Original Research Article

Cardiac MRI in diagnosis and evaluation of hypertrophic cardiomyopathy

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Abstract

Background: Hypertrophic cardiomyopathy is the commonest genetic cardiomyopathy with varied presentations from asymptomatic incidental diagnosis, various arrhythmias, heart failure to sudden cardiac death. Cardiac MRI provides complete visualization of the LV, allowing precise localization of the distribution of hypertrophy and measurement of wall thickness and cardiac mass. Cine imaging provides assessment of additional morphological information including systolic anterior motion of the anterior mitral leaflet with dynamic outflow tract obstruction, mitral regurgitation. Late gadolinium enhancement imaging enables to detect presence of myocardial fibrosis which has prognostic implication. The **purpose** of our study is to evaluate the use and applications of Cardiac MRI in diagnosis and risk stratification of hypertrophic cardiomyopathy. **Materials and Methods:** This retrospective study was performed in our department of Radio-diagnosis in a tertiary health care institute including 23 patients referred for confirmation of diagnosis of hypertrophic cardiomyopathy and risk stratification. The patients were evaluated with Cardiac MRI using standard body coil and dedicated cardiac phase array coil on 3T MRI 32 channel Siemens-Verio MRI machine with ECG and respiratory gating. **Results:** In our study, HCM is more common in males (78.26%), most common in 41-80 years age group, most common presentation being Dyspnoea on exertion (69.5%), most common phenotype asymmetrical septal hypertrophic cardiomyopathy (82.6%). Late gadolinium enhancement (52.1%), LVOT gradient more than 30mmHg at rest (43.4%), LV maximal wall thickness of 30mm or more (13%), LV dilatation with depressed ejection fraction (00%) and Myocardial perfusion defect (00%) suggest high risk group for sudden cardiac death among cases of HCM. **Conclusion:** Cardiac MRI is an excellent non invasive modality for diagnosis and risk stratification of hypertrophic cardiomyopathy.
Key Words: Hypertrophic cardiomyopathy Cardiac MR Late gadolinium enhancement LVOT gradient.

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INTRODUCTION

Hypertrophic cardiomyopathy is the commonest genetic cardiomyopathy. It is morphologically characterized by myocardial fibrosis, myocyte disarray and hypertrophy of the myocardium. Its clinical presentation is varied. It varies from asymptomatic incidental diagnosis, to various arrhythmias, to heart failure to sudden cardiac death. With excellent spatial resolution and border definition, Cardiac MRI provides complete visualization of the LV chamber, allowing precise localization of the distribution of hypertrophy and measurement of wall thickness and cardiac mass[2]. Cardiac MRI is also superior to echocardiography in the detection of apical and focal basal antero-septal variants and in recognizing non-contiguous areas of hypertrophy[3]. Cardiac MRI cine imaging provides assessment of additional morphological information including systolic anterior motion of the anterior mitral leaflet with dynamic outflow tract obstruction, mitral regurgitation, apical aneurysms, myocardial clefts, and papillary muscle abnormalities. Late gadolinium enhancement imaging enables to detect presence of myocardial fibrosis which has prognostic implication.

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AIMS AND OBJECTIVES: To evaluate the use and applications of Cardiac MRI in diagnosis and risk stratification of hypertrophic cardiomyopathy.

MATERIALS AND METHODS

This retrospective study was performed in our department of Radio-diagnosis in a tertiary health care institute including 23 patients referred with indication for confirmation of diagnosis of hypertrophic cardiomyopathy and risk stratification. Patients with any absolute contraindication for MRI like metal implants and pacemakers were excluded from the study. The study group patients were evaluated with clinical examinations and 2-D echocardiography and subsequently with Cardiac MRI using standard body coil and dedicated cardiac phase array coil on 3 T MRI 32 channel Siemens Verio MRI machine. ECG gating using retrospective synchronization was used for cardiac motion compensation. Respiratory gating was used for respiratory compensation. Routine dedicated cardiac protocol was done using localizers (3 plane FISP, 2-chamber, short axis and 4-chamber), dark blood axial (HASTE), Cine SSFP images (2-chamber, short axis and 4-chamber) sequences and Gadolinium enhanced study (dynamic first pass perfusion and delayed post contrast PSIR sequences). Additional imaging needed was Cine SSFP LVOT view and velocity/ VENC (velocity/flow imaging) sequences. Syngo.via VA30A analysis software was used for post processing the data. The diagnostic criteria used was maximal LV wall thickness greater than or equal to 15mm in end-diastolic phase and in asymmetric HCM, ratio of hypertrophied segment to non hypertrophied segment more than 1.5 [1]. Cardiac MRI findings used for stratification of risk of sudden cardiac death in hypertrophic cardio-myopathy are[1] (a) Late gadolinium enhancement, (b) LV maximal wall thickness of 30mm or more (c) LV dilatation with depressed ejection fraction, (d) Myocardial perfusion defect (e) LVOT gradient more than 30mmHg at rest.

