Takayasu's Disease with Severe Heart and Arterial Involvement in a Preschool-Age Child

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The authors describe a case of Takayasu's arteritis in a child of only 3 years of age, emphasizing the rarity of this disease in this age group. The child was admitted to the emergency room in a post-convulsive state after a tonic-clonic seizure. After a detailed clinical examination, extensive diagnostic tests, and observation of the clinical evolution, the diagnosis was Takayasu's disease with severe cardiac and arterial involvement. The report warns pediatricians and cardiologists to awareness of the possibility of this disease in very small infants, in countries where it is underdiagnosed.

Introduction

Takayasu's arteritis (TA) is a chronic granulomatous vasculitis of unknown etiology involving the aorta and its branches, which produces vascular sequelae with stenotic lesions and/or thrombus formation. The lesions are usually severe, leading to organ dysfunction, and most require a surgical approach. It is underdiagnosed in Brazil probably due to diagnostic difficulty, but Brazilian studies estimate that half of the affected individuals are in the age group under 20 years[1]. The case described in this study stands out because of the extensive vascular involvement with severe sequelae in a very young child.

Case report

JEFS, 3 years and 7 months old, male, weighing 12 kg, was admitted to the emergency room in a post-convulsive state after a tonic-clonic seizure. The family reported that this was his first convulsive episode and denied fever, gastrointestinal or respiratory changes, and use of medication. The child was in attendance because of low weight gain. There was no family history of epilepsy, and the patient was the only child of a healthy couple. The patient had negative epidemiology for tuberculosis (TB) and a visible BCG vaccine scar.

Clinical examination showed signs of malnutrition, light mucocutaneous pallor, absence of edema or jaundice, good general condition, no fever, and mild tachypnea. The cardiovascular examination showed lower deviation of the apex beat, slight muffling of the heart sounds, tachycardia, and no murmurs on auscultation of the chest, abdomen and back. BP ranged from 125/70 to 130/80 mmHg (percentile> 95 for age, according to Task Force, 1996). The pulses were reduced in the lower limbs, which showed lower temperature than the upper limbs. The liver was located at 3 cm from the RCM, and there were no enlarged lymph nodes or abdominal masses.

The blood count showed normal white blood cell count and platelet count, mild hypochromic anemia with microcytosis. CRP was negative; HHS was low; ANF was negative; rheumatoid factor was negative; blood cultures were negative. The ECG showed sinus rhythm, left ventricular hypertrophy, and ventricular repolarization changes. The X-ray of the chest showed enlarged heart area due to left ventricle (LV) hypertrophy, and signs of pulmonary congestion (Fig. 1). The computed tomography of the brain showed no changes. The Doppler echocardiogram showed LVEF = 38%, large increase in the left chambers, and the presence of large thrombi within the LV cavity (Fig. 1).

Abdominal ultrasound found a reduction in the volume of the right kidney and changes suggesting a cortical infarct area in its upper pole. Duplex scanning of carotid arteries was normal, and Doppler scanning of the abdominal aorta revealed an aortic aneurysm, with a thrombus within it, as well as poor visualization of iliac and renal vessels. The abdominal angiotomography showed diffuse concentric thickening of the proximal and suprarenal portions of the abdominal aorta, and fusiform aneurysms of the common iliac arteries (larger on the right side), with an intraluminal thrombus, and right renal hypotrophy (Fig. 2). The coronary cineangiography was not performed because of the risks associated with the placement of a vascular access device in the presence of large aneurysms of the iliac arteries.

Evolution

An improvement of the CHF was observed with the use of furosemide, spironolactone, digoxin and carvedilol. The child received heparin and, afterwards, dicumarol; no intracavitary

Key words

Takayasu arteritis/complications; vasculitis; child preschool; angiography/methods.

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Manuscript received August 12, 2009; revised manuscript received September 07, 2009; accepted October 21, 2009.

