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PICTORIAL ESSAY

Magnetic Resonance Imaging of Hypertrophic Cardiomyopathy

KH Tsang[1], WSW Chan[2], CK Shiu[1], JCY Lee[1], MK Chan[1]

[1]Department of Radiology and Imaging, Queen Elizabeth Hospital, Jordan, Hong Kong
[2]Department of Radiology and Organ Imaging, United Christian Hospital, Kwun Tong, Hong Kong

ABSTRACT

Hypertrophic cardiomyopathy is the most common genetically inherited cardiac disorder. It is characterised by a diffuse or segmental left ventricular hypertrophy. Its diagnosis is based largely on magnetic resonance imaging owing to its precise determination of myocardial anatomy for phenotype classification and risk stratification. This study discusses different phenotypes of hypertrophic cardiomyopathy on magnetic resonance imaging.

Key Words: *Cardiomyopathy, hypertrophic; Heart; Magnetic resonance imaging*

中文摘要

肥厚性心肌病的磁共振成像

曾劍鴻、陳施媛、蕭俊傑、李俊賢、陳文光

肥厚性心肌病（HCM）是最常見的基因遺傳心臟疾病。其特徵為擴散性或節段性左心室肥大，其診斷主要基於磁共振成像（MRI），因其能精確測定心肌解剖並作表型分類和風險分層。本文討論HCM在MRI上的不同表型。

INTRODUCTION

Hypertrophic cardiomyopathy (HCM) is the most common genetically inherited cardiac disorder with an autosomal dominant inheritance.[1-3] It is characterised by a diffuse or segmental hypertrophy of the left ventricle (LV), usually in the absence of other cardiac or systemic disease that leads to myocardial hypertrophy.[1] The main histological features are myocyte and myofibrillar disarray.[4,5] Diagnosis of HCM is based largely on magnetic resonance imaging (MRI), which can provide precise determination of the myocardial anatomy for phenotype classification and risk stratification (Figure 1). Delayed gadolinium-enhanced MRI of the myocardium shows tissue characterisation, specifically for identification of myocardial scar or fibrosis,[1,3] which is important in risk stratification. This study discusses different phenotypes of HCM on MRI.

PHENOTYPES

The incidence of HCM is about 0.2%.[6] Clinical presentation varies from asymptomatic to dyspnoea, chest pain, syncope, or sudden cardiac death. The usual diagnostic criterion for HCM is an LV wall thickness of ≥15 mm measured in the end-diastolic phase (Figure

Correspondence: *Dr KH Tsang, Department of Radiology and Imaging, Queen Elizabeth Hospital, Jordan, Hong Kong.*
Email: tsang_kh@yahoo.com.hk

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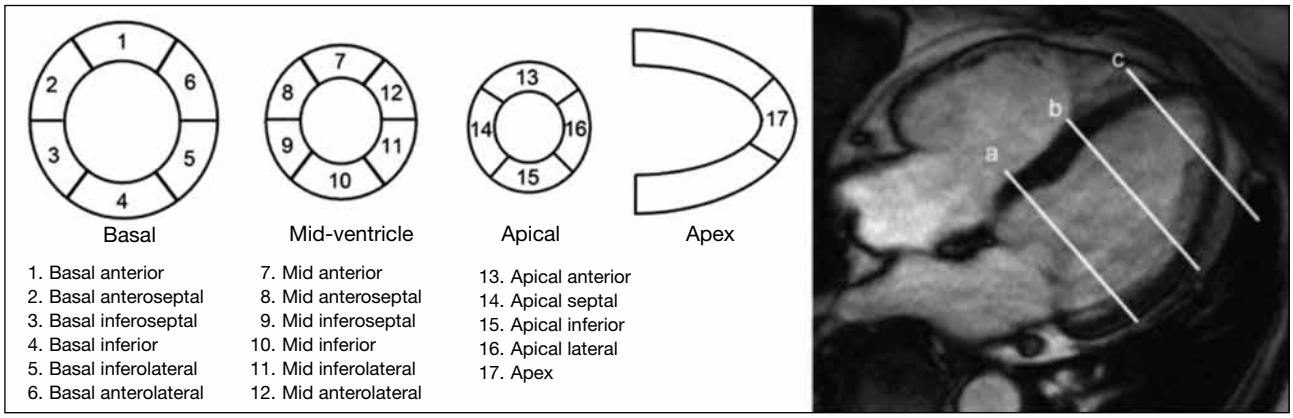

Figure 1. 17 cardiac segments obtained in short-axis view of the left ventricle. Lines a, b, and c correspond to basal ventricular level, mid-ventricular level, and apical level, respectively.

