Apical hypertrophic cardiomyopathy treated as ST-elevation myocardial infarction

Christopher J. Parr, MD*; Rajat Sharma, MD†; Philip J. Garber, MD†

ABSTRACT

Electrocardiographic changes resulting from apical hypertrophic cardiomyopathy may mimic an acute coronary syndrome. A 67-year-old Sudanese male without cardiac risk factors presented to hospital with chest pain and electrocardiographic findings of septal ST-segment elevation, ST-segment depression in V4-V6, and diffuse T-wave inversion. He was treated as an acute ST-elevation myocardial infarction with thrombolytics. There was no cardiac biomarker rise and coronary angiography did not reveal evidence of significant coronary arterial disease. Ventriculography, transthoracic echocardiography, and cardiac magnetic resonance imaging were consistent with apical hypertrophic cardiomyopathy. The patient was discharged three days later with outpatient cardiology follow-up. We highlight the clinical and electrocardiographic findings of apical hypertrophic cardiomyopathy, with an emphasis on distinguishing this from acute myocardial infarction.

RÉSUMÉ

Les modifications électrocardiographiques liées à une myocardiopathie hypertrophique apicale peuvent simuler celles d'un syndrome coronarien aigu. Un homme de 67 ans d'origine soudanaise, sans facteurs de risque de maladie cardiaque a consulté pour des douleurs thoraciques, et différentes modifications électrocardiographiques ont été observées, notamment un sus-décalage du segment ST en territoire septal, un sous-décalage du segment ST en V4-V6 et une inversion diffuse de l'onde T. Aussi a-t-il été traité par des thrombolytiques pour un infarctus aigu du myocarde avec sus-décalage du segment ST, mais l'accident cardiaque ne s'est pas accompagné d'une élévation des biomarqueurs cardiaques, et la coronarographie n'a pas montré de signes d'une coronaropathie importante. Par contre, les résultats de la ventriculographie, de l'échographie transthoracique et de l'imagerie par résonance magnétiques, eux, convergeaient vers une myocardiopathie hypertrophique apicale. Le patient a recu son congé au bout de trois jours, puis a été suivi en cardiologie, en consultation externe. Il sera donc question, dans l'article, des aspects cliniques de la myocardiopathie hypertrophique apicale et des modifications électrocardiographiques qui y sont liées, et surtout des éléments qui permettent de distinguer cette affection et de l'infarctus aigu du myocarde.

Keywords: cardiology, STEMI, myocardial infarction, apical hypertrophic cardiomyopathy, thrombolytics, PCI

INTRODUCTION

Myocardial hypertrophy commonly involves the ventricular septum[1] but may occasionally predominantly involve the cardiac apex. Since the initial characterization by Japanese researchers Sakamoto[2] and Yamaguchi,[3] apical hypertrophic cardiomyopathy (ApHCM) has been recognized as a clinical entity with distinct electrocardiographic (ECG) findings including deep precordial T-wave inversion and echocardiographic findings involving a left ventricular (LV) ace-of-spades morphology.[3] Patients are largely asymptomatic and have had a favourable prognosis in both a Japanese population[4,5] (in which it is more common) and a non-Japanese North American population.[6] However, if these characteristic ECG findings are present with chest pain, patients may be inadvertently treated for an acute coronary syndrome (ACS).[7-11] Given the time-sensitive nature and possible iatrogenic risks of reperfusion therapy, it is important to recognize ApHCM as a potential imitator of an acute myocardial infarction.

CASE REPORT

A 67-year-old man of Sudanese origin with no standard cardiac risk factors presented to our emergency department (ED) 30 minutes after awakening with new-onset continuously severe retrosternal chest pain and diaphoresis. There was no antecedent angina at baseline. Initial vital signs were as follows: oral temperature of 36.7°C (98.1°F), non-invasive blood pressure of 169/77, heart rate of 78, respiratory rate of 16, and oxygen saturation of 98% by pulse oximetry. A 12-lead ECG was performed within 15 minutes of arrival, showing a sinus rhythm with 2 mm of concave

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ST-segment elevation in leads V1-V2; ST-segment depression in leads V4-V6; and T-wave inversion in leads I, II, III, aVF, and V3-V6. In lead V4, the T wave was symmetrically inverted to a depth of 10 mm and the R wave was prominent, measuring at least 30 mm. Chest radiographs were unremarkable.

