

Hypertrophic Cardiomyopathy With Progression From Apical Hypertrophy to Asymmetrical Septal Hypertrophy: A Case Report

Hiroaki KITAOKA, MD, FJCC
Tomoo KUWAHARA, MD
Koji NISHIDA, MD
Keisuke MORIMOTO, MD
Toru KUBO, MD
Makoto OKAWA, MD
Takashi FURUNO, MD
Yoshinori DOI, MD, FJCC

Abstract

A 41-year-old man was referred to our hospital for further examination because of abnormal electrocardiography findings at a health-check examination. Transthoracic echocardiography showed left ventricular hypertrophy confined to the most distal portion of the left ventricle, which is a typical feature of apical hypertrophic cardiomyopathy. Ten years later, he was again admitted for the evaluation of chest pain. Echocardiography showed asymmetrical septal hypertrophy in addition to apical hypertrophy. These findings demonstrate morphologic evolution in hypertrophic cardiomyopathy from apical hypertrophy to asymmetrical septal hypertrophy.

J Cardiol 2005 Apr; 45(4): 155 - 159

Key Words

■Cardiomyopathies, hypertrophic (apical)
■Ventricular remodeling

■Hypertrophy (asymmetrical septal)

INTRODUCTION

Apical hypertrophic cardiomyopathy (HCM) is characterized by wall thickening confined to the most distal portion of the left ventricle^{1,2}. The morphologic evolution of HCM from apical hypertrophy to typical asymmetric septal hypertrophy has rarely been documented. We report a patient with HCM who presented initially with apical hypertrophy which progressed to asymmetrical septal hypertrophy 10 years later.

CASE REPORT

A 41-year-old man was referred to our hospital

for further examination because of abnormal electrocardiography findings at a health-check examination. He had no cardiac symptoms or family history of cardiac disease. On examination, his blood pressure was 150/80 mmHg, and pulse was regular at 60 beats/min. There was no significant heart murmur. Electrocardiography showed normal sinus rhythm and marked left ventricular hypertrophy with markedly inverted T waves, so-called giant negative T waves, in the left precordial leads (Fig. 1). Chest radiography showed no cardiomegaly with a cardiothoracic ratio of 45%. Transthoracic echocardiography showed left ventricular hypertrophy confined to the most distal portion of the left

高知大学医学部 老年病科・循環器科: 〒783-8505 高知県南国市岡豊町小蓮

Cardiology Division, Department of Medicine and Geriatrics, Kochi Medical School, Kochi

Address for correspondence: KITAOKA H, MD, FJCC, Cardiology Division, Department of Medicine and Geriatrics, Kochi Medical School, Kohasu, Oko-cho, Nankoku, Kochi 783-8505; E-mail: kitaokah@med.kochi-u.ac.jp

Manuscript received August 20, 2004; revised October 27, 2004; accepted October 27, 2004

