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A Rare Presentation of Dilated Cardiomyopathy Along with Takayasu Arteritis in a Preschool Child

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ABSTRACT

Dilated cardiomyopathy is the most common form of cardiomyopathy in children, is the cause of significant morbidity and mortality. Although the most common etiology of DCM remains idiopathic, it is estimated that upto 50% of cases are genetic. The pathogenesis of ventricular dilation and altered contractility seen in DCM varies depending on the underlying etiology; systolic dysfunction and myocyte injury are common.

Takayasu arteritis also known as “pulseless disease”, is a chronic large vessel vasculitis of unknown etiology and predominantly involves the aorta and its major branches. This disease is most common in Asians and mostly diagnosed in adolescent age group, on average at the age of 13 years. TA is characterized by inflammation of the vessel wall starting from vasa vasorum. Persistent inflammation leads to progressive scarring and intimal proliferation and can result in stenotic or occluded vessels leading to systemic manifestations.

We have reported a rare case of dilated cardiomyopathy along with Takayasu arteritis in a preschool child. The causality and association between both the conditions has not been fully understood yet. However DCM has been reported in only 5-6% cases of TA.

KEYWORDS: Takayasu arteritis, Cardiomyopathy, Cardiac failure, Vasculitis, Genetic, Systolic dysfunction

ARTICLE DETAILS

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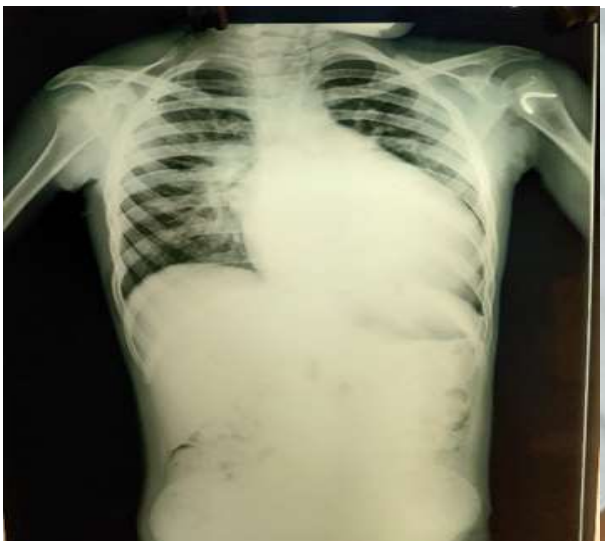
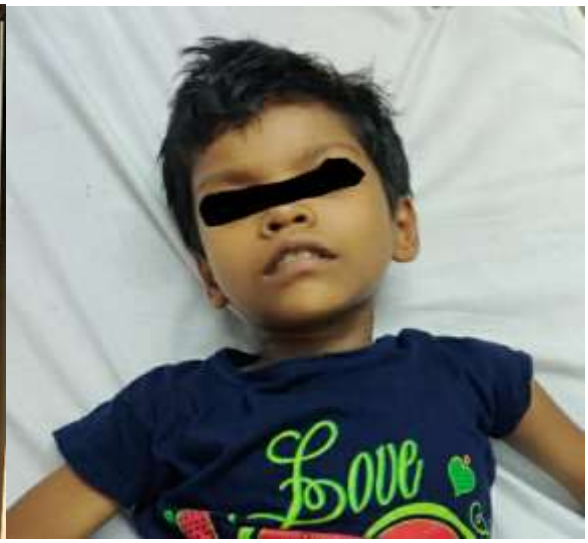


Picture.1 showing cardiomyopathy (C:T ratio =0.7)



Picture.2 showing the patient at 12 years of age in 2023

Table-1:

RADIOLOGICAL INVESTIGATIONS	FINDINGS 2016	2019	2023
1.2D ECHO	Global hypokinesia, Cardiomyopathy, Mild MR,**EF=33%** Mod LV dysfunction Grade-3 diastolic dysfunction	Dilated cardiomyopathy **EF=36%,FS=17%** Mod MR Mild TR Mod LV dysfunction	Dilated cardiomyopathy Severe MR, **EF=20%,FS=10%** Severe PH/TR Severe LV dysfunction Grade-3 diastolic dysfunction
2.CT AORTOGRAPHY		Multiple focal areas of narrowing of aorta at T4 -T8 Vetb level T12-L1, From T8 to Origin of celiac trunk, From origin of SMA to lt renal artery TAKAYASU ARTERITIS(Type-3)	
3.USG DOPPLER OF B/L LL & RT UL		Biphasic wave form in rt ul and b/l ll arteries Low resistance flow in rt ul and ll arteries.	
4.NCCT BRAIN		Left caudate and gangliocapsular lacunar infarct	