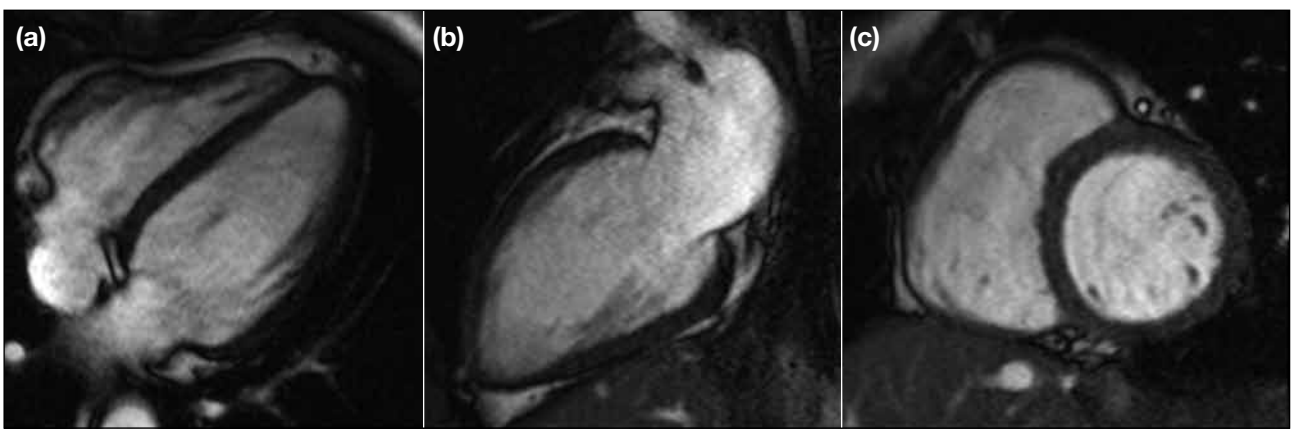

Figure 2. Normal left ventricular myocardial wall thickness at the end-diastolic phase in the (a) 4-chamber view, (b) 2-chamber view, and (c) vertical short-axis view.

2).[1-3] Phenotypes are widely variable and heterogeneous; different segments of the LV can be affected.

Asymmetrical (septal) HCM is most common, accounting for 60% to 70% of all HCM.[1,3,7] Hypertrophy of the anteroseptal LV myocardium is the most common pattern (Figure 3). The diagnosis is made when the septal wall thickness is ≥15 mm or when the ratio of the septal wall thickness to the thickness of the inferior wall of the LV is >1.5 at the mid-ventricular level.[1] Distinguishing the obstructive from non-obstructive forms is clinically important. The altered haemodynamic force along the LV outflow tract causes systolic anterior motion of the anterior mitral valve leaflet and the resulting leaflet-septal contact and obstructive physiology.[8] Concomitant mitral regurgitation with the regurgitant jet directed posteriorly into the left atrium occurs secondary to incomplete leaflet apposition.

Apical HCM is characterised by myocardial hypertrophy predominantly affecting the LV apex. This results in a spade-like configuration of the LV cavity on vertical long-axis view at the end-diastolic phase (Figure 4). The diagnosis is made when the apical wall thickness is >15 mm or the ratio of apical to basal LV wall thickness is 1.3 to 1.5.[1] This variant is seen more frequently in Japanese than in Western populations (about 25% vs. 2% of all HCM).[1,3,9] It is frequently complicated by hypertension but rarely associated with sudden cardiac death.[10]

Concentric (symmetrical) HCM accounts for 42% of all HCM,[1,3] and is characterised by concentric or symmetrical LV hypertrophy in the absence of