RESULTS

Age and sex distribution of patients: Out of 23 patients, there were 18 males (78.26%) and 5 females (21.7%) in this study. Male patients exceeded the number of female patients in all the age groups. Patients were aged from 15 to 76 years (mean 53 years), most of them were from 41-80 years age group.

Symptoms: Dyspnoea on exertion with chest pain was the commonest presentation in our study group present in 9 patients (39.1%). Dyspnoea on exertion (with or without chest pain) was the commonest symptom present in 16 patients (69.5%).

Table 1:

SYMPTOMS	No	Percentages
Dyspnoea on exertion with chest pain	9	39.1%
Only dyspnoeal on exertion.	7	30.4%
Only chest pain	3	13.0%
Decompensated cardiogenic shock	2	8.69%
Recurrent syncope	1	4.34%
Generalized weakness and giddiness	1	4.34%

Left ventricular ejection fraction, volumes and mass estimated by cardiac MRI:

Left ventricular ejection fraction was on higher side in our study group with mean ejection fraction of 71.9%. The mean Left ventricular end diastolic volume and end systolic volumes found to be 76ml and 22.9ml respectively. Mean myocardial mass was 163.8 grams.

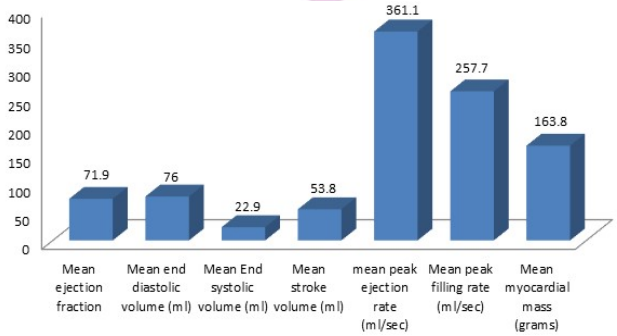


Figure 1: Left ventricular ejection fraction, volumes and mass in HCM

Table 2: Cardiac MRI findings used for risk stratification of patients with high risk of sudden cardiac death

Sr. No.	Imaging features	Number of cases	Percentage
1.	Late gadolinium enhancement suggesting presence of fibrosis	12	52.1%
2.	LVOT gradient more than 30mmHg at rest	10	43.4%
3.	LV maximal wall thickness of 30mm or more	3	13.0%
4.	LV dilatation with depressed ejection fraction	0	00%
5.	Myocardial perfusion defect	0	00%

Additional findings

Systolic anterior motion was found in 47.8% (11 patients) cases and mitral regurgitation also found in 47.8% (11 patients) cases.

Phenotypes

In our study group asymmetrical septal hypertrophic cardiomyopathy was most common phenotype in 19 patients (82.6%) followed by apical type in 4 patients (17.3%)

DISCUSSION

Hypertrophic cardiomyopathy (HCM) is the most frequent genetic cardiac disease. With excellent spatial resolution and border definition, CMR provides complete visualization of the LV chamber, allowing precise localization of the distribution of hypertrophy and measurement of wall thickness and cardiac mass[2]. CMR is also superior to echocardiography in the detection of apical and focal basal antero-septal variants and in recognizing non-contiguous areas of hypertrophy[3]. CMR cine imaging provides assessment of additional morphological information including systolic anterior motion of the anterior mitral leaflet with dynamic outflow tract obstruction, mitral regurgitation, apical aneurysms, myocardial clefts, and papillary muscle abnormalities. This study includes 23 patients referred for confirmation of HCM and risk stratification. We analysed the cardiac MR imaging findings of 23 cases of hypertrophic cardiomyopathy with respect to following parameters: Left ventricular ejection fraction, volumes, mass. Risk stratification done using presence or absence of late gadolinium enhancement, maximum wall thickness more than 30mm, LV dilatation with depressed ejection fraction, Myocardial perfusion defect and LVOT gradient more than 30mmHg[1]. We also analysed clinical features, age and gender distribution of the hypertrophic cardiomyopathy.

Age and sex distribution:

In our study hypertrophic cardiomyopathy affected middle and old aged patients most commonly with a mean age of 53 years. It was more common in males (78.2%). In study conducted by to Rickers C et al.[3], mean age was 34+/-16, range 8 to 69 and 71% patients were males.

Symptoms:

In our study most common symptom was dyspnoea on exertion (69.5 % - 16 patients), followed by chest pain (52.1 %- 12 patients). Dyspnoea on exertion with chest pain was the commonest presentation (39.1%- 9 patients), followed by only dyspnoea on exertion (30.4%- 7 patients) and only chest pain (13%- 3 patients). Chun EJ et al.[1] in their study mentioned dyspnoea with exertion as most common symptom.

