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Case Series

Noncompaction of left ventricle - a rare cardiomyopathy

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ABSTRACT

Non-compaction of left ventricular (NCLV) is a rare cardiomyopathy of unknown origin characterized by prominent ventricular trabeculations and deep intertrabecular sinusoids, in communication with the left ventricular cavity. The NCLV results due to failure of compaction during foetal development. In this study, five children were diagnosed with NCLV out of 135 cases of cardiomyopathy and the youngest case was diagnosed at 27th day of life. Congestive cardiac failure presenting as tachycardia and tachypnea were common clinical manifestations. Early detection of NCLV helps patients to get timely treatment. NCLV should be considered as one of the differential diagnosis amongst cases of cardiomyopathy.

Keywords: Non-compaction of left ventricle, Cardiomyopathy, Congestive heart failure

INTRODUCTION

Non-compaction of left ventricular (NCLV) is a rare congenital cardiomyopathy of unknown etiology, occurs due to the failure of left ventricle compaction during embryogenesis. In a few cases, various genetic inheritance patterns have been reported with 30% cases of genetic origin may be due to mutations in the MYH7 and MYBPC3 genes.[1] Some cases are associated with neuromuscular disorders. It may be detected as an isolated case or may be associated with congenital heart diseases or syndromes.

NCLV is a structural cardiac muscle disorder which is characterized by prominent ventricular trabeculations and deep intertrabecular recesses in communication with the left ventricular cavity. It is having two layers at apical and left lateral portion of ventricle with an outer compact thinner epicardial layer and an inner spongy endocardial layer. Even though the reported incidence is 8-12/million/year, the condition is likely to be more common due to occurrence of asymptomatic cases and applying different diagnostic criteria.[1,2]

It may present at any age from infancy to adulthood. Clinical manifestations may present as congestive heart failure (CCF), arrhythmia as supraventricular tachycardia, embolization, or occasionally sudden cardiac death.[3,4] The objective of this study is to report case series of rare cardiomyopathy in children so that timely diagnosis and treatment can be offered.

CASE SERIES

A case studied retrospectively and was done at K. G. P. children hospital from 2003 to 2020 on children aged 0-18 years. Two-dimensional color Doppler echocardiography and standard M-mode measurements were performed for all the suspected cardiomyopathy patients. NCLV was diagnosed based on two criteria: more than three trabeculations protruding from the ventricular wall, apically to the papillary muscles and visible in a single image plane; intertrabecular spaces perfused from the ventricular cavity and visualized on color Doppler imaging.[5] Patients with associated congenital heart diseases were excluded.

During this study period, five cases were newly diagnosed as NCLV from 135 cases of cardiomyopathy of which 4 were males and one female. Age of presentation varied from 27th day of life to 7 years with mean age was 2½ years. The Common clinical presentations were tachycardia (5/5), tachypnea (5/5), dyspnea on exertion (1/5), CCF (4/5), oedema of feet (2/5), arrhythmia as supraventricular tachycardia (1/5). All five patients fulfilled the above diagnostic criteria

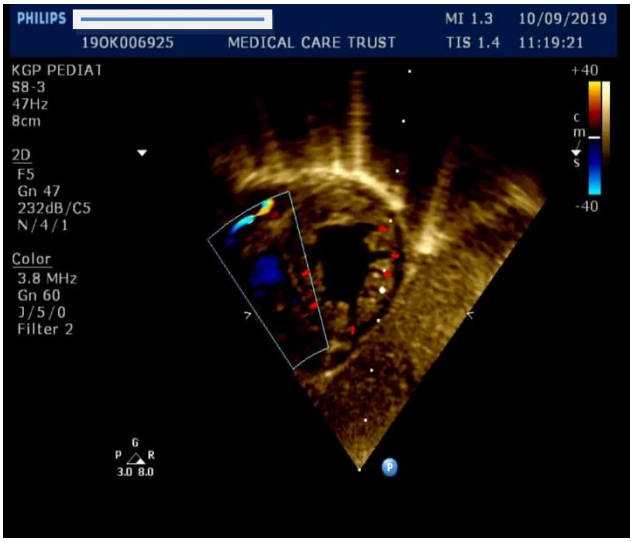

Figure 1: Still image of echocardiography reveals LV wall with multiple sinusoids accompanied by increased thickness both in end-systole and end-diastole.

DISCUSSION

NCLV has a sporadic occurrence in 60–70% of the cases, and familial occurrence in 30% with autosomal dominant is predominant pattern of inheritance. There were multiple genes identified for development of noncompaction.[5,6] As per WHO coding system, it is classified under unclassified type of cardiomyopathy. In our study, incidence of NCLV type cardiomyopathy was 4% while a study done by Andrews et al reported 9% of NCLV from 104 children with a primary cardiomyopathy.[6] Normally, ventricular myocardial trabeculations are evident in the heart by the end of the first trimester, it further develops into proper compaction of the trabecular layers in the embryo by 12 weeks to 16 weeks of gestation, when almost full compaction of myocardium occurs. Without the completion of compaction, there is myocardial dysfunction and failure of the spiral pattern of myocardial fibres which leads to defective contractile ventricular function.[7] Various nomenclatures like foetal myocardium/honeycomb myocardium/hyper trabeculation syndrome/spongy myocardium have been given to this condition. Noncompaction can affect both ventricles separately or together but left ventricle is affected more commonly.[6,7]

Speckle tracking and real time 3-dimensional echocardiography can be performed for a better visualization of structure and function of the heart.[8] This condition should be differentiated from dilated cardiomyopathy, hypertrophic cardiomyopathy and endocardial fibroelastosis. There is no specific therapy. Standard treatment of CCF may be added with beta-blockers and anticoagulation drugs. Implantation of Internal Cardiac Defibrillator may be considered for primary or secondary prevention of sudden cardiac death. Heart Transplantation is the ultimate cure.[9,10]

CONCLUSION

NCLV is a type of cardiomyopathy having poor prognosis without treatment. Early detection helps to improve the quality of life and increase the life expectancy till second to third decades.

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