

# Takayasu Disease Revealed by Arterial Hypertension in a 47-Year-Old Man

Bah Mamadou Bassirou<sup>1\*</sup>, Baldé Thierno Hamidou<sup>2</sup>, Wann Thierno Amadou<sup>3</sup>, Baldé Elhadj Yaya<sup>1</sup>, Baldé Aissatou Tiguidanké<sup>1</sup>, Koné Alpha<sup>4</sup>, Doumbouya Amadou Diouldé<sup>1</sup>, Diallo Hafsadou<sup>1</sup>, Barry Ibrahima Sory<sup>1</sup>, Samoura Sana<sup>1</sup>, Diakité Souleymane<sup>1</sup>, Béavogui Mariam<sup>1</sup>, Baldé Mamadou Dadhi<sup>1</sup>

<sup>1</sup>Cardiology Department, Ignace Deen Hospital, Conakry, Guinea

<sup>2</sup>Department of Imaging, Ignace Deen Hospital, Conakry, Guinea

<sup>3</sup>Department of Internal Medicine, Donka Hospital, Conakry, Guinea

<sup>4</sup>Cardiology Department, Donka Hospital, Conakry, Guinea

Email: \*bachirov6@gmail.com

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## Abstract

Takayasu disease is a chronic vasculitis of the large vessels. It is rare in sub-Saharan Africa and probably under-diagnosed. Arterial hypertension (AH) is a frequent finding. We report the case of a 46-year-old man with Takayasu disease revealed by hypertension. The American College of Rheumatology (ACR) criteria were used for diagnosis, with 4 out of 6 criteria. Echodoppler and angioscanner showed signs of left subclavian stenosis and circumferential, regular arterial thickening. He had a biological inflammatory syndrome. He was treated with prednisone and amlodipine. After 6 months of follow-up, he was diagnosed with tuberculosis and put on anti-tuberculosis treatment.

## Keywords

Takayasu Disease, Hypertension, Man

## 1. Introduction

Takayasu disease is an inflammatory arteritis of large- and medium-caliber vessels mainly affecting the aorta, its main dividing branches and the pulmonary arteries [1].

Its incidence varies between 1.2 and 2.6 cases/million/year. The disease is more common in Japan, South-East Asia and Latin America [1] [2] [3]. A few hospital series have been reported in North Africa [4] [5]. In Africa south of the Sahara, it occurs but is rarely described, and is probably under-diagnosed. A few

clinical cases have been described [6] [7] [8] [9] [10]. The condition affects women in almost 90% of cases, usually before the age of 40 [1].

Thickening of the vascular wall is the most characteristic early sign of the disease, progressively leading to stenosis, thrombosis and sometimes the development of aneurysms [11].

Arterial hypertension (AH) is common, sometimes revealing the disease (33% to 76% in the literature) [12].

We report the case of Takayasu disease revealed by hypertension in a 46-year-old man.

## 2. Observation

Mr. ISS, aged 46, with no previous history of cardiovascular disease, was referred from Siguiri prefectural hospital (726.4 km from the capital Conakry) for treatment of newly diagnosed hypertension. The questioning revealed tingling in the left arm, muscle cramps, headaches, dizziness and physical asthenia. The tingling and muscle cramps in the left arm began at the age of 38.

Physical examination revealed asymmetric blood pressure. Blood pressure was 160/100 mmhg in the right arm and 85/50 mmhg in the left arm. The heart rate was 68 beats/min. Cardiac auscultation was normal. The humeral and radial pulses were clearly felt on the right and decreased on the left. Auscultation of the great vessels did not reveal any murmur. Examination of the other equipment was unremarkable. Electrocardiogram and transthoracic echocardiography were normal.

The arterial ultrasound of the supra-aortic trunks (**Figure 1**) showed regular circumferential thickening of the left common carotid artery, without haemodynamic stenosis. The arterial Doppler ultrasound of the upper limbs (**Figure 1**) showed a demodulated, damped Doppler flow at the expense of the axillary, humeral, radial and ulnar arteries; indirect signs of stenosis of the left subclavian artery.

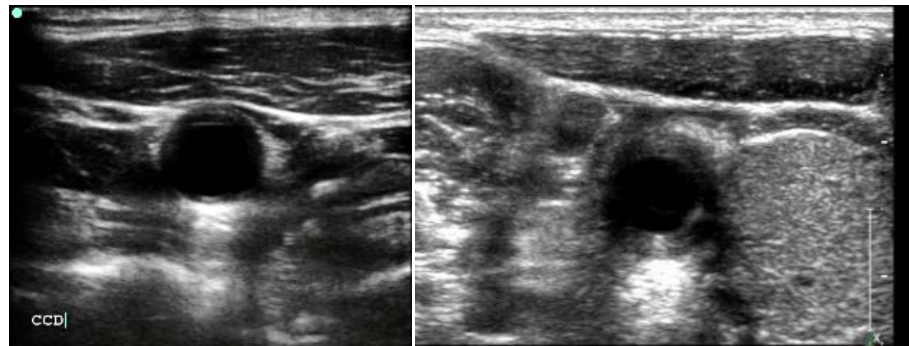
The angioscan (**Figure 2**) showed circumferential and regular parietal thickening involving the aorta and the 2 subclavian arteries, resulting in long and tight stenosis of the left subclavian artery extending over 31 mm.

