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Case Report

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

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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.

Arteritis de Takayasu tipo III en un paciente pediátrico. Reporte de caso y revisión de la literatura

Palabras clave:
Arteritis de Takayasu
Angioplastia
Tratamiento inmunosupresor

RESUMEN

La arteritis de Takayasu (AT) o «enfermedad sin pulsos» es la tercera vasculitis más frecuente en la infancia. Es crónica, idiopática, granulomatosa y afecta a vasos grandes. Afecta a las mujeres, con una relación 4:1; con una edad promedio de 26 años. Su causa es desconocida.

Presentamos el caso de una niña de 7 años y 7 meses de edad, con AT tipo III de la clasificación de Numano, en fase isquémica, a la que se le inició tratamiento con glucocorticoides e inmunosupresores, así como angioplastia temprana, por la severidad del cuadro clínico. Tuvo una evolución satisfactoria.

El diagnóstico de AT antes de los 10 años se realiza en el 2% de los pacientes; el retraso en el diagnóstico es en promedio de 19 meses; el curso de la enfermedad es variable a pesar del tratamiento inmunosupresor y quirúrgico.

Introduction

Takayasu's arteritis (TA) is the third most common vasculitis in childhood after Henoch Schönlein and Kawasaki's[1] 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.[1]

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.[2]

In childhood, the clinical picture is nonspecific, with possible fever, malaise, anorexia, myalgia, joint pain, abdominal pain,

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Table 1
Diagnostic Approach to the Patient.

Test	Result	Reference Value
Ophthalmic examination	No evidence of vasculitis	No evidence of vasculitis
Gastric bacilloscopy	Negative	Negative
Echocardiogram	Reduced cardiac motility; dilatation of left cavities, moderate mitral insufficiency, trivalve aorta with normal coronary pattern, left aortic arch. EF 40%, AF 19%, LV mass index 165 g/m^2 s	LV Mass Index 64 g/m^2 s
Electrocardiogram	Sinus rhythm, HR 140 beats per minute, aP +50, aQRS +45, PR 0.12, QRS 0.04, QTc 0.4, Left ventricle hypertrophy	Mean HR 109–169 QRS +60 Interval +20 to + 120
Chest X-ray	Situs solitus, levocardia, ICT 0.61, normal pulmonary flow	CTI 0.6–0.7
Laboratory	**Blood count:** Hb 11.8 g/dl, Hto 35.8%, platelets 405 000, leucocytes 11 700/m^3, neutrophils 9240/m^3, lymphocytes 2460/m^3 **Acute phase reactants:** ESR 10 mm/h, CRP 0.302 mg/dl, procalcitonin 0.51 ng/ml **Renal function tests:** BUN 6 mg/dl, Cr 0.6 mg/dl **Other:** 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	**Blood count:** Hb: 12–18 g/dl, Hto: 37%–47%, platelets: 145 000–450 000 leucocytes: 4000–11 000, neutrophils:4500–7500, lymphocytes: 1000–4800. **Acute phase reactants:** ESR: 0–10 mm/h, CRP: 0–0.300 mg/dl, procalcitonin: 2.0 ng/dl **Renal function tests:** BUN: 2–23 mg/dl, Cr: 0.4 1.5 mg/dl **Other:** renin: 0.5–5.7 ng/ml/h, C3: 75–135 mg/dl, C4: 12–75 mg/dl, RF negative, IgA: 124–170 mg/dl, Ig G: 923–1.110 mg/dl, IgM: 65–90 mg/dl, ANA and anti-DNA negative
Renal ultrasound	Right kidney: 83 mm × 44 mm × 46 mm, left 82 mm × 39 mm × 35 mm Doppler with adequate vascular permeability	Right kidney: 85 mm × 44 mm × 47 mm, left 81 mm × 40 mm × 35 mm

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 mofetil) 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 stenotic[4] sites. Early diagnosis and appropriate treatment prevents complications related to the disease.

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, tinnitus, 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