Left ventricular ejection fraction, volumes and mass estimated by cardiac MRI in HCM:

Left ventricular ejection fraction was on higher side in our study group with mean ejection fraction of 71.9%. The mean Left ventricular end diastolic volume and end systolic volumes found to be 76ml and 22.9ml respectively. Mean myocardial mass was 163.8 grams. Mean ejection fraction was on higher side.

Cardiac MRI findings used for risk stratification of patients with high risk of sudden cardiac death:

a. Late gadolinium enhancement (LGE) suggesting presence of fibrosis:

In our study late gadolinium enhancement was present in 52% (12 patients) cases of HCM. Study conducted by *Chun EJ et al.*[1] shows late gadolinium enhancement in 80% cases of HCM. *He D et al.*[7] and *Briasoulis et al.*[8] in their study showed that LGE is significantly associated with sudden cardiac death.

b. LVOT gradient more than 30mmHg at rest:

In our study LVOT gradient more than 30mmHg at rest was present in 43.4% (10 patients) cases. *Maron MS et al.*[9] in their study showed that patients with LVOT obstruction (defined as a basal gradient of ≥30 mm Hg) have four times increased risk of sudden cardiac death from HCM or progression to severe congestive symptoms.

c. LV maximal wall thickness of 30mm or more:

In our study LV maximal wall thickness of 30mm or more was present in 13% (3 patients) cases of HCM. *Chun EJ et al.* [(1)] and *Brenes JC et al.*[6] in their study mentioned maximum LV wall thickness of 30mm or more as strong predictor of the risk of sudden cardiac death in patients with HCM.

d. LV dilatation with depressed ejection fraction:

In our study there was no any patient with LV dilatation with depressed ejection fraction in cases of HCM. *Chun EJ et al.*[1] in their study mentioned the unfavourable prognosis in patients of HCM who developed ventricular dilatation and depressed ejection fraction called as Burned out phase.

e. Myocardial perfusion defect:

In our study there was no patient with myocardial perfusion defect in cases of HCM. *Chun EJ et al.*[1] in their study mentioned that the identification of myocardial ischemia in patients with HCM is a powerful independent predictor of cardiovascular mortality. *Maron MS et al.*[9] in their study mentioned that stress perfusion MR imaging now permits accurate qualitative and quantitative assessment of myocardial blood flow at rest and during pharmacologic stress, with superior spatial resolution to that of PET.

Phenotype:

In our study most common phenotype was asymmetrical septal hypertrophic cardiomyopathy in 82.6% (19 patients)

cases. *Brenes JC et al.*[6] in their study mentioned asymmetrical septal hypertrophic cardiomyopathy as most common phenotype in 70% patients.

CONCLUSION

MRI is an excellent, non-invasive, radiation free precise imaging modality with multi-planar capabilities and excellent soft tissue delineation. With excellent spatial resolution and border definition, Cardiac MRI provides complete visualization of the LV chamber, allowing precise localization of the distribution of hypertrophy and measurement of wall thickness and cardiac mass. Because of availability of late gadolinium enhancement imaging, velocity encoding imaging and accurate measurement of ventricular wall thickness CMR is very useful in risk stratification in cases of HCM.

Case 1. Asymmetric septal Hypertrophic Cardiomyopathy:

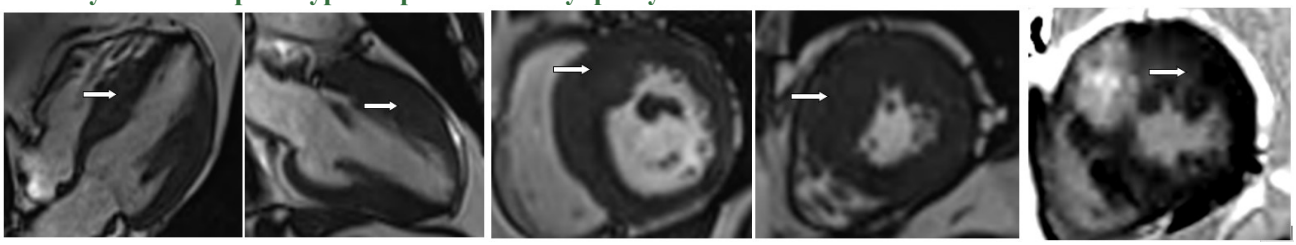


Figure 2: (a)SSFP-4 Chamber, (b) 2 Chamber, (c) Short axis mid cavity region and (d) short axis apical region show asymmetrical concentric hypertrophy of mid and apical portions of LV. (e) Post contrast short axis PSIR image shows trans mural LGE in antero-septal segment indicating poor prognosis.

