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CASE REPORTS

Apical Hypertrophic Cardiomyopathy: Clinical, Electrocardiographic, Scintigraphic, Echocardiographic, and Magnetic Resonance Imaging Findings of a Case

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ABSTRACT

The apical variant of nonobstructive hypertrophic cardiomyopathy (HCM) constitutes a minority of all cases of HCM and generally carries a favorable clinical outcome. We describe a 68 year-old Caucasian woman who presented with exertional dyspnea. The patient underwent stress testing with electrocardiogram-gated single-photon emission computed tomography imaging and resting transthoracic echocardiography. The patient also underwent cardiac magnetic resonance imaging at rest, including conventional structural and functional imaging and cine complementary spatial modulation of magnetization-tagged imaging (CSPAMM). The noninvasive evaluation of the heart demonstrated apical hypertrophy with regional systolic dysfunction, establishing the diagnosis of apical HCM. This case suggests a potential value of CSPAMM in characterizing apical HCM.

Key Words: *Apical hypertrophic cardiomyopathy; Magnetic resonance imaging; CSPAMM tagging*

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INTRODUCTION

Apical hypertrophic cardiomyopathy (HCM) is an uncommon variant of nonobstructive HCM. In one of the first case series reported by Yamaguchi et al. (1), a spadelike configuration ("ace of spades") of the left ventricular cavity at end-diastole in the right anterior oblique projection of the contrast ventriculogram was described as a characteristic finding. Currently, noninvasive imaging with surface echocardiography has largely replaced cardiac catheterization for establishing the diagnosis of apical HCM. Cardiac magnetic resonance imaging (MRI) can accurately evaluate left ventricular structure and function, and its utility for diagnosis of HCM has been reported (2,3). The use of tagging to assess regional contractility in apical HCM has not been reported. We hereby report a case of apical HCM that demonstrates the utility of cine-tagged cardiac MRI using complementary spatial modulation of magnetization (CSPAMM) in this disease.

REPORT OF A CASE

A 68 year-old Caucasian woman presented with exertional dyspnea. The physical exam was unremarkable. Resting electrocardiogram (ECG) showed marked left ventricular hypertrophy with diffuse ST segment depressions and prominent T wave inversions. The patient was evaluated with a dual-isotope myocardial perfusion stress test [rest Tl-201, stress Tc-99m Sestamibi (4)], during which she exercised for 8 min and 45 sec of the Bruce protocol and stopped due to fatigue. Stress ECG was notable for diffuse ST-segment depression beyond baseline repolarization abnormalities. Single-photon emission computed tomography (SPECT) images demonstrated a normal left ventricular cavity size with no significant fixed or reversible perfusion defects. A focal increase of photon counts was observed in both stress and rest images at the apex (Fig. 1). The ECG-gated SPECT images showed preserved basal ventricular systolic function with focal severe apical hypokinesis. Two-dimensional transthoracic echocardiography demonstrated increased left ventricular apical wall thickness with regional systolic dysfunction, but visualization of the ventricular apex was suboptimal (Fig. 2). Cardiac MRI was also performed to establish the diagnosis. Dual inversion recovery and T2-weighted fast spin echo images were obtained in the 4-chamber and transverse orientations, respectively, and demonstrated focal apical hypertrophy (Fig. 3). Standard fast ECG-triggered gradient echo images (FFE-EPI) were obtained in the short axis and 4-chamber orientation. These demonstrated a normal left ventricular cavity (end-diastolic volume 120 mL) with preserved overall systolic function (ejection fraction 62%) but a severe focal wall motion abnormality at the apex. The left ventricular mass was significantly increased (197 g; 133 g/m^2). To better define the contraction of the apical myocardium and to exclude the possibility of eosinophilic deposits or thrombus formation at the apical left ventricular cavity, we performed breathhold two-dimensional cine CSPAMM-tagged cardiac MRI (FOV: 250 mm, matrix: 44×128, slice thickness: 8 mm, tag line spacing: 8 mm, TE 6.5 msec, EPI factor 11, temporal resolution 38 msec) (5). This technique

