

## Apical Hypertrophic Cardiomyopathy in a Case with Chest Pain and Family History of Sudden Cardiac Death: A Case Report

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### ABSTRACT

Hypertrophic cardiomyopathy (HCM) is the most common genetic cardiovascular disease, which is caused by a multitude of mutations in genes encoding proteins of the cardiac sarcomere (1). Apical hypertrophic cardiomyopathy (AHCM) is an uncommon type of HCM. The sudden cardiac death is less likely to occur in the patients inflicted with AHCM (2). Herein, we presented the case of a 29-year-old man with AHCM, who had typical exertional chest pain without any cardiovascular risk factors, except for a sudden cardiac death in his older brother at the age of 28 years. After performing complete clinical and paraclinical evaluations, the patient underwent optimal medical treatment with beta-blocker agents without any symptoms.

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### Introduction

Hypertrophic cardiomyopathy (HCM) is associated with heterogeneous clinical expression, unique pathophysiology, and diverse natural history. This condition is the most common cause of sudden death in the young, including competitive athletes, and accounts for heart failure-related disability at virtually any age. The HCM is characterized by a thickened and non-dilated left ventricle (LV) in the absence of other cardiac or systemic conditions (1).

Apical hypertrophic cardiomyopathy (AHCM) is an uncommon type of HCM. This disease was initially reported in Japan. And its prevalence is different in the worldwide, which is reported as 15% of the all form of HCM in Japan, and in United State of America it is only 3% (2). Importantly, sudden cardiac death is less likely to occur in the patients suffering from AHCM (3). In AHCM, thickened apical segments produce a "spade-shaped" small apical cavity.

This can be detected by any of the noninvasive cardiac imaging techniques during the investigation of abnormally inverted precordial T waves on

electrocardiogram (ECG). In the current report, we presented the case of AHCM with the aim of attracting more attention to this uncommon disease.

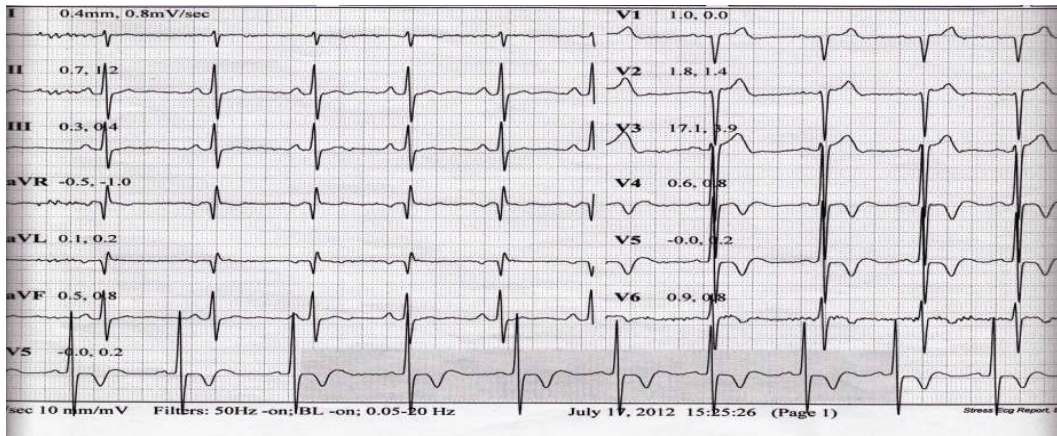
### Case Presentation

A 29-year-old man presented with typical chest pain, which was aggravated with exertion and initiated over two months before his referral. The symptoms of the patient gradually worsened to the New York Heart Association (NYHA) class III. He reported to have no lightheadedness, orthopnea, pedal edema, or family history of cardiac disease. He also had no risk factors or significant past medical history, except for a history of a sudden cardiac death in his 28-year-old brother, who had suddenly died three years ago.

