Type II Takayasu's Arteritis

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Abstract

We report a case of Takayasu's arteritis in a young Portuguese woman with severe hypertension who failed to respond to medical treatment, and who presented renal failure. A left nephrectomy was performed, a prosthesis was placed between the thoracic aorta and the aortic bifurcation, and an aorto-renal graft was inserted. The renal function remained impaired, the patient is in chronic hemodialysis, but normotensive with no additional therapy.

Key words: arteritis, Takayasu's arteritis, arterial hypertension.

Introduction

Takayasu's arteritis is an arterial inflammatory disease of unknown cause, which affects the aorta and its principal branches, as well as the pulmonary artery. Granulomatous lesions and cell infiltrations in the vascular wall lead to stenosis and obstructions of the arteries involved.

The disease was identified in 1908, and has received various names, such as aortic arch syndrome, pulseless disease, and reversed coarctation, among others. Ueno and colleagues [1] defined three varieties of the disease: type I, involving the aortic arch and its branches; type II, involving the thoraco-abdominal aorta and its branches; and type III, which is a combination of types I and II. Lupi-Herrera proposed a type IV, with involvement of the pulmonary artery, and in a sample of 107 patients,[2] he found 80%, 11%, 65%, and 45% cases of the four abovementioned types, respectively. Type IV Takayasu's arteritis is thought to be responsible for many cases of so-called primary pulmonary hypertension.[3]

In 1990, Arend and co-workers published the Takayasu's arteritis classification criteria, which were accepted by the American School of Rheumatology.[4]

Takayasu's arteritis occurs worldwide, although the majority of cases have been described in Asia and Africa. It shows a clear prevalence in women, and most diagnoses are made during adolescence, although various cases have been described in childhood and adulthood,[1,5,6] as it is the case with our patient.

Clinical case

A thirty-six year old woman, with arterial hypertension since the age of 19. At that age, surgery was proposed due to suspected renovascular hypertension, but the patient refused.

In 1988, after a long absence, she consulted her family doctor, who introduced anti-hypertensive treatment, and etiologic evaluation was reinitiated. On this date, there was a worsening of the tiredness and headaches, onset of tinnitus, visual alterations and weight loss.

Blood pressure was 182/120 mmHg, under therapy with a diuretic, a beta-blocker, a converting enzyme inhibitor, and a calcium channel blocker. The patient presented severe signs of repercussion of the disease: grade III retinopathy, left ventricular hypertrophy, and creatinine level of 2.0 mg/dL.

Chest x-ray presented a cardiac shadow of enlarged dimensions and prolapse of the ascending aorta. Electrocardiogram revealed pressure overload left ventricular hypertrophy, and echocardiogram showed concentric hypertrophy of the left ventricle and dilation of the left auricle and aortic root.

In the abdominal ultrasound, the right kidney presented a diameter 10 cm larger, total loss of corticomedullary differentiation and significant increase in echogenicity; the left kidney measured 7 cm, with corticomedullary differentiation and a cortical ratio of 1:1.

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Arteriography showed multiple stenoses of the thoracic and abdominal aorta, post-stenotic dilations, saccular aneurysms, irregular intimal surface, and stenosis of the left renal artery (*Fig. 1*).

It was decided to carry out surgery, consisting of left nephrectomy, aorto-thoracic-biliary primitive bypass with a bifurcated prosthesis of 18 cm, and right aortic-renal bypass with an 8 cm prosthesis (*Fig. 2*).

The histological examination showed: "1 – Left kidney with marked lesions of chronic nonspecific pyelonephritis, with glomerulosclerosis. Arteries of the renal hilum with preserved structure, no signs of arteritis. 2 – Right external iliac artery wall with alterations in the three layers: slack fibromuscular intimal thickening, mucinous imbibition and mild mononuclear inflammatory infiltrate; internal elastic lamina straightened and unfolded in some portions, media with loss of muscular fiber, focal inflammatory infiltrate with mononuclear cells and a multinuclear giant cell, neovascularization, fibrosis and mucinous imbibition; adventitia with focal inflammatory infiltrate, mainly by mononuclear cells. The alterations observed are compatible with the clinical diagnosis of Takayasu's arteritis.

