VIA MEDICA

CASE REPORT

Cardiology Journal
2010, Vol. 17, No. 3, pp. 306–311

ISSN 1897–5593

Dual-single photon emission computed tomography and contrast-enhanced magnetic resonance imaging to evaluate dissimilar features of apical hypertrophic cardiomyopathy

Satoshi Okayama, Hiroyuki Kawata, Ji Hee Sung, Sadanori Okada, Taku Nishida, Kenji Onoue, Tsunenari Soeda, Shiro Uemura, Yoshihiko Saito

Nara Medical University, First Department of Internal Medicine, Nara, Japan

Abstract

Apical hypertrophic cardiomyopathy (HCM) is an uncommon variant of HCM characterized by hypertrophy located in the left ventricular apex that occurs at a rate of about 30% in the Japanese population.

Although the prognosis of most patients with apical HCM is relatively benign, it can be poor if apical left ventricular aneurysms develop. However, the mechanism of aneurysmal formation is unclear. We describe two patients with apical HCM and dissimilar findings in ^{201}Thallous chloride (^{201}TlCl) and ^{123}I-betamethyl-p-iodophenyl-pentadecanoic acid (^{123}I-BMIPP) dual single-photon emission computed tomography (dual-SPECT), but no myocardial fibrosis on contrast-enhanced magnetic resonance images (MRI). One had apparently normal myocardial perfusion and metabolism, whereas the other had exercise-induced myocardial ischemia and impaired myocardial metabolism. These findings indicated that even apical HCM without myocardial fibrosis is pathophysiologically heterogeneous. Apical HCM has been evaluated by either dual-SPECT or cardiac MRI, but not by both. Thus, a combination of imaging modalities is apparently essential for elucidating the pathophysiology of apical HCM. These dissimilar findings in dual-SPECT might be important in identifying patients with apical HCM who are at high risk of forming aneurysms. (Cardiol J 2010; 17, 3: 306–311)

Key words: SPECT, MRI, TlCl, BMIPP, echocardiography

Introduction

Apical hypertrophic cardiomyopathy (apical HCM) is a rare variant of HCM characterized by localized apical hypertrophy and a giant T-wave inversion on electrocardiograms [1, 2]. Although the prognosis of most patients with apical HCM is relatively benign [3], it can be quite poor if the apical left ventricle contains aneurysms, as they are closely associated with intraventricular thrombosis and life-threatening arrhythmia [4]. However, the mechanism of aneurysm formation is unclear.

We describe two patients with apical HCM but with different findings in ^{201}Thallous chloride (^{201}TlCl) and ^{123}I-betamethyl-p-iodophenyl-pentadecanoic acid (^{123}I-BMIPP) dual single-photon emission computed tomography (dual-SPECT), although contrast-enhanced magnetic resonance imaging (MRI) did not identify myocardial fibrosis. We discuss the potential of combined dual-SPECT and

Address for correspondence: Satoshi Okayama, Nara Medical University, First Department of Internal Medicine, Nara, Japan, e-mail: satosi01@naramed-u.ac.jp

Received: 16.07.2009 Accepted: 7.09.2009

contrast-enhanced MRI to evaluate the pathophysiology of apical HCM.

Methods

We performed cardiac MRI and dual-SPECT in two patients who were diagnosed with apical HCM based on giant T-wave inversions on 12--lead rest electrocardiograms and apical hypertrophy on transthoracic echocardiograms. Apical hypertrophy was confirmed on magnetic resonance cine images, and myocardial fibrosis was investigated on gadolinium late enhancement images. We then performed ^{201}TlCl and ^{123}I-BMIPP dual--SPECT to simultaneously evaluate myocardial perfusion and metabolism. We compared cardiac MRI and dual-SPECT on arbitrary cross-sectional images.

