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BEYOND PULSELESSNESS- A CASE OF TAKAYASU ARTERITIS REVEALED BY ACUTE HEART FAILURE AND HYPERTENSION CRISIS

Paediatrics

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ABSTRACT

Takayasu arteritis or pulseless disease is the most common type of granulomatous large vessel vasculitis in adolescent females, associated with chronic, idiopathic, inflammation of large vessels predominantly aorta and its major branches, with resultant significant morbidity and mortality. Here we report the case of a 12.5 years old adolescent girl with Numano classification type III Takayasu arteritis who presented with acute onset of heart failure associated with stage 2 hypertension and an incidental finding of right contracted kidney. Paucity of specific symptoms and lab biomarkers in assessing disease activity and progression, makes the disease often unrecognized at its onset, and its activity is frequently underestimated.

KEYWORDS

Takayasu Arteritis, Vasculitis, Pediatric Rheumatology, Heart Failure, Hypertension

INTRODUCTION

Takayasu arteritis (TA) is a type of granulomatous large vessel systemic vasculitis, characterized by chronic, autoimmune inflammation of aorta and its major branches at their origin. The resultant vascular stenosis, occlusion, aneurysm or rupture can cause tissue ischemia and injury. TA is the 3rd most common vasculitis in children, preceded by IgA/ Henoch-Schönlein purpura (HSP) vasculitis and Kawasaki disease [1] and is recognized as the most common cause of renovascular hypertension in Asian children [2].

Childhood or Pediatric onset TA (c-TA/ p-TA), a distinct subset of Takayasu arteritis encompasses a wide range of age group from early infancy to adolescence. The female:male preponderance is around 2:1, and it is recognized worldwide with ethnic differences in the pattern of involvement of the aorta.

The insidious course of the disease and diagnostic lag leads to irreversible vascular damage and pulseless phase, hindering successful therapy. Higher rates of mortality, about 35-40% by five years is attributable to the progressive course of disease leading to end organ damage, congestive cardiac failure or renal failure, and secondary to drug related complications and immunosuppression [3].

Case Report

A 12.5 years old girl resident of rural Maharashtra, India 1st born to non-consanguineous parents, with an unremarkable family history was brought with complaints of low grade intermittent fever and headache for 1 month, palpitations and progressive dyspnoea for last 15 days. Child had no significant past history or any prior hospital admissions. On presentation patient had stage 2 hypertension, tachycardia, tachypnea and a decrescendo early diastolic murmur. In the acute stage of presentation, a water hammer pulse was appreciated as well. 2D-ECHO revealed global Left ventricular hypokinesia with Left Ventricular Ejection Fraction (LVEF) of 20%, with preserved myocardial thickness and end systolic, end diastolic ventricular dimensions. Incidentally a quadricuspid aortic valve with mild Aortic Regurgitation, mild Tricuspid Regurgitation without Pulmonary Arterial Hypertension was also detected. With suspicion of acute myocarditis, child was started on diuretics and ionotropic support. Further work up for persistent hypertension revealed a right contracted kidney with maintained cortico-medullary differentiation and preserved renal function. A confirmatory Computed Tomography with Angiography concluded the presence of aorto-arteritis of descending thoracic and abdominal aorta with right renal artery stenosis. Inflammatory markers, i.e. C-Reactive Protein and Erythrocyte Sedimentation Rate were elevated as well, and Creatine Kinase-Myocardial Band was moderately elevated. Thus, satisfying the mandatory as well as 2 of the 5 signs and symptoms of EULAR/ PRINTO/ PRES (European League Against Rheumatism / Pediatric Rheumatology International Trials Organization / Pediatric Rheumatology European Society) classification criteria for c-TA. Child was hence started on steroids and Methotrexate after Rheumatologist consult.

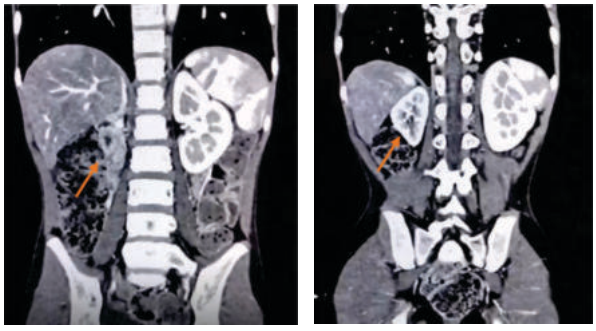

Fig.1 & 2: CT- Abdomen and Pelvis Shows Contracted Right Kidney (Red Arrow).

