

Cardiac magnetic resonance imaging in a young patient with left ventricular spongiform cardiomyopathy: A case report and review of literature

Bilal Hussain,¹ Fateh Ali Tipoo Sultan²

Abstract

Left ventricular spongiform or non compaction cardiomyopathy is one of the rarer forms of cardiomyopathy with a reported prevalence of 0.014-0.05% in adults. It is characterized by severe left ventricular systolic dysfunction occurring secondary to the failure of the myocardium to fuse resulting in non-compacted myocardium with a trabeculated appearance of the myocardium. Initially described on the basis of 2-D echocardiography, the identification of left ventricular non compaction has now improved with the introduction of sophisticated imaging modalities such as cardiac magnetic resonance imaging that allows better visualization of the non-compacted myocardium assisting in accurate diagnosis. We report a case of isolated left ventricular spongiform or non-compaction cardiomyopathy that was diagnosed on cardiac magnetic resonance imaging with classical features. The patient was started on standard heart failure medications along with anticoagulation and continues to do well on follow up.

Keywords: Case Report, Cardiomyopathies, Magnetic Resonance Imaging, Isolated Non-compaction of the Ventricular Myocardium.

Introduction

Left ventricular non compaction cardiomyopathy (LVNC) also termed as 'spongiform cardiomyopathy' due to the extensively trabeculated appearance of myocardium is a rare entity described as a primary genetic cardiomyopathy by the American Heart Association. It is postulated to result from the arrest of the embryonic development of myocardium resulting in an un-fused layer of myocardium projecting into the left ventricular cavity causing severe left ventricular systolic dysfunction.¹

Cardiac imaging is pivotal to the diagnosis of LVNC and advancements in technology have increased the sensitivity and specificity of Cardiac magnetic resonance imaging (CMR) for the diagnosis of LVNC.²

.....
¹Adult Cardiology, ²Section of Cardiology, Department of Medicine, Aga Khan University Hospital, Karachi, Pakistan.

Correspondence: Bilal Hussain. Email: bilal_observer@hotmail.com

We present the case of a young man who came to the clinic with symptom of dyspnoea. His workup including a 2-D echo showed a dilated left ventricle with severe systolic dysfunction and suspicion of non-compaction. Suspecting possible left ventricular non-compaction, CMR was done that showed classical findings consistent with the diagnosis of left ventricular non-compaction or spongiform cardiomyopathy.

To the best of our knowledge this is the first case report from Pakistan in which spongiform or non-compaction cardiomyopathy was diagnosed on CMR with classical features. The relative lack of literature from Pakistan on LVNC is largely because of a lack of resources and expertise in the field of cardiac imaging.

Case Report

A 25 year old man from Larkana, presented to The Aga Khan University Hospital clinics in August, 2015 with a history of worsening dyspnoea over the last 2 years. Due to worsening symptoms his functionality had been severely restricted. He gave no history of chest pain, syncope or palpitations. He was born of a consanguineous marriage and did not report any sudden cardiac death in the family. He was a cigarette smoker but had no past history of a major medical illness.

For his current symptoms, he had been started on various therapies including inhalers, diuretics and multivitamins. He was non compliant to medications and reported 'no benefit from treatment' as the cause of non compliance.

On examination, he had a regular pulse but was tachycardiac. Physical examination revealed jugular venous distension, pedal oedema and bi-basilar chest crepitations on chest auscultation. On precordial examination, he had a displaced apex beat, first and second heart sounds were normal, no added heart sounds or murmurs were audible.

On the basis of his history and examination, a diagnosis of heart failure was postulated. He was started on diuretics, Beta blockers and angiotensin converting enzyme inhibitors. An electrocardiogram (ECG) and echocardiogram were done.

