Usefulness of Delayed Enhancement Magnetic Resonance Imaging to Differentiate Dilated Phase of Hypertrophic Cardiomyopathy and Dilated Cardiomyopathy

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ABSTRACT

Background: The dilated phase of hypertrophic cardiomyopathy (HCM) has a poor prognosis. For correct recognition of such patients, we compared the findings in cardiac delayed enhancement (DE)-magnetic resonance imaging (MRI) between HCM and dilated cardiomyopathy (DCM) patients.

Methods and Results: Sixty-five patients (HCM 39, DCM 26) underwent gadolinium-DTPA−enhanced MRI. The HCM patients were divided into those with preserved (HCM-P, n = 30) and those with impaired systolic function (HCM-I, n = 9). DE-MRI demonstrated focal or diffuse DE at the left ventricular (LV) wall in 60% of HCM-P and 100% of HCM-I, but in only 12% of DCM. The DE distributed mainly septal to the anterior wall of LV, but the DE volume against whole LV muscle volume was much larger in HCM-I than in HCM-P and DCM (4.1 ± 6.1% in HCM-P, 14.6 ± 11.9% in HCM-I, and 0.8 ± 2.4% in DCM, means ± SD, $P < .05$). In HCM, there were weak but significant correlations between DE volume, and LV end-diastolic volume and LV end-systolic volume. In HCM-P, the percent of length shortening in the segments with DE was lower than that without DE.

Conclusions: The HCM patients had more DE than the DCM patients, and DE volume correlated to lower global and local LV function. DE-MRI may be useful to evaluate myocardial damage in HCM patients, and to differentiate the dilated phase of HCM from DCM. (*J Cardiac Fail 2007;13:372−379*)

Key Words: MRI, delayed enhancement, hypertrophic cardiomyopathy, dilated cardiomyopathy.

Hypertrophic cardiomyopathy (HCM) is a relatively common primary cardiac disease with various morphologic, functional, and clinical features.[1] The dilated phase of HCM, characterized by systolic dysfunction, is recognized as a part of HCM disease spectrum, but the clinical profile and treatment strategies in the dilated phase of HCM patients remain incompletely defined.[2] The abnormal myocardial substrate seems to be a key determinant of left ventricular (LV) impairment, consisting of interstitial fibrosis, myocardial disarray, necrosis, and scarring.[3,4] However, the clinical condition in the dilated phase of HCM resembles that in dilated cardiomyopathy (DCM) or other diseases that show LV dilation and systolic dysfunction. Therefore, if the hypertrophy was undiagnosed or underestimated during the natural course of the disease, the differential diagnosis of them becomes difficult. Because the patients with dilated phase of HCM are reported to have severe heart failure symptoms, high risk for fatal arrhythmias, and high mortality rate,[2] the early and correct recognition of those patients is necessary to determine appropriate therapeutic strategies.

Myocardial magnetic resonance imaging (MRI) is noninvasive, uses no ionizing radiation, and has high spatial resolution. Thus the value of cardiac MRI is becoming established in the assessment of cardiac function.[5,6] Recently, delayed gadolinium (Gd) enhancement (DE)-MRI is shown to be able to detect a small and focal myocardial abnormality and be useful to diagnose various cardiac diseases.[7,8] The myocardial DE visualized by DE-MRI is a common feature of HCM, and predominantly localized

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in the interventricular septum.[9–15] However, it is still unclear whether the extent of DE is related to the global and local LV dysfunction, disease progression, and clinical risk for sudden death in HCM patients. In addition, although less frequent DE was reported in DCM patients,[16–19] there was no study comparing the extent of DE between DCM and HCM.

The purpose of this study was to determine the localization and extent of DE in HCM patients, and the relationship between the DE and global and local LV function. We also compared the extent of DE between HCM and DCM patients.

Patients and Methods

Patients

We studied 65 patients (HCM 39, DCM 26) who underwent cardiac MRI between May 2003 and August 2006. HCM and DCM were diagnosed according to the World Health Organization/International Society and Federation of Cardiology definition of cardiomyopathies.[20] The diagnosis of HCM was based on echocardiographic documentation of a hypertrophied nondilated LV in the absence of other cardiac systemic diseases that could produce the magnitude of hypertrophy evident at some time during the natural course of the disease. We then divided the HCM patients into those with preserved systolic function (HCM-P, n = 30) and those with impaired systolic function (HCM-I, n = 9). The HCM-I patients were defined as LV ejection fraction lower than 45% by cine MRI, reflecting global systolic dysfunction, at the study entry. These patients were proven to have a period of hypertrophy during the natural time course of the disease. The HCM-P patients consisted of 13 cases with asymmetrical septal hypertrophy, 6 cases with apical hypertrophy, and 11 cases with concentric hypertrophy. Among them, 8 cases were diagnosed as an obstructive type of HCM.

