limb from brain (Figures 1 to 5). With extensive vascular lesions, pulselessness, carotid bruit and blood pressure discrepancies, a diagnosis of Takayasu's arteritis (TA) was proposed. She was placed on steroids and at two months follow-up, reported marked amelioration of symptoms, but was later lost to follow-up.

## Discussion

Takayasu's disease, variably known as 'Takayasu's arteritis', 'pulseless disease', 'occlusive thromboarteriopathy', 'Martorell syndrome' or 'non-specific aortoarteritis', is a chronic vascular inflammatory condition affecting large arteries predominantly the aorta and its branches.<sup>1</sup> The International Chapel Hill Consensus conference of 2012 nomenclature system divided vasculitides into large-vessel, medium-vessel and small vessel vasculitis; TA was grouped as large-vessel vasculitis together with giant cell arteritis.<sup>2</sup> A female (in their second and third decades) predominance has generally been recorded in a majority of the cases with female-to-male ratios as high as 9:1 in Japan.<sup>3</sup> In fact, in a majority of patients younger

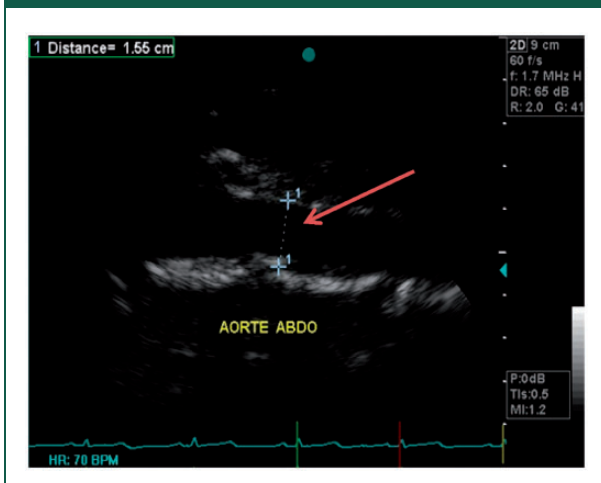
than 50 years, TA has been shown to be the most frequent cause of systemic vasculitis affecting the aorta.<sup>4</sup> Despite limited epidemiological studies with scant reports from Africa, TA is relatively more common in Asia with estimated population prevalence of 0.004% in Japan.<sup>5</sup>

TA generally involves two stages: the pre-pulseless (pre-stenotic) phase, in which the patient has non-specific symptoms, followed by the pulseless (stenotic/occlusive) phase, in which there is significant vascular insufficiency with diminished to impalpable pulses (commonly upper limb), and bruits may be

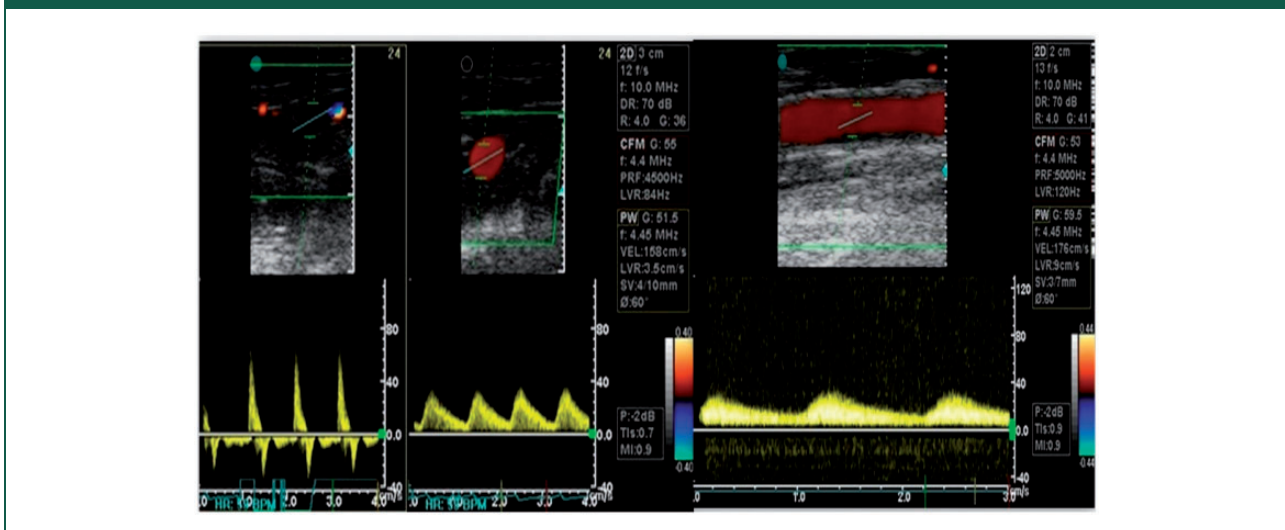
heard over the affected vessels.<sup>1</sup> The distribution of vascular affection of the disease also varies by region, with cervical and thoracic arterial lesions being more common in Japan and South America contrary to abdominal lesions in other Asian countries.<sup>5</sup> The coronary ostia, pulmonary arteries and renal arteries may also be involved. Thus, clinical manifestations are variable, ranging from being asymptomatic to subclavian steal syndrome and even cerebrovascular accidents.<sup>6</sup>

