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Case Report

Abdominal Pain, a Rare Manifestation Revealing Takayasu's Disease: A Case Report

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Abstract

Takayasu's disease is a vasculitis of the large vessels that affects the aorta and its branches, it is rare in the pediatric population. We report the case of an 8-year-old girl admitted for subacute abdominal pain and in whom the clinical examination had objectified a blood pressure asymmetry with hypertension of the upper limbs and a positive inflammatory assessment with significant stenosis of the aortic isthmus/descending aorta and significant parietal thickening of the descending thoracic aorta and the abdominal aorta allowing the diagnosis of Takayasu's disease, the management consisted of putting the patient on corticosteroid therapy and the evolution was marked by the installation of stenosis of the right peroneal artery, and occlusion of the left anterior tibial artery for which the patient was put on corticosteroid therapy and immunosuppressant with good clinical and biological evolution.

Keywords: *Takayasu's Disease (TD); Abdominal Pain; Corticosteroid Therapy*

Introduction

Takayasu's disease (TD) is a chronic inflammatory arteriopathy affecting the aorta, its main branches and the pulmonary arteries. Its name comes from the Japanese ophthalmologist Mikito Takayasu who published the first description of the disease in 1908. It thus causes lesions such as stenoses, occlusions or aneurysms in the affected areas [1]. While observations of this disease are more frequent in Japan, Asia and South America, Takayasu's disease exists in Africa, but is rarely reported in its medical literature, particularly in sub-Saharan Africa [2].

This disease is rare in children and is most often revealed by asthenia with high blood pressure (HBP). We report a case diagnosed by the Pediatrics department of the University Hospital of Tangier revealed by abdominal pain.

Observation

We report the case of an 8-year-old girl, from a 2nd degree consanguineous marriage, with a family history of a father followed for sickle cell disease and a notion of albinism in the family (paternal sister and aunt), presented for 10 days diffuse abdominal pain associated with food vomiting, without other associated signs, all evolving in a context of apyrexia and alteration of the general state made of weight loss and anorexia, the clinical examination had objectified a girl in pain, stable on the respiratory level, apyretic at 37.3°, BP in the upper limbs:

right upper limb: 110/60 mmHg, left upper limb: 130/80 mmHg (HTA), right lower limb: 90/60 mmHg, left lower limb: 90/60 mmHg, HR: 110 bpm, weight: 20 kg (-1.5DS), size: 128 cm (M).

Soft abdomen, no defense or abdominal contracture, the rest of the clinical examination was unremarkable.

An inflammatory assessment was requested, returning positive: VS: H1: 95 mm, H2: 120 mm, CRP: 156 mg/l.

Blood count: GB: 20200/mm^3, PNN: 17978/mm^3, Lymphocytes: 1656/mm^3, Hb: 13.7 g/dl, Platelets: 399000/mm^3.

Lipasemia was negative at 21U/l.

An abdominal CT scan had objectified a periaortic hypodense aspect at the suprarenal level, then a thoraco-abdominal CT angiography was requested objectifying a significant parietal thickening of the descending thoracic aorta and the abdominal aorta up to the celiac trunk/superior mesenteric artery in favor of Takayasu arteritis type III. As well as the presence of a significant stenosis of the aortic isthmus/descending aorta is consistent with a late/chronic phase of the disease.

The patient was put on a corticosteroid bolus at a dose of 30 mg/kg/day for 5 days then switched to oral therapy at a dose of 1 mg/kg/day as well as antihypertensive treatment based on amlodipine 5 mg/day.

The initial evolution was marked by a clear clinical improvement with disappearance of abdominal pain and normalization of blood pressure in both upper limbs.

Fifteen days after the start of treatment, the patient consulted for intermittent claudication of the lower limbs which worsened with exercise; a CT angiography of the lower limbs revealed an occlusion of the distal third of the right peroneal artery with an occlusion of the proximal third of the left anterior tibial artery.

The patient had received 3 boluses of corticosteroid therapy and was put on azathioprine at a dose of 3 mg/kg/day with corticosteroid therapy.

The evolution was favorable: disappearance of clinical signs of claudication.

10 months back.

Discussion

Takayasu's disease (TD), also called "aortic arteritis", "aortic arch syndrome" and "pulseless women's disease", is a vasculitis affecting large vessels, particularly the aorta and its main branches (subclavian, carotid, vertebral, renal, digestive and iliac arteries), but also the coronary and pulmonary arteries, and can be complicated by aneurysm(s) and/or arterial stenosis(es) [2].

