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Cardiology

LEFT VENTRICULAR NON-COMPACTION CARDIOMYOPATHY IN THE REAL LIFE

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ABSTRACT Left ventricular non-compaction (LVNC) is a very rare congenital cardiomyopathy which is treated recently as an unclassified cardiomyopathy by the European Society of Cardiology while the American Heart Association includes it among genetic cardiomyopathies.

It is characterized by prominent endomyocardial trabeculations that increase in number and deep intertrabecular recesses.

In this article we will report the case of a 55 years old man patient diagnosed since 2014 with an isolated LVNC with ascites and edema of the lower limbs despite high doses of loop diuretcs illustrating the limited therapeutic options of this kind of cardiomyopathy.

KEYWORDS : Left ventricular non-compaction ; heart failure ; echocardiogram ; therapeutic management.

INTRODUCTION

The Left ventricular non-compaction (LVNC) is an unclassified cardiomyopathy characterized by an abnormally thick trabeculated non-compacted myocardial layer with adjacent deep intra-trabecular recesses and a thin compacted myocardial layer [1].

Although the precise cause is not presently known, it is thought to be due to a developmental disorder involving arrest of compaction of loose myocardial meshwork during fetal ontogenesis. The emerging evidence suggests that the excessive trabeculation may result from a disturbance in the compaction process during early myocardial development [2].

It has been known to be a part of various syndromes like the Barth, Noonan, Roifman or Toriello Carey syndrome.[3][4]

Patients with LVNC may lead a normal life as the normal left ventricle functions. Patients may present at any age from infancy to old age. The clinical manifestations may involve heart pump failure, arrhythmias, and thromboembolic phenomenon. The most common complaint at admission is pump failure like our patient.[5]

The Case

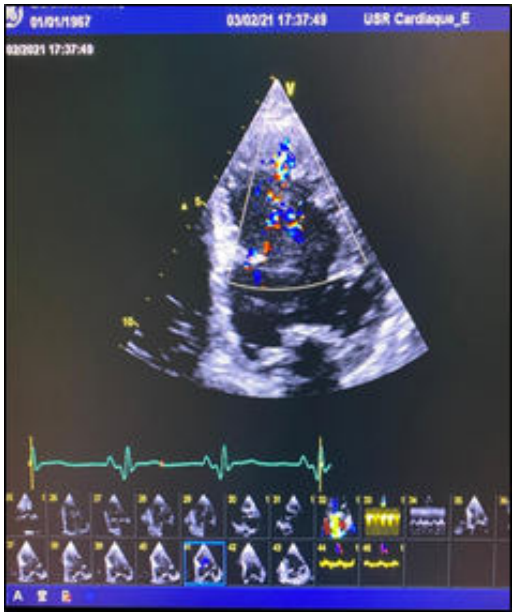
A 55 years old man followed for LV dysfunction due to LVNC diagnosed since 2014 was admitted for an impairment of his dyspnea, edema of lower limb and abdominal distencion in relation with an ascites despite of high doses of oral loop diuretics. At the clinical examination we noticed a global congestive heart failure hemodinamically stable and performed a symptomatic management : ascite and pleural punctures in addition to intraveinus loop diuretics were performed adapted to kidney function and electrolytic concentratoin.

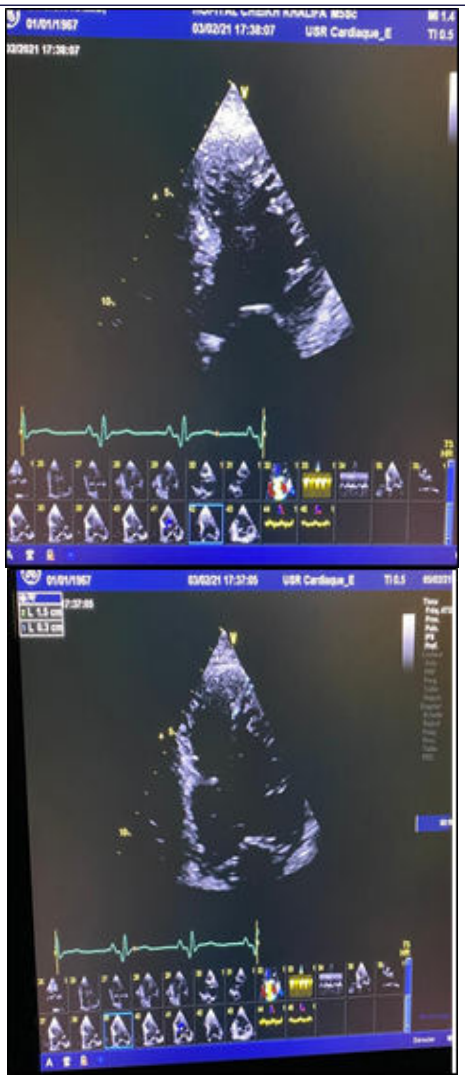
The control of his automatic implantable defibrillator found short episodes of non sustained ventricular tachycardia but recorded a long episode of 8 minutes of atrial fibrillation indicating long term oral anticoagulation.