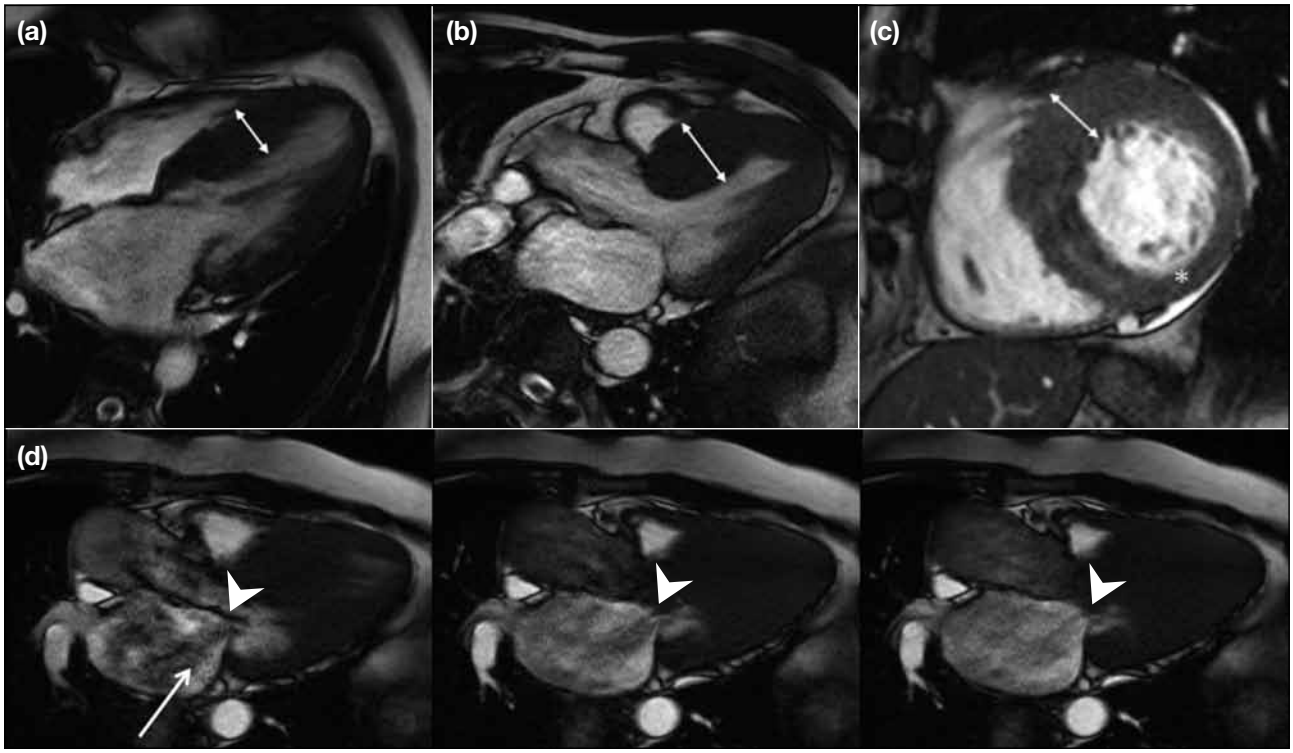

Figure 3. Asymmetrical (septal) hypertrophic cardiomyopathy in the (a) 4-chamber view, (b) 3-chamber view, (c) vertical short-axis view, and (d) cine steady-state free precession obtained in the 3-chamber view showing asymmetrical septal wall thickening up to 20 mm (double-headed arrows). The ratio of the septal thickness to the thickness of the inferior wall of the left ventricle (asterisk) at the mid-ventricular level is >1.5. Systolic anterior motion of the anterior mitral valve leaflet (arrowheads) results in narrowing of the subaortic left ventricular outflow tract with turbulence flow and mitral regurgitant flow (arrow).

