CASE REPORT

SEVERE POSTDUCTAL COARCTATION OF AORTA PRESENTING LIKE IDIOPATHIC DILATED CARDIOMYOPATHY

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ABSTRACT

Left ventricular systolic dysfunction in childhood can be due to many causes. It may be because of sequel of myocarditis, metabolic causes, storage disorder, congenital heart disease or idiopathic dilated cardiomyopathy. Severe left ventricular outflow obstruction can cause reversible severe left ventricular systolic dysfunction. Here we report a case of severe post-ductal coarctation of aorta presenting like dilated cardiomyopathy.

KEY WORDS: Dilated cardiomyopathy; Reversible Left ventricular systolic dysfunction; Coarctation of aorta.

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INTRODUCTION

Left ventricular systolic dysfunction in childhood can occur due to many causes. It may be because of sequel of myocarditis, metabolic causes, storage disorders, congenital heart disease or idiopathic dilated cardiomyopathy (DCM). Severe left ventricular outflow obstruction at the level of left ventricular outflow tract (LVOT) or aorta can cause reversible severe left ventricular systolic dysfunction.

Here we report a case of severe post-ductal coarctation of aorta presenting like dilated cardiomyopathy.

CASE REPORT

A two-years old male child presented to our institution with history of progressive breathlessness and failure to thrive for last one and half year. He was diagnosed with dilated cardiomyopathy following echocardiography evaluation done from outside and was put on medical therapy since six months of his age. As his symptoms did not improve he presented to our institute for re-evaluation. On examination pulse rate was 102 per minute with feeble bilateral femoral artery pulsation. Blood pressure in both upper limbs was 110/68 mm Hg and in lower limb was 90/66 mm Hg. Apex beat was in fifth left intercostal space just inside midclavicular line, forceful in nature. There was left ventricular S_3. Jugular venous pressure was raised with tender hepatomegaly. Chest X-ray showed cardiomegaly and on ECG there was biventricular hypertrophy.

Repeat echocardiographic evaluation showed dilated left atrium and ventricle with left ventricular hypertrophy. (Fig. 1)

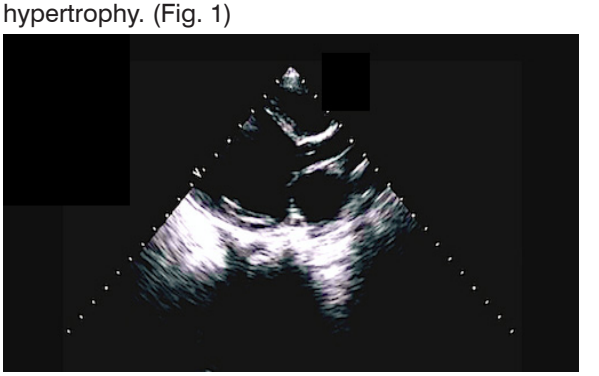

Figure 1: Transthoracic echocardiography parasternal long axis view showing the dilated Left Ventricular cavity in diastole.

There was global left ventricular hypokinesia with severe systolic dysfunction (Ejection Fraction 20%). Suprasternal view showed severe aortic narrowing just after the origin of left subclavian with peak gradient of 80 mmHg (Fig. 2).

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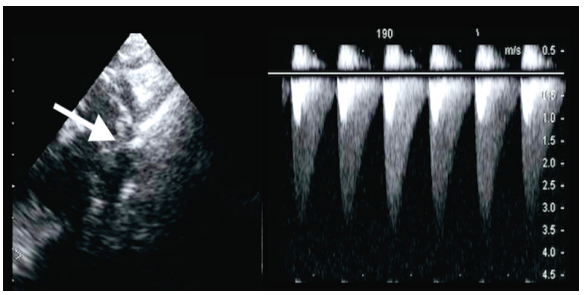

Figure 2: Transthoracic echocardiography suprasternal view showing coarctation of aorta distal to right subclavian artery. Doppler interrogation at coarcted segment shows peak gradient of 4 m/s.

A diagnosis of severe post ductal coarctation of aorta was made and Balloon aortoplasty was planned for the patient.

DISCUSSION

Coarctation of aorta can cause severe left ventricular dysfunction[1] and it can be missed in small child if meticulous echocardiographic evaluation is not done.[2,3] Presence of feeble femoral artery pulsation with left ventricular dysfunction and hypertrophy should raise the suspicion of coarctation of aorta. Other important cause of dilated cardiomyopathy like presentation in young child is anomalous origin left coronary artery from pulmonary artery. Presence of angina, severe mitral regurgitation and continuous flow in pulmonary artery in echo helps in arriving at a diagnosis. In ECG presence of deep q wave is also helpful. Aorto-arteritis is also an important cause of dilated cardiomyopathy like presentation in infancy. Absence of pulse in upper limb(s) helps usually to spot the diagnosis. Storage disorder like Gaucher's disease can present like DCM but usually associated with hepatosplenomegaly.

The management consists of relieving of the coarctation with or without stenting. Agac et al reported a case of dilated cardiomyopathy secondary to coarctation who was successfully managed with balloon expandable stent resulting in normalization of cardiac function over 6 months.[4] Raffel et al reports a 37-year-old man presenting with severe dilated cardiomyopathy secondary to occult aortic coarctation who was managed successfully with combined orthotopic heart transplantation and aortic coarctation repair.[5] A case of severe reversible dilated cardiomyopathy associated with a large left ventricular thrombus in a young child possibly due to non-specific aortitis who successfully underwent stent angioplasty with no residual coarctation, with normal upper extremity blood pressures and improved LV size and function was reported by Ponniah et al.[6] Our case highlights the importance of excluding the potential reversible cause like coarctation, critical aortic stenosis, and aortitis before diagnosing Dilated Cardiomyopathy.[7]

CONCLUSION

Detailed physical examination and echocardiographic evaluation, particularly the thoracic aorta is must while evaluating a child with dilated left ventricle with severe systolic dysfunction because missing coarctation in such cases might have deadly consequences.

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CONFLICT OF INTEREST
Authors declare no conflict of interest.
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AUTHORS' CONTRIBUTION

Conception and Design:	SG,
Data collection, analysis & interpretation:	SG, BM, VT
Manuscript writing:	SG, PS, PCB