ORIGINAL RESEARCH PAPER
Volume - 14 | Issue - 11 | November - 2025 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

AN UNVEILING CHILDHOOD TAKAYASU ARTERITIS: A RARE PRESENTATION WITH ISOLATED CHEST PAIN

Rheumatology

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ABSTRACT

Takayasu arteritis (TA) is a rare, chronic, large-vessel vasculitis affecting the aorta and its major branches, typically seen in young females. Childhood-onset Takayasu arteritis (cTAK) poses significant diagnostic challenges due to its heterogeneous presentation and nonspecific systemic symptoms. We report the case of a 14-year-old girl who presented with isolated retrosternal chest pain of one-month duration, unrelieved by empirical therapy. Clinical examination revealed absent upper limb pulses with non-recordable blood pressure, while lower limb pressures were normal. CT aortogram demonstrated diffuse mural thickening involving the aortic arch and its major branches, with occlusion of the right subclavian and vertebral arteries, consistent with type V Takayasu arteritis in the active stage. The patient was treated with high-dose corticosteroids and intravenous tocilizumab (8 mg/kg monthly), resulting in rapid resolution of symptoms and normalization of inflammatory markers. This case highlights the diagnostic importance of a high index of suspicion in adolescents presenting with atypical chest pain. Early vascular imaging and prompt initiation of immunosuppressive therapy, including biologics, are crucial in preventing disease progression and achieving sustained remission. Recognition of such atypical presentations can improve outcomes in pediatric large-vessel vasculitis.

KEYWORDS

Childhood Takayasu arteritis, chest pain, large vessel vasculitis, tocilizumab, pulse deficit

INTRODUCTION

Takayasu arteritis (TA) is a rare, chronic, relapsing granulomatous large vessel vasculitis affecting aorta, its branches, and pulmonary artery commonly observed in young females. Herein, we presenting a young female presented with isolated chest pain.

CASE STUDY

A 14-year-old, fully vaccinated, well-nourished female without comorbidities, presented with one-month retrosternal chest pain at rest, radiating to the neck not associated with dyspnoea, tachypnoea, sweating & palpitation. No diurnal variation and not aggravation by food intake. Multiple hospital visits and H2 blocker therapy failed to alleviate the symptoms.

Physical examination revealed absent pulse in both upper limbs hence patient referred to further evaluation. Detailed examination showed absent brachial, radial pulses and non-recordable blood pressure in arms. Lower limb blood pressure in right and left leg were 130/90 and 120/80, respectively. She had bilateral carotid and subclavian artery bruit. No renal bruit, carotidynia noticed. Cardiovascular examination showed early diastolic murmur at aortic area.

Laboratory investigations showed Hb- 9.6g/dL, WBC- 18600/mm^3 with neutrophilic predominance, raised CRP, ESR and mildly elevated liver enzymes. ECG- sinus tachycardia. Trop T card test was negative. Echocardiography showed mild aortic regurgitation with normal LV systolic function.

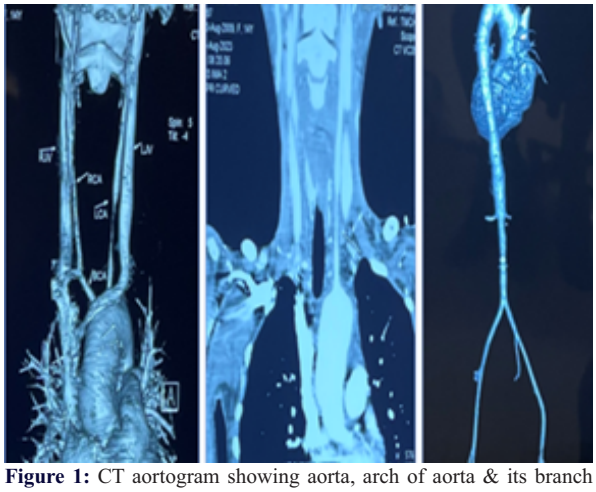

Figure 1: CT aortogram showing aorta, arch of aorta & its branch involvement and Abdominal aorta involvement

CT aorta angiogram showed diffuse mural thickening involving branches of arch of aorta causing significant luminal narrowing. Concentric mural thickening noted in suprarenal, infrarenal abdominal aorta, proximal part of SMA and right renal artery. Concentric mural thickening at coeliac artery causing 40-50% luminal occlusion. Left subclavian artery showed 80-90% luminal narrowing and complete occlusion of right vertebral & subclavian artery. All features are suggestive of Takayasu arteritis type V in the active stage (ITAS 2010 - 7).

The patient was initiated on high-dose prednisolone and IV tocilizumab 8mg/kg, resulting in the resolution of chest pain. She was maintained on monthly tocilizumab infusions, with prednisolone tapered to a low dose. Follow-up assessments showed sustained clinical improvement and disease control.

DISCUSSION:

Diagnosing Takayasu arteritis in children is challenging because of its heterogenous presentation depends upon the vessel involvement and nonspecific systemic symptoms. [1] It can be even more challenging when child presenting with isolated chest pain. Childhood chest pain mostly due to musculoskeletal causes (1-89%) and cardiac causes in 3-7% cases.[2] Aeschlimann et al study reported cardiac diseases in cTAK 3-18% from varies cohort.[1] Zhu et al reported chest pain in 21% of children (n = 3).[3]

Chest pain causes reported in cTAK includes cardiomyopathy, heart failure, valvular disease, and ischemic heart disease, and rarely coronary artery involvement (11%). Danda et al reported type 5 disease is more common in cTAK up to 56.5% compared to aTAK (47.5%).[4] Presenting symptoms not always correlate with type of TAK hence detailed workup warranted in all TAK. Treatment of childhood TA reflects adult protocol as there is no controlled therapeutic trials in children.[5]

CONCLUSIONS

This case highlights TA's diverse clinical presentations and underscores the importance of maintaining a high index of suspicion, particularly in adolescents presenting with chest pain. Early diagnosis, appropriate immunosuppressive therapy prevents disease progression and favourable outcome.

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