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Original Research Article

Paediatric Vasculitis: A Comprehensive Case Study of Takayasu's Arteritis with PRES Syndrome

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Abstract:

Background: Takayasu's arteritis is a chronic idiopathic vasculitis of medium and large vessels with a chronic continuous or waxing-waning course. The aorta and its primary branches are most involved vessel. Systemic inflammation and distal ischemia give rise to the predominant clinical features.

Clinical Description: A 11-year-old male presented with abnormal body movements, abdominal pain, vomiting and headache. Examination revealed pulselessness of upper limb but blood pressure was persistently raised with difference in upper and lower limb BP over the course of illness, the child also suffered from PRES (posterior reversible encephalopathy syndrome). MRI brain reveals mild hyperintensity of bilateral posterior parietal lobes. Colour Doppler findings of upper limb revealed bilateral subclavian arteries narrowing and wall thickening. Computed tomography angiography suggested aortoarteritis. Correlation of clinical history, physical examination, investigation reports were helpful to reach the final diagnosis as Takayasu arteritis with PRES syndrome.

Management: The patient was treated with antiepileptics, antihypertensives, steroids and other supportive measures were taken. The patient showed improvement but B.P was persistently high

Conclusion: Takayasu arteritis as a differential diagnosis of hypertensive encephalopathy should be kept.

Keywords: Takayasu's arteritis, PRES, Pulselessness, Doppler.

Introduction

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. The pathologic course of the disease begins with pan arteritis in the adventitia which progresses to the intima, eventually causing vascular narrowing, occlusion, and later in disease, aneurismal formation [1].

The disease is more prevalent in females, with a ratio of around 2.5:1 in the paediatric population [2]. Onset of the disease in children commonly occurs at around 12 years of age, although there have been documented cases of Takayasu arteritis presenting in infancy [3].

The described incidence of the disease ranges from 1-2 per million populations per year. We report a case of Takayasu arteritis with PRES syndrome in a 11-year-old male child. The purpose of this report is to create awareness among clinicians so that they consider Takayasu arteritis as a differential diagnosis of hypertensive encephalopathy.

Clinical Description:

A 11-year-old male child born out of non-consanguineous marriage presented with abnormal body movements, abdominal pain, vomiting, and headache for 2 days. On enquiry, there was history of low-grade fever, intermittent in nature, maximum recorded temperature of 38.5 degree Celsius, without diurnal variation, not associated with nigh-sweats. There was no history of cough, weight loss, or any other constitutional symptoms. There was no history of local trauma. No history of similar complaint in the past or in the family was reported.

On physical examination child weighted 27 kg (10th -25th centile), height 148 cm (75th-90th centile), BMI (<3SD) and heart rate 104/min, with absent pulsation on radial side of both upper limbs there was a 4-limb discrepancy of blood pressure (BP) along with inequality in the pulses felt at different arterial markings. The blood pressure in upper limb was 140/90 mm of Hg and 170/105mm of Hg in lower limb was noted. Electrocardiography showed sinus rhythm (SR), normal QRS duration, no ST

segment or T wave abnormality. Routine blood test was normal with low M.C. V and M.C.H. indicative of microcytic hypochromic with normocytic normochromic blood picture with a positive CRP quantitative value (32-53 mg/dl), B/L fundal examination was also done which showed B/L choroiditis (posterior uveitis). Chest Xray showed aneurysmal dilation (fig: 1). CSF routine and EEG was normal subsequently MRI brain was done which reveal mild hyperintensity within sulcal spaces of bilateral posterior parietal lobes on FLAIR sequence (fig: 2). Subsequently patient BP was persistently raised above 140/90 mm of Hg for which Colour doppler was advised, which revealed findings of bilateral subclavian arteries lumen narrowing and wall thickening with reduced patchy colour flow due to proximal bilateral subclavian arteries narrowing in upper limb. Colour Doppler findings of lower limb were normal. USG abdomen was also done which suggest smaller left kidney. CT angiography was suggestive of Aortoarteritis with stenosis of 1st & 2nd part of left subclavian artery distal to origin of left vertebral artery (fig:3) with intimal wall thickening along with right subclavian artery narrowing. Luminal narrowing of celiac trunk, abdominal aorta at L1 vertebra and left renal artery was also observed.