Cardiac MRI

Patients underwent cardiac MRI using a 1.5-T magnetic resonance imaging scanner (Avanto; Siemens, Erlangen, Germany). Cine images were acquired using the segmented ECG-triggered True FISP sequence [5]. Gadolinium late enhancement images were acquired using a segmented two-dimensional inversion recovery True FISP sequence about ten minutes after an intravenous injection of 0.15 mmol/kg gadolinium diethylenetriamine penta-acetic acid (Magnevist; Bayer Schering Pharma AG, Berlin, Germany) [6].

Dual-SPECT

Within one month of cardiac MRI evaluation, the patients underwent dual-SPECT evaluation using a double-headed gamma camera (PRISM-AXIS, Shimadzu, Kyoto, Japan) after a fast of at least six hours. A dose of 111 MBq of ^{123}I-BMIPP was intravenously injected and flushed with 10 mL of saline at rest. The patient rested for 20 minutes after the injection to ensure that ^{123}I-BMIPP adequately reflected fatty acid metabolism in the myocardium. Thereafter, an upright bicycle ergometer exercise test was started according to the ramp incremental protocol. At peak exercise, 111 MBq of ^{201}TlCl was intravenously injected and flushed with 10 mL of saline. The test continued for an additional 60 seconds to allow adequate circulation of the isotope. Both ^{201}TlCl and ^{123}I-BMIPP-SPECT data were simultaneously acquired according to the method of Nishimura et al. [7] within ten minutes of (early phase) and four hours after (late phase) exercise completion.

Case 1

A 69 year-old male was admitted to our hospital with abnormal electrocardiographic findings that were discovered at a routine health examination. Hyperlipidemia and hyperuricemia had been treated with pravastatin sodium and allopurinol respectively. Cardiac symptoms such as chest pain, dyspnea, palpitation, faintness and syncope were absent. A family history of heart disease, sudden death, or premature death was insignificant. Twelve-lead rest electrocardiography revealed atrial fibrillation and giant T-wave inversion in leads II, III, aVF, and V3––V6 (Fig. 1, left row), and transthoracic echocardiography revealed left ventricular apical myocardial hypertrophy with normal wall motion (Fig. 1, right row). These findings indicated apical HCM.

He was thus examined by cardiac MRI and dual-SPECT. Cardiac MRI confirmed the diagnosis of apical HCM on cine images (Fig. 2A, B), and showed no hyperintense areas indicating myocardial fibrosis on gadolinium late enhancement images (Fig. 2C). Next, the dual-SPECT exercise test was terminated at a heart rate of 120 bpm (100 W) at the onset of leg fatigue. However, the patient did not describe any significant cardiac symptoms and ischemic electrocardiographic changes were absent. The ^{201}TlCl and ^{123}I-BMIPP images demonstrated significantly increased uptake in the left ventricular apical myocardium and no areas of decreased uptake, indicating impaired myocardial perfusion and metabolism respectively (Fig. 2D–F). The above findings indicated a diagnosis of apical HCM without myocardial fibrosis or impaired myocardial perfusion and metabolism.

Case 2

A 66 year-old female was referred to our department with abnormal electrocardiographic findings at a pre-operative examination for resection of a uterine leiomyoma. Neither she nor any family member had a significant history of cardiac conditions and she was asymptomatic for cardiac diseases. Twelve-lead rest electrocardiography showed giant T-wave inversion in leads I, II, aVl, and V3–V6 (Fig. 3, left row), and transthoracic echocardiography revealed left ventricular apical myocardial hypertrophy with normal wall motion (Fig. 3, right row), indicating apical HCM. Cardiac MRI confirmed apical HCM on cine images (Fig. 4A, B), and showed no hyperintense areas on gadolinium late enhancement images (Fig. 4C). A dual-SPECT exercise test was terminated at a heart rate of 100 bpm (80 W) at the onset of leg