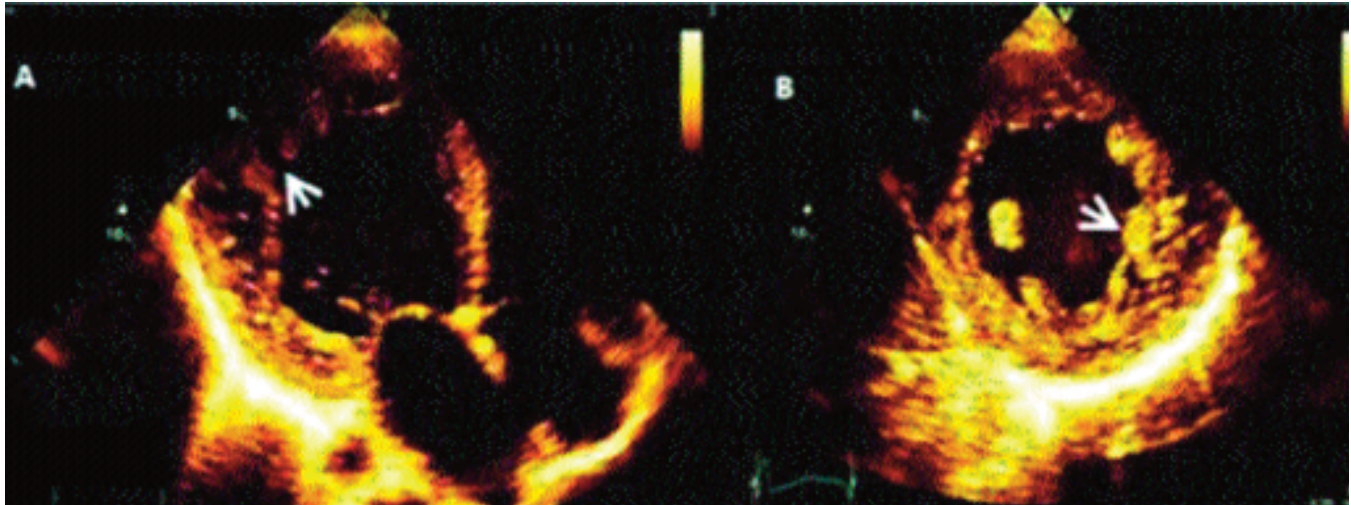


Figure-1: 2-D echo images showing (A) Apical two chamber view and (B) Parasternal short axis view of left ventricle with spongiform appearance of left ventricle wall and prominent recesses in the endocardium marked by white arrows.

