Images in Cardiovascular Medicine

SPECT Myocardial Perfusion Imaging

in the Diagnosis of Apical Hypertrophic Cardiomyopathy

Shihab Masrur, MD
Sarfaraz Memon, MD
Lovely Chhabra, MD
Matthew W. Parker, MD
Lane Duvall, MD

A 52-year-old man presented at our emergency department after suffering a fall that was possibly caused by syncope. In recollecting the episode, the patient reported a substernal burning sensation with exertion, but no chest pain. He recalled no palpitations, dizziness, presyncopal aura, or post-ictal confusion. He was a nonsmoker and reported no alcohol or illicit-drug abuse. His father had sustained a myocardial infarction, but there was no other family history of cardiomyopathy. Auscultation revealed normally split S_1 and S_2 sounds, along with a grade 2/6 systolic murmur, loudest at the right sternal border.

An electrocardiogram revealed giant T-wave inversions in the precordial and high lateral leads, without septal Q waves (Fig. 1). Exercise stress testing revealed no ischemia or arrhythmia. Single-photon-emission computed tomographic (SPECT) myocardial perfusion images suggested left ventricular (LV) hypertrophy (confined to the apical portion of the ventricle and more apparent at rest), and mild ischemia in the true apex of the ventricle (Fig. 2). Two-dimensional transthoracic echocardiography with microbubble contrast revealed marked hypertrophy of the apical third of the LV and complete annihilation of the apical portion of the LV cavity during systole (Fig. 3).

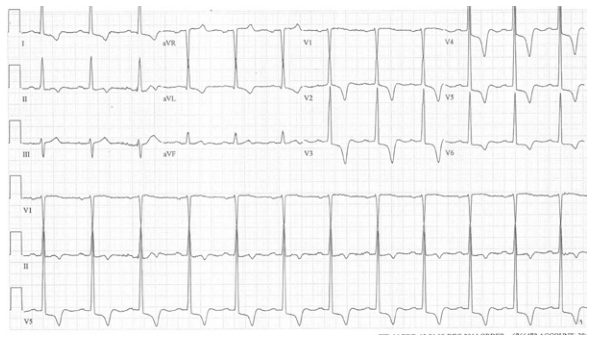

Fig. 1 *Electrocardiogram shows sinus rhythm, a prolonged PR interval, left ventricular hypertrophy, ST-segment depressions in the anterior precordial leads, and giant T-wave inversions in the precordial and high lateral leads.*

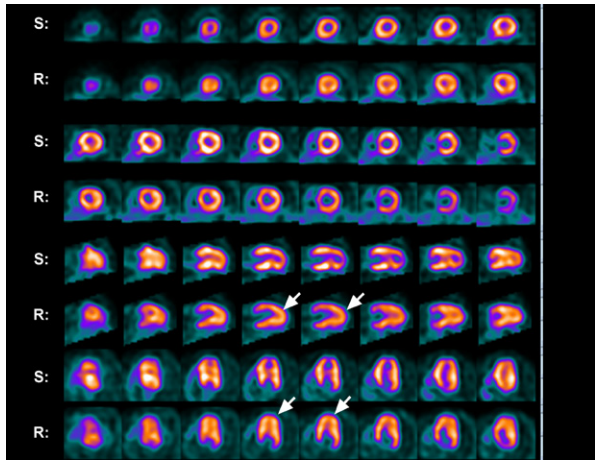

Fig. 2 *Myocardial perfusion single-photon-emission computed tomogram shows increased tracer uptake in the apex and a spade-like image of the left ventricle. Perfusion images during stress (S) and the corresponding images at rest (R) are shown; arrows indicate an ace-of-spades appearance.*

Section Editor: *Raymond F. Stainback, MD, Department of Adult Cardiology, Texas Heart Institute, 6624 Fannin St., Suite 2480, Houston, TX 77030*

From: *Department of Cardiology (Drs. Chhabra, Duvall, Masrur, Memon, and Parker), Hartford Hospital, Hartford, Connecticut 06201; and Department of Medicine (Drs. Duvall and Parker), University of Connecticut School of Medicine, Farmington, Connecticut 06030*

Address for reprints: *Shihab Masrur, MD, 80 Seymour St., Hartford, CT 06201*

E-mail: *shihabmasrur@hotmail.com*

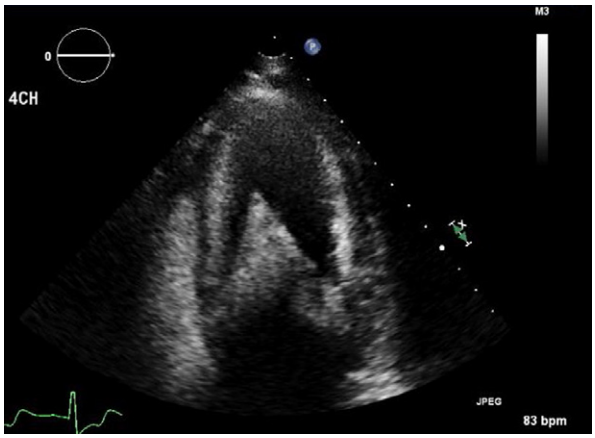

Fig. 3 *Two-dimensional transthoracic echocardiogram (apical 4-chamber view), enhanced by microbubble contrast, shows the ace-of-spades appearance of the left ventricle.*

Comment

Apical hypertrophic cardiomyopathy (HCM) is a variant of HCM characterized by local hypertrophy of the LV apex. Echocardiography usually reveals an "ace of spades" appearance of the ventricle, but echocardiographic imaging can be limited by suboptimal acoustic windows or technique.[1] Findings on SPECT myocardial perfusion imaging are also characteristic of apical HCM.[2] In presenting this case of apical HCM, we correlate the echocardiographic and SPECT images.

Because apical HCM can mimic symptomatic obstructive coronary artery disease, many patients undergo cardiac stress testing for possible ischemia. In regard to the electrocardiogram of apical HCM, the image is characteristic almost to the point of being pathognomonic—so the level of suspicion should be high.

Another useful imaging tool in the diagnosis of apical HCM is myocardial SPECT imaging. Nuclear cardiologists should be aware of myocardial SPECT findings in order to avoid inappropriate downstream testing—apical HCM can masquerade on SPECT as significant myocardial ischemia, probably secondary to closure of the epicardial coronary vasculature during systole. In our patient, a perfusion defect is seen in the correlated stress image. This defect might be due to severe mid/apical obliteration during stress, which can cause subendocardial ischemia and thereby lead to the rather paradoxical enlargement of the apical cavity during stress SPECT imaging. Stress echocardiography can be useful in correlating images in such cases. (In our patient, stress echocardiography was not performed.) Another possible explanation of the apical perfusion defect in our patient could be the demand–supply mismatch during stress: in cases of apical HCM, the myocardial mass is relatively more abundant at the apex.

The prognosis of apical HCM is benign. However, some patients, over time, will develop cardiac complications like diastolic dysfunction, left atrial enlargement, apical thrombi, and ventricular aneurysms. Therefore, periodic follow-up evaluation with echocardiography is recommended.

References

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2. Cianciulli TF, Saccheri MC, Masoli OH, Redruello MF, Lax JA, Morita LA, et al. Myocardial perfusion SPECT in the diagnosis of apical hypertrophic cardiomyopathy. J Nucl Cardiol 2009;16(3):391-5.