

A Five Years Old Girl Child with Takayasu Arteritis

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ABSTRACT

Takayasu arteritis is a systemic vasculitis of large vessels that mainly involves the aorta and its branches. It normally presents in third decade of life and has rarely been reported in children under 10 years of age. We report here a case of Takayasu arteritis in a 5 years old girl who presented with headache, generalized body swelling, severe hypertension, proteinuria and minimal functioning kidneys. Conventional angiography demonstrated narrowing of descending aorta, right subclavian artery and right common iliac artery. She responded steroids, diuretics, antiplatelets and digoxin and discharged home on maintenance therapy.

Key Words: *Takayasu arteritis. Headache. Descending aortic. Subclavian artery stenosis. Common iliac artery stenosis. Conventional angiography. Hypertension.*

INTRODUCTION

Takayasu arteritis (TA) is a systemic granulomatous vasculitis of the large vessels that involves the aorta and its large branches. It is the third most common cause of vasculitis after Henoch Schonlein purpura and Kawasaki's disease.¹ It has been rarely reported in children due to its uncommon presentation and rarity of occurrence in children less than 10 years of age. It most commonly presents in women in third decade of life with a female to male ratio of 8.5 : 1.² However, cases have been reported in children in age less than 10 years.³

European League Against Rheumatism/Paediatric Rheumatology International Trials Organisation/Paediatrics Rheumatology European Society (EULAR/PRINTO/PRES) proposed validated classification criteria for the diagnosis of TA include angiographic abnormalities, alterations in peripheral arterial pulses, systolic blood pressure (BP) discrepancy in any limb, arterial hypertension, large artery bruits, and elevated acute phase reactant.⁴

TA responds well to corticosteroids treatment; although in some cases immunosuppressive therapy and anti-hypertensive drugs need to be given and even some patients may benefit from surgical intervention.³

Here, we report the case of a young girl child with TA identified with the above diagnostic criteria.

CASE REPORT

A 5 years old girl child, resident of Skardu, was referred to our hospital for the work-up of uncontrolled hypertension. The symptoms had started 30 days back

when she developed low grade intermittent fever of sudden onset associated with sweating but not with rigours or chills. The patient complained of severe throbbing headache which was not relieved by analgesics. The patient also complained of mild, non-productive cough mostly during daytime. After 15 days, the patient developed facial puffiness which progressed to generalized swelling of the body. The patient had several episodes of headache, shortness of breath and generalized body swelling since one year of age for which she had several admissions in Skardu. The child had remained admitted first at the Skardu Hospital for about one week and was then referred to Military Hospital, Rawalpindi.

On admission, she was afebrile. Her heart rate was 128 beats per minute and respiratory rate was 55 breaths per-minute. She had generalized body swelling and her height and weight fell on the 50th centile for the age. Her right radial and femoral pulses were very weak while the radial pulse on the left side was bounding. Her blood pressure in the left arm was 160/90 mmHg which was above 95th centile according to age and height-centile charts and the right arm blood pressure was 150/80 mmHg. The blood pressures in the left and right leg were 90/60 mmHg and 115/67 mmHg, respectively. She had a systolic bruit in the right common carotid artery. Her respiratory examination showed bilateral vesicular breathing with end inspiratory basal crepitations on the back uptill middle zone. Her neurological and abdominal examination was normal. An eye examination revealed normal visual acuity. Her fundoscopy showed clear cornea and normal anterior chambers, sharp disc margins and normal retinal arteries with no indentation or new vessel formation.

A urinalysis revealed trace proteinuria. A 24 hours urine examination revealed urinary protein loss of 11.14 mg/m²/24 hours. Blood urea was elevated at 20 mmol/l with a serum creatinine of 120 µmol/l. The Westergren erythrocyte sedimentation rate was elevated at 36

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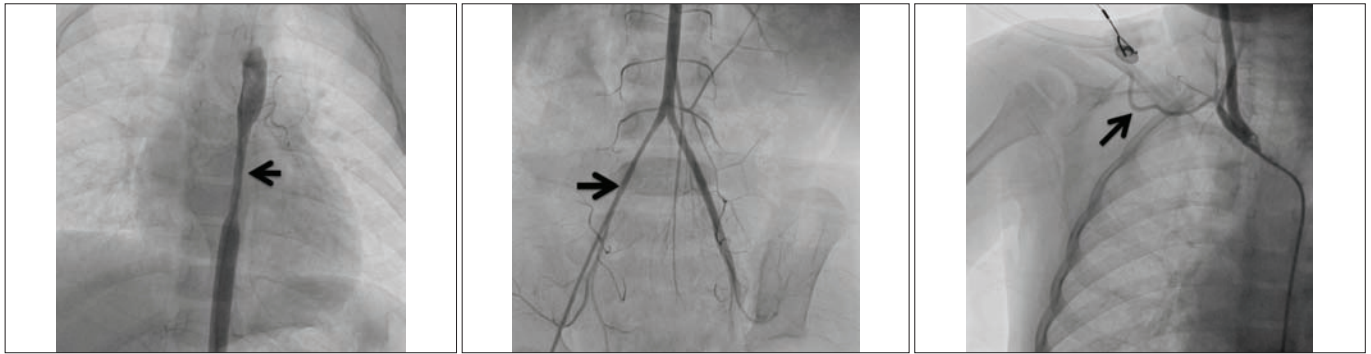


Figure 1: Arteriogram showing tubular narrowing of descending aorta (arrow).

