

Severe Aortorenal Involvement Revealing Takayasu's Disease: A Case Report

Mrich Hayate^{1*}, Sqalli Salma¹, Abounouh Soumia¹

¹Nephrology, Hemodialysis and Kidney Transplantation Department, Laayoune Regional Hospital, Morocco

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*Corresponding author: Mrich Hayate

Nephrology, Hemodialysis and Kidney Transplantation Department, Laayoune Regional Hospital, Morocco

Abstract

Case Report

Takayasu's disease is a rare inflammatory arteritis that mainly affects the aorta and its branches, most commonly affecting young women between the ages of 20 and 40. Renal involvement is common in this disease and, in some cases, can compromise its progression. It is most often linked to renal artery stenosis, which causes renovascular hypertension. Angiography is the gold standard test for establishing the diagnosis. Treatment is mainly based on corticosteroid therapy, sometimes combined with immunosuppressants, and in some cases, biotherapy or surgical intervention may be considered. We report the case of a 38-year-old female patient in whom investigation of high blood pressure associated with renal asymmetry confirmed the diagnosis of Takayasu's disease.

Keywords: High blood pressure, abdominal aortic occlusion, renal artery stenosis, large vessel vasculitis.

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INTRODUCTION

Takayasu's disease is a rare chronic inflammatory vasculitis of the large vessels, primarily affecting the aorta and its main branches. Its slow and insidious progression and long period of few symptoms explain the delay in diagnosis due to nonspecific clinical manifestations [1].

This condition mainly affects young women, with an estimated incidence of between 1 and 3 cases per million inhabitants per year in Europe. It is a rare but potentially serious disease [2], requiring early and appropriate management.

Inflammatory vascular lesions can progress to stenosis or occlusion, leading to multivisceral ischemic manifestations, particularly neurological and renal, as well as peripheral [3]. This diversity of clinical presentations is illustrated by the case of our patient.

The diagnosis is based on a comparison of clinical, biological, and imaging data, while management requires a multidisciplinary therapeutic strategy to limit complications and improve prognosis [3].

CASE REPORT

We followed a 38-year-old female patient for arterial hypertension treated with conversion enzyme

inhibitors for one year. She also had a history of COVID-19 infection treated at home a year and a half ago. The patient was admitted to the emergency department for severe renal failure complicated by hyperkalemia. The interview revealed two episodes of paroxysmal abdominal pain, relieved by painkillers, the last of which occurred one month ago.

On clinical examination, the patient was conscious, with a Glasgow score of 15/15, afebrile, eupneic, but had tachycardia at 105 bpm. Blood pressure was measured at 170/90 mmHg in the right arm and 156/78 mmHg in the left arm (asymmetry > 10 mmHg). An abdominal murmur and absence of femoral pulses were also noted, with coldness in the lower extremities.

Biologically, urea was noted at 0.72 mg/L and plasma creatinine at 48 mg/L with an estimated MDRD creatinine clearance of 12 mL/min/1.73 m². Potassium levels were elevated at 7.4 mEq/L, associated with metabolic acidosis at 17 mmol/L and sodium levels at 133 mEq/L. Twenty-four-hour proteinuria was measured at 0.64 g/day. CRP was negative with a sedimentation rate of 34 mm/h.

Renal ultrasound revealed renal asymmetry with the right kidney measuring 9.4 cm and the left kidney measuring 12.56 cm. Abdominal CT angiography revealed total occlusion of the subrenal abdominal aorta,

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associated with stenosis of the right renal artery (**Figure 1**).



Figure 1: Total occlusion of the abdominal aorta below the kidneys

Angiography of the supra-aortic trunks revealed severe stenosis of the left vertebral artery with an estimated reduction in caliber of more than 50%. Cardiac transthoracic echocardiography revealed dilated cardiomyopathy with preserved ventricular ejection fraction associated with aortic insufficiency. The etiological workup, including testing for antiphospholipid antibodies, was negative, as were

serological tests for syphilis, hepatitis, and retroviral infections.

