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Original Research Paper

Cardiology

CARDIAC MAGNETIC RESONANCE FINDINGS IN LEFT VENTRICLE NON COMPACTION: A MONOCENTRIC STUDY

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ABSTRACT

BACKGROUND: The impact of LVNC in dilated cardiomyopathy and left ventricle systolic dysfunction is controversial .

METHODS: A retrospective study of 46 patients, over a period of 09 years between September 2009 and December 2018, performed in the cardiology department "B", IBN SINA hospital, Rabat, Morocco.

RESULTS: The mean age was 44 ± 16 years with extremes ranging from 10 years to 97 years. A slight predominance of male with 54% was observed. In TTE only 17,4% of patients had a suspected LVNC. The mean LVEDD was 68.6 ± 8.4 mm. The mean LVEDV was 223.1 ± 76.3 ml with a maximum of 443 ml and a minimum of 122 ml. The mean LVEF was 37.1 ± 12.7. Akinesia was observed in 28% of patients involving the infero-septal wall in 97% of cases. The non compaction was located in anterolateral wall was involved in 97.8 % of cases, and the inferoseptal wall was always spared. The median NC/C ratio was 2.8 [2.5-3], with extremes ranging from 2.3 to 5. LGE was present in 59 % of our patients; mostly located in the infero-septal. In multivariate analysis, NC/C ratio correlates significantly with the presence of LGE in the infero-lateral wall and the apex respectively (p: 0.04 and p: 0.002). NC / C ratio correlates with LVEF (OR: 0.03, CI [0.01-0.04], p: 0.02).

CONCLUSION: This study shows the CMR profile of LVNC Moroccan patients. Our results affirm that the NC/C ratio in LVNC is a major determinant of LV fibrosis and systolic dysfunction.

KEYWORDS : LVNC; CMR; NC/C ratio

INTRODUCTION

Left Ventricle Non Compaction (LVNC) is a rare form of cardiomyopathy, defined by the presence of prominent left ventricular (LV) trabeculae, deep intertrabecular recesses, and the thin compacted layer [1]. Classified by the American Heart Association as a genetic cardiomyopathy [2] and by the European Society of Cardiology (ESC) and the World Heart Organization (WHO) as an unclassified cardiomyopathy [3]. Two entities are described, the familial (inherited) form and the non familial (sporadic) form.

Although Cardiac Magnetic Resonance (CMR) remains the gold standard in the diagnosis of NCVG, few studies have described LVNC's CMR characteristics.

We expose in this work our experience in CMR diagnosis of the NCVG, and especially the correlation between non compaction and left ventricle systolic dysfunction.

MATERIAL AND METHODS

This is a retrospective study over a period of 09 years between September 2009 and December 2018, performed in the cardiology department "B", IBN SINA hospital, Rabat, Morocco. Data were collected of 46 patients CMR reports.

CMR was performed using a 1.5-Tesla magnetic resonance scanner (Siemens Medical Systems). Images were obtained in two chamber, four-chamber, and short-axis planes from base to apex left ventricule. After baseline imaging, a bolus injection of gadolinium was administered. Ten minutes later, delayed enhancement CMR was performed. The late gadolinium enhancement (LGE) images were acquired in end-systole in the same views used for cine images.

The presence of a two-layer structure : a thick non-compacted endocardial layer with excessive trabeculations and deep intertrabecular recesses, and a thin epicardial compacted layer was screened.

The NC/C ratio was measured in end of the diastole on short axis. A ratio of NC/C > 2.3 was suggestive of LVNC.

Segmental analysis was evaluated using a standard seventeen-segment cardiac model as defined by the American Heart Association/American College of Cardiology (AHA/ACC) for standardized myocardial segmentation.

Data on LV diameters, volumes, and ejection fraction (FE) were collected. LV systolic dysfunction was defined as a LVEF < 50%.