The patient was treated for an acute ST-elevation myocardial infarction (STEMI) in the ED and was later admitted to the coronary care unit (CCU) after the initial treatment. He was initiated on aspirin, clopidogrel, intravenous heparin infusion, and atorvastatin. As the patient presented to a non–percutaneous coronary intervention (PCI) capable hospital, the anticipated first medical contact to device time was >120 minutes; tenecteplase 50 mg was administered intravenously 60 minutes into his presentation. The patient was free of chest discomfort 90 minutes into admission. During admission, serial ECGs showed no resolution of the ST-segment elevation and persistent T-wave inversion (Figure 1). High-sensitivity cardiac troponin T was within normal limits, and both troponin and creatine kinase remained stable over several days. A coronary angiogram performed 13 hours into his presentation revealed normal coronary arteries without evidence of obstructive coronary disease with a left ventriculography demonstrating obliteration of the apical cavity at end-systole (Figure 2). Transthoracic echocardiography revealed a progressive increase in LV wall thickness toward the apex without evidence of LV systolic dysfunction or outflow tract obstruction (Figure 3).

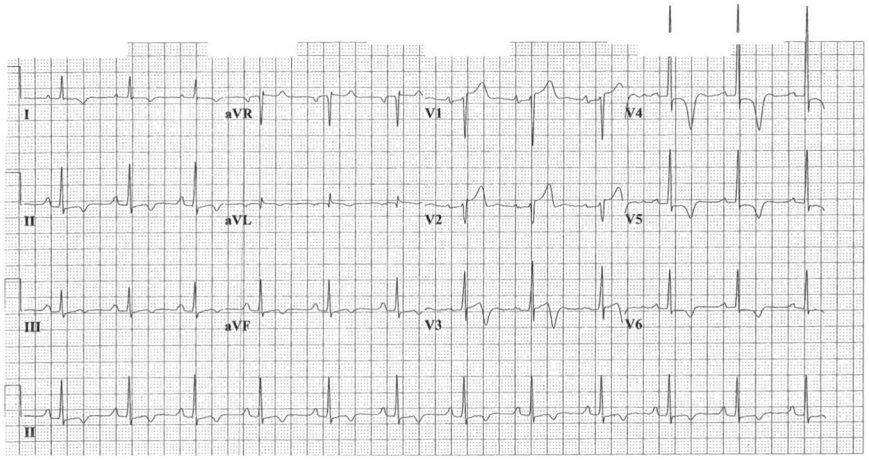

Figure 1. Index 12-lead electrocardiogram showing concave ST-segment elevation in leads V1-V2, ST-segment depression in leads V4-V6, and diffuse T wave inversion. In lead V4, the T wave was symmetrically inverted to a depth of 10 mm and the R wave was prominent, measuring at least 30 mm.