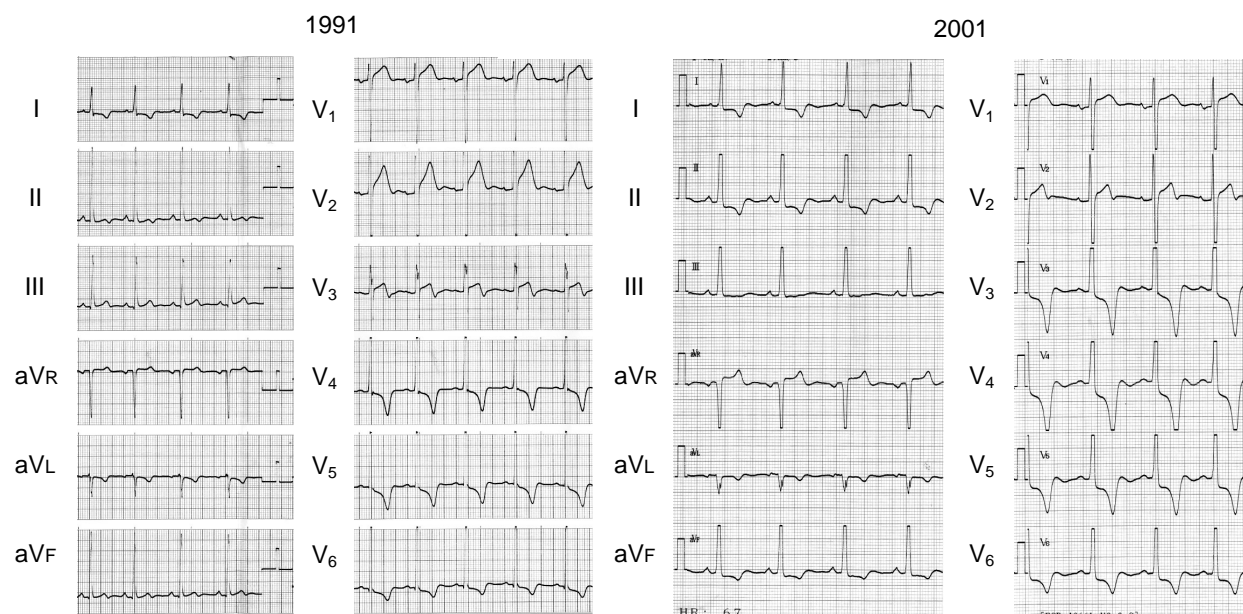


Fig. 1 Electrocardiograms recorded in 1991 and 2001

Both electrocardiograms show high voltage and deep negative T waves in the left precordial leads. In particular, negative T wave was progressive from 1991 to 2001.

ventricle, which is a typical feature of apical hypertrophic cardiomyopathy(**Fig. 2**). There was no basal hypertrophy with normal intraventricular septal wall thickness and posterior wall thickness (12mm). No serious ventricular arrhythmia was observed by 24-hour electrocardiogram monitoring. Laboratory examinations showed no abnormality with negative hepatitis C virus antibody. The diagnosis was apical HCM. He was followed up by a primary care physician.

Ten years later, he was again admitted for evaluation of chest pain. Electrocardiography again showed left ventricular hypertrophy with giant negative T waves(**Fig. 1**). Echocardiography showed asymmetrical septal hypertrophy(intraventricular septal wall thickness: 23mm, posterior wall thickness: 13mm), which was not present in 1991, in addition to apical hypertrophy(**Fig. 2**). Left ventriculography showed a distorted left ventricular configuration, apparently different from the ' spade-like left ventricular configuration ' usually seen in apical HCM. These findings demonstrate morphologic evolution in HCM from apical hypertrophy to asymmetrical septal hypertrophy.

DISCUSSION

Apical HCM is a morphologic variant of HCM characterized by a striking electrocardiographic

pattern of particularly deep T-wave inversion in the precordial leads(" giant negative T-waves ") and distinctive angiographic spade-shaped appearance of the left ventricle at end-diastole^{1,2}). Several reports of apical HCM later appeared from other countries, but the prevalence and clinical characteristics of apical HCM in Western countries seem to be markedly different from those of apical HCM in Japan³⁻⁹). Therefore, the definition, etiology, genetic background, and long-term prognosis of apical HCM remain controversial⁹⁻¹²).

