Takayasu Arteritis: A Rare Cause of Fever in a Teenager

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Keypoints

What is known:

- Takayasu arteritis is an idiopathic large-vessel vasculitis that affects the aorta and its major branches.
- Takayasu arteritis often presents with an insidious clinical course and unspecific symptoms.

What is added:

- Constitutional features, such as fever and weight loss, are the most common signs found in pediatric patients.
- Abnormal radiography and computed tomography may raise suspicion of a large vessel vasculitis, but later angioresonance imaging is needed to confirm the diagnosis.

Introduction

A 17-year-old female, previously healthy, was admitted with a three-month history of non-periodic intermittent low-grade fever. She complained of asthenia, a weight loss of 8 kg in six months, dry cough, and headache. Physical examination showed skin pallor and abdominal murmur. There was no differential blood pressure on all four limbs. Laboratory studies revealed an hypoproliferative hypochromic microcytic anemia with hemoglobin 9.7 x 10 g/L, platelet count 400 x 10^9 cells/L, hypergammaglobulinemia, elevated sedimentation rate 120 mm/h, C-reactive protein 85 mg/L, and serum amyloid A 159 mg/L and blood cultures were negative.

Laboratory investigations showed that Epstein-Barr virus, cytomegalovirus, and human immunodeficiency virus serologies were negative as well. Bone marrow biopsy revealed a hypercellular bone marrow with erythroid hyperplasia. A chest radiography showed a round hypotransparency in the mediastinum and the interferon-gamma release assay was negative. A thoraco-abdominopelvic computed tomography indicated a parietal aortic thickening extending from the ascending to the abdominal aorta (Fig. 1).

Magnetic resonance angiography confirmed these findings (Figs. 2 and 3) and detected mild stenosis of the left renal artery (Fig. 4) and celiac trunk, suggestive of a large vessel vasculitis, such as Takayasu arteritis (angiographic type V). The pediatric vasculitis activity score was 7/63. Treatment was started with acetyl salicylic acid, oral prednisolone 1 mg/kg/day, subcutaneous methotrexate 20 mg/week, and subcutaneous adalimumab 40 mg fortnightly.

There was a reduction in the pediatric vasculitis activity score to 1/63 after a year of follow-up. However, the lab tests still revealed high inflammatory marks. Treatment was switched to prednisolone 10 mg/day and adalimumab administration every week.

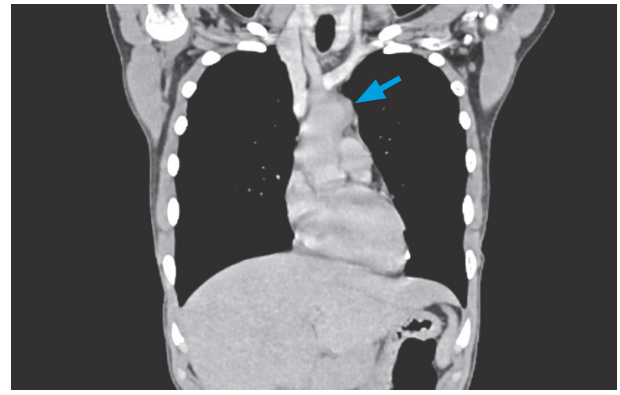

Figure 1. Thoraco-abdominopelvic computed tomography scan revealing a concentric parietal aortic thickening of the ascending aorta and aortic arch (arrow).

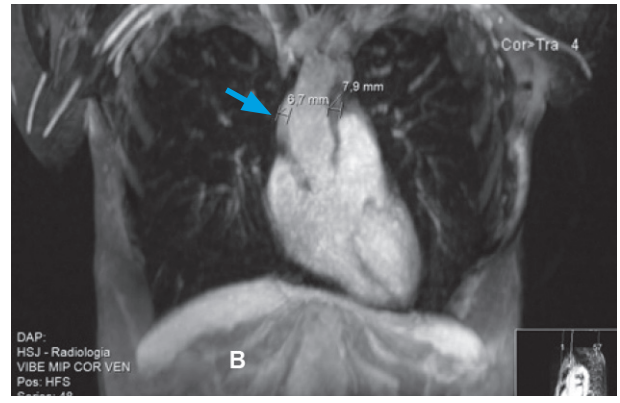

Figure 2. Magnetic resonance angiography (coronal view) showing the thickening of the ascending aorta (arrow).

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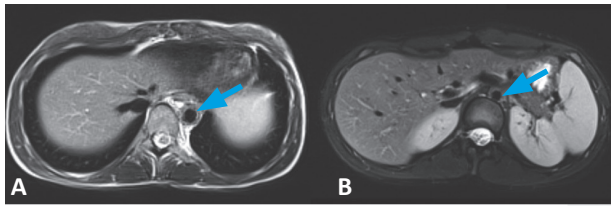

Figure 3. Magnetic resonance angiography (axial view, T2-weighted image) showing the thickening of the thoracic aorta (arrow) (A). Magnetic resonance angiography (axial view, T2-weighted image) showing the thickening of the abdominal aorta (arrow) (B).

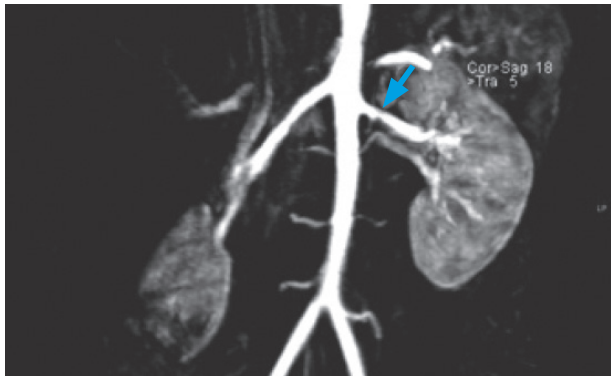

Figure 4. Magnetic resonance angiography (coronal view, reconstruction) showing the mild stenosis of the left renal artery (arrow).

Takayasu arteritis is an idiopathic large-vessel vasculitis that affects the aorta and its major branches.[1-3] Chronic granulomatous inflammation leads to stenosis and, occasionally, aneurysms of the involved portions of the arteries, producing a wide variety of symptoms.[1,2] The diagnosis of Takayasu arteritis remains a challenge mainly due to the insidious clinical course and the lack of specificity of the presenting symptoms.[3,4] Constitutional features, such as low-grade fever and weight loss, are frequently found in pediatric patients.[3] In this case, the investigation was carried out based on a change in chest radiography and a computed tomography that revealed the suspicion of vasculitis of the great arteries, which was later confirmed by angioresonance imaging. Although angiographic type V has been associated with an unfavorable outcome,[3] the prompt diagnosis and treatment tend to improve symptoms and quality of life.[2,3] Nonetheless, it is important to bear in mind the need for careful follow up and continuous screening for organ damage.[3,5]

Keywords: Adolescent; Fever/etiology; Takayasu Arteritis/diagnosis; Takayasu Arteritis/diagnostic imaging; Takayasu Arteritis/drug therapy

Author Contribuitions

DHS and MJB participated in the study conception or design. DHS, AMG and IM participated in acquisition of data. DHS, IM and MJB participated in the analysis or interpretation of data. DHS participated in the drafting of the manuscript. AMG, IM and MJB participated in the critical revision of the manuscript. All authors approved the final manuscript and are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

Conflicts of Interest

The authors declare that there were no conflicts of interest in conducting this study.

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Confidentiality of data

The authors declare that they have followed the protocols of their work center on the publication of patient data.

Consent for publication

Consent for publication was obtained.

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