A Rare Association of Takayasu Vasculitis and Pyoderma Gangrenosum in Children: About A Case.

Souad Maher, Abdelmoughit Hosni, Khadija Ben Elhossni, Ittimade Nassar and Nabil Moatassim Billah
Central Radiology Department, CHU IBN SINA–Rabat University Hospital Center, Faculty of Medicine and Pharmacy, Mohamed V University, Rabat, Morocco.

ARTICLE INFO
Article history:
Received: 25 June 2020;
Received in revised form: 18 July 2020;
Accepted: 29 July 2020;

Keywords
Takayasu, Child, Imaging.

ABSTRACT
Takayasu's disease is an inflammatory arteritis particularly rare in children and is often difficult to diagnose due to the large clinical polymorphism and the absence of specific biological criteria. We report a rare case of takayasu disease revealed by pyoderma gangrenosum in a child. The diagnosis was confirmed by imaging, which showed multifocal arterial damage suggestive of vasculitis.

© 2020 Elixir All rights reserved.

Introduction
Takayasu vasculitis is an inflammatory arteritis affecting large vessels and remains a rare entity in children, with variable clinical manifestations and life-threatening complications.

However, pyoderma gangrenosum is an inflammatory neutrophilic dermatosis that may be a rare complication of takayasu arteritis in children.

Through our case, we will illustrate the clinical particularities and the value of imaging in the diagnosis of this vasculitis as well as in the monitoring during treatment.

Patient and Observation
We report the case of a 10-year-old boy, with a pyoderma gangrenosum antecedent under treatment, consulted for hypertensive peaks in an apyretic context. Clinical examination found an upper extremity arterial pressure estimated at 200/10 mm Hg. The blood pressure in the lower limbs is measured at 88/45 mm Hg. The clinical examination is unremarkable. On the biological assessment, we have a CRP at 56 mg/l with a hypochromic microcytic anemia at 8 g/dl. A trans thoracic cardiac ultrasound was normal, which eliminated the diagnosis of a coartation of the aorta in the face of strong clinical suspicion.

A thoracoabdominal angioscan was performed and revealed a parietal thickening of the arch and descending aorta that was circumferential, irregular, in places, increasing in venous time and reducing the aortic lumen. This thickening extends to the supra aortic trunk and is responsible for complete stenosis of the left supra clavicular artery. It is associated with a parietal thickening of the primitive carotid arteries, significantly reducing the lumen with significant stenosis on the left.

All these imaging elements are consistent with takayasu arteritis.

In view of these findings, the patient was put under medical treatment based on immunosuppressive drugs (Methotrexate 10.5mg/m2/week) and high-dose corticosteroids (prednisone 1mg/kg/day) for 3 months and was marked by the regression of the inflammatory syndrome. In the face of hypertensive peaks that were resistant to treatment, the child was treated surgically with bypass surgery with a prosthesis. The evolution was favourable with a well-balanced blood pressure in a 3-year follow-up.

Discussion
Takayasu arteritis in children is a rare granulomatous vasculitis characterized by inflammation of the walls of medium and large arteries. It mainly affects the aorta and its branches, leading to several changes in the caliber of the arteries: stenosis, thrombosis, and even an aneurysm.

The thickening of the walls of the aorta and its branches is the main sign that the disease is calling.

The disease usually affects people under 40 years of age and women are more frequently affected than men.

Circumstances of discovery
Clinical
Takayasu disease is characterized by two phases:
- Pre-occlusive phase is manifested by non-specific symptoms such as:
  - Fever and/or night sweats.
  - Arthromyalgia or weight loss.
  - Skin rashes such as pyoderma gangrenosum revealing Takayasu arteritis as in the case of our patient.
- Occlusive phase: the disease can evolve and lead to arterial stenosis and/or aneurysm, after more than 3 years without treatment (2,3).