The diagnosis of TA requires at least three of the American College of Rheumatology criteria (Table 1)<sup>7</sup> to be present. This criterion is said to have a sensitivity of 91% and specificity of up to 98%. Our patient fulfilled all criteria except arteriographic evidence. Magnetic resonance angiography, CT imaging and histology are generally required for confirmation of the disease. Histology often shows granulomatous inflammatory infiltrates with macrophages, B- and T-lymphocytes, eosinophils and plasma cells with presence of giant cells. This granulomatous inflammation in vessel adventitia eventually progresses to panarteritis.<sup>6</sup> Blood tests also show raised inflammatory markers. In the late phases of the disease, four types have been described<sup>8</sup>; type 1 – classic pulselessness involving brachiocephalic trunk, carotid and subclavian arteries; type 2 – a combination of type 1 and 3; type 3 – atypical coarctation involving thoracic and abdominal aortas distal to the arch and its major branches; type 4 – extensive dilation of the length of the aorta and its major branches. The most frequent of them is type 3 (65% of cases) commonly involving subclavian, left common carotid, brachiocephalic trunk, renal,

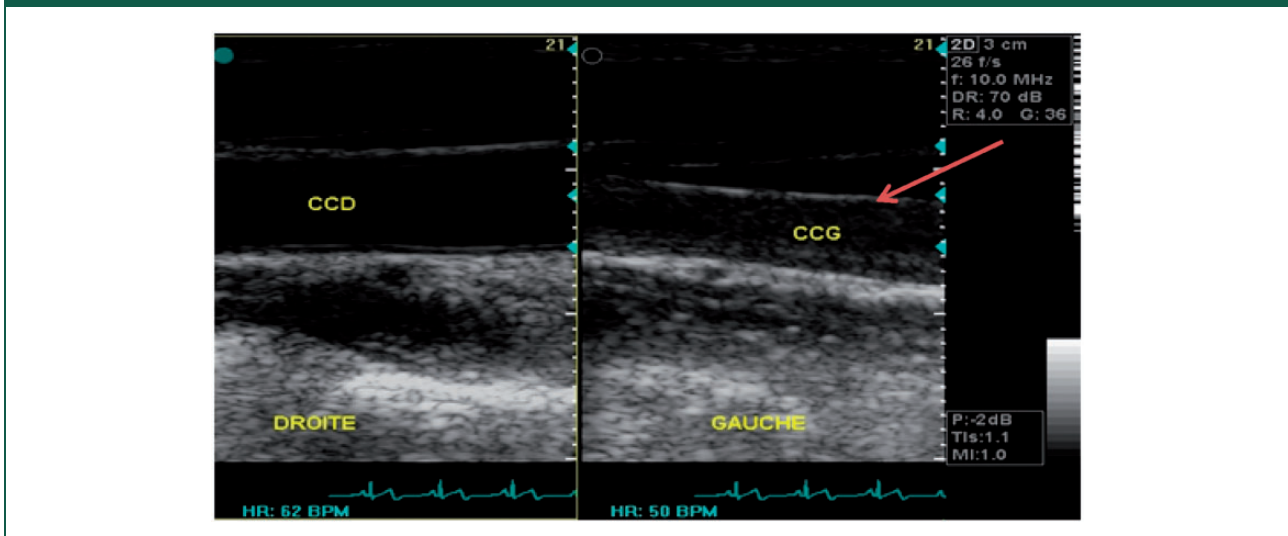
**Figure 1.** Echographic image of the abdominal aorta showing reduction in lumen from inflammation.