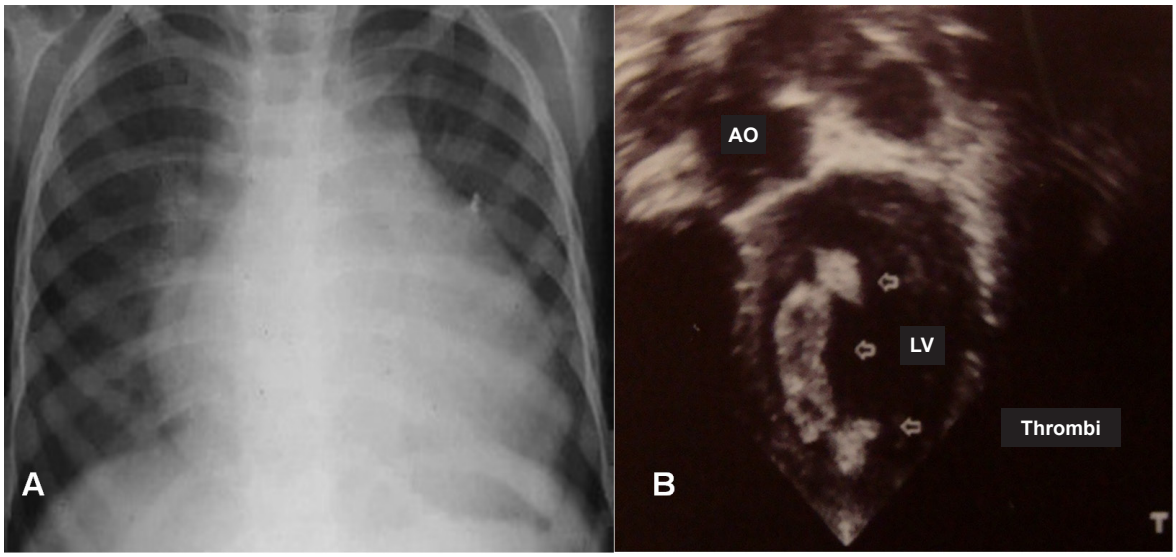

Figure 1 - Chest radiograph (A) showing cardiomegaly, and Doppler echocardiography (B) showing left ventricular enlargement and large thrombi.

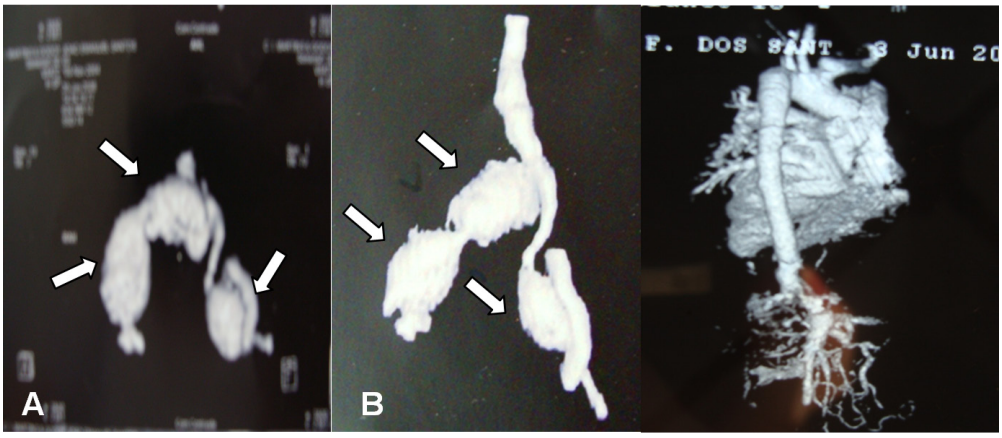

Figure 2 - CT angiography showing three large aneurysms in the iliac arteries bilaterally (A) and (B) study of the aortic arch and the innominate artery with no complications.

thrombus was visualized on the echocardiography, and there was no new thromboembolic event. The child underwent surgical resection of iliac artery aneurysms with stent placements in the right iliac artery and the abdominal aorta. Histopathological analysis of the resected vascular segment showed non-specific chronic periarteritis with areas of atherosclerosis.