Although the angiographic spade-shaped appearance of the left ventricle was reported as a hallmark of apical HCM, a new subtype of apical HCM with a nonspade apical HCM was detected using magnetic resonance imaging¹³). This subtype had hypertrophic myocardium confined to a narrow region of the septum or the anterior or lateral wall at the apical level but no hypertrophy at the basal level. Although our patient did not undergo left ventriculography or magnetic resonance imaging in 1991, the 4-chamber view of echocardiography was compatible with nonspade apical HCM. The electrocardiographic and echocardiographic changes were described in 29 patients with apical HCM (Japanese type) during follow-up period of 10.9 ± 3.7 years¹⁴). Only one patient who developed asymmetrical septal hypertrophy. Therefore, morpholog-

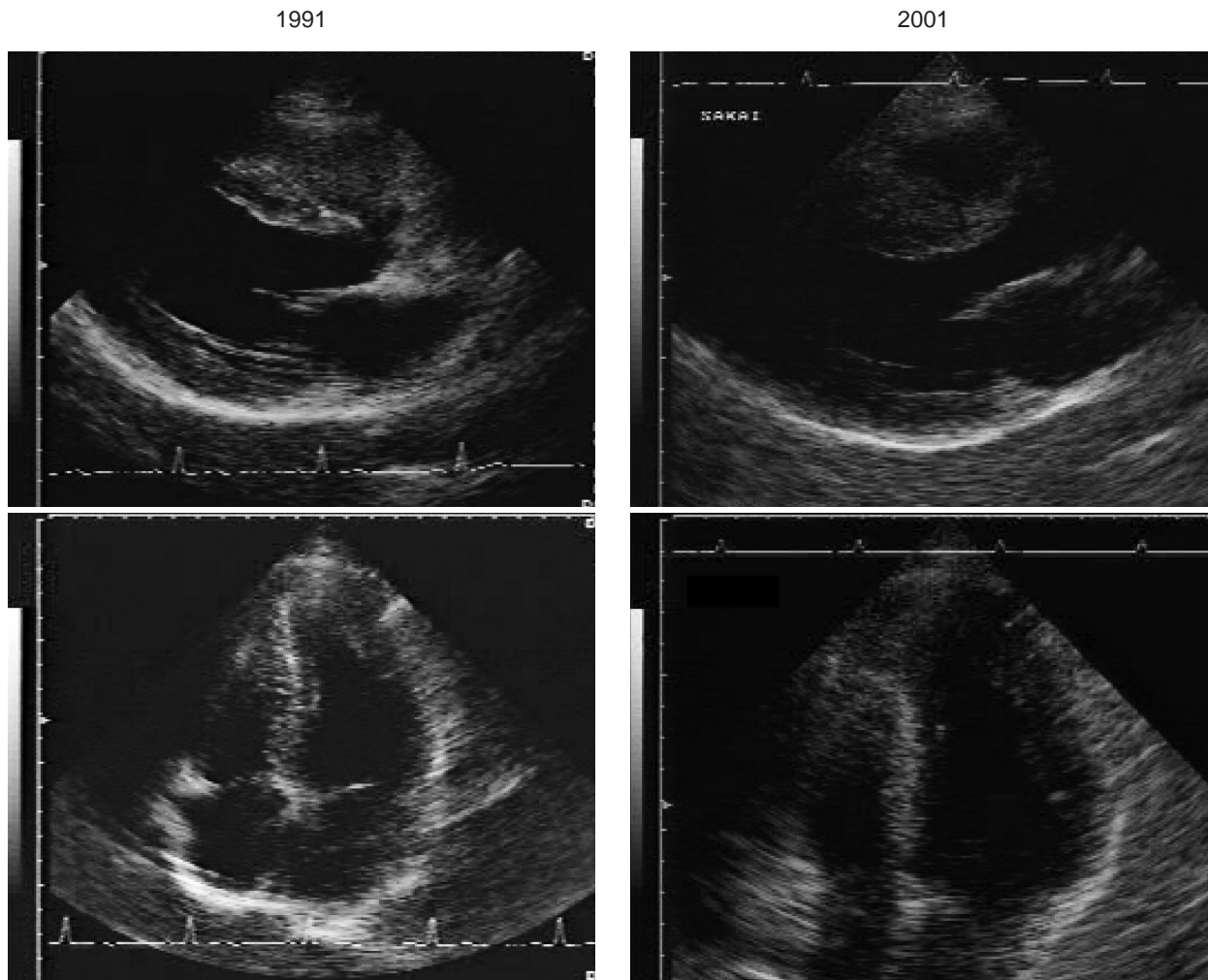


Fig. 2 Echocardiograms recorded in 1991(left column)and 2001(right column)