Physical examination revealed a displaced apical impulse and a prominent fourth heart sound. The ECG showed markedly increased QRS voltage in the precordial leads (left ventricular hypertrophy [LVH]) with ST depressions and giant inverted T waves suggestive of the apical

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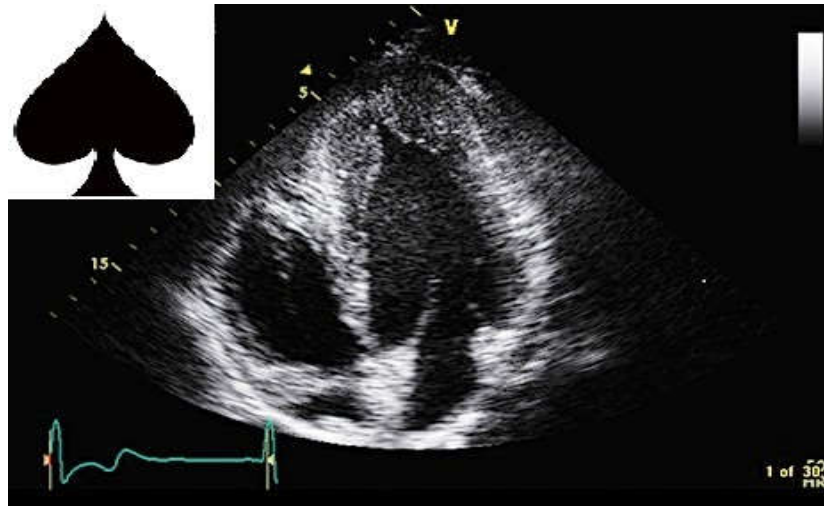


**Figure 1.** Left ventricular hypertrophy with ST depressions and giant inverted T waves in precordial leads on electrocardiogram

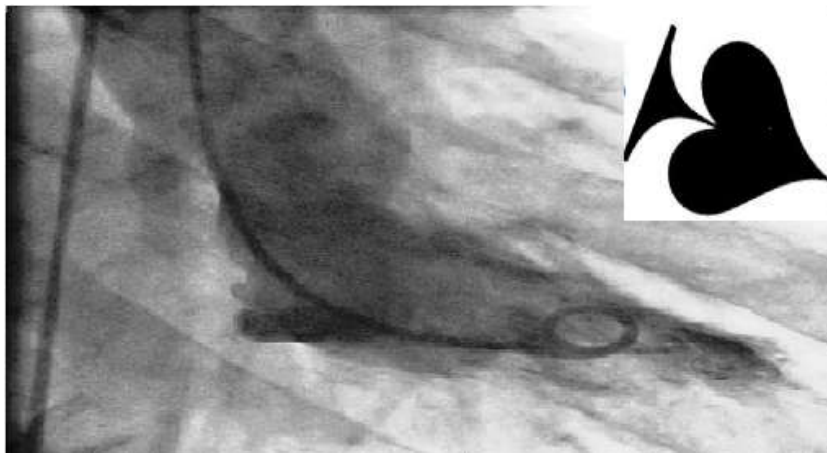
variant of HCM (Figure 1). Furthermore, the results of the complete blood count as well as renal, electrolyte, hepatic, and thyroid panels were within normal limits.

On two-dimensional transthoracic echocardi-

graphy (TTE), an apical 4-chamber view of the LV revealed the hypertrophy of the apex in an “ace-of-spades” configuration (Figure 2) as noted in his left ventriculography during the left heart catheterization (Figure 3). The slow flow of the



**Figure 2.** Massive thickening of the left ventricle apex with relative sparing of the basal septum and left ventricular free walls on two-dimensional echocardiography



**Figure 3.** A distinctive left ventricle “spade-like” configuration on contrast ventriculography

coronary arteries could explain the patient's chest pain, which was due to the imbalance between the supply and demand of oxygen.

The ambulatory 48 h rhythm monitoring did not show any arrhythmia. No AHCM was indicated in the TTE performed on the patient's first-degree relatives. Therefore, he received medical treatment. The patient became totally asymptomatic with optimal dosage of beta-blocker drugs (i.e., metoprolol succinate 47.5 mg twice a day).