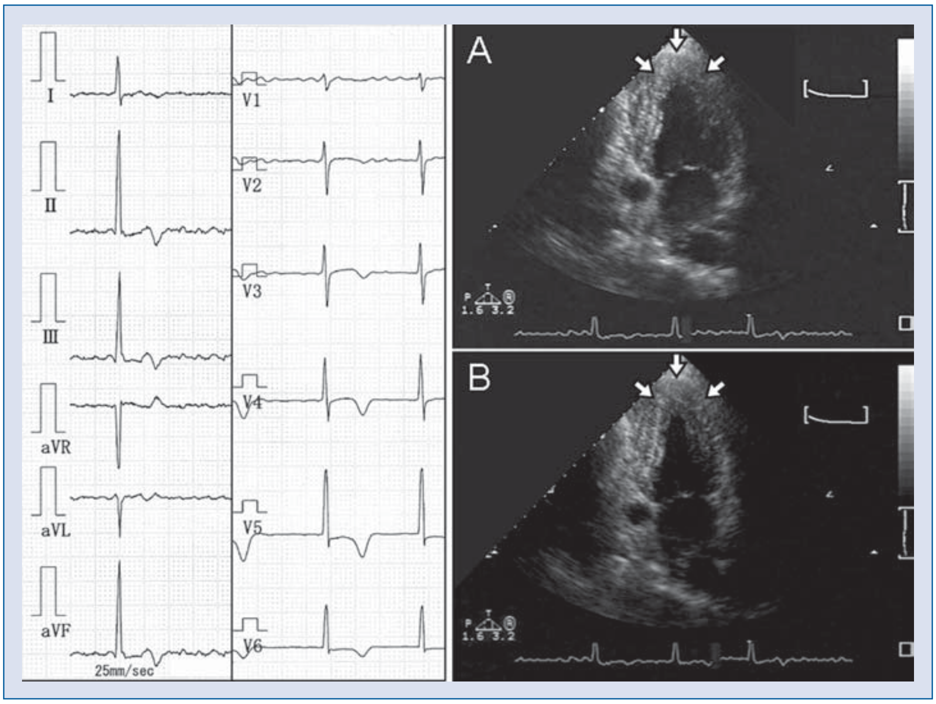

Figure 1. Electrocardiogram (left row) shows atrial fibrillation and giant T-wave inversion in leads II, III, aVF, and V3–V6. Transthoracic echocardiography (right row) at diastole (**A**) and at systole (**B**) reveals left ventricular apical myocardial hypertrophy with normal wall motion in the apical two-chamber view (arrows).

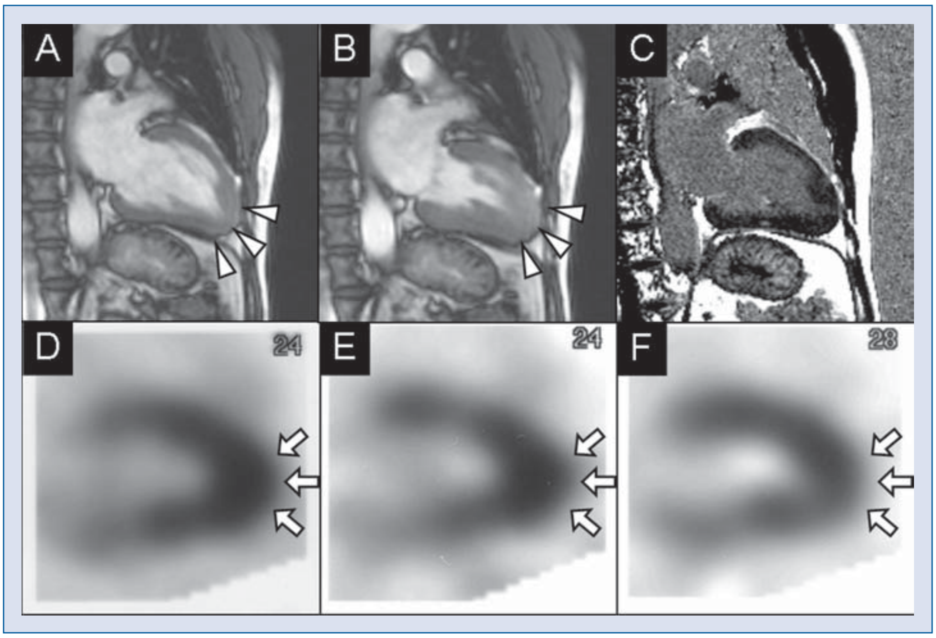

Figure 2. Cardiac magnetic resonance imaging cine imaging at diastole (**A**) and at systole (**B**) confirms left ventricular apical myocardial hypertrophy with normal wall motion (arrowheads). Gadolinium late enhancement imaging (**C**) shows no hyperintense areas. ^{201}TlCl images at early (**D**) and late (**E**) phases and ^{123}I-BMIPP image (**F**) show significantly increased uptake in left ventricular apical myocardium (arrows).

