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Case Report

Takayasu's arteritis: a diagnostic dilemma

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ABSTRACT

Takayasu`s arteritis is an uncommon form of chronic vasculitis that primarily affects medium and large blood vessels, including the aorta and its major branches. The disease is mainly seen in young women. This is marked by mononuclear cell infiltration and granulomatous inflammation within the vascular media. These pathological changes result in thickening of the arterial wall, which can lead to stenosis, occlusion, or the formation of aneurysms. It can lead to severe complications such as stroke, ischaemic heart disease, pulmonary hypertension, secondary hypertension and aneurysm formation. Here we present a case of Takayasu`s arteritis in a 10-year-old girl who presented with low grade fever, headache and systemic hypertension with thickened walls of left subclavian artery and left renal artery in computed tomography angiogram. Early diagnosis and prompt treatment are crucial for managing Takayasu`s arteritis. When the disease is in a dormant phase, the prognosis tends to be more favourable.

Keywords: Hypertension, Vascultis, Inflammation, Headache, Hyperkalaemia

INTRODUCTION

Takayasu`s arteritis is a specific chronic inflammatory vasculitis that affects large and medium sized arteries, including the aorta and its branches. The disease affects Asian women under the age of 40.[1] The incidence range from 0.3 to 3.3 million per year and prevalence range from 0.9 to 360 cases per million depending upon the geographic region studied, with higher rates observed particularly in Asia.[2] Mean age at disease onset in the paediatric population is 12 years but it is seen in all ages.[3,4] The precise cause of Takayasu's arteritis is believed to involve a cell-mediated inflammatory process within the all layers of blood vessels including adventitia inflammation, destruction of media elastic tissue and neovascularisation of intima.[5] This inflammation leads to mononuclear and granulomatous infiltrates, which can cause vessel occlusion, aneurysmal dilation, or narrowing of affected segments.[6] These changes result in a range of clinical symptoms depending on the areas involved. Patients may present with symptoms such as limb pain or weakness, headaches, syncopal episodes, and unequal blood pressure. Constitutional symptoms, including fever, weight loss, and fatigue, may appear initially.[7] Due to the frequent obstruction of major arteries branching from the aorta, the condition is also referred to as pulseless disease. The differential diagnosis involves arteritis associated with syphilis, systemic lupus erythematosus, rheumatoid arthritis, sarcoidosis, and Marfan syndrome.[8] This is a distinct form of vasculitis that requires a multidisciplinary approach for diagnosis and management. The care team typically includes specialists such as a rheumatologist, imaging experts, cardiologists, interventional cardiologists, cardiac surgeons, and pathologists, each contributing to different aspects of the disease evaluation and treatment.[9] Magnetic resonance arteriography and computed tomography angiography are non-invasive imaging techniques of choice.[10] Treatment involves medical, interventional, or surgical approaches. Steroids and methotrexate are the first-line medications.[11] For patients with refractory or severe Takayasu's arteritis, tumour necrosis factor (TNF) inhibitors may be used.[12] For

arterial stenosis and arterial dilation interventional methods are used. when interventional therapy fails, surgical therapy used. The prognosis is uncertain due to relapses and complications.[13,14] The purpose of this case report is to highlight unique manifestations of this vasculitis, including a notable combination of stable and active disease, as well as severe complications that developed after disease onset.

CASE REPORT

A 10-year-old girl who experienced low grade fever from the last 6 months, headache and systemic hypertension from the last 3 months was hospitalised at our hospital. She also had weakness, myalgia and decreased appetite. Over a 3 months of symptoms were present, 1 day before admission patient had hypertensive emergency and admitted in our hospital. The physical examination showed a significant discrepancy (>10 mmHg) in blood pressure between the right and left arms. Her blood pressure in right arm was 160/110 mmHg and 146/100 in the left arm. She had impalpable brachial and radial pulses on her left side. She had a body temperature of 37.8 C, heart rate of 96 beats/min and respiratory rate of 26 breaths/min. Anthropometry shows height 126 cm (-1.6 SD), weight 25 kg (-1.0 SD). Examination of the chest and precordial showed lateral down ward shift of apex beat and systolic murmur at mitral area. Laboratory evaluations, including blood cultures, tuberculosis skin and sputum tests, troponin I, CPK-MB and autoimmune serological assessments (p-ANCA and c-ANCA) all yielded negative results. The erythrocyte sedimentation rate level was 35 mm/h (high), and the C reactive protein level was 7.8 mg/L (low) and haemoglobin 10.6 g/dl (low) all are positive findings. Arterial blood gas analysis shows metabolic alkalosis and serum biochemistry shows hyponatremia and hypokalaemia. Aldosterone and renin levels sent before starting any antihypertensive medication, these levels come within normal limits so primary hyperaldosteronism ruled out. Echocardiography done showed dilated left ventricle, global left ventricle hypokinesia, moderate left ventricle systolic dysfunction with ejection fraction of 35%, eccentric hypertrophy of left ventricle, mild valvular aortic regurgitation and less flow velocity in left subclavian and axillary arteries suggestive of obstruction. Electrocardiogram done shows changes in wave pattern corresponds to left ventricular hypertrophy. Fundoscopy showed papilledema. Chest radiography done showed no abnormalities.

A Doppler ultrasound of the all 4 limbs revealed a significant narrowing of the proximal left subclavian artery, rest other 3 limbs normal. Contrast enhanced computed tomography (CECT) abdomen was carried out and the results showed that left kidney was relatively smaller in size measuring approx. 72×38 mm and right kidney measuring approx. 108×50 mm, no contrast opacification seen in left artery-possibility of stenosis. Computed tomography renal angiography done which demonstrated significant narrowing of the distal left renal artery as shown in Figure 1.