Management and Outcome

The child was started on anti-epileptics to control seizure, for raised Blood pressure anti-hypertensive was given. The anti-emetics, NSAIDS, steroids and supplements (calcium, iron and multivitamin) were given for symptomatic management. Subsequently the complaint of child was alleviated. Child was discharged with anti-epileptics and anti-hypertensives and kept on follow up. Further advised to follow up with paediatric nephrology for raised blood pressure.

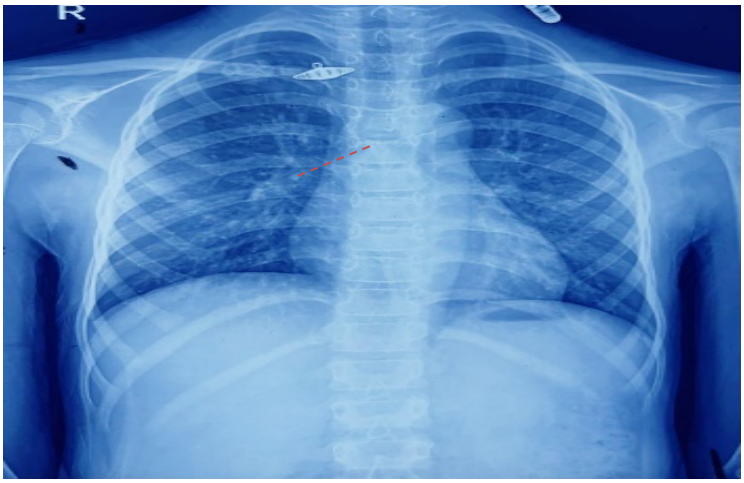

Figure 1: Xray revealed aneurysmal dilatation

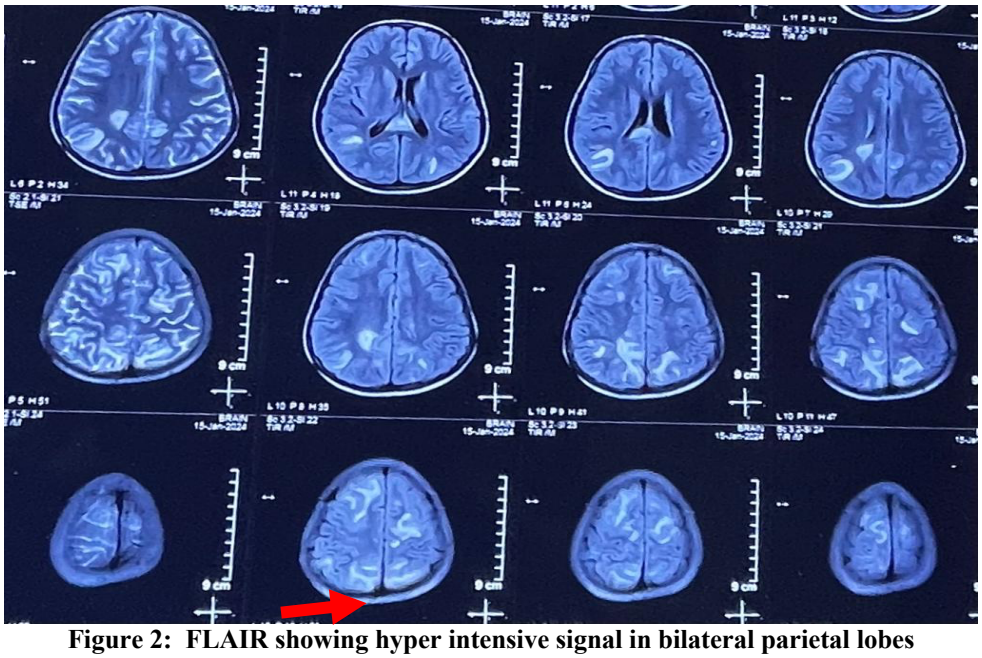

Figure 2: FLAIR showing hyper intensive signal in bilateral parietal lobes

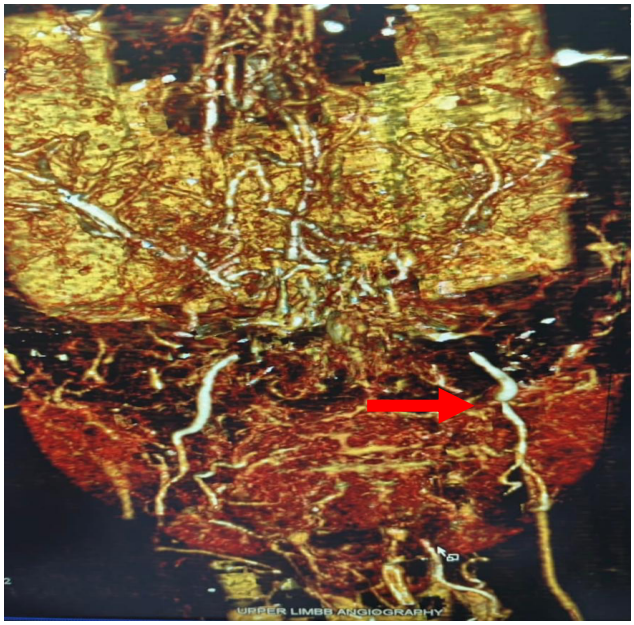

Figure 3: CT -angiography showed stenosis of left subclavian artery

Discussion

Takayasu arteritis also known as pulseless disease is a chronic large vessel vasculitis of unknown etiology that involves the aorta and its major branches. It typically presents between ages of 10 and 20yrs [4].

The average age of diagnosis in children is usually at 13 years of age. A predominantly affects female with a 2.5:1 female/male ratio in children. The etiology remains unknown, presence of abundant T cell with a restricted repertoire of T cell receptor points to cellular immunity and suggests an unknown aortic tissue antigen. Expression of IL-1, IL-6 and TNF is reported to be higher in patient of TA, inflammation of the vessel wall starting from the vasa vasorum is seen. Persistent inflammation damages the elastic lamina and muscular media leading to blood vessel dilation and aneurysm formation ultimately leading to stenotic or occluded vessels [5]. The subclavian, renal and carotid arteries are most