To determine the impact of LVNC in dilated cardiomyopathy and LV systolic dysfunction, correlations of the number of noncompacted LV segments and the NC/C ratio were analyzed.

LGE was classified according to its location in the myocardial wall: subendocardial, subepicardial, intramyocardial, or transmural.

Statistical analyzes:

All analyzes were performed using SPSS software (version 22.0). Quantitative variables of normal distribution are expressed as mean +/- standard deviation, quantitative variables of asymmetric

distribution in median and quartiles, and qualitative variables in numbers and percentages. The univariate and multivariate analyzes were performed using binary logistic regression. A"p" <0.05 was considered as statistically significant.

RESULTS

1. Clinical characteristics:

The mean age was 44 ± 16 years with extremes ranging from 10 years to 97 years. 25% of patients were under 30 years old. A slight predominance of male with 54% was observed. 95% of the population do not have a cardiovascular risk factor.

Sudden death occured in one or more first-degree relatives in 4.4% of cases.

The ECG was normal in 80% of cases, an atrial fibrillation and a left bundle branch block were observed each in 5% of cases.

After performing a thransthoracic echocardiography 50% of patients was sent to the CMR laboratory with a dilated cardiomyopathy, 17,4% only with a suspected LVNC.

2. CMR findings:

- ***Diamteres, volumes and EF:***

The mean Left Ventricular End Diastolic Diameter (LVEDD) was 68.6 ± 8.4 mm. The mean Left Ventricular End Systolic Diameter (LVESD) was 54.3 ± 9.4 mm. The mean Left Ventricular End Diastolic Volume (LVEDV) was 223.1 ± 76.3 ml with a maximum of 443 ml and a minimum of 122 ml. The mean Left Ventricular End Systolic Volume (LVESV) was 143.9 ± 68.1 ml.

The LV mass of VG was 145.1 ± 41.4 g.

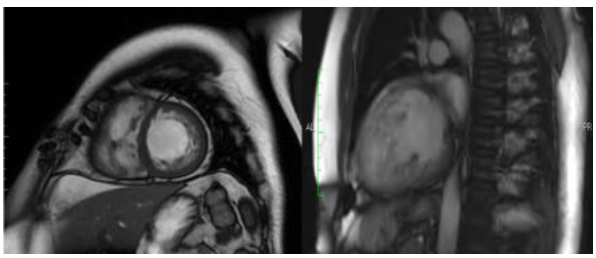
81% of patients had reduced EF, 64% with severe LV dysfunction ≤ 35%. The mean LV ejection fraction (LVEF) was 37.1 ± 12.7.

- ***LV wall motion abnormalities:***

Hypokinesia was observed in 63% of cases. Akinesia was observed in 28% of patients involving the infero-septal wall in 97% of cases, the anterolateral wall in 8.3% of cases and spared the infero-lateral wall.

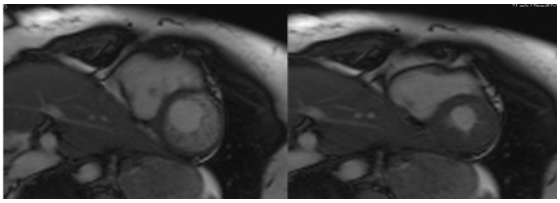
- ***Non compacted LV segments:***

The median of non compacted LV segments reached was 9[4-14], with a minimum of 2 segments and a maximum of 14. The anterolateral wall was involved in 97.8% of cases, and the inferoseptal wall was always spared. The median NC / C ratio was 2.8 [2.5-3], with extremes ranging from 2.3 to 5.



Short axis view of the LV base at diastol showing the non compaction of the anterior, lateral and inferior wall.

CMR two cavities plane showing the non compacted segments in the anterior and the inferior wall.



CMR short axis view showing trabeculation in the apical segments of the LV.

- ***Late Gadolinium Enhancement (LGE):***

LGE was present in 59 % of our patients; mostly located in the intramyocardial layer (73%). The infero-septal wall was involved in 66.7% of cases.