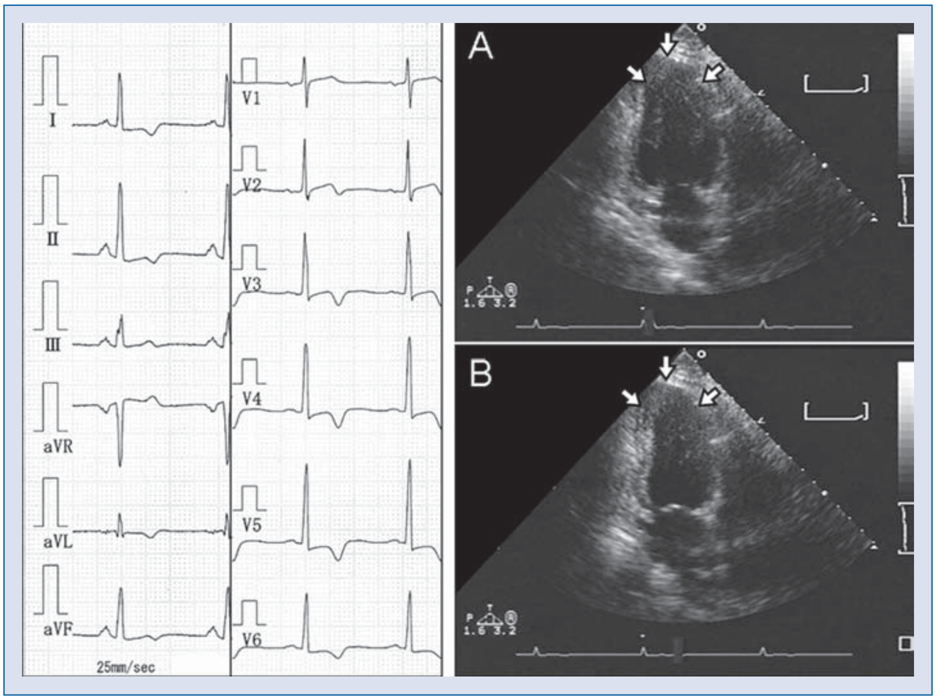

Figure 3. Electrocardiogram (left row) shows giant T-wave inversion in leads I, II, aVl, and V3–V6. Transthoracic echocardiography (right row) at diastole (**A**) and at systole (**B**) reveals left ventricular apical myocardial hypertrophy with normal wall motion in the apical two-chamber view (arrows).

fatigue, after which the patient did not describe any significant cardiac symptoms. No ischemic electrocardiographic changes were evident. The ^{201}TlCl images demonstrated a significantly decreased uptake area in the left ventricular apical myocardium during the early phase (Fig. 4D) and complete redistribution at the late phase (Fig. 4E), indicating exercise-induced myocardial ischemia. The ^{123}I--BMIPP images also demonstrated a significantly decreased uptake area in the apical myocardium during the early phase (Fig. 4F), indicating myocardial metabolism impaired by a microcirculatory disorder or delayed recovery from impaired myocardial metabolism induced by past ischemia.

However, the decreased uptake area on ^{123}I--BMIPP images was consistent with the exercise-induced ischemic myocardial area identified by ^{201}TlCl-SPECT. We thus considered that these findings reflected delayed recovery from impaired myocardial metabolism induced by past exercise-induced ischemia. Coronary computed tomography angiography confirmed the absence of significant coronary stenosis. These findings indicated a diagnosis of apical HCM with exercise-induced myocardial ischemia and impaired myocardial metabolism, but no myocardial fibrosis.