DISCUSSION

Takayasu arteritis is an inflammatory disease of large vessels commonly occurring in females in their 2nd or 3rd decade with an average age of presentation being 11.4 years, with 20% of cases diagnosed before age 19 and 2% before age 10 [4]. C-TA is a subset of TA and represents 1.5% of vasculitis in childhood. It may present as early as infancy and upto adolescent age group, with significant differences in clinical presentation compared to the adult counterpart. Ladhani et al. reported a case of c-TA in 2001 in a 2 years old child [5]. Pathogenesis of c-TA may start early in life, but often remains unrecognized due to non-specific symptoms and lack of definite biomarkers required for diagnosis. A high index of suspicion and a low threshold for diagnostic evaluation is required in doubtful cases especially those with unexplained arterial hypertension, for early diagnosis and intervention to prevent morbid complications.

Our patient had presented with an acute history of NYHA (New York Heart Association) grade 3 cardiac failure with no preceding risk factors. On examination child was found to have stage 2 hypertension with urgency. Considering renal system to be the most common cause for arterial hypertension in pediatrics, led to the discovery of right contracted kidney, subsequent work up of which revealed renal artery stenosis and aorto-arteritis of thoracic and descending abdominal aorta. Congestive heart failure in TA can be ascribed to either hypertension, myocarditis or aortic regurgitation. Talwar et al. reported myocarditis by endo-myocardial biopsies in 8 out of 11 TA patients with active disease [6]. Probability of myocarditis being

responsible for heart failure in our patient was higher owing to the acute presentation of symptoms and elevated levels of acute phase reactants. After ruling out any significant structural cardiac anomaly that could be attributed to heart failure, a diagnosis of Takayasu arteritis associated with myocarditis was made.

Factors that limited early clinical diagnosis of TA were absence of certain hallmark features like abdominal bruit, absence of peripheral pulses, blood pressure discrepancy of >10mmHg in the arms and claudication. In a study of clinical profile of a cohort of 40 c-TA patients in a tertiary care centre in India conducted by Goel et al., a similar presenting feature of hypertension (73%), headache (53%) and fever (45%) was found. Unlike our case, absent peripheral pulses (62.5%) was also a common sign. The median age of presentation in the case series was 12.5 years (range 1-16 years) and median diagnostic delay was 11.3 months (range 1-60 months) [3].

C-TA diagnosis requires a two-pronged approach, which includes measuring the acute phase reactants to monitor disease activity and confirmatory gold standard imaging to see the extent of vessel involvement. Colour-coded Doppler Sonography can accurately identify homogenous circumferential intima-media thickening of large vessels in TA, however it remains subjective [7]. Magnetic Resonance- Angiography on the other side, has a higher sensitivity and can visualize early stage of smooth muscle thickened vessel walls in TA as well as show signs of active inflammation such as mural edema on T2 weighted imaging [8]. In our patient, CT- angiography of abdominal vessels accurately correlated with the disease presentation hence, further full body MR-Angiography was planned for follow-up assessment of disease activity after a 12 weeks course of steroids and immunosuppressants or if any incidence of clinical worsening. Various novel biomarkers proposed to correlate with disease activity in TA that have been recently discovered include- MMP-2, 3 & 9; IL-6; RANTES; VCAM; PTX-3 [9-11].

With regards to treatment, a combination therapy of Prednisolone 20mg with fortnight tapering and Methotrexate 10mg per week was started with Folinic acid supplementation. Carvedilol, Furosemide, Aspirin and Nifedipine was continued for heart failure and hypertension. Other immunosuppressants commonly used with steroid is MMF (Mycophenolate Mofetil) with documented improved outcomes on follow up [3]. Ozen et al. in their series of 6 children with c-TA successfully and safely used Cyclophosphamide and steroid for induction followed by MTX for maintenance [12]. In another case series, an upgrade of immunosuppressant from MTX to MMF was required for relapse of disease [1]. However, due to limitations in clinical trial, superiority of a particular regime of immunosuppressants over another is not yet proved. Clinical trials with biologics such as Rituximab, Tocilizumab and Infliximab are in the pipeline for long term remission [13]. PTA with stenting and surgical bypass is reserved for cases with severe vaso-occlusion and end organ damage. An interesting new possibility of use of Sildenafil as a vasodilatory and antiproliferative agent in Takayasu arteritis has emerged after a case of 8 years old girl with TA showed dramatic improvement in symptoms of claudication and absent peripheral pulses after the use of Sildenafil [14].

CONCLUSION

Takayasu arteritis is often encountered in pediatric rheumatology setting, however non-specific nature of symptoms frequently diverts the diagnosis to commoner differentials. Keeping an eye out for covert features like absent peripheral pulses and abdominal bruit, and a thorough physical examination in cases of unexplained arterial hypertension and unexplained inflammatory syndromes, can improve diagnostic accuracy manifolds. An early recognition of disease is vital in order to start timely immunosuppression to prevent irreversible vascular damage and related morbidity and mortality.

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