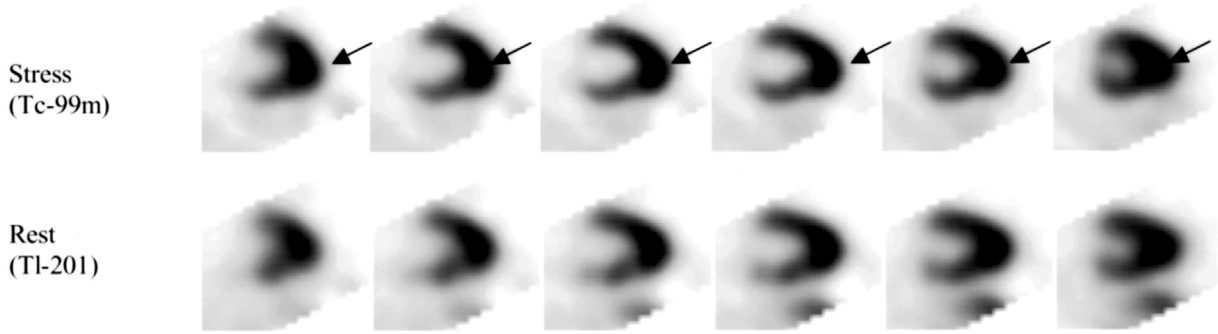

Figure 1. The SPECT images of the left ventricle in the vertical long axis orientations at stress and rest. The left ventricular cavity size is normal, and there are no perfusion defects in either stress or rest images. A focal increase of photon counts is seen at the left ventricular apex (arrows).

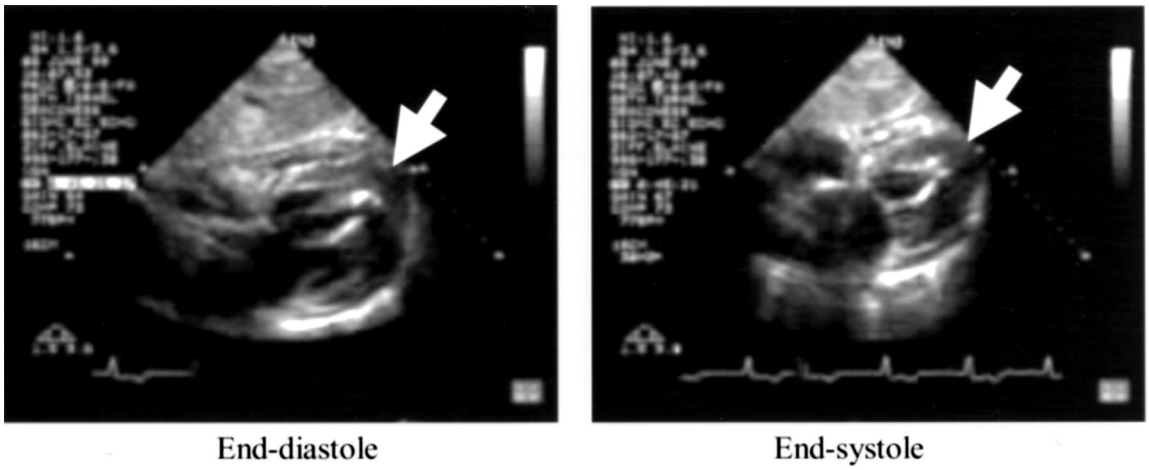

Figure 2. Echocardiographic images of the left ventricle from the subcostal view. Image quality is suboptimal, a common limitation of echocardiography, particularly for evaluation of the apical left ventricle. Focal apical hypertrophy with regional hypokinesis can be identified (arrows).