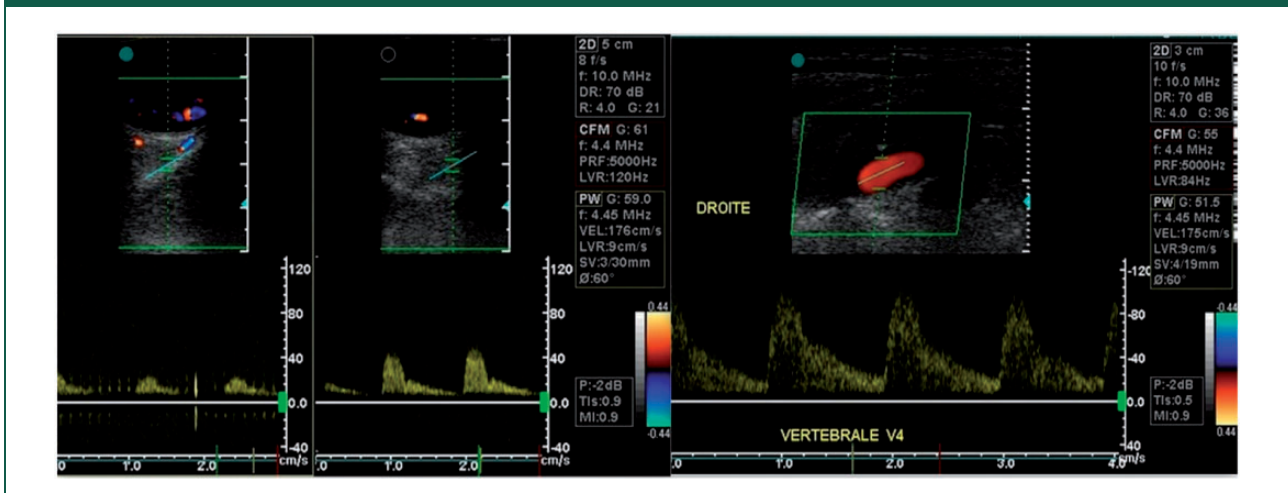
**Figure 2.** Doppler pulsed wave image showing luminal reduction and decreased flow velocity in axillary (left panel) and Brachial (right panel) arteries.



**Figure 3.** Echographic image showing severe occlusion of the left common carotid artery (arrow).



**Figure 4.** Doppler pulsed wave image showing decreased flow velocities in left & right ophthalmic arteries (left panel) and vertebral artery (right panel).