Parasternal long-axis(upper row)and apical 4-chamber(lower row)views showed left ventricular hypertrophy confined to the most distal portion of the left ventricle in 1991. However, marked hypertrophy was shown in the basal left ventricle in 2001(intraventricular septal wall thickness 23 mm, posterior wall thickness 13 mm)

ic documentation from apical hypertrophy to asymmetrical septal hypertrophy is indeed rare.

The etiology of apical HCM is unresolved. Hepatitis C viral infection and other environmental factors have been proposed as causes of apical HCM because family history of HCM is rare in patients with apical HCM^{11,15}). Although genetic involvement seems to be uncommon in apical HCM compared to other types of HCM, various mutations have recently been reported to be responsible for apical HCM in a few Japanese patients^{16,17}). These include Arg162Trp, Gly203Ser and Lys183del mutations in the cardiac troponin I

gene, and mutations in the cardiac myosin binding protein-C gene, beta-myosin heavy chain gene (Lys953Glu), and cardiac troponin T gene(Phe110 Ile). Co-existence of patients with apical HCM and those with “ usual HCM ”(*i.e.* asymmetrical septal hypertrophy)has been reported in the same family¹⁸). Our patient showed progressive remodeling from apical hypertrophy to asymmetrical septal hypertrophy, indicating that patients with apical HCM are a part of the broad spectrum of familial HCM with etiology due to abnormality in the sarcomeric protein.

要 約

心尖部に限局した肥大が非対称性心室中隔肥大へ変化した
肥大型心筋症の1例北岡 裕章 桑原 朋 西田 幸司 森本 啓介
久保 亨 大川 真理 古野 貴志 土居 義典

症例は41歳，男性．健康診断で心電図異常を指摘され，精査目的で紹介された．心電図で左側胸部誘導に巨大陰性T波が認められ，心エコー図法では心基部には肥大が認められず，心尖部に限局した左室肥大が認められた．明らかな肥大型心筋症の家族歴は認められず，心尖部肥大型心筋症と診断した．10年後，安静時胸痛の精査目的で再度当科へ来院した．心電図では，以前と同様に巨大陰性T波が認められたが，心エコー図法では，心尖部肥大に加え，以前には認められなかった心基部の非対称性心室中隔肥大が認められた．冠動脈造影では有意狭窄は認められなかった．これらの所見は心尖部肥大型心筋症の病因論を考察するうえで重要と考えられる．

