Case Report

Swiss Cheese Myocardium: A Diagnostic and Prognostic Sign of HCM by Newer CMR Techniques

Atul Kapoor[1], Harinderpal Singh[2], Aprajita Kapoor[3], Goldaa Mahajan[1]

[1]Consultant Radiologist Advanced Diagnostics, Amritsar, Punjab, INDIA.
[2]Director Cardiology Fortis Hospital, Amritsar, Punjab, INDIA.
[3]Consultant Advanced Diagnostics, Amritsar, Punjab, INDIA.

ABSTRACT

Cardiac MR(CMR) has an important role in the evaluation of patients of suspected cardiomyopathies and also helps to differentiate between the various types based on the type of late gadolinium enhancements(LGE's). We describe a characteristic Swiss cheese myocardium appearance in a patient with hypertrophic cardiomyopathy (HCM) using automatic scar analysis algorithm using low < 2SD scale threshold weighting to assess the replacement and interstitial fibrosis along with T1 mapping to quantify it.

Key words: Cardiac MRI, Hypertrophic cardiomyopathy, Extracellular volume.

INTRODUCTION

Cardiac magnetic resonance imaging (CMR) is increasingly being used for the evaluation of patients with suspected cardiomyopathies as it provides detailed myocardial morphology including fibrosis evaluation which has diagnostic and prognostic implications for the patient.[1,2] The diagnosis of hypertrophiccardiomyopathy (HCM) traditionally is based on the echocardiographic findings of asymmetric left ventricular hypertrophy and systolic anterior motion of mitral leaflet. CMR due to superior spatial resolution is being used as adjunctive technique in patients of HCM with phenotypic heterogeneity. Due to improvement in techniques, CMR is now being tried for prognostic evaluation of HOCM. Late gadolinium enhancement (LGE) sequences enable the identification of myocardial fibrosis, which is associated with poor outcome.[3]

The scarring seen in hypertrophic obstructive cardiomyopathy(HCM) is of mixed type i.e. replacement, interstitial and plexiform type with overlapping patterns which alone cannot be differentiated from scarring seen in other types of cardiomyopathies seen as late gadolinium enhancements(LGE's).

We describe the use of an automated scar algorithm technique using signal weighting using low gray scale threshold on late gadolinium enhanced Images which gives a characteristic Sweese Cheese appearance of the myocardium due to mixture of fibrosis seen in the HCM patient. The quantification of fibrosis was done using T1 mapping on MRI to estimate extracellular volume (ECV) of the myocardium. The use of these novel techniques can not only be diagnostic but also give prognostic implications.

CASE REPORT

A 35 year old non hypertensive male born of a non consanginous marriage with no prior family history of heart disease presented with history of palpitations. His general physical examination and electrocardiogram werenormal. Echocardiogram done revealedasymmetric septalthickening with normal left ventricle, systolic and diastolic functions. No systolic anterior motion of mitral leaflet was seen. CMR was done on 1.5 T Siemens Amira system using body surface matrix coil. Cine gated images were taken from base to apex of heart in short axis views and revealed thickened basal interventricular septum with normal wall motion and left ventricle function(Figure 1). Native T1 images of the myocardium were taken using EKGgated ShMOLLI sequence and revealed

global prolongation of T1 values with mean T1 of 1150 msec. Post intravenous gadolinium enhanced images obtained in same positions using same sequence to estimate T1 shortening and to determine extracellular volume(ECV).It revealed globallyincreased ECV from 31-35% in all the myocardial segments(Figure 2). LGE's images using inversion recovery sequences after nulling of myocardium revealed small patchy areas of LGE's opposite right ventricleinsertions (Figure 3). Automated scar analysis evaluation was done as shown by Kim *et al*[4] using low grayscale threshold of 1.8 SD. It revealed a classical Swiss cheese appearance of myocardium with patchy areas of replacement fibrosis interspersed with diffuse areas ofinterstitial fibrosis (Figure 4a,b)