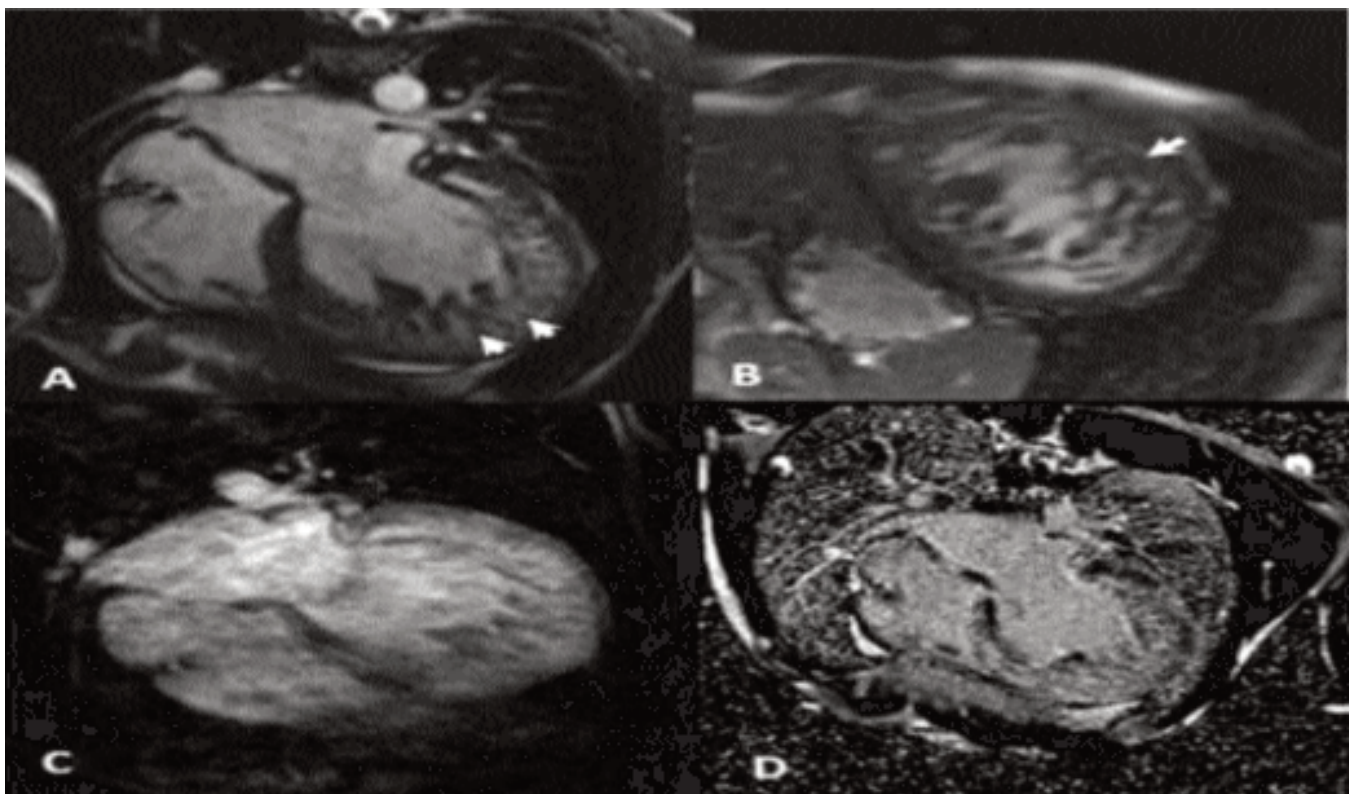


Figure-2: Cardiac MRI steady state free precession images (SSFP) showing (A) Four chamber view and (B) short axis view of the left ventricle with classical spongiformmomon compaction pattern marked with white arrows. Early gadolinium images of CMR(C) showing lack of hypo-enhancement to suggest intra-cavitary thrombus, delayed enhanced images with gadolinium (D) showing no definite area of hyper-enhancement to suggest myocardial fibrosis.

His ECG showed intra-ventricular conduction delay pattern and echocardiography showed grossly dilated and enlarged left ventricle with severely reduced left

ventricular systolic function and global hypokinesia. The left ventricular myocardium appeared to have a rugged appearance with prominent trabeculations seen

projecting into the left ventricle cavity (Figure-1). A possible diagnosis of LVNC was made on the basis of echo findings. To confirm the diagnosis a contrast enhanced CMR was ordered.

CMR was done on Siemens Avanto 1.5 tesla machine using 10cc gadolinium as contrast. Steady state free precession images (SSFP) showed severe generalized global hypokinesia of the left ventricle (Ejection Fraction 17%). Prominent recesses with spongiform pattern were seen in the left ventricle, especially in the apical and antero-lateral region. The compacted myocardium was thin, especially in the apical and antero-lateral region with visible compacted and non-compacted zones. The non-compacted to compacted ratio was more than 4:1 during diastole. These findings were consistent with the diagnosis of left ventricular non-compaction cardiomyopathy.

Early gadolinium images showed no area of hypo-enhancement to suggest intra-cavitary thrombus. Delayed enhanced imaging with gadolinium showed no definite area of hyperenhancement to suggest myocardial fibrosis (Figure-2).

On the basis of these findings the patient was diagnosed to have isolated left ventricular non-compaction or spongiform cardiomyopathy. He was continued on optimized heart failure medications and was started on anticoagulation for prevention of thrombo-embolic complications. He was advised device therapy for primary prevention. On follow up after a month, he was doing fine on medical therapy.

Discussion

LVNC is a rare cardiomyopathy that is characterized by the presence of a trabeculated myocardium layer, with deep inter-trabecular recesses adherent to a thin compact layer of myocardium resulting in severe left ventricular dysfunction.¹

The etiology of LVNC is assumed to be due to errors in embryonic development.¹ First described on echocardiography in 1984 by Engberding and Bender as the persistence of isolated myocardial sinusoids. LVNC is a rare, yet recognized cause of cardiomyopathy, with a prevalence of 0.014% in adult population.³ Our patient was a male and it is recognized that LVNC occurs more often in men than in women.^{2,4} LVNC has been described occurring alone and as a part of complex congenital heart diseases and other forms of cardiomyopathies. In our case, LVNC was an isolated finding without other congenital cardiac malformations.

The presentation of patients with LVNC depends on the

stage of disease. Most patients presenting at later stages have symptoms of heart failure, as was the case with our patient. No ECG findings are specific for the diagnosis of LVNC. Our patient had intra-ventricular conduction delay pattern on ECG.