During this phase, the clinical signs are variable depending on the affected organ and can be distinguished:
- Cardiovascular signs:
  - Vascular murmurs, arterial hypertension in 70%.
  - Heart failure of valvular origin (fatigue, dyspnea ...).
  - Damage to the pulmonary arteries (dyspnea, hemoptysis ...).
- Neurosensory signs: manifested by visual disturbances, headaches and/or ischemic stroke.
Biologically
An inflammatory syndrome characterized by an acceleration of biological markers of inflammation, particularly sedimentation rate (1,2).

It is associated with microcytic hypochromic anemia in 50% of cases.

Etiological diagnosis
The etiologies of Takayasu disease are numerous. The most frequently mentioned is the infectious origin, due to the frequency of associated tuberculosis. Levels of antibodies to mycobacterial antigens have been noted in patients with Takayasu disease (2,3,5).

There are no specific risk factors, however certain phenomena can aggravate takayasu arteritis such as:
- High blood pressure.
- Diabetes
- Smoking
- Obesity

Imaging means
1. Doppler Ultrasound
This is a non-invasive method to visualize vascular abnormalities in the wall of the aorta and its branches during inflammatory flare-ups.

It provides accurate information about the arterial wall, especially during inflammatory flare-ups, showing regular circumferential hypoechoic thickening of the affected areas and the presence of long, regular stenoses (4,6).

It shows stenotic lesions, ectatic lesions and/or signs of aortic insufficiency.

In the case of takayasu arteritis, ultrasonography detects hypoechoic, circumferential and regular thickening of the affected areas associated with regular and long stenoses.

It is also a good test for monitoring the progression of vasculitis as well as for post-treatment surveillance.

2. Computed Tomography scan (CT scan)
CT scans allow a precise exploration of the arterial wall in inflammatory pathology.

The visualization of a thickening of more than 2mm of the aortic wall, hypodense, which increases homogeneously at a late stage is specific of an acute-looking takayasu arteritis (7,8).

However, irregular thickening of the arterial wall with associated calcifications are in favour of chronicity.

3. Magnetic resonance imaging (MRI)
MRI is as useful as CT scan in the positive diagnosis and follow-up of takayasu disease.

It shows a regular diffuse thickening of the aortic wall with a contrast shot showing the acute phase of the disease (9).

MRI angiography is used to map arterial lesions and to detect occlusive damage observed in the chronic phase.

4. Positron Emission Tomography (PET)
This is an expensive and unavailable test that confirms the activity of vasculitis and its evolution during treatment.

Decreased 18FDG binding to the arterial wall is a factor in favour of arteritis (10).

Differential diagnosis
- Infectiousortisitis (tuberculosis, syphilis...)
- Inflammatory aortitis (Horton's disease, Behcet's disease, systemic lupus erythematosus...)
- Fibromuscular dysplasia and atherosclerosis.

Therapeutic management
- Medical treatment:
  To limit arterial inflammation, treatment is based on corticosteroid therapy, taking into consideration contraindications (6).

Immunosuppressive treatment may be used in the rebellious forms and in the case of active arteritis (11).

It is combined with symptomatic treatment with a cardiovascular aim with regular monitoring to prevent vascular complications.

- Surgical treatment
  Vascular prostheses, de-obstructions, endarterectomies and interventional therapy have a place in the management of takayasu vasculitis (11,12).

Conclusion
Takayasu vasculitis is a rare condition in children and is multifactorial affecting the aorta and its branches. The clinical signs of this disease are non-specific and may be revealed by rashes such as pyoderma gangrenosum. Imaging remains the key examination for the detection of early inflammatory signs of the arterial walls and also in post-therapy monitoring.

Conflicts of interest
The authors do not declare any conflict of interest.

Contributions by Authors
All the authors have contributed to this work. All authors have read and approved the final version of the manuscript.
Thoracic angioscanner after injection of the contrast product at late time in axial (c) and sagittal (d) sections in the same case: showing an increase in the parietal thickening of the arch and the descending aorta reducing the aortic lumen and responsible for a complete stenosis of the artery above the left clavicle with significant reduction of the left primitive carotid artery, all of which may be part of an arteritis of the Takayasu type.

References


