CASE REPORT

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Takayasu Arteritis

Şule GÖKÇE,[a]
Deniz GÜNEŞ,[a]
Gizem ALTUNORDU,[a]
Günay AMANOVA,[a]
Sadık AKŞİT[a]

[a]Department of Child Health and Diseases,
General Pediatrics Unit,
Ege University Faculty of Medicine
İzmir

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Correspondence:
Şule GÖKÇE
Ege University Faculty of Medicine,
Department of Child Health and Diseases,
General Pediatrics Unit,
İzmir, TURKEY
sule.gokce@ege.edu.tr

ABSTRACT Takayasu arteritis is a rare systemic vasculitis. It is seen in young female adolescents as two-four times more than males. The etiology of this vasculitis also called "Japanese Disease" due to its high incidence in Japan, has not yet been clarified. Patients might present with non-specific findings in the early phase of the disease. In this article we present a case of a 16-year-old girl who has been referred to our clinic for weakness and fatigue. She had been diagnosed with iron deficiency many times in other centers. However, she applied to our clinic because her complaints did not decrease. The patient was diagnosed with Takayasu arteritis with the history and clinical findings. In this article we aimed to call attention to the Takayasu arteritis and to emphasise once again the importance of detailed history and physical examination.

Keywords: Takayasu arteritis; medical history taking; physical examination

Takayasu arteritis (TA) is a chronic autoimmune, large-vessel vasculitis and generally affects the aorta and branches, rarely pulmonary arteries. It was first described in 1908, in a Japanese patient with retinal abnormalities.[1] The disease most commonly seen in young women, especially in Far Eastern countries, causes granulomatous inflammation and circulatory disturbances by forming stenosis, thrombosis and aneurysms in the large arteries, especially the aorta. As a result of these pathologies, besides non-specific symptoms such as fever, fatigue, myalgia, arthralgia, headache, abdominal pain, severe clinical signs associated with hypoxia may occur in organs that are fed by the affected artery.[2] Here, we present the case of a young girl referred to our clinic with claudication accompanying non-specific features, and diagnosed with delayed Takayasu arteritis.

CASE REPORT

A 16-year-old girl feeling fatigue for seven months applied to our clinic with complaints of weakness that was associated with anemia. The detailed medical history was positive for intermittent and unexplained fever, weight loss, occasional headache, numbness and pain in the lower extremities particularly on the left leg that had worsened within the past 3 months, particularly while climbing stairs, and resembling ischemic pain (claudication), and which decreased after a short rest. Her past medical history and family history were unremarkable. As our patient had features of claudication, a detailed systemic examination was done which revealed the absence of arterial pulses in the left arm. The arterial pulses in the right arm were a little feeble. Her blood pressure was 135/76.9 mmHg in right arm, however blood pressure could not be

measured and non recordable on her left arm and legs. A systolic murmur was heard on the abdominal aorta. Laboratory findings showed an elevated erythrocyte sedimentation rate (ESR) of 60 mm/h (normal value <20 mm/1 h) and a serum C-reactive protein level of 4.6 mg/dl (normal value <0.5 mg/dl). The results of other laboratory tests were normal. Upper extremity Doppler Ultrasound showed irregularity and stenosis with diffuse thickening on the long segment wall in the left subclavian, axillary and brachial proximal part. Thorax angio computerized tomography of the patient showed wall thickening and lumen narrowing in the aortic arch, descending aorta and abdominal aorta, complete occlusion of the two cm segment in the infrarenal abdominal aorta, complete occlusion of the left axillary artery in the subclavian artery (Figure 1 A, B, C). On the basis of clinical manifestations, Doppler Ultrasonografic and angiographic findings, the diagnosis of TA was made. Interventional radiology indicated that the patient's endovascular recanalization was impossible. In the Cardiovascular Surgery council, it was stated that the distance of cladication of the patient was over 50 meters without night time leg pain, so the intervention was not necessary for the time being and clopidogrel treatment would be appropriate. Two mg/kg/day methylprednisolone therapy was started and she was transferred to the our rheumatology clinic.

DISCUSSION

Takayasu arteritis (TA), also called pulseless disease is a chronic inflammatory vasculitis that was first described by Mihito Takayasu, a Japan Ophthalmologist in 1908. TA is rare in childhood, but up to 20% of the patients are diagnosed under the age of 19 years. It is widely seen in women of childbearing age and in the Asian region mainly Southeast Asia, India, Turkey and Africa . It commonly involves the aorta and its major branches.[3,4] In Japan, the prevalence is reported as 4/100.000, but according to autopsy reports this rate is one third / 3,000.[5] A study conducted by Sarıtaş et al. from Turkey stated the incidence as 3,4 /1,000,000.[6] The etiopathogenesis is not clear, as is the case in many other vasculitis and autoimmune diseases. However, infections, environmental factors, abnormal cell-mediated immunity, other molecular and genetic abnormalities may contribute to it.[7] It is known that the disease is mediated by T-cell and causes destruction by some inflammatory cytokines in the large arteries, especially in the aorta and its branches. Progressive granulamatous inflammation leads to stenosis, thrombosis, and aneurysms in the affected large arteries, and clinical manifestations of the disease develop due to these pathologies.

