ISSN: 0975-8585

Research Journal of Pharmaceutical, Biological and Chemical Sciences

Left Ventricular Non – Compaction and Ventricular Septal Defect – A Rare Association.

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ABSTRACT

We report a case of 11 month old male child who presented to emergency department with generalised tonic clonic movements. Cardiac examination showed a systolic murmur in left parasternal area. Echocardiogram revealed a 2 mm small perimembranous ventricular septal defect along with LV non - compaction. LV systolic function was normal. He was managed with anti-epileptics and is on regular follow up. This case demonstrates the rare association of ventricular septal defect with LV non compaction.

Keywords: Left ventricular non-compaction, Ventricular septal defect

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ISSN: 0975-8585

INTRODUCTION

Left ventricular non compaction (LVNC) is an under-recognized cardiac abnormality characterized by prominent ventricular trabeculations and deep intertrabecular recesses usually associated with impaired LV systolic function. The true prevalence is unclear however the estimated prevalence is around 0.014% in patients undergoing echocardiography. Symptoms of LVNC may vary from heart failure, arrhythmias to thromboembolic manifestations. Progression may vary according to the amount of non-compaction, however regression of non - compaction has been rarely reported [1].

Here we report a case of 11 month old male child who presented to us with seizure disorder however was found to have LV non-compaction with associated ventricular septal defect.

CASE REPORT

A 11 month old male child presented to Paediatric ED with 2 episodes of generalized tonic –clonic movements. Born out of a non- consanguineous marriage there was no significant family history. General physical examination was normal. There were no neurological deficits. Cardiovascular examination revealed a grade 3//6 systolic murmur in left parasternal area. Laboratory investigations were normal. Chest X-ray was normal. Computed tomography of brain was essentially normal. Electroencephalography demonstrated no epileptic waveforms. In view of audible murmur he was referred to cardiology department for further evaluation. Electrocardiogram was normal. Echocardiography revealed a 2 mm small peri-membranous ventricular septal defect with left to right shunt and multiple trabeculations with inter-trabecular recess findings suggestive of left ventricular non compaction (LVNC)(Figure 1 & 2) . Colour Doppler showed flow within the inter – trabecular recesses consistent with LVNC(Figure 3). Left ventricular systolic function was normal. He was treated conservatively with anti – epileptics. As the child was asymptomatic for LV non-compaction, he was advised close follow up.

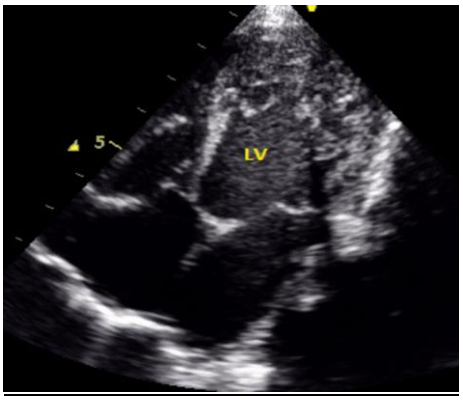

Figure 1: Two-Dimensional Echocardiogram In Apical Four Chamber View Showing Left Ventricle And Prominent Trabeculations With Sinuses Consistent With Noncompacted Myocardium Of The Lateral Wall.

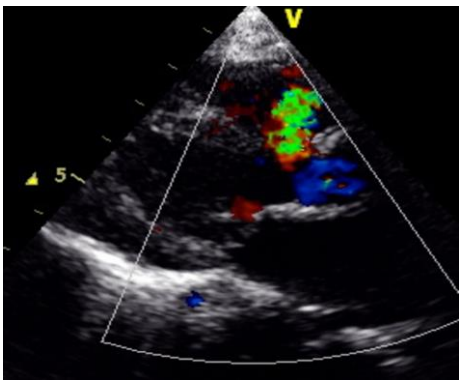

Figure 2: Echocardiogram In Parasternal Long Axis View Showing Perimembranous VSD With Colour Flow Demonstrating Left To Right Shunt.