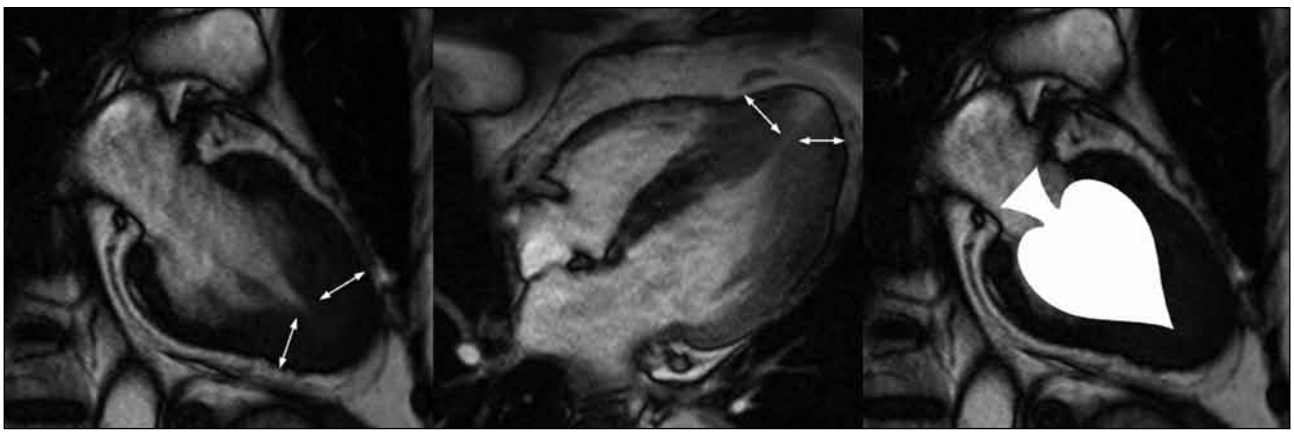

Figure 4. Apical hypertrophic cardiomyopathy with thickening of the apical myocardial wall up to 18 mm (double-headed arrows) and a spade-shaped left ventricular cavity. The apical wall thickness is best measured at a long-axis view to prevent foreshortening.

secondary causes (Figure 5). The myocardial thickness is >15 mm and is less commonly seen in other causes that result in diffuse hypertrophy such as hypertensive disease. The LV cavity dimension is reduced in a concentric manner.

Mid-ventricular HCM is a rare variant of asymmetric HCM and is characterised by hypertrophy of the middle third of the LV wall (Figure 6), with mid–LV cavity narrowing with a dumbbell configuration.[3,11] In severe cases, it is associated with an apical dilatation

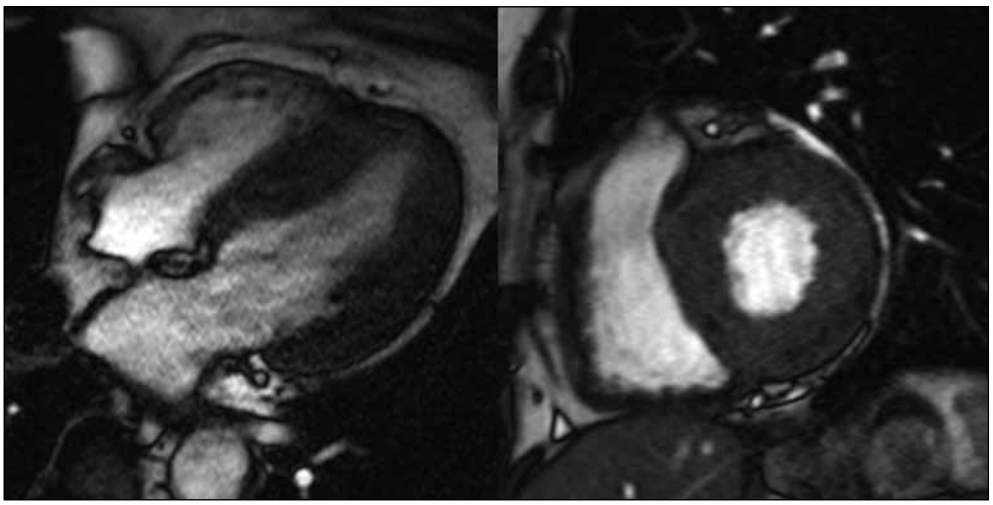

Figure 5. Concentric (symmetrical) hypertrophic cardiomyopathy with diffuse myocardial wall thickening without segmental preference.

