Case Report

Type III Takayasu's Arteritis in a Pediatric Patient. Case Report and Review of the Literature

Karla Mendiola Ramírez, Astrid Cristina Portillo Rivera, Abraham Galicia Reyes, José Antonio García Montes, María del Rocio Maldonado Velázquez, Enrique Faugier Fuentes

Servicio de Reumatología, Hospital Infantil de México Federico Gómez, Mexico City, Mexico
Servicio de Cardiología y Hemodinamia, Instituto Nacional de Cardiología Ignacio Chávez, Mexico City, Mexico

ABSTRACT

Takayasu's arteritis (TA), also known as “pulseless disease”, is the third most common vasculitis in childhood. It is a chronic, idiopathic, granulomatous vasculitis that involves large vessels. It occurs most commonly in females with a 4:1 ratio over males; the average age of appearance is 26 years. Its cause is unknown.

Here we report the case of a 7-year-old girl, with type III TA according to the Numano classification, in the ischemic phase, treated with corticosteroids and immunosuppressive agents and early angioplasty due to the severity of the disease. The outcome was satisfactory.

The diagnosis of TA in children under 10 years of age is made only in 2% of them; the delay in diagnosis reaches a mean of 19 months. The course of the disease is variable despite surgical and immunosuppressive treatment.

Introduction

Takayasu's arteritis (TA) is the third most common vasculitis in childhood after Henoch Schönlein and Kawasaki's disease. Its cause is unknown but it is known to be mediated by T cells and antibodies which are not organ-specific, although antiaorta and antiendothelium antiannexin V antibodies have been occasionally reported. There is a possible intervention of tuberculosis as a cause, with granulomas and Langhan's giant cells found, whose morphology resembles tuberculous lesions in patients with TA. There is a high incidence of positive intradermal tuberculin.2

The average age of presentation is 11.4 years, 20% of cases were diagnosed before age 19 and 2% before age 10. There is a delay in diagnosis of 19 months.

In childhood, the clinical picture is nonspecific, with possible fever, malaise, anorexia, myalgia, joint pain, abdominal pain,
hypertension, hypertensive retinopathy, heart failure, headache and seizures. The presence of murmurs and absence of pulses are present in the ischemic stage of the disease.2

The course of the TA is variable, despite the use of corticosteroids, which reduce by 50% the progression of the lesions; immunosuppressive therapy (methotrexate/azathioprine/mycophenolate moftel) leads to a better control of the disease and prevents restenosis.3

Stenosis is not reversible and early angioplasty is required in patients with renovascular hypertension, severe claudication, stroke, myocardial infarction, renal artery stenosis, moderate regurgitation of the aortic valve and the presence of more than 3 stenotic4 sites. Early diagnosis and appropriate treatment prevents complications related to the disease.

<table>
<thead>
<tr>
<th>Test</th>
<th>Result</th>
<th>Reference Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ophthalmic examination</td>
<td>No evidence of vasculitis</td>
<td>No evidence of vasculitis</td>
</tr>
<tr>
<td>Gastric bacilloscopy</td>
<td>Negative</td>
<td>Negative</td>
</tr>
<tr>
<td>Echocardiogram</td>
<td>Reduced cardiac motility; dilatation of left cavities, moderate</td>
<td>LV Mass Index 64 g/m² s</td>
</tr>
<tr>
<td></td>
<td>mitral insufficiency, trivalve aorta with normal coronary pattern, left aortic arch.</td>
<td></td>
</tr>
<tr>
<td>Electrocardiogram</td>
<td>Sinus rhythm, HR 140 beats per minute, aP +50, aQRS +45, PR 0.12, QRS 0.04, QTc 0.4, Left ventricle hypertrophy</td>
<td>Mean HR 109–169</td>
</tr>
<tr>
<td>Chest X-ray Laboratory</td>
<td>Situs solitus, levocardia, ICT 0.61, normal pulmonary flow</td>
<td>Interval +20 to +120</td>
</tr>
<tr>
<td>Blood count:</td>
<td>Hb 11.8 g/dl, Hto 35.8%, platelets 405 000, leucocytes 11 700/m³, neutrophils 9240/m³, lymphocytes 2460/m³³</td>
<td>CTI 0.6–0.7</td>
</tr>
<tr>
<td>Acute phase reactants:</td>
<td>ESR 10 mm/h, CRP 0.302 mg/dl, procalciitonin 0.51 ng/ml</td>
<td></td>
</tr>
<tr>
<td>Renal function tests:</td>
<td>BUN 6 mg/dl, Cr 0.6 mg/dl</td>
<td></td>
</tr>
<tr>
<td>Other:</td>
<td>renin 4.6 ng/ml/h, C3 135 mg/dl, C4 18.3 mg/dl, RF negative, IgA 409 mg/dl, IgG 1260 mg/dl, IgM 262 mg/dl, ANA and anti-DNA negative</td>
<td></td>
</tr>
<tr>
<td>Renal ultrasound</td>
<td>Right kidney: 83 mm × 44 mm × 46 mm, short, left</td>
<td></td>
</tr>
<tr>
<td></td>
<td>82 mm × 39 mm × 35 mm</td>
<td></td>
</tr>
</tbody>
</table>