It represents the most common vasculitis of the large vessels in children. Thickening of the vascular wall is the most characteristic early sign of the disease, gradually leading to stenoses, thromboses and sometimes the development of aneurysms [3] which is the case for our patient, the disease was revealed by abdominal pain following thickening of the abdominal aorta.

The incidence and prevalence of Takayasu's disease remains more marked in adults compared to children [4].

Epidemiological data on Takayasu's disease are poor, the annual incidence reported in South Korea has been 0.4 (95% CI 0, 1.1) per million children with Takayasu's arteritis [5], according to a national health insurance database in South Korea the prevalence has been

estimated at 0.04 (95% CI 0.00, 0.08) for children aged 0 to 4 years and 0.63 (95% CI 0.36, 0.91) per 100,000 for those aged 15 to 19 years [6].

The clinical signs of Takayasu's arteritis generally vary according to the location and extent of the vascular lesion; the onset of the disease, known as the pre-occlusive phase, is generally characterized by the onset of an inflammatory syndrome associated with non-specific clinical signs, which explains the diagnostic delay [7]. The diagnosis of the disease is generally only made in the occlusive phase where ischemic symptoms become clear, with delays that can reach ten years [8].

In our case, the girl presented the same thing at the beginning; she had an inflammatory syndrome associated with non-specific abdominal pain developing in a feverish context.

The diagnosis of Takayasu's disease is based on a bundle of clinical and imaging arguments; in children, European consensus criteria validated on an international cohort were established and published in 2010 - PRINTO/EULAR/PReS - with a sensitivity of 100% and a specificity of 99.9%. The diagnosis of pediatric Takayasu's arteritis is made in a case starting at 18 years of age or younger with:

- An angiographic abnormality (CT angiography or MRI angiography or arteriography) of the aorta or its main or pulmonary branches, such as aneurysm/dilation, stenosis, occlusion, parietal, segmental or focal thickening; and this without argument for fibromuscular dysplasia or other vascular cause
- + 1/5 criteria among:
- Abolishing, lowering or asymmetry of arterial pulse; claudication of a limb on exertion.
- Blood pressure differential > 10 mm Hg.
- Vascular murmur of large vessels, audible or palpable.
- HTA > 95th percentile for height.
- VS > 20 mm or CRP > laboratory norm [9].

In our case, the diagnostic criterion was hypertension associated with an inflammatory syndrome, with stenosis of the aortic isthmus/descending aorta and thickening of the abdominal aorta, followed by claudication of both lower limbs during exercise.

Management of active Takayasu's disease is based on glucocorticoids 1 to 2 mg/kg/day maintained for 2 to 4 weeks then tapered.

The indication for methylprednisolone boluses, more readily at a dose of 15 mg/kg over 3 days, is broader than in adults, to be discussed in active forms at diagnosis and/or at risk.

During Takayasu's arteritis, either immediately or in the event of non-response or relapse during the tapering of corticosteroid therapy, immunosuppressive treatment is usually proposed (methotrexate, azathioprine. +/-TNFi) [9,10].

Concerning our patient, she was initially put on a corticosteroid bolus for 5 days then switched to oral therapy with good initial progress, but in view of the development of an occlusion of the right peroneal and left anterior tibial arteries revealed by intermittent claudication of both lower limbs, a corticosteroid bolus was received with the addition of an immunosuppressant (azathioprine 5 mg/kg/day) with good progress, the girl had not presented any clinical signs with a negative inflammatory assessment over a 10-month follow-up.

The evolution of the disease is generally favorable if early and adequate management has been well initiated, which helps to reduce pediatric morbidity and mortality [11,12].

According to a pediatric study, pediatric mortality is estimated between 0 and 27% but there are certain factors that have been identified that can be incriminated in particular: access to care, geographical origin, recruitment bias, etc [13,14].

The unfavorable evolution according to some pediatric studies can be due to cardiovascular and renal complications essentially, therefore the prevention of serious organ damage can be done by the establishment of an adapted treatment [14].

Conclusion

Takayasu's disease remains a rare disease in pediatrics, the symptomatology depends on the location of the vascular lesion, the paraclinical confirmation depends on the angiogram which shows the type and location of the vascular lesion and also depends on an inflammatory assessment which is most often positive, the management is essentially based on corticosteroid therapy and immunosuppressants.

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