

An Abdominal Aortic Aneurysm Revealing a Fatal Takayasu Disease

—A Case Report from the Libreville University Hospital Centre (CHUL)

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Abstract

Takayasu disease is inflammatory arteritis that predominantly affects large-calibre arteries. Its aetiology remains poorly understood. In sub-Saharan Africa, it remains an underdiagnosed condition. We report a case of Takayasu disease revealed by an abdominal aortic aneurysm. The patient was a 54-year-old woman admitted to hospital with severe abdominal pain, localised in the epigastric region and rated at 6/10 on the visual analogue scale, which was unresponsive to standard analgesics. Physical examination revealed asymmetric blood pressure readings. The electrocardiogram showed a regular sinus rhythm with a heart rate of 100 beats per minute and the presence of T-waves indicative of subepicardial ischaemia in the lateral region. The complete blood count showed pancytopenia, with white blood cells at 200/ μ l, haemoglobin at 7.9 g/dl, and thrombocytopenia at 5000/ μ l. C-reactive protein was positive at 160 mg/l and the Coombs test was positive. A chest and abdominal CT angiogram revealed an enlargement of the aortic arch extending into the descending and abdominal aorta (57 mm), with significant parietal thickening reducing the vascular lumen and diffuse parietal microcalcifications. The patient was immediately started on prednisone at a dose of 40 - 60 mg per day. The course of the illness was marked 72 hours later by the onset of heart failure with haemodynamic instability. Takayasu disease is being reported with increasing frequency in sub-Saharan Africa. It generally presents cardiovascular symptoms, and thoraco-abdominal-pelvic CT angiography is of considerable value in the diagnosis.

Keywords

Aneurysm, Abdominal, Takayasu, Fatal Outcome, CHUL

1. Introduction

Takayasu disease is an inflammatory arteritis that primarily affects large-calibre arteries [1]. It is a non-specific panarteritis with an adventitial origin, the aetiology of which remains poorly understood [2]. The incidence of the disease varies worldwide, ranging from 100 cases per million per year (Japan) to 2.6 cases per million per year (United States) [3] [4].

In sub-Saharan Africa, it remains an underdiagnosed condition, making it a rare disorder [5], with only a few cases described in the literature. Nevertheless, aneurysmal involvement in Takayasu arteritis remains rare. We report a case of Takayasu disease presenting with an abdominal aortic aneurysm.

2. Clinical Presentation

The patient was a 54-year-old woman with no significant past medical history and no cardiovascular risk factors, who was admitted to hospital with severe abdominal pain. The pain was localised in the epigastric region, rated at 6/10 on the visual analogue scale, unresponsive to standard analgesics and without radiation. The clinical examination revealed a blood pressure of 134/86mmHg in the left arm and 105/75mmHg in the right arm, with a more prominent and throbbing radial pulse on the left. Auscultation revealed a left para-aortic murmur. The electrocardiogram showed a regular sinus rhythm with a heart rate of 100 beats per minute and the presence of subepicardial ischaemic T-waves in the lateral region (**Figure 1**). Myocardial enzymes, notably troponin, and D-dimer levels were negative. The complete blood count revealed pancytopenia with a white blood cell count of 200/ μ l, a haemoglobin level of 7.9 g/dl, mean corpuscular volume at 75 fl and mean corpuscular volume at 22 pg. This represented hypochromic microcytic anaemia and thrombocytopenia at 5000/ μ l. C-reactive protein was positive at 160 mg/l and the Coombs' test was positive. Renal function tests revealed impaired renal function with a glomerular filtration rate of 25 ml/minute. Cardiac Doppler ultrasound showed a normal aortic root diameter of 27 mm (**Figure 2**).

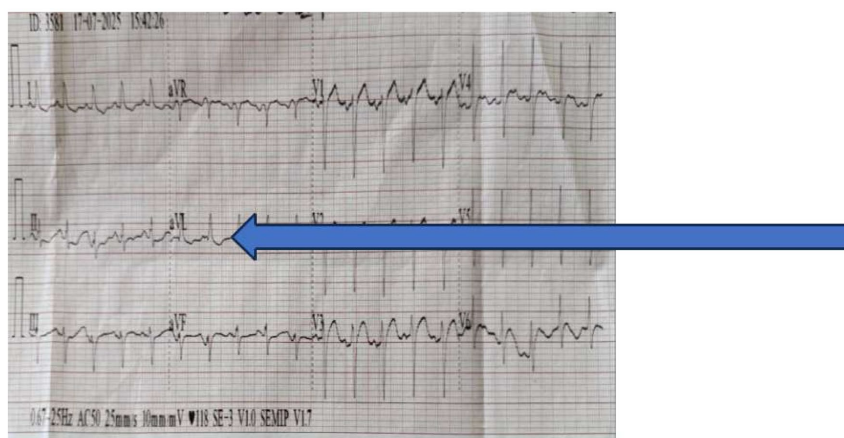


Figure 1. An electrocardiogram showing T-waves indicative of subepicardial ischaemia in the lateral region.

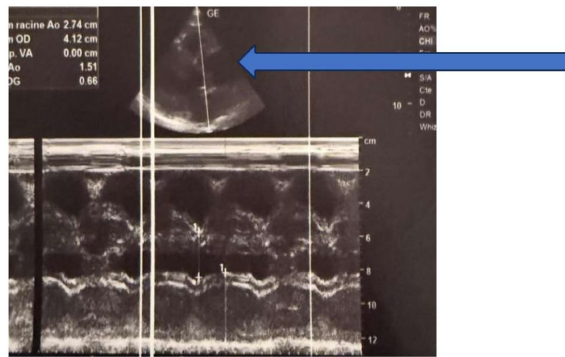


Figure 2. Cardiac Doppler ultrasound image showing an aortic diameter of 27 mm at the level of the Valsalva sinus.

