

Takayasu arteritis: a rare case scenario

Singh M.¹, Dayal R.², Kumar N.³, Singh S.P.⁴, Gupta L.K.⁵, Neelam⁶

¹Dr. Madhu Singh, Lecturer, Department of Paediatrics, ²Prof (Dr). R. Dayal, Professor and Head, Department of Paediatrics, ³Prof (Dr). Neeraj Kumar, Professor, Department of Paediatrics, ⁴Dr. Sheo Pratap Singh, Assistant Professor, Department of Paediatrics, ⁵Dr. L.K. Gupta, Associate Professor, Department of Pediatrics, Government Medical College, Firozabad, ⁶Dr. Neelam, Junior Resident, Department of Paediatrics, ^{1,2,3,4,6}authors are affiliated with S.N. Medical College, Agra, Uttar Pradesh, India.

Corresponding Author: Dr. L.K. Gupta, Associate Professor, Department of Pediatrics, Government Medical College, Firozabad (U.P.) India. E-mail: guptaneeta98@gmail.com

Abstract

Takayasu's arteritis (TA), commonly known as "Pulseless disease", is rare granulomatous arteritis, which affects large and medium-sized arteries, especially the aorta and its branches. Other arteries that can be affected are proximal portions of pulmonary, coronary, and renal arteries. Initially, mononuclear cell infiltrates in the adventitia leading to granuloma formation, followed by disruption of the elastic layer and subsequent massive medial and intimal fibrosis. These lesions result in segmental stenosis, occlusion, dilatation, and aneurysmal formation in the affected vessels.

Diminished or absent pulses are present in 84-96% of patients, associated with limb claudication and blood pressure discrepancies between the two arms. The symptoms are secondary to the involved artery, and it may evolve into a life-threatening condition. The involvement of the carotid artery leads to ophthalmic artery hypoperfusion and causes the ocular ischemic syndrome. It usually affects young females during the second and the third decades of life but rare cases have been reported in children as young as 24 months of age. Although females are more likely to be affected in TA, males are also affected. In adults approximately 80% of patients with TA are women, although the female-to-male ratio varied from 9:1 in reports from Japan, 6.9:1 in Mexico to 1.2:1 in Israel.

Aim of our report is to describe the case of a young female of 14 years of age. She presented with arterial hypertension and respiratory distress. CT Angiography showed Type IIb Takayasu Arteritis. ECHO revealed dilated cardiomyopathy with grade III diastolic dysfunction with decreased ejection fraction up to 20-25%. She was treated with steroids, anti-hypertensive drugs, and diuretics.

Key words: Takayasu's arteritis, Pulseless disease, absent pulses disease.

Introduction

Takayasu arteritis is a large vessel vasculitis affecting mainly aorta and its major branches. It occurs most commonly in female patients in the second and third decades of life, but also has been reported in children as young as 24 months of age [1-4]. The disease is more prevalent in Asian countries but cases are reported from all ethnicities. Clinical manifestations of TA are nonspecific. The course of the vasculitis is divided into an early active inflammatory phase and late chronic phase. The active phase lasts for weeks to months with

a remitting and relapsing course. It is characterized by the systemic disease with symptoms of fever, general malaise, night sweats, loss of appetite, weight loss, headaches, dizziness, arthralgia, skin rashes, etc. The acute phase does not occur in all patients, but constitutional symptoms are often seen in children with TA. TA is seldom diagnosed in the early phase.

Feature suggestive of vessel inflammation esp. tenderness along arteries, bruits, and an aneurysm may point to the diagnosis of TA. The late chronic phase occurs due to arterial stenosis and/or occlusion and ischemia of organs. Its clinical manifestations are varied and related to the location of arterial lesions. Suspected

Manuscript received: 4th September 2019

Reviewed: 10th September 2019

Author Corrected: 14th September 2019

Accepted for Publication: 17th September 2019

Case Report

TA mandates vascular imaging. Intra-arterial angiography is the gold standard for diagnosis and evaluation of Takayasu arteritis. With recent advances, it has been largely replaced by computed tomography angiography or magnetic resonance angiography (MRA). Immunosuppressants such as prednisone and/or methotrexate to decrease or eliminate inflammatory activity are the mainstay of treatment. The 5-year survival rate in adults is as high as 94% [5].