Based on the clinical, biological, and radiological data, as well as the American College of Rheumatology (ACR) criteria (**Table I**), the diagnosis of Takayasu's disease was confirmed.

Table 1: Sharma Criteria for Diagnosis of Takayasu. Presence of two major, or one major and two minor criteria, or four minor criteria suggests a high probability of Takayasu [9]

Major Criteria	Left mid-subclavian artery lesion
	Right mid-subclavian artery lesion
	Characteristic signs and symptoms of at least one month duration (limb claudication, pulselessness or pulse differences in limbs, fever, neck pain, transient amaurosis, blurred vision, syncope, dyspnea or palpitations)
Minor Criteria	High erythrocyte sedimentation ($> 20\text{mm h}^{-1}$)
	Carotid artery tenderness
	Hypertension ($> 140/90\text{mmHg}$ brachial or $160/90\text{mmHg}$ popliteal)
	Aortic regurgitation or annuloaortic ectasis
	Pulmonary artery lesion
	Left mid-common carotid lesion
	Distal brachiocephalic trunk lesion
	Descending thoracic aorta lesion
	Abdominal aorta lesion
	Coronary artery lesion

The patient underwent hemodialysis due to life-threatening hyperkalemia and received corticosteroid therapy at a dose of 1 mg/kg/day, gradually reduced over six months. Antihypertensive treatment was initiated, including a beta-blocker and a calcium channel blocker instead of angiotensin-converting enzyme inhibitors. A surgical consultation was requested, but the family refused amputation of both lower limbs up to the thighs.

The patient's condition worsened, with a deterioration in blood pressure and the onset of impaired consciousness, with a Glasgow Coma Scale score of 9/15. A brain CT scan revealed extensive ischemic strokes, which caused the patient's death on the same day.

DISCUSSION

Renal involvement is a common and potentially serious complication of Takayasu's disease. It is most often associated with stenosis or occlusion of the renal arteries, leading to renovascular hypertension and progressive deterioration of renal function [1,2], thus representing a major factor in poor prognosis, especially when it progresses to advanced chronic renal failure [2].

In this patient, the combination of abdominal aortic stenosis and renal artery stenosis reflects an extensive form of the disease, associated with severe and resistant hypertension, progressing to end-stage renal failure complicated by severe fluid and electrolyte disturbances [3]. The life-threatening hyperkalemia observed in our patient can be explained by decreased renal potassium excretion, resulting in a life-threatening emergency due to the high risk of cardiac arrhythmias [4].

The primary treatment for Takayasu's disease is to control vascular inflammation, mainly through corticosteroid therapy, which achieves clinical remission in approximately 25 to 50% of cases [5]. However, in cases of insufficient therapeutic response or corticosteroid dependence, conventional immunosuppressants such as methotrexate, azathioprine, and cyclophosphamide must be introduced [5]. Biotherapies, particularly tocilizumab and rituximab, have been shown to be effective in refractory forms, improving inflammatory control and reducing relapses [6,7].

In patients with significant renal impairment, revascularization of the renal arteries or abdominal aorta is one of the treatment options to consider. Endovascular

or surgical techniques can improve blood pressure control and preserve renal function, but their effectiveness depends heavily on the stage of disease progression and the level of control of inflammatory activity [8]. Forms associated with extensive aortic occlusion, as in our patient, are associated with a high risk of complications and mortality despite optimal management [3,8].

The fatal outcome observed in our patient highlights the importance of early diagnosis of Takayasu's disease in cases of unexplained hypertension in young women, hence the need for rigorous monitoring of renal involvement, which largely determines the prognosis [1,2].

CONCLUSION

Takayasu's disease is a rare but potentially serious vasculitis; its prognosis is closely linked to the extent of vascular lesions and the involvement of target organs, particularly those of renal origin, which is a major factor in morbidity and mortality.

This reality requires early management and rigorous monitoring in order to limit the progression to severe complications, as illustrated in our patient.

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