An echocardiogram was performed to evaluate the current left and right ventricle functions. We found an hypokinetic dilated cardiomyopathy in severe left ventricle (LV) systolic dysfunction with LV ejection fraction LVEF at 15-20% secondary to a LVNC. Trabeculations were well seen, and we performed Chin method to confirm the diagnosis (Figures 1-2-3). There was no significant valve disease and the right ventricle's size and function were preserved.

According to new guidelines of heart failure, before discharge of the patient, we introduced salcubitril/valsartan therapy and SGLT2 antagonist with strict hygieno-dietetic rules.

A clinical improvement was noted and regular controls were decided to increase gradually doses of therapies.





Figures 1-2-3 Showing Four Chamber View With Aspect Of Left Ventricle Trabeculation

DISCUSSION

Our observation remain to challenging therapeutic in a rare diagnosed cardiomyopathy.

Due to its low cost and widespread availability, 2D-echo is usually the first investigation in the evaluation of LV hyper-trabeculation. Presently, there are four 2D-echo-based criteria that are commonly used, but none are considered as the gold standard (Table 1 [6]). The first criterion was developed by Chin et al and defines LVNC as an epicardial compacted myocardium layer (X) to endocardial non-compacted layer ratio (Y) ≤0.5 in the end-diastole [7].

Jenni and co-worker also proposed a method that involves detection of the two myocardial layers, non-compacted and compacted, in short axis views of the LV in end systole. This method describes the ratio >2 between non compacted to compacted myocardium. The third definition proposed by Stollberger et al. determines the number of prominent trabeculations visible in the apical views of the LV in diastole. Regarding diagnosis, there is a debate going on with regards to features displaced by angiography, echocardiography, computed tomography, and magnetic resonance imaging. One can use any of imaging for confirmation.[8]

Table 1. Echocardiographic-based Left Ventricular Non-Compaction (LVNC) Diagnostic Criteria [6].

	Chin	Jenni	Stollberger	Gebhard
Year	1990	2001	2002	2012
Total Patients; Patients with LVNC	8; 8	34; 34	62; 62	123; 41
Selection Criteria	Patients referred for echo and satisfied criteria	Patients referred for transthoracic echo	Patients referred for echo demonstrating >3 trabeculations distal to papillary muscle in the four-chamber view	Patients with LVNC by other echo criteria, severe aortic stenosis and matched controls
Age Range	11 months to 22.5 years	16 to 75 years	18 to 75 years	20 to 52 years
Description of Criteria	1. 2-layered structure with an epicardial compacted (C) and endocardial noncompacted (NC) layer ≤0.5	1. 2-layered structure 2. Noncompacted endocardial layer Colour Doppler evidence of inter-trabecular recesses supplied by intraventricular blood 3. NC/C ≥2 4. No coexisting cardiac abnormalities	1. 2-layered structure 2. >3 trabeculations protruding from LV wall apically to papillary muscle in 1 imaging plane 3. NC/C ≥2	1. 2-layered myocardium 2. Maximal systolic compacted thickness <8 mm
View	Parasternal Short-axis view	Parasternal Short-axis view	Non-standard views	Parasternal Short-axis view
Phase	End-systole	End-diastole	End-diastole	End-systole
Correlation with Clinical	No	No	No	No
Strengths	Widely available Cost-effective Short scanning time	Weaknesses	Participants had wide age range Results based on small cohorts Studies not prospectively derived Image quality dependant on body habitus Oversensitive in certain populations Non-specific in low-risk populations	

Management of symptomatic patients with congestive heart failure is with digoxin, diuretics, angiotensin-converting enzyme inhibitors, beta-blockers and afterload reducing agents. Some patients undergo cardiac transplantation. Cardiac rhythm abnormalities are managed with standard protocol, while some patients may benefit from an implanted cardiac defibrillator for severe ventricular tachyarrhythmias to prevent sudden death.[8]

Managing LVNC presents a significant clinical challenge given the variability in manifestations and the limited long-term efficacy of specific treatments.

In particular, those with reduced LV function should be reviewed frequently and treated with evidence-based, guideline-directed pharmacologic therapy. As per guidelines, an intracardiac defibrillator should be offered to those who survive an episode of sustained ventricular tachycardia (VT) or sudden cardiac arrest [9]. Successful cardiac transplantation has been reported in some patients and should be considered for those with end-stage heart failure [10].

Even though the event rate of stroke is 1–2% per year, the optimal medical strategy in those who do not meet the standard criteria for anticoagulation remains uncertain given the scarcity of data [11]. However, patients with a prior cardioembolic event, evidence of an intracardiac thrombus and/or documented atrial fibrillation should be treated with anticoagulation consistent with standard recommendations for cardiogenic embolism [12].

CONCLUSION

Isolated non-compaction of the left ventricle is a rare congenital cardiomyopathy characterized by abnormally thick trabeculated non-compacted myocardial generally located at the level of the apex of the left ventricle.

The echocardiography and magnetic resonance imaging make diagnosis which may be difficult in the atypical forms.

This cardiomyopathy can lead to cardiac failure, lethal ventricular arrhythmias, mural thrombi responsible of cerebrovascular stroke.

For those with impaired LV function treatement should be based on guideline-directed pharmacologic therapy.

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