Symptomatic stenotic or occlusive lesions can be effectively treated with endovascular revascularization procedures like bypass grafts, patch angioplasty, endarterectomy, percutaneous transluminal angioplasty, or stent placement [6]. Despite providing a short-term benefit, endovascular revascularization procedures are associated with a high failure rate in patients with Takayasu's arteritis. Both, surgical and endovascular, treatments become risky and achieve poorer outcomes if they are undertaken during a period of inflammatory activity [7]. Descriptions of Takayasu arteritis in pediatric age are scanty.

Case Report

Patient Bharti, 14 years female child with unremarkable family or past medical history was admitted for fever and cough with respiratory distress, in the form of fast breathing from the past two months. These symptoms were on and off in nature. Her parents also reported that she had fatigue, myalgia for the past two months.

On admission, she was alert, afebrile with acute respiratory distress. Her Blood Pressure was 122/74mmHg in the right upper arm in the supine position and not felt in other limbs. Other pulsations like brachial and radial of the left hand and femoral, popliteal, posterior tibial and dorsalis pedis of both sides were absent. She was also having crept in both sides of the lungs and hepatomegaly of 3-4 cms below the right subcostal margin in midclavicular line with icterus present in eyes. Her urine output was also decreased by up to 0.7ml/kg/hr. The rest of the physical examination was normal.

Investigations on admission revealed mild anemia (Hb-9.6gm/dl) and jaundice (S.bilirubin total-3mg/dl, direct -1.2mg/dl, indirect-1.8mg/dl). The rest of the laboratory investigations like TLC, platelets, S.creatinine, S.urea, and electrolytes was normal. USG abdomen and thorax reports having thickened and contracted gall bladder with mild to moderate ascites with minimal pleural effusion.

2 D ECHO report revealed all cardiac chambers dilate with global hypokinesia with an ejection fraction of 20%-25% only (dilated cardiomyopathy with grade III diastolic dysfunction). CT Angiography showed Type IIB Takayasu Arteritis.

On treatment with antihypertensives, diuretics, and steroids; she got relieved of respiratory distress and her urine output increased.

Discussion

Takayasu arteritis has been rarely reported in childhood. It is an autoimmune disease involving the arterial walls of large arteries, causing panarteritis. The American Rheumatological Society considers three of the following six criteria necessary for a definite diagnosis of Takayasu's disease:

1. Onset before 40 years
2. Claudication of the extremities
3. A decrease in the brachial pulse in one or both arms
4. A difference of 10 mm Hg or more in blood pressure measured in both arms
5. Audible bruit on auscultation of the aorta or subclavian artery
6. Narrowing at the aorta or its primary branches on an arteriogram

We evaluated a case of 14 years old female meeting 5 of the 6 criteria required to diagnose TA. A common clinical mode of presentation in our patient was acute respiratory distress with arterial hypertension, together with other nonspecific symptoms like fever, fatigue, myalgia. The disease is also called 'pulseless disease' since peripheral pulses are often absent due to vascular obstruction; however, in this patient only right upper limb pulsations were felt. In another report of 31 children with TA from South Africa [8], arterial hypertension was the most common presenting feature, followed by cardiac failure, bruits, and absent pulses.

The diagnosis of TA is based on characteristic findings of the diseased aorta and its major branches seen on angiography. This is demonstrated by luminal abnormalities such as stenosis or aneurysmal dilatation of the aorta, its major branches, and the pulmonary arteries. Concerning imaging studies, traditionally the angiographic patterns have been divided in: type I, affecting the aortic arch; type II, the thoracic and abdominal aorta; type III, the aorta both above and below the diaphragm; and type IV, the aorta and the pulmonary arteries [9-11]. In our case, it was type II.