The biological work-up showed a biological inflammatory syndrome with a positive CRP of 18 mg/l, and an accelerated SV of 85 mm/hour, fasting blood glucose was 0.9 g/l, and the lipid, renal and liver work-up was normal.

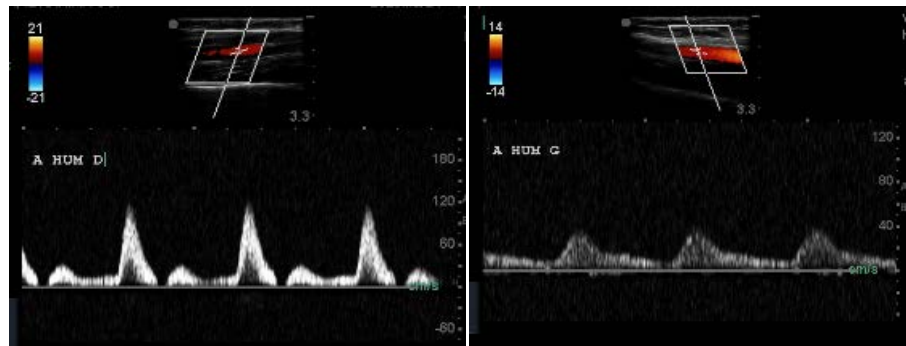
The diagnosis of Takayasu's disease was based on the presence of 4 out of 6 American College of Rheumatology (ACR) criteria: age of onset less than 40 years; vascular claudication of the left upper limb; decreased brachial pulse; blood pressure difference greater than 10 mmHg.

The patient was put on medication: Prednisone 40 mg/day as initial treatment, then gradually reduced to 10 mg/day. He also took Amlodipine 10 mg/day, Aspirin 100 mg/day and Atorvastatin 40 mg/day.

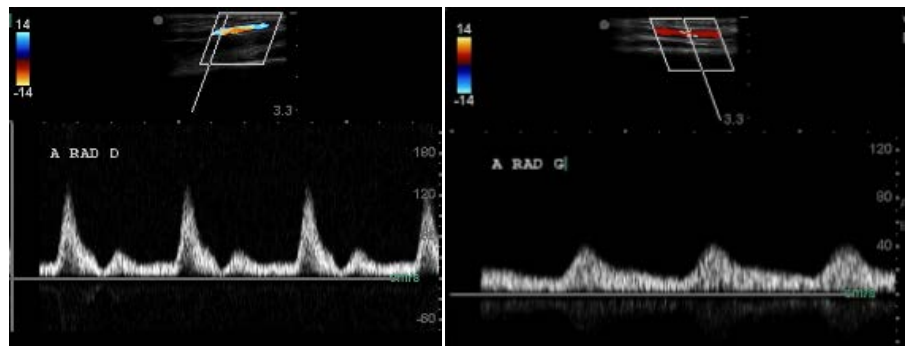
At 6 months, the patient developed a right pleural effusion which was analyzed as extrapulmonary tuberculosis. He was put on anti-tuberculosis treatment.



(a)

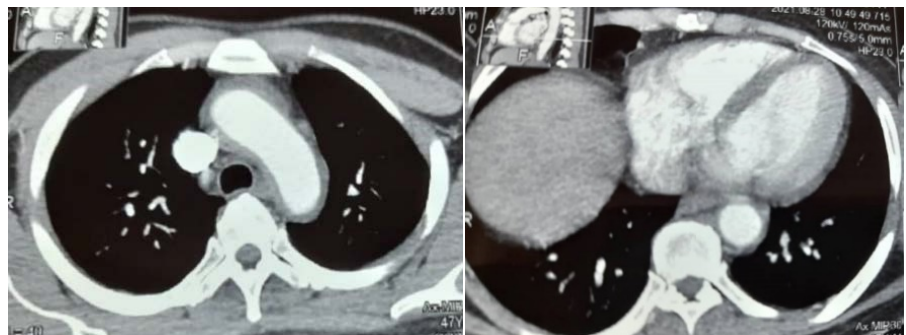


(b)



(c)

**Figure 1.** Comparative right and left upper limb Doppler ultrasound showing circumferential thickening of the left common carotid artery (a) demodulated and damped Doppler flow of the humeral (b) and left radial (c) arteries. (a) Left and right common carotid arteries; (b) Right and left humeral arteries; (c) Right and left radial arteries.



(a)

(b)



(c)

**Figure 2.** Thoracic angioscan axial sections and 3D reconstruction showing (a) and (b) circumferential thickening of the aorta and (c) long stenosis of the proximal left subclavian artery (arrow).

### 3. Discussion

Takayasu disease is a rare vascular disease [12]. In Guinea, only one case of Takayasu disease has been reported in the literature [10].

This disease predominates in the female and oriental populations [5]. Its occurrence in our patient is therefore a rare case.