Discussion

Dual ^{201}TlCl and ^{123}I-BMIPP SPECT has been widely applied in Europe and Japan to evaluate patients with various types of heart disease. Myocardial perfusion is usually evaluated by ^{201}TlCl, whereas ^{123}I-BMIPP reflects fatty acid metabolism in the heart. A decrease in ^{123}I-BMIPP accumulation indicates myocardial areas where fatty acid metabolism is suppressed and the source of ATP production switches from fatty acid to glucose because of current and past ischemia as well as microcirculatory disorders [8]. Moreover, a reduction in BMIPP is the most sensitive indicator of metabolic abnormalities in patients with HCM [9].

Thus, dual-SPECT can simultaneously evaluate both myocardial perfusion and metabolism, whereas cardiac MRI is one of the most rigorous and accurate methods of evaluating cardiac morphology and fibrosis. Comparisons are facilitated because both dual-SPECT and cardiac MRI can provide several arbitrary cross-sectional images of the heart.

Here, dual-SPECT classified two apical HCM patients without myocardial fibrosis in contrast-enhanced MRI into subtypes with and without im-

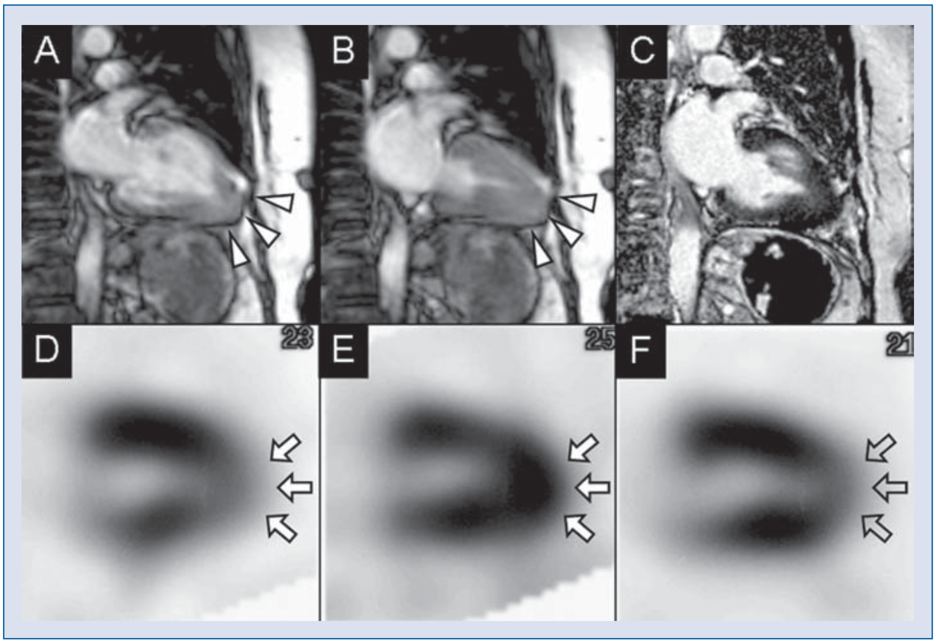

Figure 4. Cardiac magnetic resonance imaging cine images at diastole (**A**) and at systole (**B**) confirm left ventricular apical myocardial hypertrophy with normal wall motion (solid arrowheads). Gadolinium late enhancement image (**C**) shows no hyperintense areas. ^{201}TlCl image at early phase (**D**) shows area of significantly decreased uptake in left ventricular apical myocardium, and ^{201}TlCl image at late phase (**E**) shows complete redistribution (solid arrows). ^{123}I-BMIPP image at early phase (**F**) shows area of significantly decreased uptake in apical myocardium (unfilled arrows).