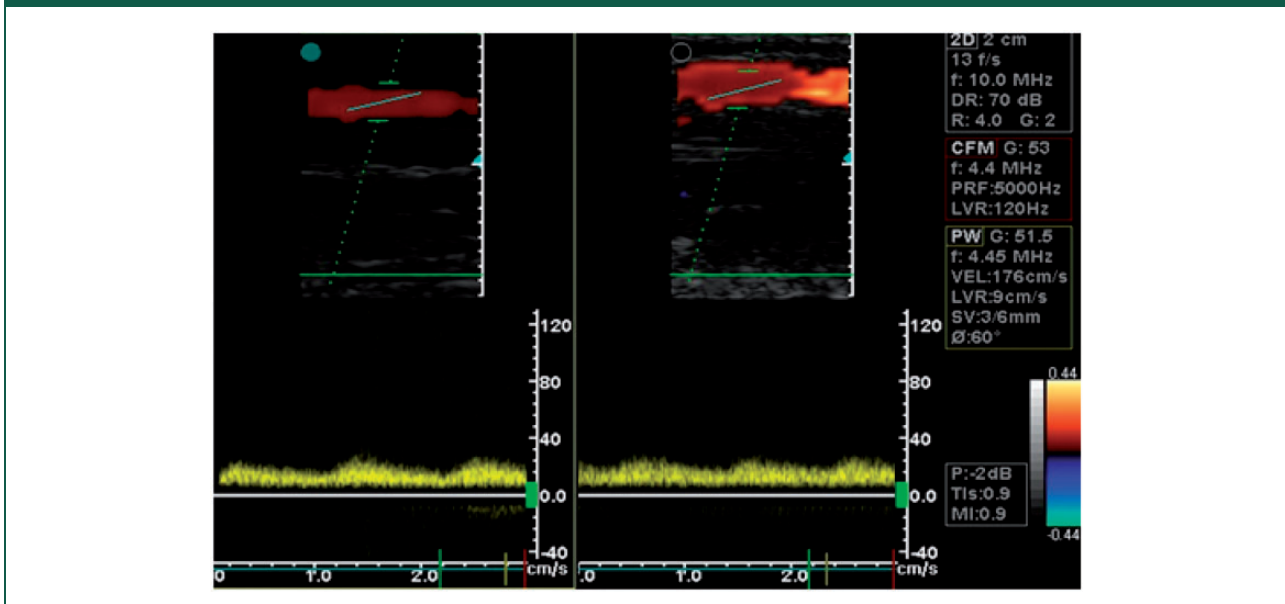


coeliac, superior mesenteric and pulmonary arteries. Quite rarely it may involve the axillary, brachial, vertebral, coronary and iliac arteries.<sup>8</sup>

Recently, doppler echocardiography has been demonstrated to be of high diagnostic use for even early (pre-stenotic) TA.<sup>9</sup> Schmidt et al. demonstrated good correlation of colour doppler sonography with angiographic features of patients with non-specific symptoms and features of systemic inflammation. They suggested that colour doppler imaging of primary extra cranial branches of aortic arch (subclavian, carotid) could be very

crucial for the diagnosis of TA even in the pre-stenotic phase of the disease. The differential diagnoses for TA are very wide but closely related are giant cell arteritis (with similar histologic features) and sarcoidosis.<sup>6</sup>

Generally, management involves the use of corticosteroids. However, some studies have suggested IL-6 receptor inhibitors, B-cell depletion, cytotoxic drugs (especially for steroid resistant or relapsing cases) and anti-tumor necrosis factor agents.<sup>10</sup> Open and endovascular graft and bypass surgery have proven a long-term beneficial effect especially

**Figure 5.** Doppler image showing significant reduction in flow velocities in left radial (left panel) and ulnar (right panel) arteries.**Table 1.** American College of Rheumatology (ACR) 1990 criteria for diagnosis of TA.<sup>7</sup>

i.	Age at onset less than or equal to 40 years
ii.	Claudication of extremities – development and worsening of fatigue and discomfort in muscles of 1 or more extremity while in use
iii.	Decreased brachial artery pulse
iv.	Systolic blood pressure difference greater than 10 mmHg between arms
v.	Bruit over subclavian arteries or abdominal aorta
vi.	Arteriographic abnormality: narrowing or occlusion of the entire aorta, its primary branches or large arteries in the proximal upper or lower extremities, not due to arteriosclerosis, fibromuscular dysplasia or similar causes; changes usually focal or segmental

in cases with severe stenosis; however, appropriate and timely pharmacologic therapy may limit vascular interventions. The follow-up of patients should include control of cardiovascular risk factors especially hypertension, dyslipidemias and also anti-platelets aggregants for stroke prevention. Diagnosis of TA in primary care facilities especially in resource-limited settings like ours is usually very challenging with late presentation to hospital, absence of state-of-the-art diagnostic imaging tools; with patients at risk of developing advanced vascular lesions and exposing them to fatal complications.

TA is rare among the Africans. In resource-limited settings, thorough clinical and doppler echocardiographic assessment is important in the diagnosis of TA with relatively good response to steroids.

#### Declarations

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