The DCM patients were also determined when the LV contraction was globally impaired (global LV ejection fraction $<45\%$), without history or signs of myocardial infarction, severe valvular heart diseases, or LV hypertrophy. Many of these patients had fixed or reversible perfusion defects on nuclide stress perfusion scanning, but the epicardial coronaries were mostly normal in coronary angiography. Eleven age- and sex-matched normal controls were also examined to obtain the normal values for LV function. This study protocol was in accordance with the Declaration of Helsinki and approved by the institutional review board (Hamamatsu University School of Medicine, Hamamatsu, Japan), and all patients gave informed consent.

MRI Protocol

All 65 patients and 11 controls underwent electrocardiogram-gated MRI. Imaging was performed on a 1.5 Tesla (T) MR system (Signa Infinity Twinspeed, GE Medical Systems, Waukesha, Wisconsin) with a gradient system performance of maximum amplitude of 40 mT/m and slew time of 150 T·m·sec. An 8-element phased array cardiac coil was used in all studies. Typically, 3 planes such as short axis, sagittal long axis, and 4-chamber view were obtained for 2-dimensional (2D) FIESTA cine images and delayed myocardial enhancement images. 2D FIESTA cine images were based on the steady state free precession sequence, and delayed myocardial enhancement imaging was based on the inversion recovery prepared fast gradient echo (IR-FGRE) sequence. The 6 to 9 slices were used to cover whole heart. The slice thickness/gap was typically 10 mm/0 mm. The matrix was 192 × 192 for 2D FIESTA cine, and 256 × 160 for IR-FGRE. The trigger delay for IR-FGRE was 300 ms; however, for the patients with tachycardia, system-dependent trigger delay time was selected, which was shorter than 300 ms. The readout data line for IR-FGRE was 160 each, where 24 data lines were acquired per segment. Field of view was 34 cm for both sequences. Sixteen data lines were acquired per each segment, and 16 phases were acquired for 2D FIESTA cine sequences. Shortest repetition time and echo time were selected; however, the values were not exactly the same for each study, because they were related to the orientation of scanning plane and slice thickness. Sequence parameters were (2D FIESTA and IR-FGRE, respectively), flip angle: 45° and 20°; readout bandwidth: 125 kHz and 31.25 kHz. Breath-hold cine MR images were obtained in contiguous short-axis planes from the apex to the base of the heart with the patient in a resting state. After the 2D FIESTA cine images were acquired, 0.2 mmol/kg of contrast material (Gd-DTPA-BMA, Daiichi Pharma, Tokyo, Japan) was injected, and after a 15-minute delay, DE images were acquired. Before the DE images, pilot DE images were acquired with 5 different inversion times. Thus optimum inversion times for the IR-FSPGR were measured right before the DE, which were between 200 and 240 ms. This process in search of optimum contrast was concluded within 3 minutes.

Analysis of MRI Images. Two experienced cardiovascular radiologists interpreted all the MRIs without knowledge of clinical findings. LV end-diastolic volume (LVEDV), end-systolic volume (LVESV), ejection fraction (LVEF), and LV mass were acquired from the 2D FIESTA cine images in short-axis view. The values for LV volume were indexed by dividing them with body surface area (LVEDVI and LVESVI). Regional analyses of DE-MRI were performed using the 20-segments model (Fig. 1).[21] To estimate DE quantitatively, the DE area was traced manually, rendered to DE volume, and the percentage against total muscle volume was calculated. Furthermore, we examined percent length shortening (%LS) to evaluate the regional LV wall contraction. The maximum distances from the center of each slice to the endocardial margin of each LV segment at end-diastole (LD) and at end-systole (LS) were measured. Then the %LS was calculated by the

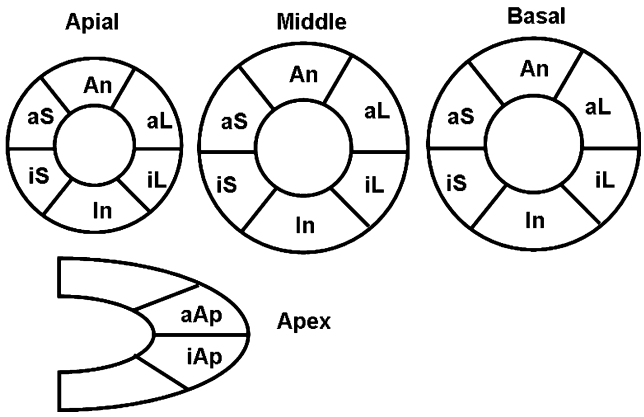

Fig. 1. Regional analyses of delayed enhancement-magnetic resonance imaging using the 20-segments model. An, anterior; aL, anterolateral; iL, inferolateral; In, inferior; iS, inferoseptal; aS, anteroseptal wall in apical, middle, and basal LV; aAP, anteroapex; iAP, inferno apex wall of the left ventricle.