Distribution of non compacted segments and wall akinesia and LGE is described in Figure 1.

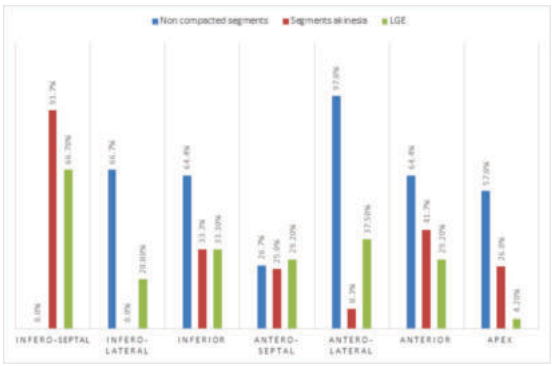
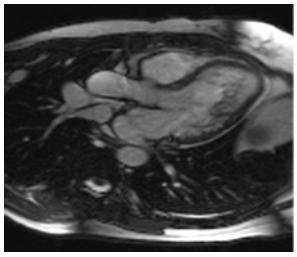
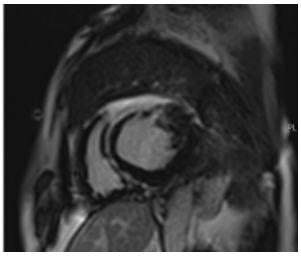


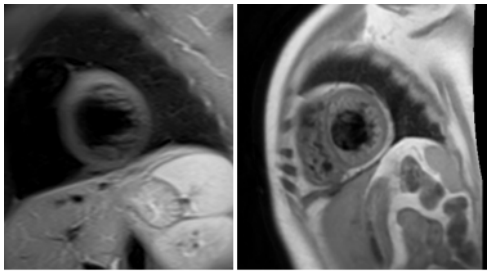
Figure 1 : Distribution of non compacted segments, wall akinesia and LGE.



CMR plane focusing in LV out tract showing the increase of wall thickness in subendocardial layer and excessive trabeculation in mid-anteroseptal and mid-inferolateral wall



Delayed enhancement images at mid short axis levels showing midwall enhancement predominantly at tne inferoseptal wall.



- ***Short axis view of a LVNC***

Dilated Cardiomyopathy (DCM) and LV dysfunction.

There wasn't any impact of the number of non compacted LV segments in the appearence of a DCM or a LV dysfunction (Table 1).

Table 1 :Non compacted LV segments correlation

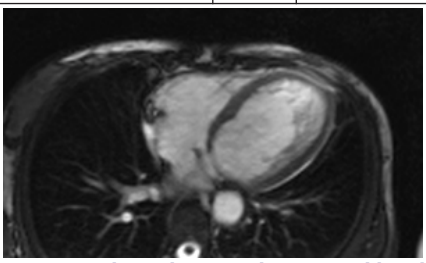
Characteristics	OR	CI	p
LVEDD	0,3	[-0,05-0,7]	0,07
LVEF	-0,07	[-0,9-4,3]	0,6
Presence of LGE	3,7	[-0,1-7,6]	0,06
LGE location	-0,6	[-2,1-0,8]	0,3

The NC/C ratio had significant impact in the apperaence of a DCM and LV dysfunction (Table 2).

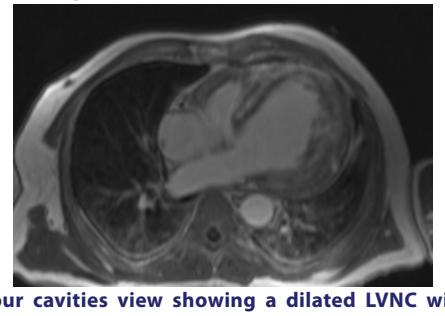
Table 2 : NC/C ratio correlation

Characteristics	OR	CI	p
LVEDD	-0,01	[-0,03-0,003]	0,09
LVEDV	0,005	[0-0,01]	0,06
Apical LGE	-1,7	[-2,6- -0,7]	0,002