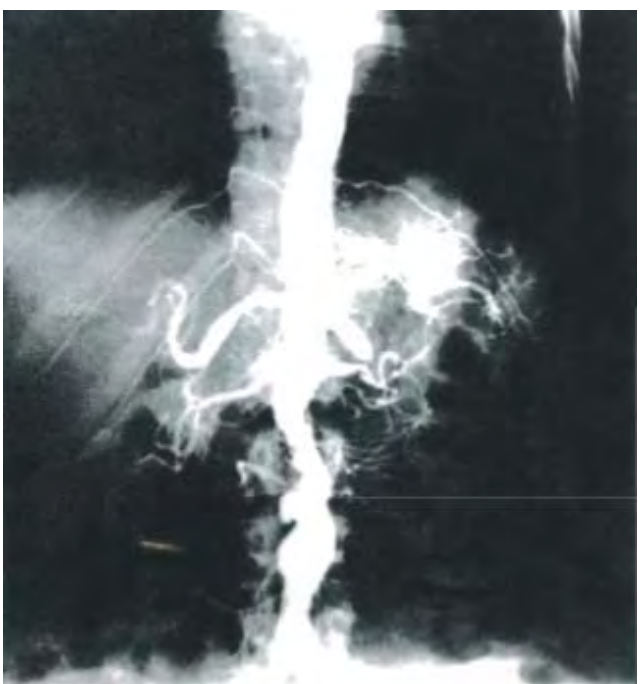
The deterioration of the renal function, irreversible by the date of surgery, led to the establishment of a chronic dialysis program, which the patient has been undergoing for four years, without requiring another treatment.

Comments

Without appropriate treatment, the clinical symptoms of the patient deteriorated until irreversible damages were caused, namely, lesions due to arterial hypertension.

In young patients, Takayasu's arteritis may present an acute, nonspecific inflammatory phase, and as a result, may pass unnoticed, with the disease manifesting itself later through cardiac, vascular and neurological signs resulting from vascular obstructions.

Types II and III are the most typical among patients, due to the involvement of the aortic arch and the resulting absence of radial pulse (reversed coarctation) and syncope, while type II arteritis is more frequently accompanied by arterial hypertension, due to coarctation of the abdominal aorta[7] and



Anteroposterior abdominal aortography showing multiple stenosis of the abdominal aorta, post-stenotic dilatations, saccular aneurysms, and stenosis of the left renal artery.

FIG. 1

Schematic representation of revascularization carried out (drawing by A. Diniz da Gama).

FIG. 2

involvement of the renal arteries.

A reduction in aortic capacity and reactivity of the baroreceptors may also be involved in the pathophysiology of arterial hypertension[8,9] which is present in 72% of patients with this type of arteritis.[2]

The etiology of Takayasu's arteritis, as well as its possible relation with collagen diseases, other vasculitis, syphilis, tuberculosis and other immunologic diseases, are not clear. The high incidence of tuberculosis (48% of Lupi-Herrera [2] patients) suggests that microbacteria are involved in the etiology of the disease.

The diagnosis was not difficult, due to the exuberance of the clinical symptoms and the arteriographic images that make up the clinical condition of our patient within the diagnosis criteria proposed by Ishikawa[10]. The therapy, however, presented severe problems for us, given the advanced stage of the disease, renal lesions, stenosis of the left renal artery and severity and resistance of the arterial hypertension[11] to medical therapy.

The decision to revascularise was based on the fact that in 75% of patients, control of arterial pressure is achieved.[12,13,14] The maintenance of the previous condition would involve a severe risk to the patient, due to risks of cardiac or cerebral complications resulting from arterial hypertension.[15] The left nephrectomy enabled a significant factor in the elevation of arterial pressure to be eliminated, and with the revascularization of the left kidney, an attempt was made to recover some residual function, however, this was not verified.

The late diagnosis of this condition, which is rare among us, had negative results on this female patient, whose renal function could not be recovered with the revascularization surgery. ■

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