paired myocardial perfusion (exercise-induced myocardial ischemia) and metabolism. These findings indicated that even apical HCM without myocardial fibrosis is pathophysiologically heterogeneous. Because the presence of myocardial fibrosis is related to cardiac dysfunction and arrhythmia in HCM, its absence is associated with a relatively benign prognosis [10, 11]. However, even without myocardial fibrosis in contrast-enhanced MRI, the long-term prognosis of patients with impaired myocardial ischemia and metabolism according to dual-SPECT is considered to be poor. This is because apical mismatch areas with impaired myocardial perfusion and metabolism, but no myocardial fibrosis, can probably predict the development of new myocardial fibrosis, and it might be longitudinally associated with the formation of apical aneurysms. Apical HCM has been evaluated by either dual-SPECT [12] or cardiac MRI alone [13], but not by both. These cases suggest that the combination of imaging modalities is essential for elucidating the pathophysiology of apical HCM.

In conclusion, Dual-SPECT uncovered dissimilar myocardial perfusion and metabolism between two patients who had apical HCM without myocardial fibrosis on contrast-enhanced MRI. This combination of imaging modalities should thus help to clarify the pathophysiology of patients with apical HCM.

Acknowledgements

The authors do not have any conflict of interest regarding this study.

References

1. 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.
2. Yamaguchi H, Ishimura T, Nishiyama S et al. Hypertrophic non-obstructive cardiomyopathy with giant negative T waves (apical hypertrophy): Ventriculographic and echocardiographic features in 30 patients. Am J Cardiol, 1979; 44: 401–412.
3. Nasermoaddeli A, Miura K, Matsumori A et al. Prognosis and prognostic factors in patients with hypertrophic cardiomyopathy in Japan: Results from a nationwide study. Heart, 2007; 93: 711–715.
4. Matsubara K, Nakamura T, Kuribayashi T, Azuma A, Nakagawa M. Sustained cavity obliteration and apical aneurysm formation in apical hypertrophic cardiomyopathy. J Am Coll Cardiol, 2003; 42: 288–295.

5. Miller S, Simonetti OP, Carr J, Kramer U, Finn JP. MR imaging of the heart with cine true fast imaging with steady-state precession: Influence of spatial and temporal resolutions on left ventricular functional parameters. Radiology, 2002; 223: 263–269.
6. Huber AM, Schoenberg SO, Hayes C et al. Phase-sensitive inversion-recovery MR imaging in the detection of myocardial infarction. Radiology, 2005; 237: 854–860.
7. Nishimura M, Hashimoto T, Kobayashi H et al. Myocardial scintigraphy using a fatty acid analogue detects coronary artery disease in hemodialysis patients. Kidney Int, 2004; 66: 811–819.
8. Taki J, Matsunari I. Metabolic imaging using SPECT. Eur J Nucl Med Mol Imag, 2007; 34 (suppl. 1): S34–S48.
9. Tadamura E, Kudoh T, Hattori N et al. Impairment of BMIPP uptake precedes abnormalities in oxygen and glucose metabolism in hypertrophic cardiomyopathy. J Nucl Med, 1998; 39: 390–396.
10. Teraoka K, Hirano M, Ookubo H et al. Delayed contrast enhancement of MRI in hypertrophic cardiomyopathy. Magn Reson Imag, 2004; 22: 155–161.
11. Moon JC, McKenna WJ, McCrohon JA, Elliott PM, Smith GC, Pennell DJ. Toward clinical risk assessment in hypertrophic cardiomyopathy with gadolinium cardiovascular magnetic resonance. J Am Coll Cardiol, 2003; 41: 1561–1567.
12. Matsuo S, Nakamura Y, Takahashi M, Mitsunami K, Kinoshita M. Myocardial metabolic abnormalities in hypertrophic cardiomyopathy assessed by iodine-123-labeled beta-methyl-branched fatty acid myocardial scintigraphy and its relation to exercise-induced ischemia. Jpn Circ J, 1998; 62: 167–172.
13. Gebker R, Neuss M, Paetsch I, Nagel E. Images in cardiovascular medicine. Progressive myocardial fibrosis in a patient with apical hypertrophic cardiomyopathy detected by cardiovascular magnetic resonance. Circulation, 2006; 1; 114: e75–e76.