Historically, immunohistochemical studies of infiltrating cells from the aortic tissue of patients with Takayasu disease showed infiltration by macrophages, CD4+ T cells, CD8+ T cells,  $\gamma\delta$ T cells, natural killer (NK) cells and neutrophils, and this hypermetabolic infiltrate determines the 18F-Fluorodesoxyglucose uptake that may be seen when positron emission tomographies are performed in Takayasu disease [13].

The etiology of this inflammatory condition remains unknown. However, close epidemiological links between tuberculosis and Takayasu disease have been reported in the literature [14] [15]. Indeed, the prevalence of tuberculosis is higher in Takayasu disease than in the general population, ranging from 6.3 to 20% for active tuberculosis and 20% to 82% for latent tuberculosis [15] [16]. However, a cause-and-effect relationship has not been established. Tuberculosis was diagnosed in our patient after 6 months of follow-up with corticoid treatment.

Takayasu disease progresses in two phases. The first phase, known as pre-occlusive, is characterized by systemic symptoms: fever, night sweats, weight loss, arthromyalgia and sometimes erythema nodosum.

The second phase, known as occlusive, is characterized by the vascular lesions of Takayasu's arteritis. It is characterized by arterial manifestations affecting the upper limbs, such as claudication in 30% to 62% of cases [12] [17], abolition of a pulse in 50% to 96% of cases [12] or asymmetric blood pressure [18].

In our patient, stenosis manifested itself mainly as tingling in the left arm associated with muscle cramps.

High blood pressure is common in Takayasu's disease (33% - 76%) and may be due to a variety of causes: renal arterial damage, aortic pseudo coarctation and parietal rigidity secondary to vascular damage, and widening of the blood pressure differential in the case of aortic insufficiency [12] [19]. Our patient presented with hypertension and anisotension. In our case, this was a sign of the disease.

Involvement of the arteries of the upper limbs is the most common, often revealing the disease, particularly that of the subclavian arteries [19]. Other locations are also possible. These include carotid, aortic and renovascular involvement [12].

The disease is diagnosed on the basis of a combination of clinical, biological, radiological and more rarely, histological criteria [20]. The diagnostic criteria used are those of the ACR [21] or those of Ishikawa modified by Sharma [22], which are mainly clinical and supplemented by arteriography. The presence of 3 or more ACR criteria allows the diagnosis to be made with a sensitivity of 90.5% and a specificity of 97.8% [23]. Our patient had 4 out of 6.

As arteriography is used less and less because of its invasive nature and inaccessibility, it is tending to give way to other examinations, in particular echodoppler, angioscanner, MRI and PET scan.

Echo Doppler provides precise information on the arterial wall, particularly during inflammatory outbreaks, showing regular hypoechoic circumferential thickening of the affected areas and the presence of long, regular stenoses. It can also provide haemodynamic information on stenotic lesions [1]. Circumferential thickening of the left common carotid artery was found in our patient and was associated with indirect signs of stenosis of the left subclavian artery, with demodulated, damped Doppler flow in the axillary, humeral, radial and left ulnar arteries.

MRI angiography and angioscan with or without 18 FDG positron emission tomography of the aorta and its branches are recommended for all cases of large-vessel vasculitis [23].

In Takayasu's disease, angioscan and MRI show circumferential thickening of the aortic wall. The coexistence of stenosis, dilatation and aneurysm of the aorta and/or its main branches is of great diagnostic interest. Involvement of the pulmonary artery may be found in 30% - 50% of cases, occurring late and also manifesting as a parietal thickening, aneurysm or stenosis, or even occlusion [1]. Our patient presented with 3.7 mm circumferential parietal thickening of the aorta creating a "double ring" appearance associated with significant stenosis of the proximal left subclavian artery.

18F-FDG-PET with CT is useful for diagnosis and to assess response to anti-inflammatory treatment.

Initial treatment options include high-dose glucocorticoid therapy (prednisone 40 - 60 mg/d), alone or in combination with immunosuppressants, with Tocilizumab or Methotrexate as alternatives [22]. Once the acute phase has been controlled, glucocorticoid reduction should be initiated to achieve a target pred-

nisone dose of 15 - 20 mg/d over 2 - 3 months, then  $\leq 10$  mg/d after 1 year [23].

Antihypertensive drugs are indicated for patients with elevated blood pressure figures.

Antiplatelet or anticoagulant therapy should not be used routinely. In specific situations such as vascular ischaemic complications or a high risk of cardiovascular disease, these may be considered on a case-by-case basis [24].

Treatment of co-morbidities, in particular active tuberculosis, as in the case of our patient, is part of the overall management of Takayasu disease.

In patients who are in remission and have complications of the aorta or its branches that do not warrant urgent intervention, endovascular interventional treatment or surgical repair should be determined by a multidisciplinary team including the vascular surgeon, vascular physician, cardiology and radiology specialists [23].

#### 4. Conclusion

Takayasu disease is a rare vasculitis that is often under-diagnosed. Although it is more common in young women, it can also affect men. High blood pressure is often the tell-tale sign of this disease. Early diagnosis, ideally at a preclinical stage, is crucial, and treatment is mainly based on the use of corticosteroids, as well as the management of associated co-morbidities.

#### Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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