Case 2. Asymmetric septal Hypertrophic Cardiomyopathy

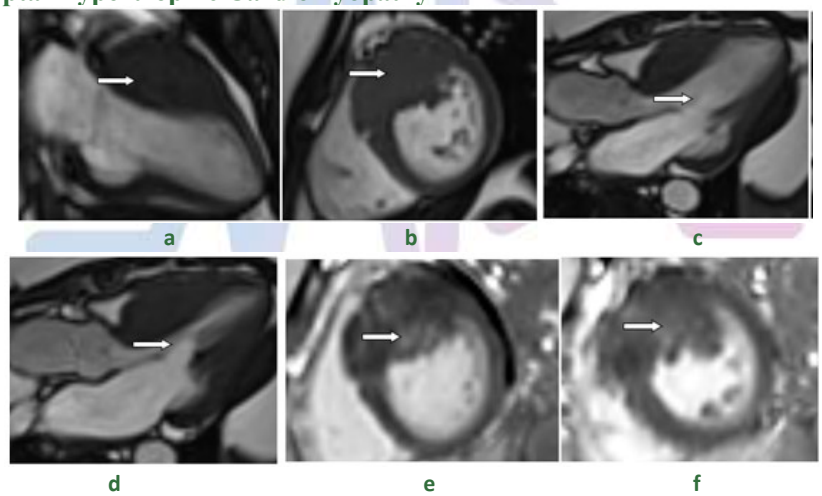


Figure 3: (a) and (b) SSFP-4 Chamber and short axis views show asymmetrical hypertrophy of the antero-septal and anterior segments of basal and mid portions of LV. (c) and (d) SSFP-3 Chamber views show SAM of anterior mitral leaflet causing LVOT compromise. (e) And (f) Post contrast SA PSIR images show patchy LGE in hypertrophied segments indicating poor prognosis.

Case 3. Asymmetric septal hypertrophic cardiomyopathy

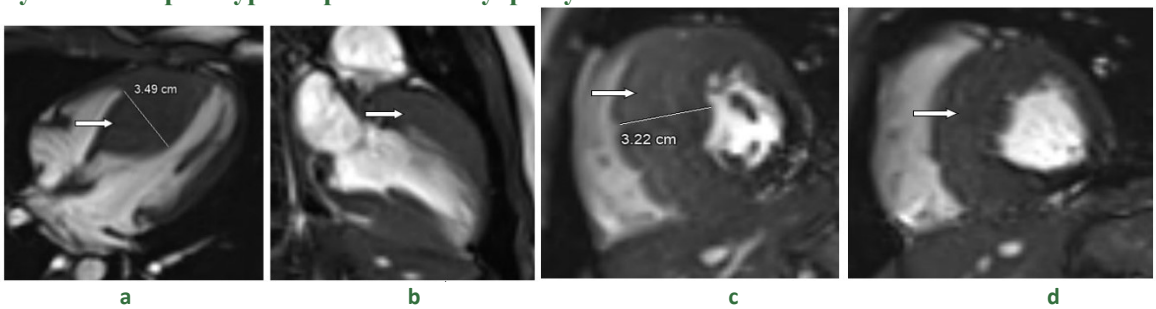
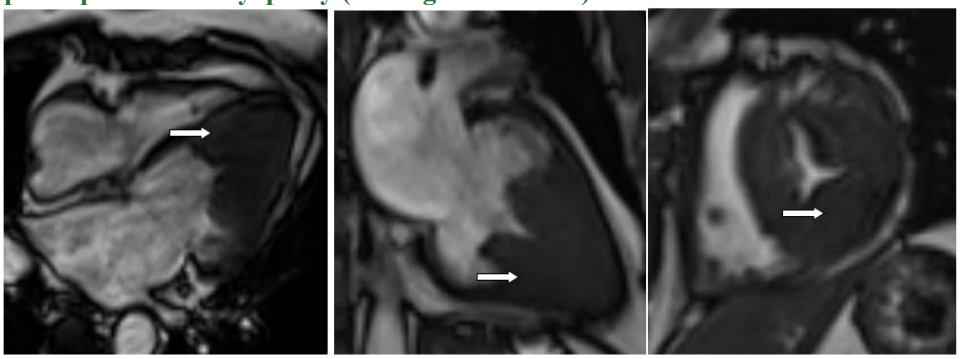


Figure 4: (a)SSFP-4 Chamber, (b) 2 Chamber, (c) Short axis mid cavity region and (d) short axis apical region show asymmetrical septal hypertrophy of mid and apical portions of LV. LV maximal wall thickness of 30mm or more indicating poor prognosis.

Case 6. Apical hypertrophic cardiomyopathy (Yamaguchi variant)



a b c

Figure 4: (a)SSFP-4 Chamber, (b) 2 Chamber, (c) short axis apical region show apical type of hypertrophy of LV with spade like configuration of the LV cavity in end diastole.

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