Infero-lateral LGE	0,5	[0,03-1]	0,04
Infero-septal LGE	0,02	[-0,3- 0,3]	0,1
Infero-septal akinesia	2,1	[1,4-2,8]	<0,001
LVEF	0,03	[0,01-0,04]	0,002



CMR four cavities plane showing showing a dilated LV with severe non compaction



CMR four cavities view showing a dilated LVNC with late gadolinium enhancement reflecting fibrosis.

DISCUSSION

CMR has the best sensitivity in identifiying myocardial trabeculations and their extensions [4]. It provides a three dimensionnal approach of heart structure which allows to screen endocardial thrombi and associated congenital heart diseases.

CMR diagnostic criteria were defined by Peterson et al. [5]

- Visual appearance of two distinct myocardial layers : a compacted epicardial layer and a non compacted endocardial layer
- Presence of marked trabeculations and deep intertrabecular recesses within the noncompacted layer
- Noncompacted-to-compacted myocardial ratio greater than 2.3 as measured in end-diastole

A quantitative diagnostic criterion introduced by Jacquier et al. [6]

- Value of the myocardial mass of the non compacted myocardium (defined at end-diastole) greater than 20% of the global left ventricular mass.

And another Quantitative diagnostic criterion introduced by Grothoff et al. [7]

- Trabeculated ventricular mass greater than 25% of the global left ventricular mass; non compacted mass greater than 15 g/m2

In our study, all the patients had the typical aspect of LVNC : marked trabeculations and deep intertrabecular recesses within the non compacted layer ; Of the 17 segments analyzed, the median of affected LV segments was 9 (4,14), with a minimum of 2 segments and a maximum of 14 uncompacted segments.

In LVNC, non compacted segments are located in the apex and the lateral wall; septal segments are rarely involved [5,8]. Similar results were observed in our study, the anterolateral wall was involved in 97.8% of cases, followed by the inferolateral wall in 66.7% of cases and the apex in 57% of cases, the inferoseptal wall was spared in all our patients.

The normal myocardial compaction process moves from the base to the apex and from the septal wall to the lateral wall [9]. Peterson et al. showed that it's the inhibition of the normal embryological myocardial compaction process which can explain the typical pattern of non compacted LV segments distribution [5].

A particular advantage of CMR is to characterize distinctly, through late gadolinium enhancement analysis, the myocardial fibrosis [10,11].

In our study, LGE was present in 59%; mostly located in the intramyocardial layer. The infero-septal wall was involved in 66.7% of cases.

This proves that the fibrosis mainly affects the septal wall, and therefore also explains wall motion abonormalities. Akinesia was observed in 28% of our patients, and it mainly concerned the infero-septal wall (97% of cases), the anterolateral wall in 8.3% of cases and spared the infero-lateral wall.

We tried to understand the correlation between the number of unon compacted LV segments and the presence of fibrosis, in multivariate then univariate analysis, there is no correlation between these two variables respectively (p: 0.06 and 0.09) .

In multivariate analysis, we found that NC / C ratio correlates significantly with the presence of LGE and therefore fibrosis in the infero-lateral wall and the apex respectively (p: 0.04 and p: 0.002). NC / C ratio correlates with LVEF (OR: 0.03, CI [0.01-0.04], p: 0.02). Therefore, we could affirm that the higher the NC / C ratio, the most important the fibrosis and the LV systolic dysfunction.

Aras et al found that the number of non compacted LV segments is a major determinant of LV systolic dysfunction [12]. Lofiego et al also found a positive correlation between the number of non compacted LV segments and the LVEF [13].

In the opposite, Fazio et al have pointed out that there is no association between non compacted LV segments and LV dysfunction [14]. These results were confirmed by Gilbert Habib et al. [15].