Figure 2: Arteriogram showing relative narrowing of the right common iliac artery.

Figure 3: Arteriogram showing narrowing of right subclavian artery.

mm/hour. The C-reactive protein was 12 mg/dl. Her serum was negative for rheumatoid factor, antinuclear factors, anti-double stranded DNA antibodies and anti-neutrophil cytoplasmic antibodies. The complement factors C3 and C4 were normal. The hepatitis B antigen was negative. The tuberculin skin test was positive. ASOT was normal. Chest radiography showed cardiomegaly. Electrocardiography showed left ventricular hypertrophy. Her initial echocardiography was done which showed mildly dilated left ventricle with moderate left ventricular function and ejection fraction of 50%. She was kept on captopril tablet (300 ug/kg) and nifedipine capsules (200 ug/kg). Her general condition improved but the blood pressure was still not under control and the difference of right and left arm blood pressure was > 10 mmHg. On the basis of strong clinical suspicion we again requested a 2D echocardiogram which showed thoracic coarctation and an arteriogram was suggested which showed tubular narrowing of descending aorta (Figure 1), narrowing of the right common iliac artery (Figure 2) and narrowing of the right subclavian artery (Figure 3). Her DTPA scan revealed diminished functioning of both kidneys with non-obstructive pelvicalyceal hold-up.

The patient was treated with prednisolone with an initial dose of 2 mg/kg/day in 3 divided doses. This dose was maintained for 30 days and then a slow tapering of the prednisolone was started. The hypertension was controlled by carvedilol 100 ug/kg 12 hourly. An antiplatelet agent (Aspirin 12.5 mg/kg 12 hourly) was prescribed to prevent thrombotic complications. The patient was also given a combination of spironolactone, frusemide and digoxin. With this regime the girl improved her blood pressure and her renal functions normalized. Her ESR which was initially 36 fell down to 20 and then 15 mm after first hour. The patient was discharged with the same medicines and was advised weekly follow-up.

DISCUSSION

Takayasu's arteritis (TA) is a chronic granulomatous vasculitis of unknown etiology involving the aorta and its

branches, which produces vascular sequelae with stenotic lesions and/or thrombus formation. Takayasu, a Japanese ophthalmologist, first described the condition in 1908 in a young female with retinal changes. Later in 1928, Shimzu and Sano described it as pulseless disease. Its estimated incidence is 2.6/100,000,0/year. The disease has worldwide distribution but is more common in Japan, India and China.⁵ In Pakistan, it has been reported in adults and children from 12 to 19 years of age.⁶ TA is very rare in children less than 10 years of age³ and has not been reported from Pakistan in this age group as per authors knowledge.

The clinical manifestations of Takayasu arteritis are very non-specific and thus this clinical entity is difficult to diagnose. However, the earlier this disease is diagnosed, the better is the prognosis.² The clinical symptoms normally present in the children are headache (31%), fatigue, claudication of the upper limbs and abdominal pain.³ However, this patient presented with generalized body swelling preceded by facial puffiness which has rarely been reported in literature.

The patient met the EULAR (European League Against Rheumatism) / PRES (Paediatric Rheumatology European Society) criteria which has been specially formulated for paediatric Takayasu arteritis patients. This criteria involves the mandatory criterion of angiographic abnormalities (conventional, CT, or MR) of the aorta or its main branches with at least one of the following four features: decreased peripheral artery pulse(s) and/or claudication of extremities, blood pressure difference > 10 mmHg, bruits over aorta and/or its major branches and hypertension (related to childhood normative data).⁴

The imaging findings in patients with TA generally have typical angiographic findings including a combination of aneurysm formation and cylindrical segmental stenosis or occlusion. Pathologic examination of affected arteries is characterized by a medial degeneration with disruption of the elastic lamina. Giant cells infiltration may be present.¹ The most commonly found lesion is stenosis (53%), followed by occlusion (21%) and aneurysm (10%).³ This patient also had tubular stenosis

of descending aorta, right common iliac artery and right subclavian artery on her arteriogram.

TA presents therapeutic challenges especially in children and many clinicians face problems while controlling the blood pressure and the symptoms of the patients. For this patient, oral corticosteroids were used as the first-line therapy as usually done in adults with TA and she showed excellent response. In cases of resistance or serious side effects related to steroid treatment, other agents including oral or intravenous cyclophosphamide, methotrexate and mycophenolate and recently, anti-tumour necrosis factor agent infliximab have been proposed as a second line treatment.¹

The indications for surgery include hypertension with critical renal artery stenosis, extremity claudication limiting activities of daily living, cerebrovascular ischaemia or critical stenosis of three or more cerebral vessels, moderate aortic regurgitation, and cardiac ischaemia.⁷ This patient had no such indications yet and she is improving and maintaining her normal blood pressure on oral medications.

Takayasu arteritis is a rare entity in children. Reporting such cases will help improve the knowledge of paediatricians and rheumatologists about TA in childhood, allowing early diagnosis and better therapeutic results.

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