J Cardiol 2005 Apr; 45(4): 155 - 159

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, Nagasaki F, Nakanishi S, 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
- 3) Wigle ED, Sasson Z, Henderson MA, Ruddy TD, Fulop J, Rakowski H, Williams WG: Hypertrophic cardiomyopathy: The importance of the site and extent of hypertrophy: A review. *Prog Cardiovasc Dis* 1985; **28**: 1 - 83
- 4) Maron BJ, Bonow RO, Seshagiri TNR, Roberts WC, Epstein SE: Hypertrophic cardiomyopathy with ventricular septal hypertrophy localized to the apical region of the left ventricle(apical hypertrophic cardiomyopathy). *Am J Cardiol* 1982; **49**: 1838 - 1848
- 5) Keren G, Belhassen B, Sherez J, Miller HI, Megidish R, Berenfeld D: Apical hypertrophic cardiomyopathy: Evaluation by noninvasive and invasive technique in 23 patients. *Circulation* 1985; **71**: 45 - 56
- 6) Louie ER, Maron BJ: Apical hypertrophic cardiomyopathy: Clinical and two-dimensional echocardiographic assessment. *Ann Intern Med* 1987; **106**: 663 - 670
- 7) Webb JG, Sasson Z, Rakowski H, Liu P, Wigle ED: Apical hypertrophic cardiomyopathy: Clinical follow-up and diagnostic correlates. *J Am Coll Cardiol* 1990; **15**: 83 - 90
- 8) Klues HG, Schiffers A, Maron BJ: Phenotypic spectrum and patterns of left ventricular hypertrophy in hypertrophic cardiomyopathy: Morphologic observations and significance as assessed by two-dimensional echocardiography in 600 patients. *J Am Coll Cardiol* 1995; **26**: 1699 - 1708
- 9) Kitaoka H, Doi Y, Casey SA, Hitomi N, Furuno T, Maron BJ: Comparison of prevalence of apical hypertrophic cardiomyopathy in Japan and the United States. *Am J Cardiol* 2003; **92**: 1183 - 1186
- 10) Eriksson MJ, Sonnenberg B, Woo A, Rakowski P, Parker TG, Wigle D, Rakowski H: Long-term outcome in patients with apical hypertrophic cardiomyopathy. *J Am Coll Cardiol* 2002; **39**: 638 - 645
- 11) Maron BJ: Apical hypertrophic cardiomyopathy: The continuing saga. *J Am Coll Cardiol* 1990; **15**: 91 - 93
- 12) Koga Y, Nohara M, Miyazaki Y, Toshima H: Two forms of apical hypertrophic cardiomyopathy: Japanese and Western forms. *in* *Cardiomyopathy Update 2: Hypertrophic Cardiomyopathy*(ed by Toshima H, Maron BJ). University of Tokyo Press, Tokyo, 1988; pp 293 - 308
- 13) Suzuki J, Watanabe F, Takenaka K, Amano K, Amano W, Igarashi T, Aoki T, Serizawa T, Sakamoto T, Sugimoto T, Nishikawa J: New subtype of apical hypertrophic cardiomyopathy identified with nuclear magnetic resonance imaging as an underlying cause of markedly inverted T waves. *J Am Coll Cardiol* 1993; **22**: 1175 - 1181
- 14) Koga Y, Katoh A, Matsuyama K, Ikeda H, Hiyamuta K, Toshima H, Imaizumi T: Disappearance of giant negative T waves in patients with the Japanese form of apical hypertrophy. *J Am Coll Cardiol* 1995; **26**: 1672 - 1678
- 15) Matsumori A, Ohashi N, Nishio R, Kakio T, Hara M, Furukawa Y, Ono K, Shioi T, Hasegawa K, Sasayama S: Apical hypertrophic cardiomyopathy and hepatitis C virus infection. *Jpn Circ J* 1999; **63**: 433 - 438
- 16) Kimura A, Harada H, Park J-E, Nishi H, Satoh M, Takahashi M, Hiroi S, Sasaoka T, Ohbuchi N, Nakamura T, Koyanagi T, Hwang TH, Choo JA, Chung KS, Hasegawa A, Nagai R, Okazaki O, Nakamura H, Matsuzaki M, Sakamoto T, Toshima H, Koga Y, Imaizumi T, Sasazuki T: Mutations in the cardiac troponin I gene associated with hypertrophic cardiomyopathy. *Nat Genet* 1997; **16**: 379 - 382

- 17) Anan R, Shono H, Kisanuki A, Arima S, Nakao S, Tanaka H: Patients with familial hypertrophic cardiomyopathy caused by a Phe110Ile missense mutation in the cardiac troponin T gene have variable cardiac morphologies and a favorable prognosis. *Circulation* 1998; **98**: 391 - 397
- 18) Ko Y, Lei MH, Chiang FT, Chen JJ, Kuan P, Lien WP: Apical hypertrophic cardiomyopathy of the Japanese type: Occurrence with familial hypertrophic cardiomyopathy in a family. *Am Heart J* 1992; **124**: 1626 - 1630