This disvergence between the different studies could be explained by the fact that LV systolic dysfunction is multifactorial, due to the advanced stage of DCM and extended fibrosis.

Conclusion:

This study shows the CMR profile of LVNC Moroccan patients. Our results confirms that the NC/C ratio in LVNC is a major determinant of LV fibrosis and systolic dysfunction.

REFERENCES :

1. Jenni R, Oechslin EN, van der Loo B. Isolated ventricular non-compaction of the myocardium in adults. Heart 2007;93:11–5.
2. Maron BJ, Towbin JA, Thiene G, et al. Contemporary definitions and classification of the cardiomyopathies: an American Heart Association Scientific Statement from the Council on Clinical Cardiology, Heart Failure and Transplantation Committee; Quality of Care and Outcomes Research and Functional Genomics and Translational Biology Interdisciplinary Working Groups; and Council on Epidemiology and Prevention. Circulation 2006;113:1807–16.
3. Elliott P, Andersson B, Arbustini E, et al. Classification of the cardiomyopathies: a position statement from the European Society Of Cardiology Working Group on Myocardial and Pericardial Diseases. Eur Heart J 2008 ;29:270–6.
4. Thuny F, Jacquier A, Jop B, et al. Assessment of left ventricular non-compaction in adults: side by- side comparison of cardiac magnetic resonance imaging with echocardiography. Arch Cardiovasc Dis 2010; 103:150–159
5. Petersen SE, Selvanayagam JB, Wiesmann F, et al. Left ventricular non-compaction: insights from cardiovascular magnetic resonance imaging. J Am Coll Cardiol 2005; 46:101–105
6. Jacquier A, Thuny F, Jop B, et al. Measurement of trabeculated left ventricular mass using cardiac magnetic resonance imaging in the diagnosis of left ventricular non-compaction. Eur Heart J 2010; 31:1098–1104
7. Grothoff M, Pachowsky M, Hoffmann J, et al. Value of cardiovascular MR in diagnosing left ventricular non-compaction cardiomyopathy and in discriminating between other cardiomyopathies. Eur Radiol 2012; 22:2699–2709.
8. Stöllberger C, Finsterer J: Septal hypertrabeculation/noncompaction: cardiac and neurologic implications. Int J Cardiol 2009, 132:173-175.
9. Sedmera D, Pexieder T, Vuillemin M, et al. Developmental patterning of the myocardium. Anat Rec 2000;2584:319- 37.
10. Ivan D, Flamm SD, Abrams J, Kindo M, Heck K, Frazier OH: Isolated Ventricular Non-Compaction in Adults with Idiopathic Cardiomyopathy: Cardiac Magnetic Resonance and Pathologic Characterization of the Anomaly. J Heart Lung Transplant 2005,

24(6):781-786.
11. Alsaileek AA, ISyed I, Seward JB, Julsrud P: Myocardial fibrosis of left ventricle: magnetic resonance imaging in non compaction. J Magn Reson Imaging 2008, 27:621-624.
12. Aras D, Tufekcioglu O, Ergun K, et coll. Clinical features of isolated ventricular non compaction in adults long-term clinical course, echocardiographic properties, and predictors of left ventricular failure. J Card Fail. 2006; 12:726-733.
13. Lofiego C, Biagini E, Ferlito M, et al: Paradoxical Contributions of Non- Compacted and Compacted Segments to Global Left Ventricular Dysfunction in Isolated Left Ventricular Noncompaction. Am J Cardiol 2006, 97(5):738-741.
14. Fazio G, Corrado G, Novo G, et al: Ventricular dysfunction and number of non compacted segments in non compaction: Non-independent predictors. Int J Cardiol 2010, 141:250-253
15. Habib G, Charron P, Eicher JC, et al. Isolated left ventricular non-compaction in adults: clinical and echocardiographic features in 105 patients. Results from a French registry. Eur J Heart Fail 2011; 13:177-85.