ECHO features suggesting LVNC include the presence of two-layered appearance of the myocardium with a thin, compacted outer layer and a thicker, non-compacted inner layer with prominent left ventricular trabeculations, predominantly in the apical and mid-ventricular areas. The widely accepted criteria for diagnosing LVNC on ECHO is that the end-systolic ratio between non-compacted and compacted myocardium should be greater than 2.0.^{5,6} The echo of our patient was suggestive of non-compaction.

ECHO has limitations in diagnosing LVNC such as the failure to visualize the LV apex during study and the inability to differentiate between normal variant and pathological trabeculations. CMR overcomes the shortcomings of ECHO in diagnosing LVNC as it visualizes the entire myocardium more clearly, adequately defining the thickness of myocardium and helping in accurately calculating LV volumes and ratios of non compacted to compacted myocardium.

The CMR diagnostic criteria for LVNC includes a non-compacted to compacted myocardial ratio greater than 2.3 as measured in end-diastole, along with the visualization of two distinct myocardial layers i.e. a compacted epicardial layer and a non-compacted endocardial layer.⁷ In our patient CMR findings were classical, fulfilling all the criteria for diagnosis of LVNC.

The superiority of CMR to ECHO is its 3-dimensional approach.⁸ As the entire heart is visualized, concomitant right ventricular non compaction can also be accurately diagnosed by CMR as opposed to conventional 2-D ECHO. If other structural heart defects are present, CMR can easily identify these anomalies.

Additional CMR features such as endocardial T2 signaling, early and delayed enhancement imaging of the subendocardial layer provide information regarding function and fibrosis of the affected segments and can, thus, help in identifying areas of arrhythmogenic activity or presence of apical LV clots.⁹

While the CMR provides a crisp image quality with high spatial resolution, its use is limited because of its limited availability and prolonged image acquisition time.

Conclusion

Left ventricular non compaction is a rare form of

cardiomyopathy and is associated with significant morbidity and mortality. Diagnosis of LVNC is based on cardiac imaging. Due to limitations of ECHO, CMR is replacing ECHO as the diagnostic modality of choice for LVNC. Recent advances in CMR have made the diagnosis of LVNC more accurate and have helped in identifying patients who can benefit from device therapy. The limitation to mass use of CMR in the diagnosis of cardiomyopathies such as LVNC, remains the limited availability of CMR across the country.

Informed Consent Obtained: Yes.

Conflicts of Interest: None.

Funds Declaration: None.

Disclaimer: None.

References

1. Maron BJ, Towbin JA, Thiene G, Antzelevitch C, Corrado D, Arnett D, 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 Apr 11; 113: 1807-16.
2. Zuccarino F, Vollmer I, Sanchez G, Navallas M, Pugliese F, Gayete A. Left ventricular noncompaction: imaging findings and diagnostic criteria. *AJR Am J Roentgenol*. 2015; 204: W519-30.
3. Aras D, Tufekcioglu O, Ergun K, Ozeke O, Yildiz A, Topaloglu S, et al. Clinical features of isolated ventricular noncompaction in adults long-term clinical course, echocardiographic properties, and predictors of left ventricular failure. *J Card Fail*. 2006; 12: 726-33.
4. Stanton C, Bruce C, Connolly H, Brady P, Syed I, Hodge D, et al. Isolated left ventricular noncompaction syndrome. *Am J Cardiol*. 2009; 104: 1135-8.
5. Jenni R, Oechslin 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.
6. Stöllberger C, Gerecke B, Finsterer J, Engberding R. Refinement of echocardiographic criteria for left ventricular noncompaction. *Int J Cardiol*. 2013; 165: 463-7.
7. Petersen SE, Selvanayagam JB, Wiesmann F, Robson MD, Francis JM, Anderson RH, et al. Left ventricular non-compaction: insights from cardiovascular magnetic resonance imaging. *J Am Coll Cardiol*. 2005; 46: 101-5.
8. Thuny F, Jacquier A, Jop B, Giorgi R, Gaubert JY, Bartoli JM, 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: 150-9.
9. Grothoff M, Pachowsky M, Hoffmann J, Posch M, Klaassen S, Lehmkuhl L, 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.