DISCUSSION

Diagnosis of HCM on CMR has been based on the patterns of asymmetric left ventricle hypertrophy alongwith demonstration of myocardial fibrosis on LGE images. The fibrosis seen in HCM is of mixed type i.e. replacement, interstitial and plexiform type with overlapping patterns which alone cannot be differentiated from scarring seen in other types of cardiomyopathies seen as late gadolinium enhancements(LGE's).The
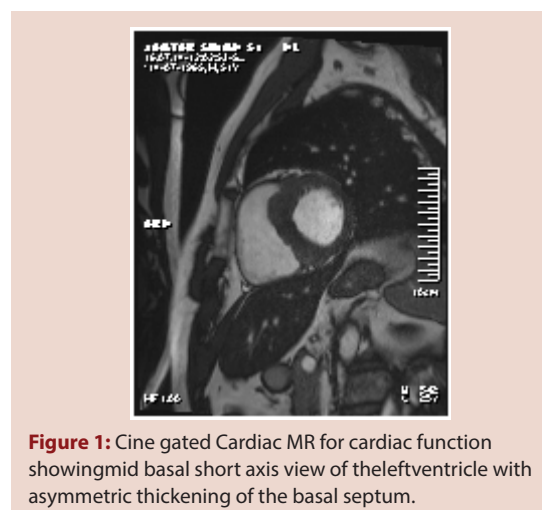

Figure 1: Cine gated Cardiac MR for cardiac function showingmid basal short axis view of theleftventricle with asymmetric thickening of the basal septum.
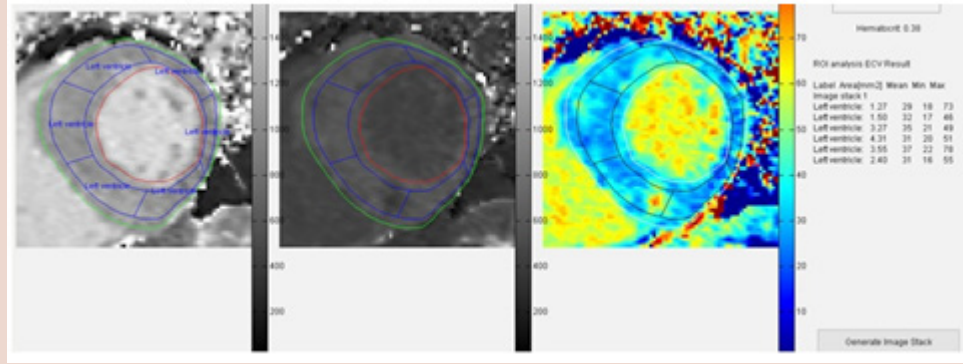
Figure 2: Extracellular volume estimation of the left ventricle using plain and post contrast IV gadolinium with global increased ECV from 31-35%.

Figure 3: Late gadolinium enhanced images a) showing patchy areas of LGE's in the thickened LV septum at RV insertion points.

Figure 4a: Scar gray scale weighting done by remote less than 2 SD method.

histological correlate of LGE in HCM seems to be increased myocardial collagen rather than myocardial disarray which is also observed on histological specimens and is postulatedto reflect microvascular ischemia and microscopic replacement fibrosis due to small intramural coronary arteriole dysplasia.[5,6] Another alternative hypothesis for LGE in HCM suggested that the causative sarcomeric gene mutations might lead to a phenotypic expression of increased myocardial connective tissue deposition,[7] Hence there is a need to detect myocardial fibrosis and quantify the excessive collagen deposition in the Myocardium. Detection of myocardial fibrosis by LGE is variable in different HCM cohorts and is between 40-80%.Traditionally found patterns are patchy mid myocardial LGE, patchy LGE at RV insertion points and diffuse confluent trans mural pattern. Detection of LGE in HCM alsohas association with ventricular arrhythmias, SCD and the pathophysiological link has been shown on Holter monitoring.[8,9,10] Therefore robust detection and characterization of collagen could form the basis for risk stratification of HCM to be considered for ICD therapy.

So far collagen has been difficult to detect and quantify on imaging mainly due to diffuse nature of the disease process and also due to limitation in processing techniques. Native T1 mapping and ECV estima-
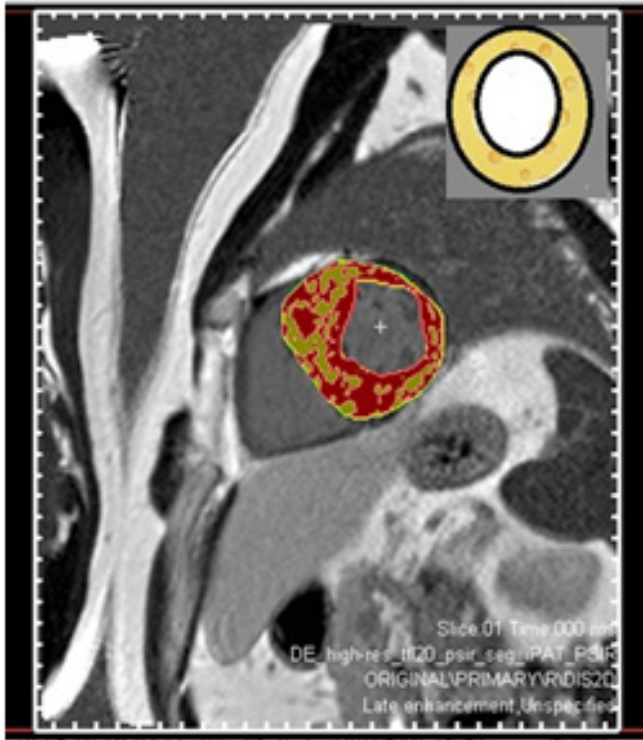
Figure 4b: Swiss Cheese appearance of the fibrosis with areas of replacement fibrosis as orange and interstitial fibrosis as yellow with inset showing pictorial depiction of swiss cheese.