## Discussion

The diagnostic criteria for AHCM included the demonstration of asymmetric LVH, confined to the LV apex, with an apical wall thickness of  $\geq 15$  mm and a ratio of maximal apical to posterior wall thickness of  $\geq 1.5$  mm (4). About 54% of the patients with AHCM are symptomatic. As in our case, chest pain is the most common presentation, followed by palpitations and no syncope (4, 5).

The most frequent ECG findings are negative T waves in the precordial leads, which are found in 93% of the patients, followed by LVH in 65% of the patients (6). The ECG in our patient showed LVH and negative T waves with a maximum depth of 7 mm. Furthermore, the TTE, which is the initial diagnostic tool for AHCM, revealed the hypertrophy of the LV apex.

On the other hand, ventriculography AHCM demonstrates a distinctive LV "spade-like" configuration (7). Although the initial diagnostic test for AHCM is most commonly TTE, the best diagnostic tool is considered to be cardiac magnetic resonance imaging (5-7). Unlike other variants of HCM, the prognosis of AHCM is relatively benign (4, 6). The overall mortality rate of AHCM patients was estimated to be 10.5%, while the cardiovascular mortality was 1.9% after a follow-up of  $13.6 \pm 8.3$  years (8).

Sudden death and cardiovascular events occur more commonly in the patients with asymmetric septal hypertrophy than in those with AHCM (8,9). A large LV end diastolic dimension may predict cardiac events in the AHCM patients (6). Some patients may develop sudden life-threatening complications; therefore, close follow-up of these patients is recommended. We hope that this presentation would help to the earlier diagnosis of this disease.

## Acknowledgments

Non

## Conflict of Interest

The authors declare no conflict of interest.

## References

1. Maron BJ, McKenna WJ, Danielson GK, Kappenberger LJ, Kuhn HJ, Seidman CE, et al. American college of cardiology/European society of cardiology clinical expert consensus document on hypertrophic cardiomyopathy. A report of the American college of cardiology foundation task force on clinical expert consensus documents and the European society of cardiology committee for practice guidelines. *J Am Coll Cardiol.* 2003; 42:1687-713.
2. 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; 92:1183-6.
3. Alcalai R, Seidman JG, Seidman CE. Genetic basis of hypertrophic cardiomyopathy: from bench to the clinics. *J Cardiovasc Electrophysiol.* 2008; 19:104-10.
4. Bos JM, Towbin JA, Ackerman MJ. Diagnostic, prognostic, and therapeutic implications of gene testing for hypertrophic cardiomyopathy. *J Am Coll Cardiol.* 2009; 54:201-11.
5. Maron BJ, Seidman CE, Ackerman MJ, Towbin JA, Maron MS, Ommen SR, et al. How should hypertrophic cardiomyopathy be classified? What's in a name? Dilemmas in nomenclature characterizing hypertrophic cardiomyopathy and left ventricular hypertrophy. *Circ Cardiovasc Genet.* 2009; 2:81-5.
6. Maron BJ, Doerer JJ, Haas TS, Tierney DM, Mueller FO. Sudden deaths in young competitive athletes: Analysis of 1866 deaths in the United States, 1980-2006. *Circulation.* 2009; 119:1085-92.
7. Olivetto I, Maron MS, Autore C, Lesser JR, Rega L, Casolo G, et al. Assessment and significance of left ventricular mass by cardiovascular magnetic resonance in hypertrophic cardiomyopathy. *J Am Coll Cardiol.* 2008; 52:559-66.
8. Moukarbel GV, Alam SE, Abchee AB. Contrast-enhanced echocardiography for the diagnosis of apical hypertrophic cardiomyopathy. *Echocardiography.* 2005; 22:831-3.
9. Maron BJ, Towbin JA, Thiene G, Antzelevitch C, Corrado D, Arnett D, et al. Contemporary definitions and classification of the cardiomyopathies: an American heart association scientific statement from the council on clinical cardiology, heart failure and transplantation committee; quality of care and outcomes research and functional genomics and translational biology interdisciplinary working group. *Circulation.* 2006; 113:1807-16.