Role of Myocardial Deformation to Improve Assessment of Regional and Global Function in Non-Isolated Non-Compaction Cardiomyopathy: New Insights in Cardiac Mechanics

Olivier Huttin1*, Zied Frikha1, Pierre-Yves Marie2, Yves Juillière1, and Christine Selton-Suty1

1Department of Cardiology, Institute of Lorrain Heart and Vascular Centre, University Hospital of Nancy, France
2Department of Nuclear Medicine, University Hospital of Nancy, France

Abstract

A 55-year-old man with acute congestive heart failure was referred to the echocardiography laboratory for evaluation of a dilated cardiomyopathy with severe left ventricular systolic dysfunction and criteria for non-compaction. Cardiac magnetic resonance imaging confirmed a non-compacted apical LV with segmental dysfunction and an inferior myocardial infarction. Coronary angiogram showed a chronic right coronary occlusion. Speckle tracking imaging with analysis of peak systolic strain and post systolic strain index helped to differentiate ischemia and non-compaction as mechanisms of wall motion abnormalities. These findings support the concept that non-compaction is a phenotypic marker of an underlying diffuse cardiomyopathy involving both morphologically normal and non-compacted myocardium and the association with micro and macro vascular coronary arteries disease is discussed.

ABBREVIATIONS

©LVNC: Left Ventricular Non Compaction; NC: Non Compacted; C: Compacted; CAD: Coronary Artery Disease; MRI: Magnetic Resonance Imaging; PSI: Post Systolic Strain Index, 2DLS: 2D Longitudinal Speckle Tracking; LVEF: Left Ventricular Ejection Fraction; CMR: Cardiovascular Magnetic Resonance; MI: Myocardial Infarction; MDE: Myocardial Delayed Enhancement; WMA: Wall Motion Abnormalities

INTRODUCTION

According to the ESC Working Group on Myocardial and Pericardial Disease, left ventricular non compaction (LVNC) is still an unclassified cardiomyopathy and considered as a non-ischemic myocardial disease. Association between LVNC and coronary artery disease (CAD) is not common and its frequency is not well established. Deformation imaging appears to be an interesting tool to evaluate the degree and the extent of LV dysfunction. Furthermore, it may also give interesting information regarding the mechanisms of segmental abnormalities. The analysis of post systolic shortening and the calculation of post systolic strain index (PSI) may help to identify segmental dysfunction due to the primitive cardiomyopathy and ischemic related dysfunction with or without significant CAD. The purpose of this case report was to illustrate the role of deformation imaging to discriminate both the degree and the aetiology of segmental dysfunction as well as to define a new tool to help classification of patient with similar dilated phenotype.