INTRODUCTION

Dilated cardiomyopathy (DCM) is one of the cardiomyopathies, a group of diseases that primarily affect the myocardium (the muscle of the heart). [1] Different cardiomyopathies have different causes and affect the heart in different ways.[1] The heart becomes weakened and enlarged and cannot pump blood efficiently affecting different organ systems. [1] About 50% of the cases are genetic (usually autosomal dominant) including some metabolic causes [3] with most mutations affecting genes encoding cytoskeletal proteins, [2] while some affect other proteins involved in contraction. [2]

Takayasu Arteritis (TA), also known as pulseless disease, is a chronic large vessel vasculitis of unknown etiology that predominantly involves the aorta and its major branches. The disease is most common in Asians. Age of onset is typically between 10 and 40 years. Most children are diagnosed as adolescents, on average at the age of 13 years. Younger children may be affected but diagnosis in infancy is rare. TA preferentially affects females with a reported 2-4:1 female/male ratio in children and adolescents and a 9:1 ratio in adults [3]. TA causes massive intimal fibrosis and narrowing of arteries[1].

DCM however rarely reported to be seen in only 5-6% of cases of TA[4]. But the exact correlation regarding the causality and association has not been fully understood yet. The presentation as DCM is rarely reported and is due to involvement of coronary artery, severe hypertension, and cardiac failure are bad prognostic factors. [1],[2]

CASE REPORT

4-year-old female child born out of non consanguineous marriage with normal perinatal and developmental history presented with bipedal edema, palpitation and shortness of breath in 2016. Clinical examination was suggestive of congestive cardiac failure [HR =130/min, RR=48/min, low volume pulses, tender hepatomegaly and anasarca]. Patient was managed in ICU and CCF resolved. Chest x ray was s/o cardiomegaly (C:T ratio=0.7)**[PIC-1].** 2DECHO s/o- DCM with global hypokinesia of LV with ejection fraction of 33.2%. Any association with muscular dystrophy was ruled out. Patient had a negative family history as well. She was then discharged with beta blockers and diuretics.

This patient again presented at the age of 7 years in 2019 with sudden onset right sided hemiparesis. NCCT Brain showed Left caudate head and gangliocapsular infarct. Blood investigations revealed CBC-Leukocytosis, elevated CRP, normal c3 & c4, ANA-negative, sickling – negative, normal LFT, RFT and electrolytes. Coagulopathy was ruled out. On detailed examination of peripheral pulses only left Radial artery pulsations were palpable. All other peripheral pulses in Right upper limb and bilateral lower limbs were absent. The BP was more than 95^{th} +12^{th} percentile for age and sex of the child. There was a difference in the systolic BP of the upper and lower limbs. Thus a possibility of Takayasu arteritis was considered .The diagnosis was confirmed by CT angiography of aorta and its main branches which was s/o TYPE-3 Takayasu Arteritis. To substantiate our findings USG doppler was done. It was s/o biphasic waveform and low resistance flow in right upper limb and bilateral lower limb arteries.

Repeat 2D-ECHO was s/o DCM with global hypokinesia with an ejection fraction of 36%. A short course of steroids was given in order to decrease the underlying vascular inflammation. Patient was discharged with steroid, beta blocker and diuretic therapy.

This patient now came for follow up to pediatric OPD at the age of 12 years.**[PICTURE-2]** Repeat 2D-ECHO was s/o DCM with EF-20% with severe MR, severe PH/TR and severe LV dysfunction. Patient was discharged with Beta blocker, PDE5 inhibitor and diuretic and advised for follow up after 1 month.**Table-2** summerised all the relevant radiological investigations.

DISCUSSION

Takayasu Arteritis is the most common cause of renovascular hypertension in India. Indian origin aortoarteritis is a chronic granulomatous, necrotizing vasculitis predominantly affecting aorta and its branches. [5]. Recently, more emphasis has been given on an immunopathological cause. [5],[6]

The disease is classified based on the site of involvement: [7]

- Type I: Aortic arch involvement
- Type II: Thoracoabdominal involvement
- Type III: Diffuse involvement
- Type IV: Pulmonary involvement
- Type V: Aneurysmal type.

The initial presentation of our patient was congestive cardiac failure secondary to DCM at the age of 4 years. Gradually the patient developed pulselessness, hypertension and acute ischemic stroke secondary to TA. CT angiogram was s/o-Type 3 TA with diffuse involvement of the aorta and its branches. Serial 2DECHO s/o declining ejection fraction due to progression of the disease and cardiac remodeling .The association between DCM and TA has not been completely understood yet. DCM however rarely reported to be seen in only 5-6% of cases of TA[4]. A few cases have been reported in adults. But the occurrence of these two conditions in a preschool child is one of a kind and needs to be reported.

Medical management is the mainstay of treatment and surgical intervention should be considered whenever possible.

CONCLUSION

Our case highlights that there might be a causal association between DCM and TA. Every case of DCM without obvious genetic association and family history should be examined properly for the absence of peripheral pulses and difference in systolic BP between upper and lower limbs. Detailed clinical examination is the key to early diagnosis and management of such cases so as to prevent morbidity and mortality due to the disease process.

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