commonly involved branches of aorta. The clinical manifestations of Takayasu's disease are commonly divided into early pre-pulseless and late pulseless phases. During the early phase, nonspecific systemic symptoms and signs predominate. The later phase is characterized by ischemia and symptoms secondary to arterial occlusion. The clinical spectrum at presentation of children with TA differs from that of adults. Children frequently have hypertension, headaches, fever, and weight loss at diagnosis of TA. Supradiaphragmatic (aortic arch) diseases presents with CNS manifestation (stroke, TIA) or CVS (heart failure, palpitations). Infradiaphragmatic (mid-aortic syndrome) presents with hypertension, abdominal bruits and pain.

Proposed Classification Criteria for Paediatric-Onset Takayasu Arteritis
Angiographic abnormalities (conventional, CT, or magnetic resonance angiography) of the aorta or its main branches and at least 1 of the following criteria:
• Decreased peripheral artery pulses) and/or claudication of extremities
• Blood pressure difference between arms or legs of >10 mm Hg
• Bruits over the aorta and/or its major branches
• Hypertension (defined by childhood normative data)
• Elevated acute-phase reactant (erythrocyte sedimentation rate or C-reactive protein)

Conclusion and Recommendation

1) A high index of suspicion and detailed diagnostic work up is required for early diagnosis of Takayasu arteritis in children presented with high blood pressure and pulselessness.

There was a 4-limb discrepancy of blood pressure along with inequality in the pulses felt at different arterial markings

2) Takayasu arteritis can present as renovascular hypertension.

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