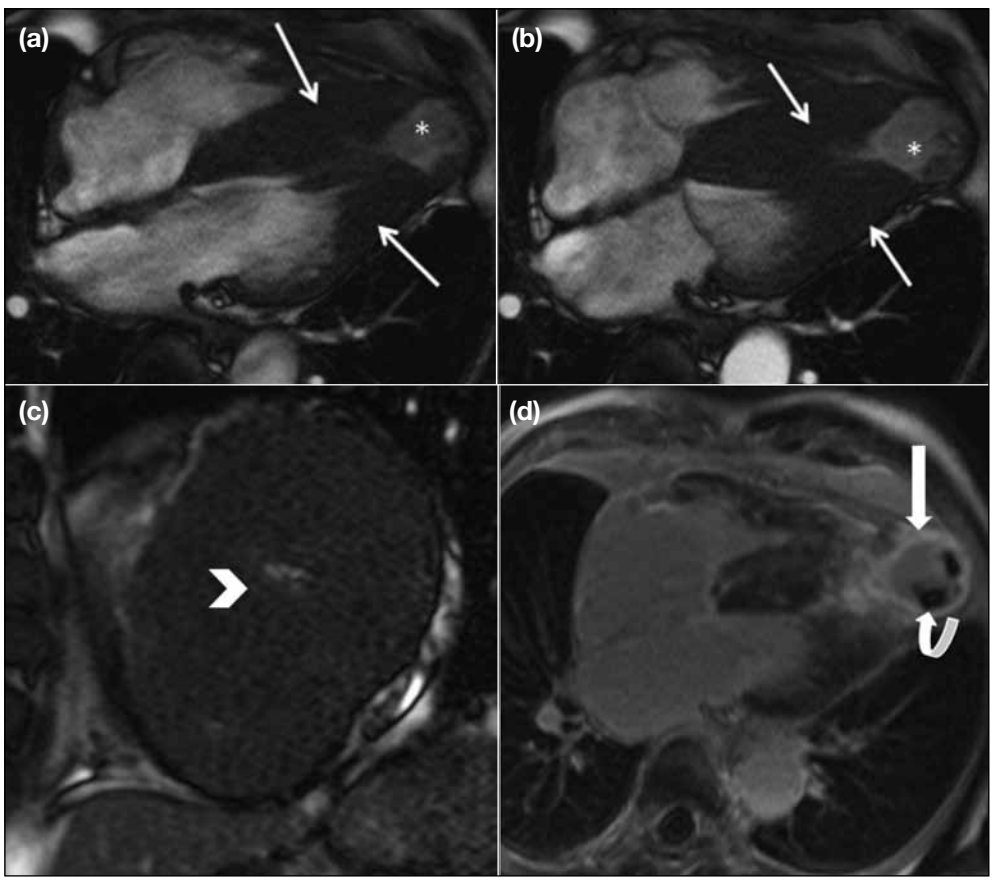

Figure 6. Mid-ventricular hypertrophic cardiomyopathy with an apical aneurysm in the (a) 4-chamber view at the end-diastolic phase, (b) 4-chamber view at the systolic phase, (c) vertical short-axis view at the mid-ventricular level at the end-systolic phase, and (d) late gadolinium enhancement long-axis view showing marked mid-ventricular wall thickening compatible with hypertrophic cardiomyopathy (thin arrows), apical wall thinning with aneurysm formation (asterisks), marked narrowing at the mid–left ventricular cavity (arrowhead) resulting in mid-ventricular obstruction with increased systolic pressure at the apex, and progressive apical dilatation and ischaemia resulting in burned-out apex with transmural delayed enhancement, characteristic of myocardial scar or fibrosis (thick arrow), with apical thrombi (curved arrow).

secondary to increased systolic pressure in the apex as a result of mid-ventricular obstruction.[1] In about 10% of patients, there is progression to a burned-out apex secondary to ischaemia with apical aneurysm formation and late myocardial enhancement after gadolinium administration.[12]

Mass-like HCM is due to the focal segmental location

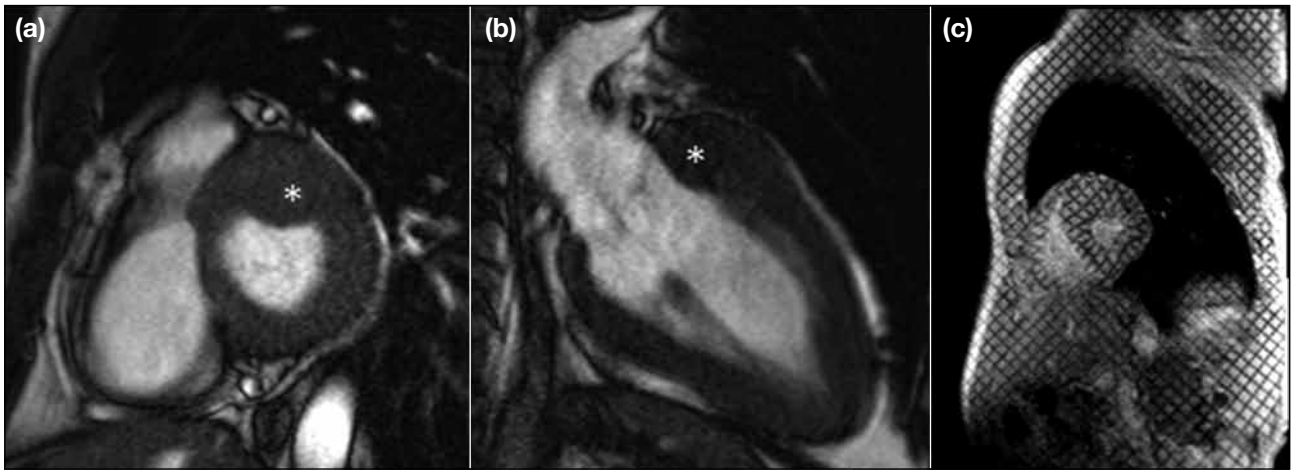

Figure 7. Mass-like hypertrophic cardiomyopathy in the (a) basal vertical short-axis view, (b) 2-chamber view, and (c) basal short-axis view with myocardial tagging technique showing a mass-like appearance of the focal myocardial wall thickening in the basal anterior segment (asterisks). It can be readily differentiated from myocardial neoplasm by the tagging technique, which shows contractility within the hypertrophied segment.