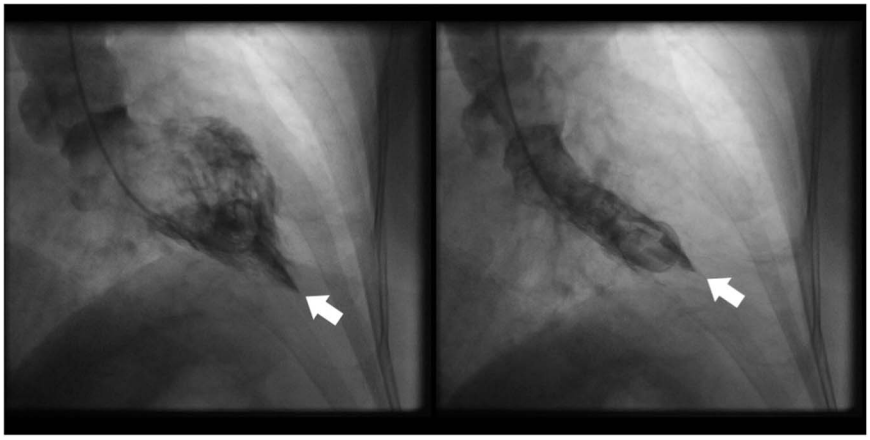

Figure 2. Left ventriculogram in right anterior oblique orientation showing (a) end-diastolic ace-of-spades configuration and (b) end-systolic obliteration of the apical cavity. White arrow indicates cardiac apex.

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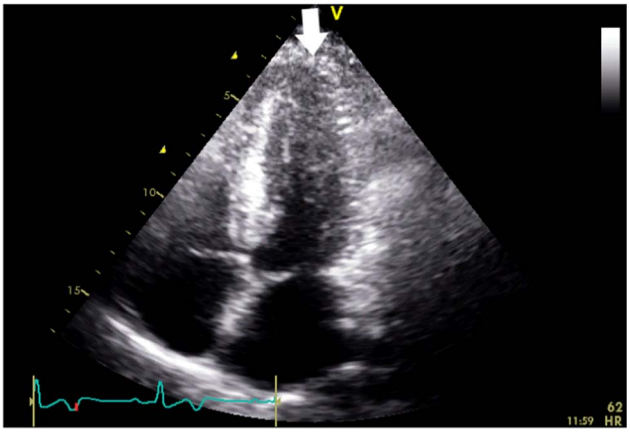

Figure 3. Transthoracic echocardiography showing progressive increase in wall thickness toward the left ventricular apex (white arrow) in the apical four-chamber view

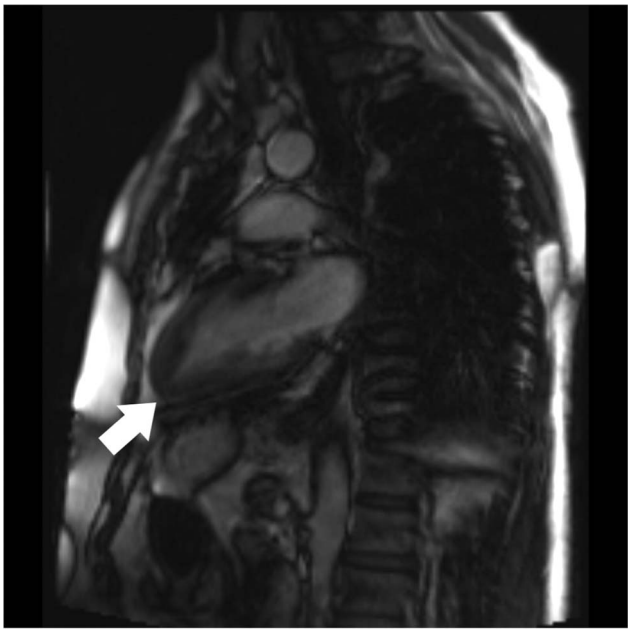

Figure 4. Cardiac magnetic resonance imaging demonstrating progressive thickening of myocardium toward the left ventricular apex (white arrow)

However, apical views were suboptimal, and no spade-like appearance of the apex was observed. On admission day three, the patient was discharged home on metoprolol.

Four months later, the patient was seen at an outpatient follow-up by his cardiologist and was well. Oral metoprolol was continued. Cardiac magnetic resonance imaging (MRI) was suboptimal but appeared to show an "ace-of-spades" configuration (Figure 4). A repeat transthoracic echocardiogram confirmed ApHCM with a progressive increase in wall thickness toward the apex with a wall thickness greater than 15 mm. A 24-hour Holter monitor showed predominantly a sinus rhythm with infrequent asymptomatic premature atrial contractions and no ventricular ectopy.