CASE PRESENTATION

A 55 years old male was referred to our hospital for a first episode of acute heart failure. His risk factors for coronary disease were smoking, hypertension and hyperlipidemia. A family history of cardiac disease with a sudden death was noted. At admission, physical examination revealed a heart rate of 120 bpm, high blood pressure of (172/83 mm Hg) with clinical...
signs of left heart failure. The 12-lead ECG showed sinus rhythm with incomplete LBBB. A negative T wave in V2 to V6, D1 and Avl was noted (Figure 1). Blood tests revealed an elevated BNP at 1000ng/ml (normal<500ng/ml) and a troponin at 0.06 (normal<0.04). Echocardiography performed after stabilization of the patient on medical treatment demonstrated a severe LV systolic dysfunction (LVEF Simpson biplane 22%), moderate LV dilatation and no LV hypertrophy. Parasternal short axis revealed the presence of numerous prominent trabeculations and deep intertrabecular recesses in 6 segments at the level of mid lateral and apical segments of the LV (Figure 2). Hypertrabeculation fulfilled echocardiography criteria of LVNC (systole Ratio NC to and apical segments of the LV (Figure 2). Hypertrabeculation fulfilled echocardiography criteria of LVNC (systole Ratio NC to compacted layer above two) [1]. There was no RV involvement and no mitral or aortic regurgitation. Pulmonary artery pressure was estimated at 45mmhg. A global hypokinesia with a slightly more pronounced LV wall motion abnormality (WMA) in the inferior wall was noted. To better elucidate global and regional LV function we measured systolic and post-systolic longitudinal deformation with the use of 2D longitudinal (2DLS) speckle tracking from the 3 apical LV chambers views. The calculation of global and regional longitudinal strain values was easily obtained by Automated Function Imaging software. There was a global longitudinal LV systolic dysfunction affecting mainly basal and mid LV segments and mildly impaired at the level of the 5 apical NC segments. Furthermore post systolic shortening study (PSS) with post systolic strain index (PSI) calculation demonstrated high values in the inferior and infero-lateral wall segments and normal values at in the anterior and antero-septal walls and also in the NC apical segments (Figure 3). These discrepancies in the results suggested a LVNC with severe LV systolic dysfunction related to the cardiomyopathy itself and another suspected ischemic mechanism in relation with the altered PSI in the inferior wall. Evolution on medical therapy was good and patient was treated as a non-STEMI by antiaggregation therapy and anticoagulation. Coronary angiogram was performed a few days later and showed a CAD with right coronary occlusion. There was no significant lesion on the left coronary artery. A cardiac magnetic resonance (3T Signa HDxt General Electric Healthcare, Milwaukee, WI) confirmed the diagnosis of LVNC. Furthermore it illustrated a severe LV dysfunction (LVEF=0.32) and severe hypokinesia associated with intermediate late Gadolinium enhancement in the LV inferior lateral wall (Figure 4). Hence, the patient was initiated on low-dose of ACE inhibitors, beta blockers and aspirin because of the CAD. There was no improvement of LVEF at 3 months follow-up on medical therapy and patient was proposed an automatic implantable cardioverter defibrillator.

DISCUSSION

This case illustrates that mechanisms of LVS dysfunction in LVNC are not clearly elucidated. Several studies demonstrated that global LVS dysfunction is common in LVNC [2-4]. The correlation between number of NC segments and LVEF remains controversial. Several papers suggested that the degree of NC and the extent of the NC myocardium was related to the severity of LV systolic dysfunction [5,6]. In contrast, Habib et al and Fazio et al reported exactly opposite findings [2,7]. Myocardial hypoperfusion of NC myocardium may be the first step of myocardial damage and possibly the basis of LV failure [8]. Impaired microcirculation can lead to a relative chronic myocardial ischemia caused by mismatch of blood demand and supply to multiple prominent trabeculae segments but also in compacted myocardium [9]. The link between LVNC and ischemic cardiomyopathy remains unclear but the diagnosis of the etiology of dysfunction is a major clinical challenging. Coronary angiography usually reveals no significant abnormalities. Stoeckl et al demonstrated a rare association between coronary arteriosclerosis and LV hypertrabeculation which does not seem to affect prognosis [10]. Positron emission tomography showed a diminished reserve of coronary blood flow in the compact and NC segments [11]. Jenni confirmed not only that LVNC is associated with coronary microcirculatory dysfunction but also that is not confined to NC segments and extends to most segments with WMA [12]. Myocardial fibrosis represents a second hypothesis to explain decreased LV function as it is observed in 50% of patients involving both compacted and NC myocardium [13]. Our patient had severe LV dysfunction. WMA were related to both the NC and to a previous MI. At this point, the controversy regarding the link between those two entities has not been settled and prevalence of coronary pathologic findings in patients with LVNC is unknown. For AMI, LVNC association is described only in several case reports [14,15].

2DSTE is now used in clinical practice to measure deformation. It allows functional assessment of the LV providing regional and

Figure 1 12-lead ECG: Sinusual tachycardia with a left QRS axis and incomplete left bundle branch block morphology, negative T wave in the anterior leads.
Figure 2 Transthoracic echocardiography at day 4; (A) 3D apical view: severe LV systolic dysfunction (LVEF 25%) and (B) 2D short axis color Doppler view demonstrating trabeculations and deep sinusoids within NC.