tion are two novel techniques to diagnose interstitial fibrosis and also to quantify it but these cannot differentiate between different causes of interstitial fibrosis. With the use of intravenous gadolinium delayed images are obtained using inversion recovery sequence with myocardium nulling where the scar tissue appears as hyper-intense due to increased uptake of contrast by the scar tissue. By modifying this technique by the use of automated scar analysis algorithm with low grey scale threshold of 2 SD or less one can differentiate between the relative signal of the nulled myocardium, increased trapping of contrast in the interstitial scar and the hyper-intense replacement scar which gives a swiss cheese appearance.

To our knowledge this sign has not been previous described and is characteristic for HCM. The use of CMR using scar weighting algorithm as described alongwith T1 mapping with ECV estimation not only gives diagnostic appearance of the condition but also by quantifying the fibrosis gives a prognostic significance of these patients due to prevalence of prolific mixed scarring in the myocardiumwhich carries increased risk for arrythmia and sudden death. Therefore by early detection and quantification of fibrosis patients can be stratified as high risk to be considered for device implants.

REFERENCES

1. Teraoka K., Hirano M., Ookubo H., *et al.* Delayed contrast enhancement of MRI in hypertrophic cardiomyopathy. MagnReson Imaging. 2004;22(2):155–61.

2. Choudhury L., Mahrholdt H.,Wagner A., *et al.* Myocardial scarring in asymptomaticor mildly symptomatic patients with hypertrophic cardiomyopathy. J Am CollCardiol 2002;40(2):2156–64.

3. Kwon D.H., Setser R.M., Popovic Z.B., *et al.* Association of myocardial fibrosis, electrocardiography and ventricular tachyarrhythmia in hypertrophic cardiomyopathy: a delayed contrast enhanced MRI study.Int J Cardiovasc Imaging 2008;24(6):617–25.

4. Kim RJ, Fieno DS, Parrish TS, Harris K, Chen EL, Simonetti O *et al.* Relationship of MRI delayed contrast enhancement to irreversible injury, infarct age, and contractile function. Circulation: 1999;100(19):1992–2002.

5. Rubinshtein R., Glockner J.F.,Ommen S.R., *et al.* Characteristicsandclinical significance of late gadolinium enhancement by contrast-enhanced magnetic resonance imaging in patients with hypertrophic cardiomyopathy. Circ Heart Fail 2010;3:51–8.

6. Hanlon R., Grasso A., Roughton M., *et al.* Prognostic significance of myocardial fibrosis in hypertrophic cardiomyopathy. J Am CollCardiol 2010;56:867–74.

7. Bruder O., Wagner A.,Jensen C.J., *et al.* Myocardial scar visualized by cardiovascular magnetic resonance imaging predicts major adverse events in patients with hypertrophic cardiomyopathy. J Am CollCardiol 2010;56(11):875–87.

8. Adabag A.S., Maron B.J., Appelbaum E., *et al.* Occurrence and frequency of arrhythmias in hypertrophic cardiomyopathy in relation to delayed enhancement on cardiovascular magnetic resonance. J Am CollCardiol 2008;51(14):1369–74.

9. 9 Moon J.C, Reed E, Sheppard M.N., *et al.* The histologic basis of late gadolinium enhancement cardiovascular magnetic resonance in hypertrophic cardiomyopathy. J Am CollCardiol 2004;43(12):2260–4.

10. 10 Kwon D.H.,Smedira N.G., Rodriguez E.R., *et al.* Cardiac magnetic resonance detection of myocardial scarring in hypertrophic cardiomyopathy: correlation with histopathology and prevalence of ventricular tachycardia. J Am CollCardiol 2009;54(3):242–49.