

Takayasu's Arteritis: Rare Cause of Hypertension in Children

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Abstract

Takayasu arteritis (TA) is a large vessel vasculitis that involves the aorta, its major branches and pulmonary arteries. Diagnosis of TA during childhood remains challenging due to the non-specific symptoms. We report a six years age boy with unresolved hypertension who was later diagnosed as childhood TA. Oral corticosteroid was started 2mg/kg/day. TA is rare in children; Childhood TA must be considered in children who present with, hypertension and increased acute phase reactants.

Key words: Children, Takayasu's arteritis, Steroids

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Introduction

Takayasu arteritis (TA) is a large vessel vasculitis that involves the aorta, its major branches and pulmonary arteries. Diagnosis of TA during childhood remains challenging due to the non-specific symptoms. The estimated worldwide incidence is 2.6 cases per million per year. The disease has been mainly studied in Japan but Western studies have also been published¹.

The first manifestation of Takayasu's arteritis was recognized between the age of 11 and 48 years. In 66 of the 84 patients (78.5%), it was between the ages of 11 and 29 years². In a retrospective review of 107 charts of patients with TA studied during a 19 year period up to May, 1974, most patients' ages ranged from 11 to 30 years (80 per cent). The youngest was four years old¹. A recent review article reported a total of 241 TA cases published in the paediatric age group³

The Case

A six years age boy came in with persistently high blood pressure since four months. He also has history of swelling of whole body which gradually started from eyes, face to whole body. No history of haematuria, loss of consciousness, sore throat, skin lesions. Patient was diagnosed as Post Streptococcal Glomerulonephritis and was admitted in other hospital. Hypertension Grade-II and discharged on Amlodipine 5mg/day Enalapril 0.08mg/kg/day. However medication was discontinued since the past four days and BP was still persistently high so was planned for renal biopsy and was referred to our hospital. On examination patient was well built, afebrile, BP: 130/90mm of Hg (>99th percentile) which was persistently high in successive days and hard to record in lower limbs. Other systems were essentially normal. Investigations were as follows.

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Day 1 of admission	Erythrocyte Sedimentation Rate was 52 (high), platelets 3,60,000/cumm. Anti Streptolysin "O" titre was positive.
Day 2 of Admission	ANA, AntiDsDNA, PT, APTT were with in Normal Limit
Day 3 of admission ECHO	moderate LV systolic dysfunction with mild MR dilated LA, LV
Day 4 of admission CT Angiogram	long segment circumferential smooth wall thickening of the thoracic and abdominal aortic wall with progressive narrowing of its lumen. Similar wall thickening of the SMA, bilateral main renal artery with marked stenosis of left renal artery. Thrombus of right common iliac artery with reformation with collaterals just before its bifurcation. Smaller left Kidney. S/O Takayasu 's Arteritis

Our patient was started on Prednisolone 2mg/kg/day. Hypertension was treated with antihypertensive agents, since aggressive therapy was necessary to prevent complications. Patient also received Amlodipine 5mg/day Enalapril 0.08mg/kg/day.

Discussion

Takayasu's disease is a chronic inflammatory disease of large and medium sized arteries, involving the aorta and its main branches, the pulmonary arteries, and the coronary tree. Since the original report of Takayasu's disease in 1908⁴, the estimated worldwide incidence is 2.6 cases per million per year, with women more commonly affected than men. Peak onset is in individuals in their 30s. The disease has been mainly studied in Japan but Western studies have also been published⁵. Cardiac features are present in up to 40% of cases. One case reported in 6years old with cardiac feature⁶. The aetiology and the precise pathogenesis of Takayasu's arteritis are still unknown but much has been learnt about the disease since its initial description by M. Takayasu, a Japanese ophthalmologist in 1908⁷. This disease is the commonest cause of reno-vascular hypertension in Asian children⁸. Arterial hypertension is the most common finding on physical examination of children which was also present in our case (82.6%), and often suggests the diagnosis but we could not find exact prevalence data. The tests of inflammatory activity are elevated in most cases⁹ which is similarly

to our results. In our case, this finding and their clinical features supported the request for imaging studies and helped to establish our patients' diagnosis.

This is diagnosed according to the criteria adopted by the American College of Rheumatology are shown in Table 1⁹.

For purposes of classification, a patient shall be said to have Takayasu's arteritis if at least three of these six criteria are present. The presence of any three or more criteria yields a sensitivity of 90.5% and a specificity of 97.8%

Treatment: The two most important goals of treatment are controlling the inflammatory process and controlling the hypertension. Corticosteroids are the most important therapeutic agents. Therapy is continued until patients achieve remission. Corticosteroids are still the mainstay of treatment¹⁰. Glucocorticoids are an effective agent for most patients with active TA. Remission has been achieved in 60% of patients treated with glucocorticoids¹¹. Our patient was also started on steroid (prednisolone) 2mg/kg/day. Antiplatelet agents and heparin may prove useful in preventing stroke. Due to the rarity of the disease, we could not find controlled studies of medical treatment of children with TA.

Anti-inflammatory therapy can lead to dramatic improvement in TA. The mortality rate in children, though, is as high as 35%¹². The outcome depends on the

Table 1: Criteria of American College of Rheumatology for the Classification of Takayasu's Arteritis

Criteria	Definition
Age at disease onset in year <40 years.	Development of symptoms or findings related to Takayasu's arteritis at age <40 years.
Claudication of extremities	Development and worsening of fatigue and discomfort in muscles of one or more extremity while in use, especially the upper extremities.
Decreased brachial artery pulse	Decreased pulsation of one or both brachial arteries.
Blood pressure difference >10 mmHg	Difference of >10 mmHg in systolic blood pressure between arms.
Bruit over subclavian arteries or aorta	Bruit audible on auscultation over one or both subclavian arteries or abdominal aorta.
Arteriogram abnormality	Arteriographic narrowing or occlusion of the entire aorta, its primary branches, or large arteries in the proximal upper or lower extremities, not due to arteriosclerosis, fibro-muscular dysplasia, or similar causes: changes usually focal or segmental.

vessel involvement and on the severity of hypertension. Hypertension is treated with antihypertensive agents, and aggressive therapy is necessary to prevent complications. It was difficult to control hypertension in this our patient, he received Amlodipine 5mg/day and Enalapril 0.08mg/kg/day at discharge. In a Turkish survey, one patient with pulmonary artery stenosis died within the first 3 years and two patients underwent nephrectomy¹³. In South Africa, the mortality rate was 22.5%; 7 of 31 patients died because of hypertension or complications after kidney transplantation. One patient died after EBV-associated haemophagocytic syndrome¹⁴. Prevention of organ damage may avoid worse outcome.

Prognosis: Takayasu's arteritis in children is a serious illness and a mortality of 10-30% has been reported on follow up. In his prospective studies on the natural history of Takayasu's arteritis, Ishikawa estimated a 5-years survival of 83% in all patients with Takayasu's

arteritis and a 10-year survival of 58% in patients with severe disease. There are few follow-up studies in children, and the mortality rate has ranged from 21% to 40% in the short term.¹⁵ More recently, the prognosis has significantly improved due to interventional procedures for the treatment of renal and aortic stenosis. We could not find out the Long term follow up data on children is not available.

Conclusion

We conclude that TA is a rare disease in a paediatric setting, and also that it has to be considered in cases of unexplained hypertension or unexplained inflammatory syndromes without signs of localization. A thorough physical examination can lead to the correct diagnosis if pulses cannot be felt, even if these are not constant findings. Since the disease can be progressive and life-threatening, an early recognition is vital in order to start immunosuppression

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