These pains were accompanied by diffuse cramps in the lower limbs, prompting a thoracic CT angiogram, which revealed an enlargement of the aortic arch extending along the descending and abdominal aorta (57 mm) as far as the bifurcation of the iliac arteries, strongly suggestive of an aneurysm (**Figure 3**) with significant parietal thickening reducing the vascular lumen and diffuse parietal microcalcifications (**Figure 4**).

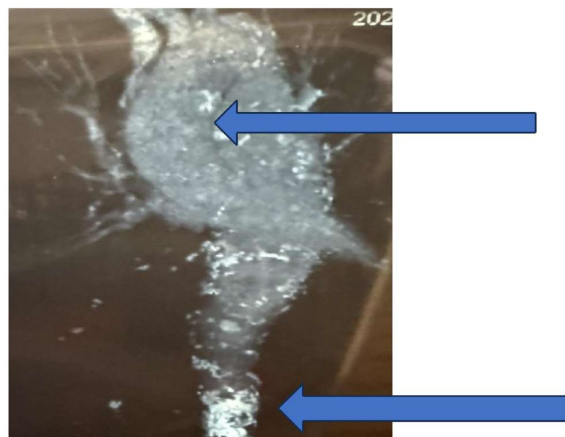


Figure 3. Thoraco-abdominal CT angiogram showing an increase in the volume of the aortic arch extending into the descending and abdominal aorta (57 mm).



Figure 4. Thoraco-abdominal CT angiogram showing diffuse parietal microcalcifications highly suggestive of Takayasu disease.

In view of these findings, a diagnosis of Takayasu arteritis was made, and the patient was immediately put on prednisone at a dose of 40 - 60 mg/day. The course of the illness was marked 72 hours later by the onset of respiratory distress with haemodynamic instability. Following resuscitation efforts, the patient died.

Ethical Considerations

Written consent was obtained from the patient for the use of her clinical and paraclinical data, as well as for the publication of this case report. The patient's confidentiality and anonymity were strictly maintained.

3. Discussion

Takayasu disease is an inflammatory arteritis that primarily affects the aorta and its main branches; its aetiology remains unknown [1]. When the condition affects the abdomen, it typically presents as severe abdominal pain, usually localised in the epigastric region. It is therefore particularly important to consider this diagnosis in a patient with no known cardiovascular risk factors. It is frequently reported in the literature that Takayasu disease predominantly affects women under the age of 40 in nearly 90 per cent of cases [6] and is thought to mainly affect patients of Asian, African or South American origin, with an incidence of between 1.2 and 3.6 cases per year per million inhabitants [7]. In our case, the patient was a 54-year-old woman. A few so-called "late-onset" forms, occurring after the age of forty, have been described in 25 per cent of cases [7]. A positive Coombs test could explain the presence of autoimmune haemolytic anaemia. According to the 1990 ACR diagnostic criteria, at least three out of six criteria were met: the age at onset of symptoms was under 40 years according to the 1990 ACR criteria, asymmetry in blood pressure between the two arms, and the presence, on a chest CT angiogram, of an enlargement of the aortic arch extending into the descending and abdominal aorta (57 mm) as far as the bifurcation of the iliac arteries, with significant parietal thickening reducing the vascular lumen and diffuse parietal microcalcifications. The presence of bicytopenia and severe thrombocytopenia can be explained by several mechanisms, the primary one being bone marrow dysfunction. Systemic inflammation caused by Takayasu disease may affect bone marrow, preventing it from producing sufficient red blood cells and platelets, or splenic sequestration leading to thrombocytopenia and causing splenomegaly. These additional clinical features may indicate a turning point in the progression of the disease.

Thickening of the vascular wall is an early and characteristic sign, but the development of an aneurysm in the affected arterial territories increases the rates of morbidity and surgical mortality. Given the cost of the investigations, we were unable to look into other possible causes of the inflammation.

Takayasu disease progresses in two phases characterised by clinical manifestations: the pre-occlusive phase, which may be absent or mild. If signs are present, they manifest as a deterioration in general health, myalgia, arthralgia, fever, night

sweats, pleural effusion, pericarditis, episcleritis and erythema nodosum [8]; and the occlusive phase, which may be the sole phase of the disease from the outset, presenting as a vascular manifestation (ischaemic, stenotic, aneurysmal or dissecting) with or without an inflammatory syndrome. As regards our patient, we believe she was seen during the occlusive phase. A thoracic CT angiogram confirms dilation of the affected segment but also reveals thickening of the arterial wall [9]. When the disease is in a flare-up phase, treatment involves the administration of prednisone at a dose of 40 - 60 per day. In the majority of case series, corticosteroid therapy achieves remission in only 50 per cent of cases, hence the need for a combination with an immunosuppressant such as methotrexate, one injection per day [8]. The patient died from severe heart failure with haemodynamic instability, probably due to an aortic dissection resulting from an aneurysm rupture.

4. Conclusion

Takayasu disease is a condition that is being reported with increasing frequency in sub-Saharan Africa. Diagnosis is often delayed due to a lack of awareness of the condition. It generally presents with cardiovascular symptoms. A thoraco-abdominal-pelvic CT angiogram is of considerable value in diagnosis.

Conflicts of Interest

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

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