Figure 3 Bulls-eye figure of peak systolic strain values in the LV recorded from 3 apical chambers views (A) 2D Longitudinal peak systolic strain values: global strain was -6.7%. Sub normal longitudinal deformation at the level of the apical NC segments; lower values of systolic deformation in the septal and inferior wall. (B) PSI profile from NC basal lateral segment abnormal inferior mechanics. High PSI values at the level of the inferior and infero lateral wall.

Figure 4 A. Coronary angiogram showed a right coronary occlusion - B. Representatives samples of CMR imaging (day 7): CMR late gadolinium enhancement images with a single, midventricular slice transmural inferior infarct with decreased LV systolic function (26% of LVEF).
global myocardial mechanics which are of great interest in LVNC. The diagnosis of LVNC is usually assessed with the use of the echocardiographic criteria of Chin, Jenni and Stöllberger [1,16]. Paterick and al tried to introduce the concept of myocardium contraction of the NC segments as an additional diagnosis criteria [17]. Abnormal myocardial mechanics with criteria of pathologic hypertrabeculation may help to diagnose LVNC. We demonstrated a global LV longitudinal dysfunction with a significant 2DLS basal-apex ratio and preserved strain values in NC segments despite an overall hypokinesia. Another study using tissue Doppler – derived strain reported preserved deformation in basal segments [18]. Other authors evocated the role of rotation as a potential quantitative functional diagnostic criterion [19]. These findings suggest that the worse WMA are localized at the level of the NC segment, but in our experience we found a mildly impaired longitudinal deformation in the apical and lateral non-compact segment compared to severe dysfunction of the overall compacted myocardium. It may be explain by the thickness of the NC segments with preserved contractile properties of the NC layer. STE curve analysis by calculation of PSI taking into accounts both systolic and early diastolic deformation provide further information on the heterogeneity of myocardial mechanics in our case. Values of 2DLS are significantly reduced in both infarcted and “non-ischemic” segments, implying a reduction in systolic deformation. But PSI was normal in non-ischemic compacted and NC segments. PSI bull-eyes analysis showed normal values in non-infarcted segments among both compacted and NC LV areas. In non-ischemic myocardium, virtually all contraction occurs during systole with very little post-systolic shortening. PSI combines systolic and early diastolic deformation values as a quantitative measure of the post systolic compression. It was calculated as 100(PS-ESS)/PS (PS=Peak Strain over whole beat). It represents the relative amount of ischemia-related segmental thickening/shortening, which is found to occur after aortic valve closure. Previous studies defined PSI as a good marker of chronic ischemic dysfunction [20] and had a sensitivity of 95% and a specificity of 89% in the identification of acutely ischemic segments during coronary occlusion [21]. It takes into account the ESS and has a much higher potential than strain values alone as marker of ischemic dysfunction. However our results did not support the chronic myocardial ischemic hypothesis to explain segmental dysfunction in LVNC. Indeed we did not found any pattern of ischemic dysfunction in the non-infarcted segment. The use of this parameter could promote a better understanding of LWMA mechanical characterization and was the only one able to dearly discriminate ischemic territories in this patient with non-isolated LVNC.

CONCLUSION

This case report highlights the importance of LVS function assessment by new tools in order to improve comprehension of global and regional mechanisms of LV dysfunction in LVNC. Myocardial dysfunction due to ischemic disease exists in LVNC both secondary to micro and macrovascular CAD. Neither visual assessment nor standard 2DLS were able to distinguish ischemic and non-ischemic compacted segment. PSI seems able to overview and detect regional ischemic disease in an overall global myocardium dysfunction. Moreover we observed a GLS base-apex gradient due to functional impact of an abnormal apical myocardial architecture enable the clinician to distinguish between normally trabeculated myocardium from LVNC. Following this first report on the interest of post systolic shortening analysis in LVNC, this parameter should be measured in various phenotypes of LVNC to assess its additional diagnostic value.

REFERENCES

15. Stöllberger C, Finsterer J. Left ventricular hypertroabeculation/