**Case Report**

The case is a female patient, 7 years and 7 months old, previously healthy, from the State of Mexico. She began her current illness a month earlier, with malaise, myalgia, vomiting, claudication, headache, tinnitus, fosfenus, abdominal and chest pain. She was admitted to a second level hospital for hypertension and heart failure and was sent to a third level hospital due to absence of pulses and left ventricular hypertrophy. She was admitted due to hypertension, absence of pulses (left brachial and lower limbs), an aortic pansystolic grade II/VI murmur, hepatomegaly and claudication. Her study protocol can be seen in Table 1.

A computed tomography scan revealed hypoplasia of the right vertebral artery (V4), left subclavian stenosis and narrowing

![Fig. 1. CT angiography. Stenosis at the left subclavian artery, thoracic aorta, abdominal aorta and renal arteries.](http://www.elsevier.es)
of the descending aorta with arteritis data on the wall, yux-
tadifragmática and transverse diameters of 4.5–5.0 mm. Were also stenosis in the origin of the superior mesenteric artery (3.3 mm) and significant stenosis in the origin of the right renal artery (Fig. 1).

The diagnosis of TA III was based on the following: decreased peripheral arterial pulses and limb claudication, pressure difference greater than 10 mmHg, murmur over the aorta, arterial hypertension and angiographic abnormalities (thoracic ascending and abdominal aorta and renal arteries).

Glucocorticoids and methotrexate were initiated. Cardiac catheterization was performed due to the presence of more than 3 sites of stenosis. We documented diastolic dysfunction, mild mitral regurgitation; severe stenosis of the thoracoabdominal aorta with a gradient of 50 mmHg and nonsevere renal artery stenosis (10 mmHg gradient) (Fig. 2A and B). Angioplasty was performed in which two stents were placed in the stenotic area, with a postsurgical gradient of 0 (Fig. 3A and B). She is currently asymptomatic with no residual gradient. She no longer receives glucocorticoids but is receiving methotrexate, folic acid, vitamins A, C and D, aspirin, furosemide, spironolactone, and captopril.

**Discussion**

TA represents 1.5% of vasculitides in childhood, with 2% of cases diagnosed before age 10. Their course is variable, depending on the degree of activity, time of diagnosis, presentation and associated symptoms and the effect on other organs. They may have multiple relapses despite treatment. The inflammatory process causes thrombosis in the affected arteries, progressive appearance of stenosis, dilation and aneurysms. Antiplatelet therapy with low dose aspirin reduces the frequency of ischemic events.

Treatment is based on the use of glucocorticoids and immunosuppressive therapy. The use of ACE inhibitors is controversial due to their renal effects, but can be considered for use in patients with normal renal function. Surgical correction with angioplasty techniques is effective, increasing 5 years survival to 80%–95%. Restenosis occurs in 31.7% during the first year. The risk decreases by 50% with the use of corticosteroids and immunosuppressants. Follow-up angiography should be performed every 12 months.

The experience in our country has been described by Lupi-Herrera in a series of 107 patients with TA, whose ages ranged from 11 to 30 years, with a mean age 26 years. The major clinical manifestations were asthenia, weight loss, headache,
claudication, and hypertension. Stenosis occurred in the thoracic aorta (25% upper/lower 67%), subclavian (85%) and renal (62%) arteries. A higher morbidity was associated with the Mexican phenotype, severity of disease expression and variations depending on the medical and surgical treatment employed.10

Worldwide experience is very similar, but with few cases reported in the pediatric literature. The studies conclude that it is a condition that must be actively suspected in order to initiate early treatment to decrease mortality. In our country, there are few publications and experience regarding treatment. In this case report, a good clinical history with an appropriate semiology made it possible to steer towards an accurate diagnosis, providing an opportunity for the patient to receive adequate medical and surgical treatment.

Conflict of Interest

The authors declare no conflict of interest.

Acknowledgement

The authors of this article would like to thank Dr. Jose Agustin Ramirez Barreto for his help in translating this article.

References