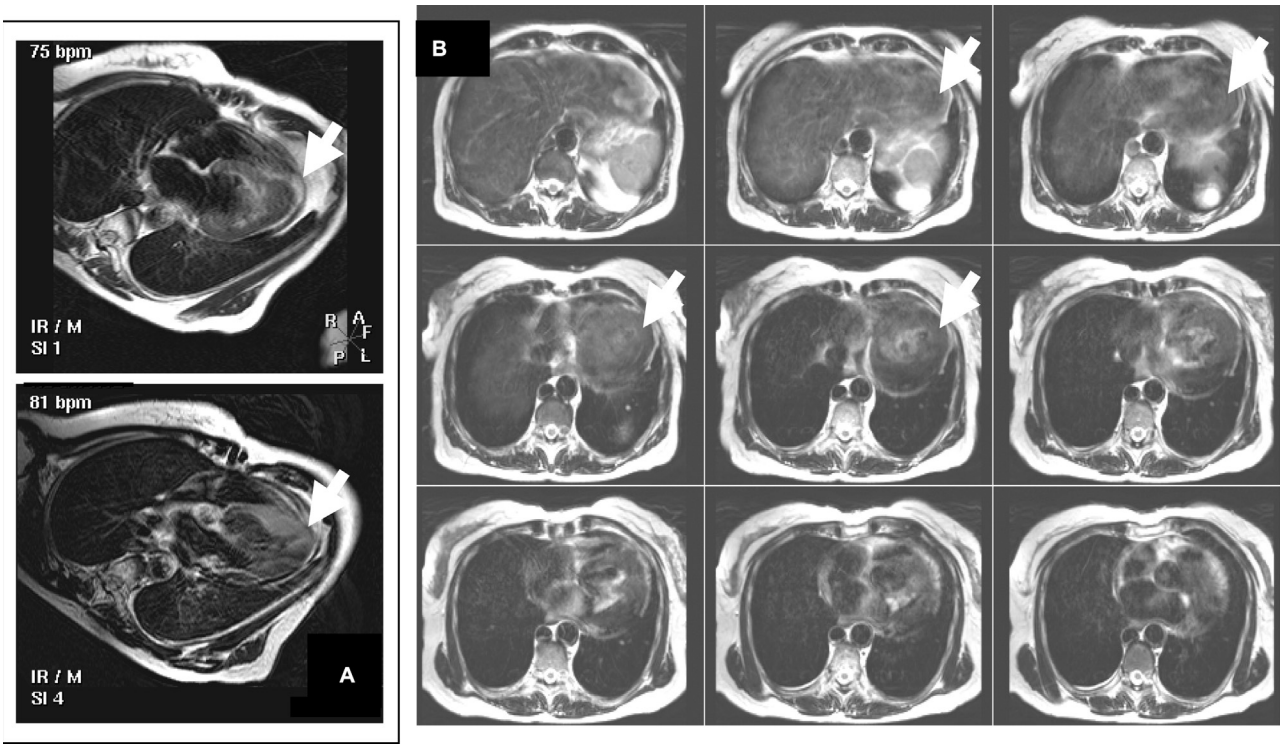

Figure 3. (A) Dual inversion spin-echo images (TI: 330 msec, TE: 100 msec, TR: 1500 msec, turbo factor: 31) of the left ventricle in the 4-chamber orientation. (B) Contiguous transverse T2-weighted fast spin echo images (TE: 120 msec, TR: 2250 msec, turbo factor: 18), covering the entire heart. With both imaging approaches, focal apical hypertrophy (arrows) can be readily identified.

can visualize the endocardial and epicardial deformation of a single slice of myocardium during both systole and diastole, extending into late diastolic phases. The visual assessment of the tagged images confirmed the diagnosis of apical HCM with corresponding apical systolic dysfunction (Fig. 4).

DISCUSSION

The HCM has a heterogeneous genetic etiology and variable phenotypic expression, with a prevalence of approximately 1/500 in the general population (6). The apical variant of HCM was initially described in Japan by Sakamoto et al. (7), and later by Yamaguchi et al. (1). Apical HCM constitutes ~25% of all cases of HCM in Japan (1) but ≤2% of cases in Western countries (8,9). Patients with apical HCM may present with typical or atypical angina, dyspnea, palpitations, or may be asymptomatic (8,10). Although apical HCM is thought to have a benign clinical course (10,11), some reports suggest significant functional limitations in patients with this disease (8).

The presence of giant negative T waves in the precordial leads on the 12-lead ECG may raise initial suspicion for apical HCM, but is neither a highly sensitive, nor a highly specific diagnostic marker (8). Scintigraphy also can detect localized apical hypertrophy (12,13), and ECG-gated SPECT may be of additional value by demonstrating focal apical systolic dysfunction, as shown in the case that we present. Two-dimensional transthoracic echocardiography is usually diagnostic (1,6–8), but may be inadequate in patients with poor acoustic windows. Cardiac MRI allows for the noninvasive evaluation of cardiac structure and function with exquisite accuracy, and its utility in assessing HCM has been reported (2,3,14–16). Although there are no studies documenting the superiority of a certain imaging modality in establishing the diagnosis of apical HCM, two reports have suggested that cardiac MRI provides a more complete morphologic assessment of the left ventricular apex in patients with HCM compared with two-dimensional transthoracic echocardiography (2,3). The identification of a focal region of myocardial hypertrophy readily distinguishes HCM from other cardiomyopathies with globally increased wall thickness, including hypertensive, amyloid, postobstructive, etc. Cine-tagged MRI may have additional utility by demonstrating the regional contractility of the hypertrophied apex. Though other conditions (such as apical thrombi or eosinophilic deposits in eosinophilic cardiomyopathy) may provide a similar anatomic image of "hypertrophied" apex, tagged MR imaging can demonstrate the endocardial and epicardial apical contraction, thereby providing tissue characterization through functional imaging. An alternative approach would be with Gd-enhancement, in which the normally perfused myocardium would enhance, as opposed to nonvascular mural tissues.