DISCUSSION

ApHCM is a mostly non-obstructive asymptomatic cardiomyopathy.[4-6] In rare situations in which patients are symptomatic, they may present with adverse sequelae including heart failure (related to LV diastolic dysfunction and elevated filling pressures),[12] angina, syncope, arrhythmia including atrial fibrillation, stroke from apical thromboembolism, and sudden cardiac death.[13,14] ApHCM was historically regarded as a phenotype largely unique to Japanese patients, in whom it was first described. It occurs more frequently in the Japanese population, with one comparative study revealing apical variants accounting for 15% and 3% of all hypertrophic cardiomyopathy within Japanese and Minnesotan cohorts, respectively.[15] Smaller studies have suggested a prevalence between 3% and 11% in North American cohorts.[1,6,16,17] The etiology of ApHCM is multifactorial, with studies suggesting variants with a genetic predisposition[18] and also demonstrating exclusive development during adulthood.[4] A family history is uncommon,[4,19] and genomic data regarding inheritance patterns, penetrance, and expressivity of known genetic mutations are presently unknown.[18]

Classically described ECG findings comprise "giant negative T waves (greater than 10 mm) associated with high QRS voltage (R wave greater than 26 mm in lead V5 or the sum of the S wave in lead V1 and the R wave in lead V5 35 mm or more)."[3] These voltage criteria may be related to both LV hypertrophy or to differences in localized wall thickness leading to disparities in the duration of repolarization.[2] Later studies emphasized any precordial giant negative T waves as the hallmark of ApHCM, although giant negative T waves may be present in as little as 11% of American Caucasian patients with ApHCM.[19] Giant negative T waves diminish with age[6] and are significantly more common in Japanese patients.[15] Notably, giant negative T waves may be seen in myocardial ischemia, right ventricular hypertrophy, electrolyte abnormalities, hypertensive heart disease, and cerebrovascular disease.[2,20,21]

Echocardiographic criteria described in research protocols include two-dimensional echocardiography

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showing asymmetric LV hypertrophy with apical predominance, an apical wall thickness of >15 mm, and a ratio of maximal apical to LV posterior wall thickness of >1.5 at end-diastole.[6] An end-diastolic "ace-of-spades" configuration on left ventriculography is considered pathognomonic for ApHCM. Moreover, this may reveal a midventricular obstruction and an apical aneurysm, two potentially overlapping morphologies with significant clinical implications.[5] Cardiac MRI may facilitate diagnosis even if echocardiography or ventriculography fail to help in the diagnosis of ApHCM initially[6] and may allow for early detection in the absence of the "ace-of-spades" configuration.[22]

Chest pain mimicking ACS in the setting of ApHCM has been previously reported,[8,10,11] including ST-segment elevation in contiguous leads resulting in the administration of thrombolytics,[7,9] as in this case. If patients with ApHCM develop angina, several mechanisms have been proposed. Apical infarction may occur because of systolic midventricular obstruction from cavitary obliteration, with subsequent diminished coronary perfusion or high wall tension from an apical aneurysm.[7] Impaired diastolic relaxation or systolic compression of vessels by a hypertrophied myocardial mass may result in inadequate coronary flow.[8,13] Lastly, there may be a component of a supply-demand mismatch with an increased apical myocardial mass without sufficient coronary perfusion.

There is no specific treatment for ApHCM.[10] Patients should be referred to a cardiologist for risk stratification, monitoring, and management. Given the heterogenous clinical spectrum, medical therapy is directed toward specific symptoms, including the judicious use of beta-blockers and diuretics for angina, arrhythmia, and heart failure. Patients with a concomitant apical aneurysm have a particularly poor prognosis.[23] Overall survival has ranged greatly from 47% at 20 years[19] to 95% at 15 years,[6] with heterogeneity in age at the first presentation playing a role in the discrepancies.