During hospitalization, the child presented with a lesion on the dorsum of the right foot, suggesting vasculitis, as well as fever, and increased HSS and CRP. The patient showed clinical and laboratorial improvement after pulse therapy with methylprednisolone. An echocardiogram performed after one year of treatment showed normalization of LV systolic function and reduction of left cardiac chambers. The thoracic and abdominal angiotomography was repeated and showed no impairment of the aortic arch or the innominate artery, but it showed little contrast in the iliac arteries and large collateral network (Fig. 2). Currently, the child has normal blood pressure levels, normal renal function, no CHF, and is gaining weight.

Discussion

AT was first described by the ophthalmologist Mikito Takayasu, in 1908; he observed aneurysms and arteriovenous anastomoses in the retina[2]. It is an inflammatory segmental chronic vasculitis, of a destructive/degenerative nature, with diffuse and multifocal extension, affecting large and medium arteries, particularly the thoracic aorta and its major branches, and also the pulmonary and coronary arteries[2]. In the late

phase, there are areas of stenosis interspersed with dilatations and normal segments, as well as occlusions by thrombi and atypical coarctations[1,2]. These changes cause ischemia in different territories, especially the cerebral, coronary, peripheral arterial and renal territories. Studies suggest that immune and/or genetic factors are involved in the genesis of the disease[3,4]. AT is considered the third most common worldwide cause of childhood vasculitis. About three quarters of the patients are between 10 and 20 years of age, with a higher incidence in males (1:8)[5].

Usually, the disease presents three phases. The first phase results from an acute inflammatory process with nonspecific symptoms, with no pulse changes. The second phase reflects the exacerbation of vascular inflammation, manifested by pain in the vessel. These phases may last from months to years. The third stage is fibrotic, and the manifestations are secondary to arterial occlusions[6-8]. An association of TA with TB and with rheumatoid arthritis; Crohn's disease; lupus erythematosus; thyroiditis; ulcerative colitis; and ankylosing spondylitis is described by several authors[4,9]. In this case, no diagnostic workup was performed in relation to these diseases because of the absence of other signs and symptoms related to them.

Angiography is the imaging method of choice[5,10], traditionally used to evaluate the affected vessels, but it is a procedure of high risk due to exposure to radiation, rupture of vessels, hemorrhages, and infections. It was not indicated in this case because of the presence of giant aneurysms at sites of vascular access for the exam. For the low risk, we opted for a thoracic and abdominal angiotomography, which revealed no lesions in the aortic arch, in the brachiocephalic trunk, and in the entire thoracic aorta, but a bilateral reduced flow was observed in the iliac arteries, with formation of a collateral vascular network.

The case-study met the diagnostic criteria developed by the American College of Rheumatology (ACR)[8,9] and was classified as type II according to the classification of Ueno et al[8,9], modified by Lupi and Sanchez-Torres[8,9]. It included involvement of the thoracic aorta (descending and abdominal segments), with no involvement of the aortic arch[8,9].

The clinical management of TA consists of the control of vascular inflammation with the use of immunosuppressants, during the inflammation activity phase[4,5]. In the chronic phase, revascularization surgery is often necessary, with a high rate of reestenosis[10]. In the case described in this study, there was myocardial dysfunction that responded well to medical treatment and control of high blood pressure. CNS changes were probably due to the detachment of intracardiac thrombi, leading to stroke, resulting in hemiplegia.

From the case presented in this study and from the literature review, we concluded that the awareness of this disease by pediatricians and cardiologists permits its diagnosis and proper treatment, thus reducing morbidity and mortality. The hypothesis of TA should be included in the differential diagnosis of cases of cardiomyopathy that occur concomitantly with high blood pressure, ischemic strokes, pulse asynchrony between limbs, and renovascular hypertension, even in very small infants. It is recommended that other diseases be investigated, especially tuberculosis, because of its high prevalence in our country.

Potential Conflict of Interest

No potential conflict of interest relevant to this article was reported.

Sources of Funding

There were no external funding sources for this study.

Study Association

This study is not associated with any post-graduation program.

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