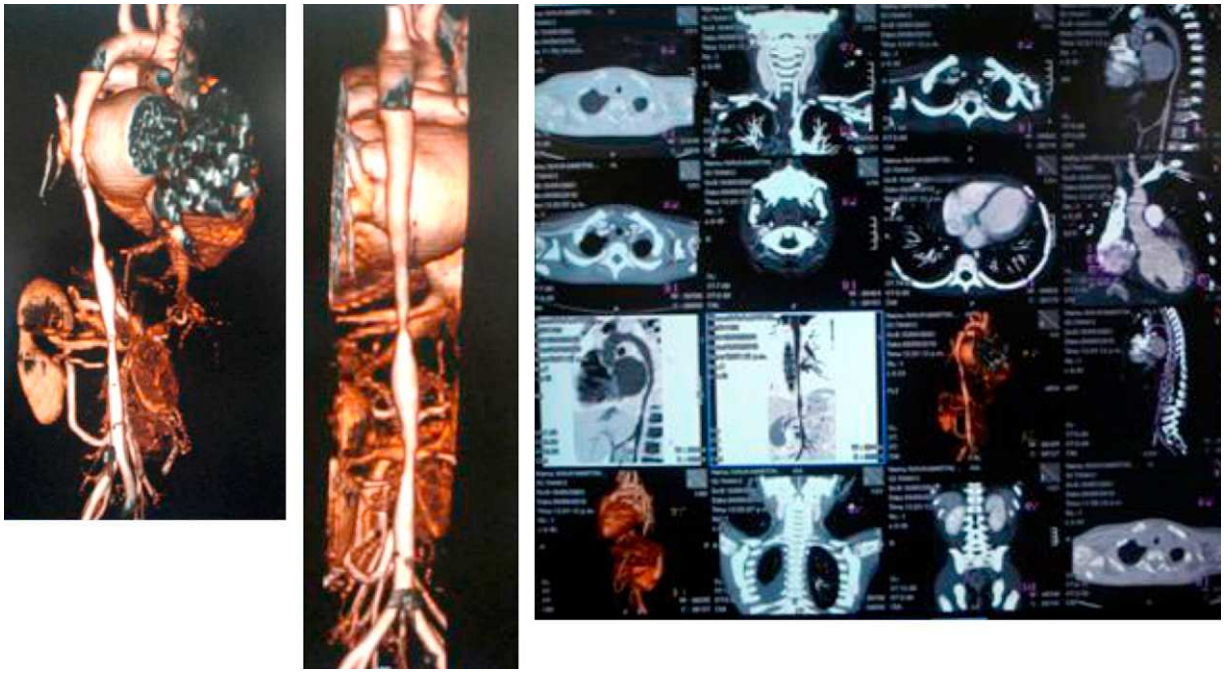

Fig. 1. CT angiography. Stenosis at the left subclavian artery, thoracic aorta, abdominal aorta and renal arteries.

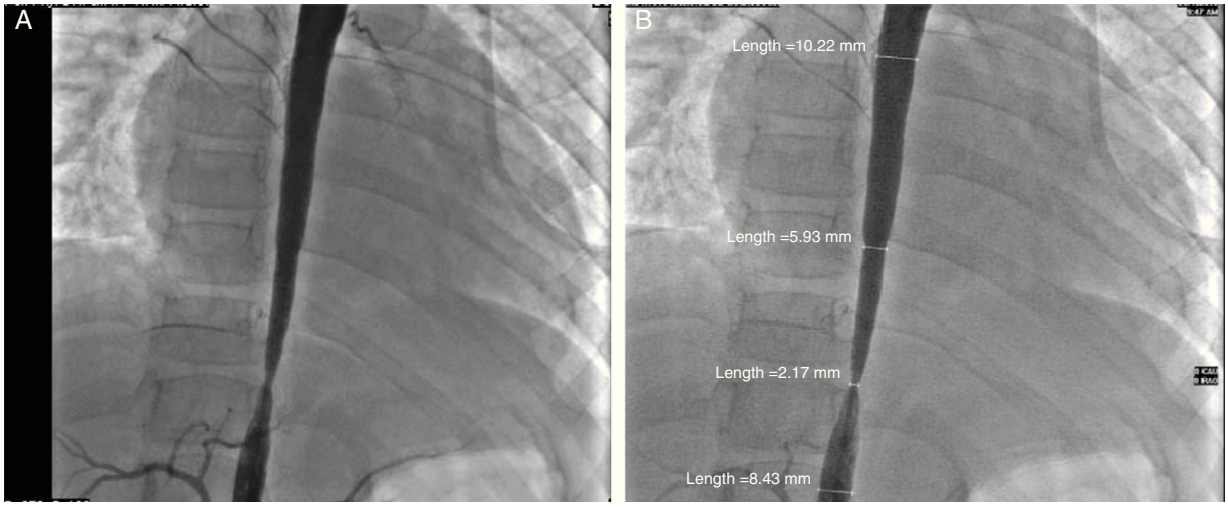

Fig. 2. (A) and (B) Cardiac catheterization showing severe stenosis of the thoraco-abdominal aorta. Pressure gradient of 50 mmHg. Stenosis below the renal arteries is not severe.

of the descending aorta with arteritis data on the wall, yuxtadiafragmática 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 vasculitidies 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.[5] 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.[6,7]

