

Biventricular Non-Compaction Cardiomyopathy

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Abstract

Left ventricular non-compaction (spongy myocardium) is one of the most misclassified cardiomyopathies.

It is characterised by an excessively prominent trabecular meshwork of myocardium and deep intertrabecular myocardium due to an arrest in the compaction process of the myocardial fibres. It could be isolated i.e. without any other structural heart defects or associated with congenital heart defects.

The clinical manifestations are variable heart failure, arrhythmia, thromboembolic phenomena depending on extent of non-compaction of cardiac segment.

Case Report

Y, 20 yrs unmarried female, presented to us with complaints of shortness of breath, palpitations and chest pain of 15 days duration. Her shortness of breath was gradually progressive from NYHA class II to NYHA IV. It was associated with pricking type of chest pain which was non-exertional, non-radiating present intermittently and would get relieved on its own.

She had similar episodes for the last 2 years.

Her family history was uneventful.

On examination, she had pulse of 88 beats/min, regular. Peripheral pulses were well felt. JVP was not raised and blood pressure was 150/80 mm Hg.

Systemic examination she had a short systolic murmur in mitral area, grade 3 with S3 gallop.

Respiratory system: Nothing contributory.

Laboratory investigations were all within normal limits.

Chest X-Ray: Cardiomegaly and lung parenchyma was clear.

ECG: Q waves in lead I, aVL, V₅, V₆.

2D-Echo : Revealed a large LA:LV with moderately depressed LV function, both systolic and diastolic with LV apex obliterated.

Cardiac MRI: LVEF 35.8% with moderate hypokinesia of all segments with significant non-compaction and end diastolic apical thickness of 13mm. Non compaction of Right ventricle was seen as well.

She was started with anti-coagulation

therapy - oral warfarin therapy, ACE inhibitors and lasilactone.

She improved symptomatically.

All family members were screened for similar picture. However, all members were found to be normal.

Discussion

Myocardial non-compaction is a rare disorder of uncertain etiology.¹

WHO has classified isolated non-compaction of left ventricle (IVNC) as an unclassified type of cardiomyopathy.² Normally between foetal life of 5th week and 8th week inter-trabecular spaces are obliterated and ventricular compaction occurs from the base towards the apex and from epicardium to endocardium. When there is an arrest in this progression results in non-compaction of myocardium.³ Failure of left ventricular compaction process is possibly caused by mutations in the genes encoding α -dystrobrevin and Cypher/ZASP which are integral parts of the complex which links the extracellular matrix of myocardial cell to the cytoskeleton.⁴ This mutation is common in patients with dilated cardiomyopathy and may explain the development of left ventricular dilatation and systolic impairment in patients with IVNC.⁴

Left ventricular non-compaction (LVNC) was first described by Chin et al,¹ has a male preponderance (56% -82%). Female preponderance has been reported in Arabs /Africans.¹

Clinical manifestations are highly variable, which may range from no symptoms to disabling heart failure (seen in 67% cases)⁵ arrhythmias : ventricular tachyarrhythmias (47% adults),⁵ atrial fibrillation (25% adults), left bundle branch block, PSVT.

Diagnosis is made by echocardiography. Jenni et al defined 4 criteria for diagnosis of IVNC⁶ :

1. Absence of co-existing cardiac abnormalities.
2. Left ventricular wall thickness with end systolic ratio of non-compacted to compacted layer of >2 is diagnostic.
3. Findings mostly in apical and mid-ventricular region.
4. Blood flow seen directly from the ventricular cavity into the deep intertrabecular recess as seen by doppler echocardiography.

TTE is the gold standard for diagnosis. Other modalities used are MRI, contrast ventriculography and CT scan.

The treatment of LVNC is mainly management of clinical manifestations. Long term prophylaxis with anti-coagulants is recommended especially in high risk patients with severe non-compaction, embolic phenomena and ejection fraction of <25%.^{1,5} In high risk patients with refractory heart failure, severely depressed left ventricular function, prolonged intraventricular conduction, cardiac transplantation, implantable defibrillator or biventricular pacemakers have been tried⁵.

A study done by Toyono et al for the effects of carvedilol on left ventricular function, showed it may have a beneficial effect on LV function, hypertrophy, metabolism and adrenergic abnormalities.

The largest series of isolated non compaction of left ventricle in adults was recently described by Oeschlin et al. As many as 47-60% of adults die or undergo cardiac transplantation within

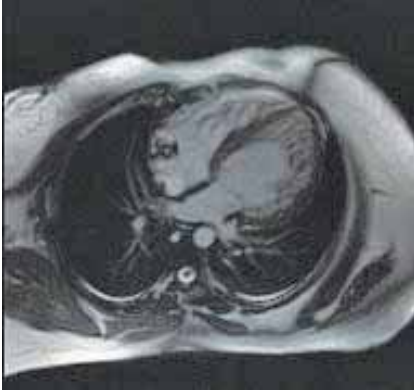


Fig. 1: Cardiac MRI showing non-compaction of both ventricles

4-6 years⁵ and most develop ventricular dysfunction over 10 years.

Due to high morbidity and mortality associated with non-compaction of myocardium the need of the hour now is to create awareness among physicians and cardiologists.

LVNC is rare cardiomyopathy and RV involvement is reported in less the half of cases of LVNC. We report a rare case of Biventricular Non-compaction Cardiomyopathy.

References

1. Chin TK, Perloff JK, Williams RG, Jue K, Mohrman R. Circulation. Isolated noncompaction of left ventricular myocardium. *A Study of Eight Cases* 1990; 82:507-13.
2. Richardson P, McKenna W, Bristow M, et al. Report of the 1995 World Health Organisation/ International society and Federation of cardiology task force on the definition and classification of cardiomyopathies. *Circulation* 1996; 93:841-2.
3. Elshershari H, Okutan V, Celiker A. Isolated non compaction of ventricular myocardium. *Cardiol Young* 2001; 11:472-5.
4. Vatta M, Mohapatra B, Jimenez S, Sanchez X, Faulkner G et al. Mutations in Cypher/ZASP in patients with dilated cardiomyopathy and left ventricular noncompaction. *J Am Coll Cardiol* 2003; 42:2014-27.
5. Oeschlin EN, Attenhofer Jost CH, Rojas JR, Kaufmann PA, Jenni R. Longterm follow up study of 34 adults with isolated left ventricular noncompaction: A distinct cardiomyopathy with poor prognosis. *J Am Coll Cardiol* 2000; 36:493-500.
6. Jenni R, Oeschlin E, Schneider J, Attenhofer Jost C, Kaufmann PA. Echocardiographic and pathoanatomical characteristics of isolated left ventricular non-compaction; a step towards classification as a distinct cardiomyopathy. *Heart* 2001; 86:666-71.