The quantitation of tagging parameters in apical HCM has not been described. As hypertrophy may involve the

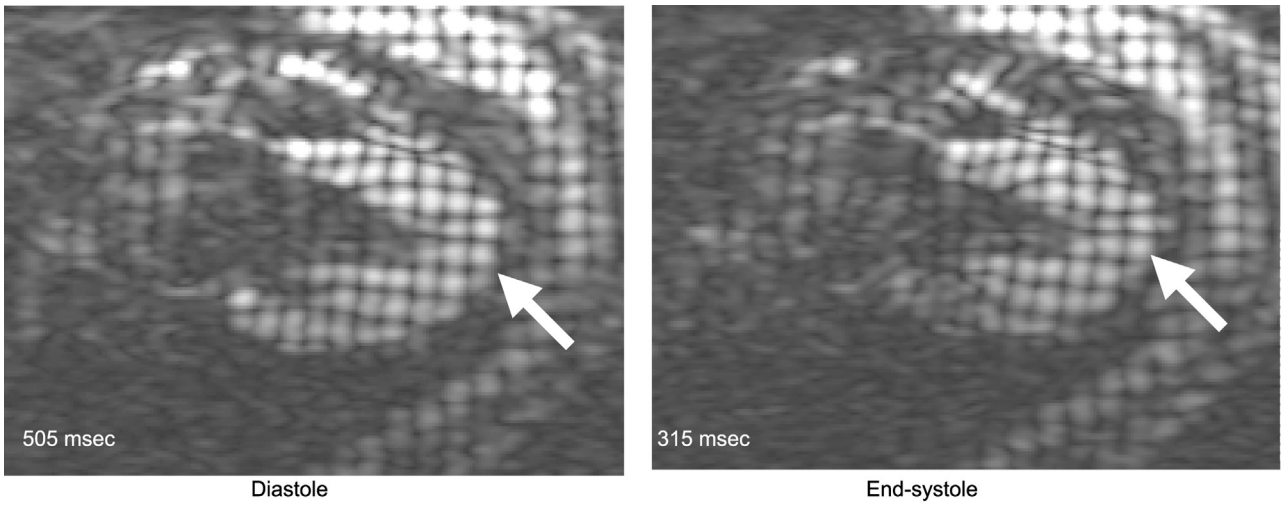

Figure 4. Diastolic (left panel) and systolic (right panel) images of the heart in a 4-chamber view. Images were acquired during a single breath hold, using cine CSPAMM MRI. A late diastolic phase (rather than the end-diastolic phase) is presented to allow visualization of the left ventricular cavity. The absence of distortion of the tag lines at the left ventricular apex confirms the abnormal contractile pattern at the area of focal apical hypertrophy (arrows).

left ventricular walls at different levels and the degree of hypertrophy and systolic dysfunction varies widely among patients, it would be very difficult to generalize findings from any single patient. The visual evaluation, and particularly in the 4-chamber orientation, planned so as to include the hypertrophied left ventricular segment, can have significant diagnostic value.

In the case that we present, findings from ECG, ECG-gated SPECT, echocardiography, and cardiac MRI all corroborated the diagnosis of apical HCM. Cardiac MRI has unique advantages, as it can provide diagnostic information in anatomy, structure, global, and regional function and contraction pattern in patients with suspected HCM and particularly those with the apical HCM variant.

ABBREVIATIONS

CSPAMM	complementary spatial modulation of magnetization
ECG	electrocardiogram
HCM	hypertrophic cardiomyopathy
MRI	magnetic resonance imaging
SPECT	single-photon emission computed tomography