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%.[8] 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.[9]

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.[6,7] The major clinical manifestations were asthenia, weight loss, headache,

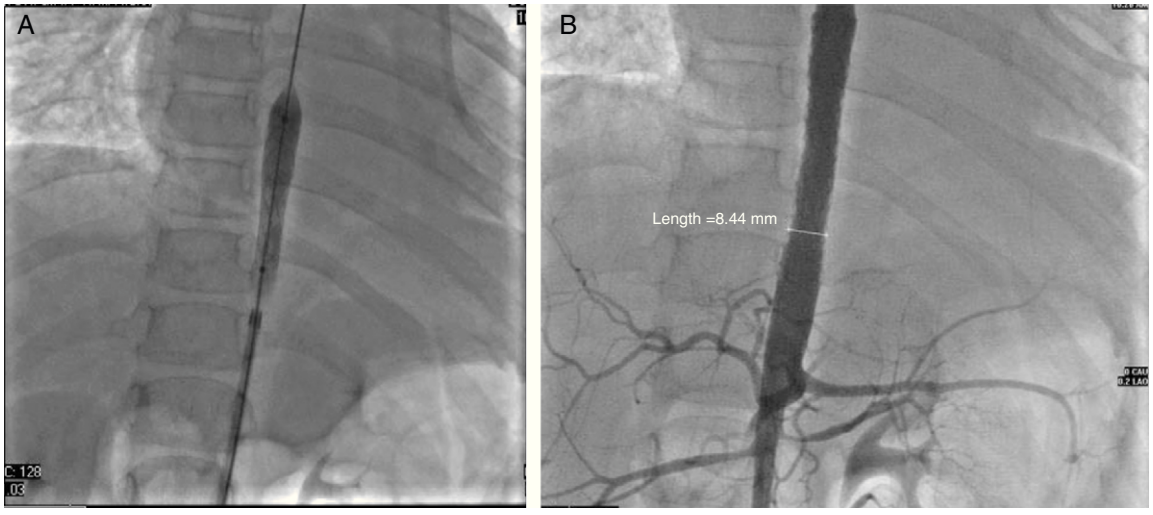

Fig. 3. (A) and (B) Angioplasty. Placement of 2 long stents, covering the area of severe stenosis. Postoperative gradient: none. Gradient of the renal stenosis: 10 mmHg. There was no procedure on the left subclavian or the renal arteries.

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.

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References

1. Al Abrawi S, Fuillet M, David L, Barral X, Cochat P, Cimaz R. Takayasu Arteritis in children. Ped Rheum. 2008;6:1–5.
2. Lupi E. Arteritis de Takayasu. In: Attie F, Zabel C, Buendía H, editors. Cardiología Pediátrica, diagnóstico y tratamiento. México, DF: Editorial Panamericana; 2001. p. 431–43.
3. Osman M, Aaron S, Noga M, Yacyshy E. Takayasuŭs arteritis progression on anti-TNF biologics: a case series. Clin Rheumatol. 2011;30:703–6.
4. Ruperto N, Ozen S, Pistorio A, Dolezalova P, Brogan P, Maldonado V, et al. EULAR/PRINTO/PRES criteria for Henoch-Schönlein purpura, childhood polyarteritis nodosa, childhood Wegener granulomatosis and childhood Takayasu arteritis: Ankara 2008. Ann Rheum Dis. 2010;69:790–7.
5. Gargah T, Harrath BM, Bachrouche H, Rajhi H, Abdallah BT, Lakhoua RM. First case of childhood Takayasu arteritis with renal artery aneurysms. Ped Rheum. 2010;8:1–4.
6. De Souza WAS, Machado PN, Pereira MV, Arrae DAE, Ries Neto ET, Mariz AH, et al. Antiplatelet therapy for prevention of arterial ischemic events in Takayasu arteritis. Circ J. 2010;74:1236–41.
7. Masafumi U. Antiplatelet therapy in the treatment of Takayasu arteritis. Circ J. 2010;74:1079–80.
8. Park MC, Lee WS, Park BY, Lee KS, Choi D, Shim HW. Post-interventional immunosuppressive treatment and vascular restenosis in Takayasuŭs arteritis. Rheumatology. 2006;45:600–5.
9. Reddy E, Robbs VJ. Surgical management of Takayasu's arteritis in children and adolescents. Cardiovasc J Africa. 2007;18:393–7.
10. McKinnon MK, Clark MT, Hoffman SG. Limitations of therapy and guarded prognosis in an American Cohort of Takayasu Arteritis Patients. Arthritis Rheum. 2007;3:1000–9.