Based on the clinical findings, inflammatory markers and imaging results patient was diagnosed with Takayasu`s arteritis. Methylprednisolone pulse dose started and then tapered methotrexate (15 mg weekly) and folic acid 5 mg/week was also started.

The cardiologist opinion was take calcium channel blockers (amlodipine at 0.5 mg/kg/day) and alpha blockers (prazosin at 0.5 mg/kg/day) for hypertension. The arterial stenting was done after the control of acute phase at subclavian and renal artery and patient discharged after a week of surgery with normal percentile of blood pressure.

Follow up

The patient followed up for 2 weeks after the discharge. Patient chief complaint of headache and palpitations resolved after the treatment. Blood pressure maintained between the normal limits.

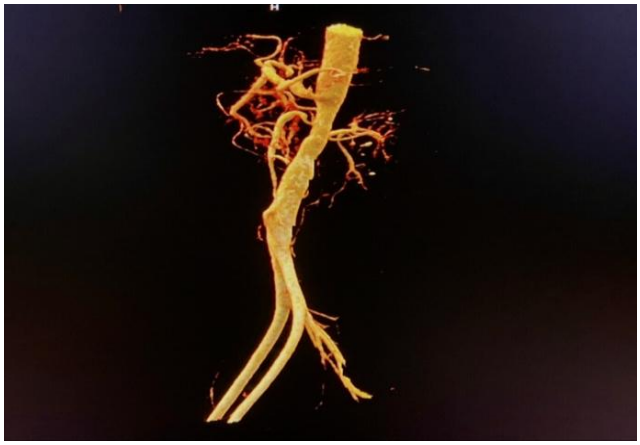

Figure 1: Marked narrowing of distal left renal artery.

DISCUSSION

Mikito Takayasu, Japanese ophthalmologist first presented this case in 1908 at the Annual Meeting of the Japan Ophthalmology Society. He observed arteriovenous anastomoses around the optic disc in a 21-year-old woman.[15] After Takayasu's observations, other ophthalmologists identified radial artery involvement, leading to the classification of the condition as a vasculitis.[16]

In the case study done by Khadka et al, 26-year-old woman was admitted for light headedness and frequent syncopal attacks followed by loss of consciousness.[7] In the another case study on Medicina 2024, 48-year-old Caucasian woman was admitted for prolonged crises of the angina pectoris.[9] In our case, the age of the patient is 10 years the patient presented with low grade fever from the last 6 months, headache and systemic hypertension from the last

3 months which is a rare presentation for paediatric population. Arterial hypertension presents in the form of discrepancy in pulses and difference in the blood pressure in 60% of the children and the authors would like to emphasize the importance of thorough physical examination which can be missed in busy clinics.

Clinically, Takayasu disease can present with a wide range of symptoms depending on the vessels involved. It often begins with constitutional symptoms such as fever, weight loss, claudication, and fatigue, before progressing to other manifestations such as weakness, headache, dizziness, hypertension, retinopathy, aortic regurgitation, and vascular bruits. Depending upon the site of occlusion caused by inflammatory infiltrates, neurological symptoms like seizures and syncope also seen.[17] Tuberculosis is a known triggering factor for Takayasu arteritis. Many patients diagnosed with this vasculitis have a history of tuberculosis. These patients either had a prior history of tuberculosis or were diagnosed with active or latent tuberculosis infection concurrently with vasculitis.[18]

Investigations such as acute phase reactants (CRP and ESR) also provide evidence in favour of diagnosis. Magnetic resonance angiography (MRA) and computed tomography angiography (CTA) are the gold-standard imaging techniques for diagnosing this disease.[10] CTA is generally more cost-effective and often offers better resolution than MRA. In our case, we used CTA to confirm the diagnosis.

Our patient met the current diagnostic criteria, according to ACR EULAR 2022: absolute requirements - age less than or equal to 60 years at time of diagnosis, and evidence of vasculitis on imaging; additional clinical criteria - female sex (+1), reduced pulse in upper extremity (+2), and systolic blood pressure difference in arms more than or equal to 20 mmHg (+1); and additional imaging criteria - two arterial territories are affected (+2), and abdominal aorta involvement with renal or mesenteric involvement.

A score more than or equal to 5 points is needed for the classification of Takayasu arteritis.[19]

Glucocorticoids remain the first-line treatment for Takayasu arteritis, achieving remission in approximately 60% of patients. Disease-modifying antirheumatic drugs (DMARDs) are of two main types: synthetic and biologic. Synthetic DMARDs include methotrexate and Janus kinase inhibitors like tofacitinib. Biologic DMARDs target specific components of the immune system like interleukin-6 (IL-6) inhibitors such as tocilizumab and tumor necrosis factor (TNF) inhibitors such as infliximab. Above medications can be used in combination with glucocorticoids or with each other to enhance therapeutic efficacy. The choice of treatment depends upon individual patient after considering factors such as disease severity, comorbidities, and potential side effects.[19]

This case study is one of the few studies performed on the subject of Takayasu arteritis in India as the disease is rare and mostly affects women under the age of 40. In this case, the patient was only followed up for 3 months, and her long-term condition was not included in the study. It is recommended that patients with Takayasu's arteritis undergo long-term observation, even after being discharged from the hospital.

CONCLUSION

The case highlights the importance of thorough medical examination in each visit just by palpating peripheral pulses and recording blood pressure can avoid catastrophic emergency admission. A multidisciplinary approach is essential for the diagnosis, follow-up, and treatment of Takayasu's arteritis.

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