ISSN: 0975-8585

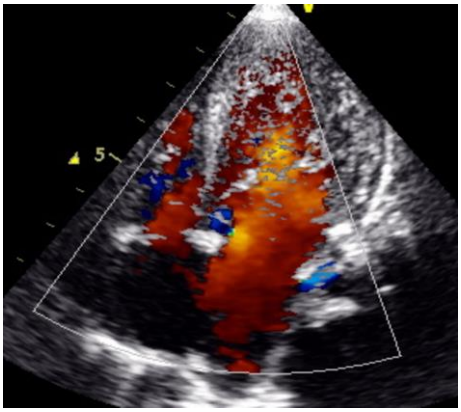

Figure 3: Two-Dimensional Echocardiogram In Apical Four Chamber View Showing Colour Doppler With Flow Within The Inter-Trabecular Recesses Consistent With LV Non-Compaction.

DISCUSSION

LVNC is a rare congenital cardiomyopathy classified under unclassified cardiomyopathies as per European society of cardiology [2]. It is characterized by numerous prominent trabeculations and deep inter-trabecular recesses in hypertrophied and hypokinetic segments of the left ventricle along with systolic dysfunction. It was thought due to defective morphogenesis of endo-myocardium resulting in prominent trabeculations and deep inter-trabecular recesses that communicate with LV cavity [3,4]. LVNC occurs in isolation or associated with congenital cardiac anomalies like Ebsteins anomaly, cc-TGA, hypo plastic left heart (HLH) syndrome, atrial septal defect, patent ductus arteriosus and ventricular septal defect [5]. The true prevalence of the disease is unclear; however the prevalence was 0.014% of patients referred to the echocardiography laboratory [6]. LVNC can manifest with heart failure, arrhythmias and thromboembolic manifestations. Echocardiography remains gold standard for the diagnosis of LVNC. The other modalities used in the diagnosis of LVNC are contrast ventriculography, computed tomography of heart and cardiac magnetic resonance imaging. The diagnostic criteria includes (1) a thickened LV wall consisting of two layers: a thin compacted epicardial layer; and a markedly thickened endocardial layer with numerous prominent trabeculations and deep recesses with a maximum ratio of non-compacted to compacted myocardium >2 : 1 at end-systole in the parasternal short-axis view, (2) Colour Doppler evidence of flow within the deep inter-trabecular recesses and (3) prominent trabecular meshwork in the LV apex or mid-ventricular segments of the inferior and lateral wall. Treatment of LVNC should be directed at monitoring and management of LV dysfunction, prevention of arrhythmias and thromboembolic manifestations. In view of high incidence sudden death reported in some series these people are advised intra-cardiac defibrillator to prevent sudden cardiac death(6). As our patient was asymptomatic he was advised follow up .The prognosis of LVNC varies with the type and the amount of non-compaction .

ACKNOWLEDGEMENT

We would like to show our gratitude to the Department of Cardiology and Team of Echo cardiographers for their immense help and support.

REFERENCES

[1] Eurlings LW, Pinto YM, Dennert RM, Bekkers SC. Int J Cardiol 2009;136(2):e35-6.
[2] Elliott P, Andersson B, Arbustini E, Bilinska Z, Cecchi F, Charron P, Dubourg O, Kühl U, Maisch B, McKenna WJ, Monserrat L, Pankuweit S, Rapezzi C, Seferovic P, Tavazzi L, Keren A. Eur Heart J 2008 ;29(2):270-6.
[3] Hussein A, Karimianpour A, Collier P, Krasuski RA. J Am Coll Cardiol 2015;66(5):578-85.
[4] Weiford BC, Subbarao VD, Mulhern KM. Circulation 2004;109(24):2965-71. Review.
[5] Tsai SF, Ebenroth ES, Hurwitz RA, Cordes TM, Schamberger MS, Batra AS. Pediatr Cardiol 2009;30(5):597-602.
[6] Oechslin EN, Attenhofer Jost CH, Rojas JR, Kaufmann PA, Jenni R. J Am Coll Cardiol 2000;36(2):493-500.