Case Report

Ultrasonography and positron emission tomography are new, promising techniques to assess large-vessel vasculitides. Color-coded Doppler sonography can facilitate an accurate diagnosis of Takayasu arteritis by the characteristic appearance. Homogeneous circumferential intima-media thickening of the common carotid arteries is a specific ultrasonographic finding in patients with Takayasu arteritis [12].

More recently MRI has been used to establish the diagnosis of TA in children, to monitor disease activity and to guide treatment. Early in the disease of TA, smooth muscle thickened vessel walls, which may be the only manifestation of vascular inflammation, maybe not detected by conventional angiography but MRI can visualize the thickened vessel wall directly, and in addition it can show other signs of active inflammation such as mural edema with T2-weighted imaging and increased wall vascularity with enhanced imaging.

TA is a disease with severe prognosis, mortality rate being reported in children from 35 to 40% by five years. It is therefore important to have a high index of suspicion and in doubtful cases a low threshold for diagnostic evaluation.

The present study underlines the possibility of TA in any young patient with unexplained arterial hypertension.

Funding: Nil, **Conflict of interest:** None initiated,
Permission from IRB: Yes

References

1. Ladhani S, Tulloh R, Anderson D. Takayasu disease masquerading as interruption of the aortic arch in a 2-year-old child. *Cardiol Young*. 2001;11(2):244-246. doi: <https://doi.org/10.1017/S104795110100021X>
2. Kothari SS. Takayasu's arteritis in children-a review. *Images Paediatr Cardiol*. 2002;3(4):4-23.

3. Rossman MG Takayasu arteritis. <http://emedicine.medscape.com/article332378-overview>. Accessed on 27 Sep 2019.
4. Hom C. Pediatric Takayasu arteritis. <http://emedicine.medscape.com/article1007566-overview>. Accessed 27 Sep 2019.
5. Koide K. Takayasu arteritis in Japan. *Heart Vessels Suppl*. 1992;7:48-54.
6. Tyagi S, Khan AA, Kaul UA, Arora R. Percutaneous transluminal angioplasty for stenosis of the aorta due to aortic arteritis in children. *Pediatr Cardiol*. 1999;20 (6): 404-410. doi: <https://doi.org/10.1007/s002469900501>
7. Tyagi S, Sharma VP, Arora R. Stenting of the aorta for recurrent, long stenosis due to Takayasu's arteritis in a child. *Pediatr Cardiol*. 1999; 20(3):215-217. doi: 10.1007/s002469900445.
8. Hahn D, Thomson PD, Kala U, Beale PG, Levin SE. A review of Takayasu's arteritis in children in Gauteng, South Africa. *Pediatr Nephrol*. 1998;12(8):668-675. doi: 10.1007/s004670050526
9. Jain S, Sharma N, Singh S, Bali HK, Kumar L, Sharma BK. Takayasu arteritis in children and young Indians. *Int J Cardiol*. 2000;75(1):S153-S157. doi: 10.1016/s0167-5273(00)00180-7.
10. Ozen S, Bakkaloglu A, Dusunsel R, Soylemezoglu O, Ozaltin F, Poyrazoglu H, et al. Childhood vasculitides in Turkey: a nationwide survey. *Clin Rheumatol*. 2007;26(2):196-200. doi: 10.1007/s10067-006-0266-6
11. Muranjan MN, Bavdekar SB, More V, Deshmukh H, Tripathi M, Vaswani R. Study of Takayasu's arteritis in children: clinical profile and management. *J Postgrad Med*. 2000;46(1):3-8.
12. Morales E, Pineda C, Martínez-Lavín M. Takayasu's arteritis in children. *J Rheumatol*. 1991;18 (7): 1081-1084.

How to cite this article?

Singh M, Dayal R, Kumar N, Singh S.P, Gupta L.K, Neelam. Takayasu arteritis: a rare case scenario. *Int J Pediatr Res*. 2019;6(09):481-483. doi:10.17511/ijpr.2019.i09.07