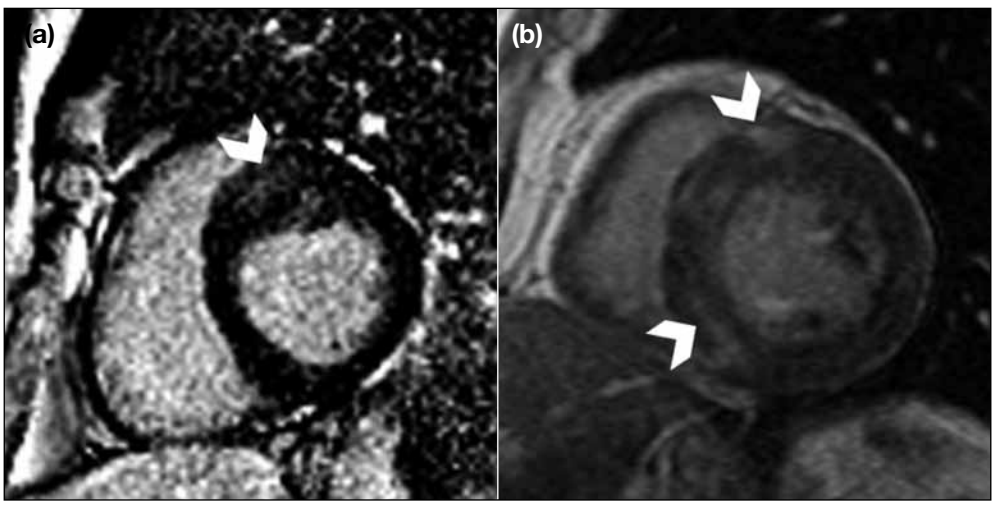

Figure 8. Hypertrophic cardiomyopathy in delayed enhancement images in short-axis views showing enhancement (arrowheads) that is typically of mid-wall distribution and punctate or patchy, with fibrosis mainly in the (a) anteroseptal segment or (b) along the whole septal wall.

of the myocardial disarray and fibrosis (Figure 7).[12] It is differentiated from a genuine myocardial mass by its homogeneous signal intensity and perfusion signal parallel to the adjacent normal myocardium. The myocardial tagging technique shows contractility in the hypertrophied segment, which is absent in myocardial neoplasm.[13]

DISCUSSION

Myocardial fibrosis or scarring can be detected by cardiac MRI in 33% to 86% of patients with HCM.[1,3,14,15] As opposed to subendocardial distribution of late gadolinium enhancement in myocardial infarcts, the pattern of late gadolinium enhancement in HCM usually appears as small punctate, patchy hyperenhancement with a mid-wall distribution (Figure 8). The regional location of late gadolinium enhancement within the hypertrophied segment is suggestive of underlying HCM.

Risk factors that can be detected by cardiac MRI include myocardial wall thickness ≥30 mm, presence of LV outflow tract obstruction or systolic anterior motion, presence of wall fibrosis, myocardial wall perfusion

defect, LV dilatation, and decreased ejection fraction.

Between January 2010 and May 2015 at Queen Elizabeth Hospital in Hong Kong, 29 men and 15 women aged 36 to 85 (mean, 63) years were diagnosed by MRI with septal HCM (n = 31), apical HCM (n = 6), symmetrical HCM (n = 5), mid-ventricular HCM (n = 1), and mass-like HCM (n = 1). The mean hypertrophied LV wall thickness was 20.1 (standard deviation, 4.2; range, 15-36) mm. Most patients had mid-wall fibrosis, probably owing to old age. Systolic anterior motion of the anterior mitral valve leaflet was identified in 10 of the 31 patients with septal HCM. Late mid-myocardial gadolinium enhancement was identified in the hypertrophied segments in 41 patients.

CONCLUSION

Cardiac MRI is useful for diagnosing and risk stratification of HCM. Familiarisation with its MRI appearance can help make a more accurate diagnosis.

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