The diagnosis of ApHCM can prove challenging if patients present with acute chest discomfort and an abnormal ECG. Integral components of the initial diagnostic process in this instance include a careful evaluation of coronary risk factors, quality of chest discomfort, ECG pattern and progression, and cardiac biomarker evolution. Despite these factors, if patients present without a known history of apical hypertrophy, activation of ACS protocols with reperfusion may be warranted based on the best evidence available to clinicians at the time. While ApHCM may interfere with the clinical diagnosis because of ECG findings indicating infarction, it is equally as important to consider alternative explanations for chest pain in this setting.

In a retrospective evaluation of this case, given the equivocal ECG and lack of cardiac biomarker elevation, point-of-care echocardiography (POCE) may have been useful in distinguishing ApHCM from ACS, leading one to consider causes of pain such as a supply-demand mismatch as opposed to plaque rupture resulting in STEMI. The use of POCE in the setting of suspected ACS to recognize regional wall-motion abnormalities has been studied,[24] including by emergency physicians using a standardized protocol.[25] Kerwin and others have demonstrated that with brief standardized training, emergency physicians can improve their ability to recognize regional wall-motion abnormalities.[26] In the ED, the absence of regional wall-motion abnormalities on POCE has been used to rule out ACS in the setting of ST-segment elevation resulting from pulmonary embolism[27] and hypercalcemia.[28] Herein, we suggest that POCE performed in the ED with the support of a cardiologist may have allowed for recognition of the apical hypertrophy as the cause of the patient's ECG findings.

Our case illustrates the complex manner in which ApHCM may present in the era of rapid early reperfusion therapy. It should reinforce the idea that not all chest pain with simultaneous ST-segment elevation is a result of myocardial infarction. For patients with ECG findings suggestive of alternate diagnoses, POCE may be useful in guiding further management.

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REFERENCES

1. Chikamori T, Doi YL, Akizawa M, et al. Comparison of clinical, morphological, and prognostic features in hypertrophic cardiomyopathy between Japanese and western patients. *Clin Cardiol* 1992;15(11):833-7.
2. Sakamoto T, Tei C, Murayama M, et al. Giant T wave inversion as a manifestation of asymmetrical apical hypertrophy (AAH) of the left ventricle. Echocardiographic and

https://doi.org/10.1017/cem.2017.409 Published online by Cambridge University Press

ultrasono-cardiotomographic study. *Jpn Heart J* 1976; 17(5):611-29.