An early active inflammatory phase and a late chronic or stenotic phase make up the clinical course of TA.[8] In early or pre-pulseless phase, non-specific findings such as fever, weight loss, fatigue, headache, night sweats are observed. In serial case studies, it is stated that the most common cause of delayed diagnosis is due to the non-specific findings in the acute phase, which last for three months or several years, and even some cases might present with stenotic phase. A large series of cases conducted by Maffei et al. reported that non-specific findings include about 40% fatigue, 20-30% fever and weight loss.[9] Another study showed 45% fever, 20% to 30% vomiting and headache. Abdominal pain, unexplained seizures, abnormal urinary excretion, joint pain, hypertension can also be observed in the early phase of TA. Another study in which fourteen cases were evaluated stated that the non-specific findings could be seen in more than 30% of patients.[5] Our patient had fatigue, intermittent and unexplained fever, weight loss, occasional headache associated with different diagnoses like anemia. The time from the onset of the findings to the diagnosis was approximately one year, which is consistent with the literature.

Progressive inflammation in the chronic or stenotic phase of the disease; stenosis, occlusion, or aneurysm develop on the affected arteries, and findings such as lack of pulse, retinopathy, extremite temperature differences, color changes occur. Kerr and colleagues reported that carotid arteries (70%) were involved most commonly, subclavian arteries (45%), and the upper extremity was more common than the lower extremity.[10] Our patient had aortic narrowing in the aortic arch, descending aorta and abdominal aorta, and complete blockage

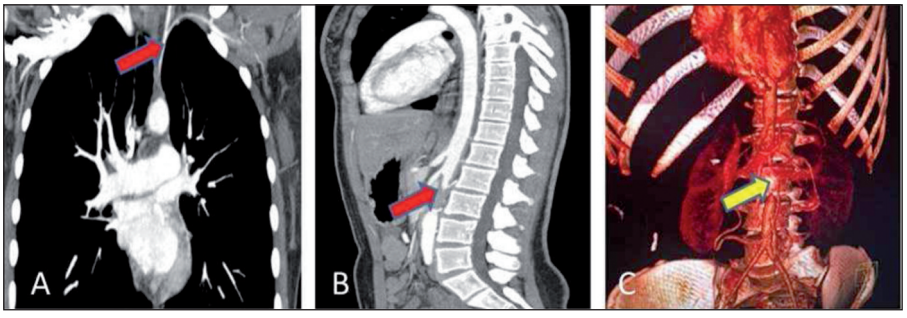

FIGURE 1: A) Complete occlusion of the left axillary artery in the subclavian artery, **B, C)** Complete occlusion of the two cm segment in the infarenal abdominal aorta.

in a two cm segment in the infrarenal abdominal aorta. The left carotid artery and left subclavian artery lumen were narrow and the left axillary artery was completely obstructed.

Claudication, which is defined as limb pain that occurs during movement and regresses with short rest, is a very important clinical finding that is frequently found in the chronic or stenotic stage of the disease. Li and colleagues have reported that they have detected 30% claudication in their studies.[5] Although there was no evidence of upper extremity claudication in the history and physical examination of our patient, there was a lower extremity pain which became evident during movement. Takayasu arteritis was thought to be caused by the claudication presentation of the case, the lack of blood pressure and pulselessness.

The diagnosis of the disease is mainly based on a detailed history and physical examination. In addition to the angiographic abnormality of the aorta and/or main branches or pulmonary arteries according to the 2010 EULAR/PRINTO/PRES criteria; 10 mmHg systolic blood pressure difference between pulmonary abnormalities and/or claudication, extremities, presence of murmur in aorta or branches, hypertension, and at least one of the high acute phase indicator criterion are observed (Table 1).[11] Systemic infections, giant cell arteritis, other inflammatory diseases and malignancies should be considered in differential diagnosis. In addition to angiographic abnormality, our case met the diagnostic criteria with pulse abnormality and claudication, hypertension, abdominal aorta murmur and high acute phase indications.

There are no specific biochemical markers for Takayasu arteritis. However, as has been pointed out in many studies, acute phase reactants are generally high during the inflammatory phase. The sedimentation rate of our patient was 60 mm / hour (high) in our clinic application.

The response to corticosteroids is usually good in standard therapy. In addition, conventional immunosuppressive agents can also be used. A by-pass may be required in some cases where the distention in the artery is advanced. However, when medical treatment is unresponsive, or when there

TABLE 1: EULAR/PRINTO/PRES criteria and classification definition of Takayasu Arteritis.

Mandatory criteria	
Angiographic abnormality	Angiography (conventional, CT, and MRI) of the aorta, its main branches or pulmonary arteries showing aneurysm/dilatation, narrowing, occlusion, or thikhened arterial wall, not due to any other causes
Additional criteria (need one of the five)	
1) Pulse deficit or claudication	Lost/decreased/unequal peripheral artery pulse
2) Blood pressure discrepancy	Discrepancy of four- limb systolic blood pressure >10 mmHg in any limb
3) Bruits	Audible murmur or palpable thrills over large arteries
4) Hypertension	Systolic/ diastolic blood pressure >95 centile for height
5) Acute phase reactant	Erythrocyte sedimentation rate (ESR) >20 mm per hour or C reactive protein (CRP) above normal

is sudden thrombosis, graft operation (ven or artificial), transluminal angioplasty or endovascular stent may be performed by by-pass.[5] In our patient, methylprednisolone therapy was given without the need for any intervention process.

In conclusion, Takayasu arteritis should be considered in the etiology of cases with non-specific systemic findings. We also wanted to emphasize once again the importance of detailed history and detailed physical examination through this case whose diagnosis was delayed.

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Conflict of Interest

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions

Concept: *Şule Gökçe;* ***Supervision:*** *Sadık Akşit;* ***Data Collection:*** *Deniz Güneş, Gizem Şenyazar; Günay Amanova;* ***Literature Search:*** *Sadık Akşit;* ***Writing Manuscript:*** *Şule Gökçe, Sadık Akşit.*

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