REFERENCES

1. Yamaguchi, H.; Ishimura, T.; Nishiyama, S.; Nagasaki, F.; Nakanishi, S.; Takatsu, F.; Takatsu, F.; Nishijo, T.; Umeda, T.; Machii, K. Hypertrophic Nonobstructive Cardiomyopathy with Giant Negative T Waves (Apical Hypertrophy): Ventriculographic and Echocardiographic Features in 30 Patients. Am. J. Cardiol. **1979**, *44*, 401–412.
2. Suzuki, Y.; Kadota, K.; Nohara, R.; Tamaki, S.; Kambara, H.; Yoshida, A.; Murakami, T.; Osakada, G.; Kawai, C.; Tamaki, N.; Mukai, T.; Torizuka, K. Recognition of Regional Hypertrophy in Hypertrophic Cardiomyopathy Using Thallium-201 Emission-Computed Tomography: Comparison with Two-Dimensional Echocardiography. Am. J. Cardiol. **1984**, *53*, 1095–1102.
3. Higgins, C.B.; Byrd, B.F., 3rd; Stark, D.; McNamara, M.; Lanzer, P.; Lipton, M.J.; Schiller, N.B.; Botvinick, E.; Chatterjee, K. Magnetic Resonance Imaging in Hypertrophic Cardiomyopathy. Am. J. Cardiol. **1985**, *55*, 1121–1126.
4. Stuber, M.; Spiegel, M.A.; Fischer, S.E.; Scheidegger, M.B.; Danias, P.G.; Pedersen, E.M.; Boesiger, P. Single Breath-Hold Slice-Following CSPAMM Myocardial Tagging. Magma **1999**, *9*, 85–91.
5. Berman, D.S.; Kiat, H.; Friedman, J.D.; Wang, F.P.; van Train, K.; Matzer, L.; Maddahi, J.; Germano, G. Separate Acquisition Rest Thallium-201/Stress Technetium-99ms Estamibi Dual-Isotope Myocardial Perfusion Single-Photon Emission Computed Tomography: A Clinical Validation Study. J. Am. Coll. Cardiol. **1993**, *22*, 1455–1464.
6. Maron, B.J.; Gardin, J.M.; Flack, J.M.; Gidding, S.S.; Kurosaki, T.T.; Bild, D.E. Prevalence of Hypertrophic Cardiomyopathy in a General Population of Young Adults. Echocardiographic Analysis of 4111 Subjects in the CARDIA Study. Coronary Artery Risk Development in (Young) Adults. Circulation **1995**, *92*, 785–789.
7. Sakamoto, T.; Tei, C.; Murayama, M.; Ichiyasu, H.; Hada, Y. Giant T Wave Inversion as a Manifestation of Asymmetrical Apical Hypertrophy (AAH) of the Left Ventricle. Echocardiographic and Ultrasono-Cardiotomographic Study. Jpn Heart J. **1976**, *17*, 611–629.
8. Louie, E.K.; Maron, B.J. Apical Hypertrophic Cardiomyopathy: Clinical and Two-Dimensional Echocardiographic Assessment. Ann. Intern. Med. **1987**, *106*, 663–670.
9. Maron, B.J. Apical Hypertrophic Cardiomyopathy: the Continuing Saga. J. Am. Coll. Cardiol. **1990**, *15*, 91–93.
10. Webb, J.G.; Sasson, Z.; Rakowski, H.; Liu, P.; Wigle, E.D. Apical Hypertrophic Cardiomyopathy: Clinical Follow-Up and Diagnostic Correlates. J. Am. Coll. Cardiol. **1990**, *15*, 83–90.
11. Koga, Y.; Itaya, K.; Toshima, H. Prognosis in Hypertrophic Cardiomyopathy. Am. Heart J. **1984**, *108*, 351–359.
12. Suzuki, Y.; Kadota, K.; Nohara, R.; Tamaki, S.; Kambara, H.; Yoshida, A.; et al. Recognition of Regional Hypertrophy in Hypertrophic Cardiomyopathy Using Thallium-201 Emission-Computed Tomography: Comparison with Two-dimensional Echocardiography. Am. J. Cardiol. **1984**, *53*, 1095–1102.
13. Reddy, V.; Korcarz, C.; Weinert, L.; Al-Sadir, J.; Spencer, K.T.; Lang, R.M. Apical Hypertrophic Cardiomyopathy. Circulation **1998**, *98*, 2354.
14. Higgins, C.B.; Byrd, B.F., 3rd. ; Stark, D.; McNamara, M.; Lanzer, P.; Lipton, M.J.; et al. Magnetic Resonance Imaging in Hypertrophic Cardiomyopathy. Am. J. Cardiol. **1985**, *55*, 1121–1126.
15. Ibrahim, T.; Schwaiger, M. Diagnosis of Apical Hypertrophic Cardiomyopathy Using Magnetic Resonance Imaging. Heart **2000**, *83*, E1.
16. Park, J.H.; Kim, Y.M.; Chung, J.W.; Park, Y.B.; Han, J.K.; Han, M.C. MR Imaging of Hypertrophic Cardiomyopathy. Radiology **1992**, *185*, 441–446.

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