3. Yamaguchi H, Ishimura T, Nishiyama S, et al. Hypertrophic nonobstructive cardiomyopathy with giant negative T waves (apical hypertrophy): ventriculographic and echocardiographic features in 30 patients. *Am J Cardiol* 1979; 44(3):401-12.
4. Sakamoto T, Amano K, Hada Y, et al. Asymmetric apical hypertrophy: ten years experience. *Postgrad Med J* 1986; 62(728):567-70.
5. Minami Y, Haruki S, Hagiwara N. Phenotypic overlap in hypertrophic cardiomyopathy: apical hypertrophy, midventricular obstruction, and apical aneurysm. *J Cardiol* 2014;64(6):463-9.
6. Eriksson MJ, Sonnenberg B, Woo A, et al. Long-term outcome in patients with apical hypertrophic cardiomyopathy. *J Am Coll Cardiol* 2002;39(4):638-45.
7. Lin CS, Chen CH, Ding PY. Apical hypertrophic cardiomyopathy mimicking acute myocardial infarction. *Int J Cardiol* 1998;64(3):305-7.
8. Cubukçu AA, Scott PJ, Williams GJ. Apical hypertrophic cardiomyopathy presenting as acute subendocardial myocardial infarction. *Int J Cardiol* 1993;38(3):329-32.
9. Sayin T, Koçum T, Kervancioglu C. Apical hypertrophic cardiomyopathy mimics acute coronary syndrome. *Int J Cardiol* 2001;80(1):77-9.
10. Olearczyk B, Gollol-Raju N, Menzies DJ. Apical hypertrophic cardiomyopathy mimicking acute coronary syndrome: a case report and review of the literature. *Angiology* 2008;59(5):629-31.
11. Diaconu CC, Dumitru N, Fruntelata AG, Lacau S, Bartos D. Apical Hypertrophic Cardiomyopathy: The Ace-of-Spades as the Disease Card. *Acta Cardiol Sin* 2015;31(1):83-6.
12. 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(1):83-90.
13. Jan MF, Todaro MC, Oreto L, Tajik AJ. Apical hypertrophic cardiomyopathy: present status. *Int J Cardiol* 2016;222:745-59.
14. Moon J, Shim CY, Ha JW, et al. Clinical and echocardiographic predictors of outcomes in patients with apical hypertrophic cardiomyopathy. *Am J Cardiol* 2011; 108(11):1614-9.
15. Kitaoka H, Doi Y, Casey SA, et al. Comparison of prevalence of apical hypertrophic cardiomyopathy in Japan and the United States. *Am J Cardiol* 2003;92(10):1183-6.
16. Wigle ED, Sasson Z, Henderson MA, et al. Hypertrophic cardiomyopathy. The importance of the site and the extent of hypertrophy. A review. *Prog Cardiovasc Dis* 1985;28(1): 1-83.
17. Cannan CR, Reeder GS, Bailey KR, Melton LJ 3rd, Gersh BJ. Natural history of hypertrophic cardiomyopathy. A population-based study, 1976 through 1990. *Circulation* 1995;92(9):2488-95.
18. Towe EC, Bos JM, Ommen SR, Gersh BJ, Ackerman MJ. Genotype-Phenotype Correlations in Apical Variant Hypertrophic Cardiomyopathy. *Congenit Heart Dis* 2015; 10(3):E139-45.
19. Klarich KW, Attenhofer Jost CH, Binder J, et al. Risk of death in long-term follow-up of patients with apical hypertrophic cardiomyopathy. *Am J Cardiol* 2013; 111(12):1784-91.
20. Yamanari H, Saito D, Mikio K, et al. Apical hypertrophy associated with rapid T wave inversion on the electrocardiogram. *Heart Vessels* 1995;10(4):221-4.
21. Jacobson D, Schrire V. Giant T wave inversion. *Br Heart J* 1966;28(6):768-75.
22. Suzuki J, Shimamoto R, Nishikawa J, et al. Morphological onset and early diagnosis in apical hypertrophic cardiomyopathy: a long term analysis with nuclear magnetic resonance imaging. *J Am Coll Cardiol* 1999;33(1): 146-51.
23. Chen CC, Lei MH, Hsu YC, Chung SL, Sung YJ. Apical hypertrophic cardiomyopathy: correlations between echocardiographic parameters, angiographic left ventricular morphology, and clinical outcomes. *Clin Cardiol* 2011; 34(4):233-8.
24. Kontos MC, Arrowood JA, Paulsen WH, Nixon JV. Early echocardiography can predict cardiac events in emergency department patients with chest pain. *Ann Emerg Med* 1998;31(5):550-7.
25. Frenkel O, Riguzzi C, Nagdev A. Identification of high-risk patients with acute coronary syndrome using point-of-care echocardiography in the ED. *Am J Emerg Med* 2014; 32(6):670-2.
26. Kerwin C, Tommaso L, Kulstad E. A brief training module improves recognition of echocardiographic wall-motion abnormalities by emergency medicine physicians. *Emergency Medicine International* 2011;2011:483242.
27. Shy BD, Gutierrez A, Strayer RJ. Bedside Ultrasound to Evaluate Pulmonary Embolism Masquerading as ST Elevation Myocardial Infarction (STEMI). *Journal of Emergency Medicine* 2015;49(5):703-4 e1.
28. Strand AO, Aung TT, Agarwal A. Not all ST-segment changes are myocardial injury: hypercalcaemia-induced ST-segment elevation. *BMJ Case Reports* 2015;2015.

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