

SPECT Myocardial Perfusion Imaging in the Diagnosis of Apical Hypertrophic Cardiomyopathy- Case Series and Literature Review

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ABSTRACT

Apical hypertrophic cardiomyopathy (AHCM) is a subtype of hypertrophic cardiomyopathy (HCM) in which hypertrophy mostly affects the apex of the left ventricle, resulting in mid-ventricular obstruction. The diagnosis is usually made when the LV apex has an apical wall thickness of ≥ 15 mm in echocardiography, though sometimes it is missed due to the poor acoustic window in two-dimensional echocardiography. Single Photon Emission Computed Tomography-Myocardial Perfusion Imaging (SPECT-MPI) can often detect apical hypertrophy. The apical hypertrophy was identified by SPECT-MPI in the reported three cases, which were not previously diagnosed by echocardiography.

Keywords: Apical hypertrophic cardiomyopathy, Single Photon Emission Computed Tomography, Myocardial Perfusion Imaging

Bangladesh J. Nucl. Med. Vol. 27 No. 1 January 2024

DOI : <https://doi.org/10.3329/bjnm.v27i1.71520>

INTRODUCTION

Apical hypertrophic cardiomyopathy is a variant of hypertrophic cardiomyopathy (HCM) in which hypertrophy predominantly involves the apex of the left ventricle, resulting in mid-ventricular obstruction, as opposed to the left ventricular outflow tract obstruction seen in HCM. Patients with apical HCM may present with angina, heart failure, myocardial infarction, syncope, or arrhythmias and are managed with medications. Here we present three cases that came to NINMAS for Single Photon Emission Computed Tomography (SPECT)-Myocardial Perfusion Imaging (MPI) and were diagnosed as apical hypertrophy by SPECT-MPI.

CASE 1

A 60 years old male presented with central chest pain on exertion and referred to the nuclear cardiology division of

National Institute of Nuclear Medicine & Allied Sciences (NINMAS) for a MPI examination. Patient was non-diabetic, normotensive with no family history of ischemic heart disease (IHD). He also denied any family history for sudden cardiac death, seizure activity, or syncopal episodes. Physical examination findings were unremarkable. The patient's electrocardiography (ECG) demonstrated normal sinus rhythm with left ventricular hypertrophy and T wave inversion in the inferolateral leads. His 2-dimensional Echocardiography revealed hypokinesia of the anterior apical segment, small left ventricular cavity with apical hypertrophy (Thickness- 19 mm). His coronary angiogram revealed no definite abnormality. He had undergone a single day stress rest ^{99m}Tc-sestamibi myocardial perfusion scan. MPI showed increased tracer uptake in the apex with dyskinesia and suspected to be having apical hypertrophy. Further echocardiography confirmed the diagnosis as the apical thickness was about 17 mm.



Figure 1: Echocardiography image of 60 years old male patient showing thickened apex (1.9cm)

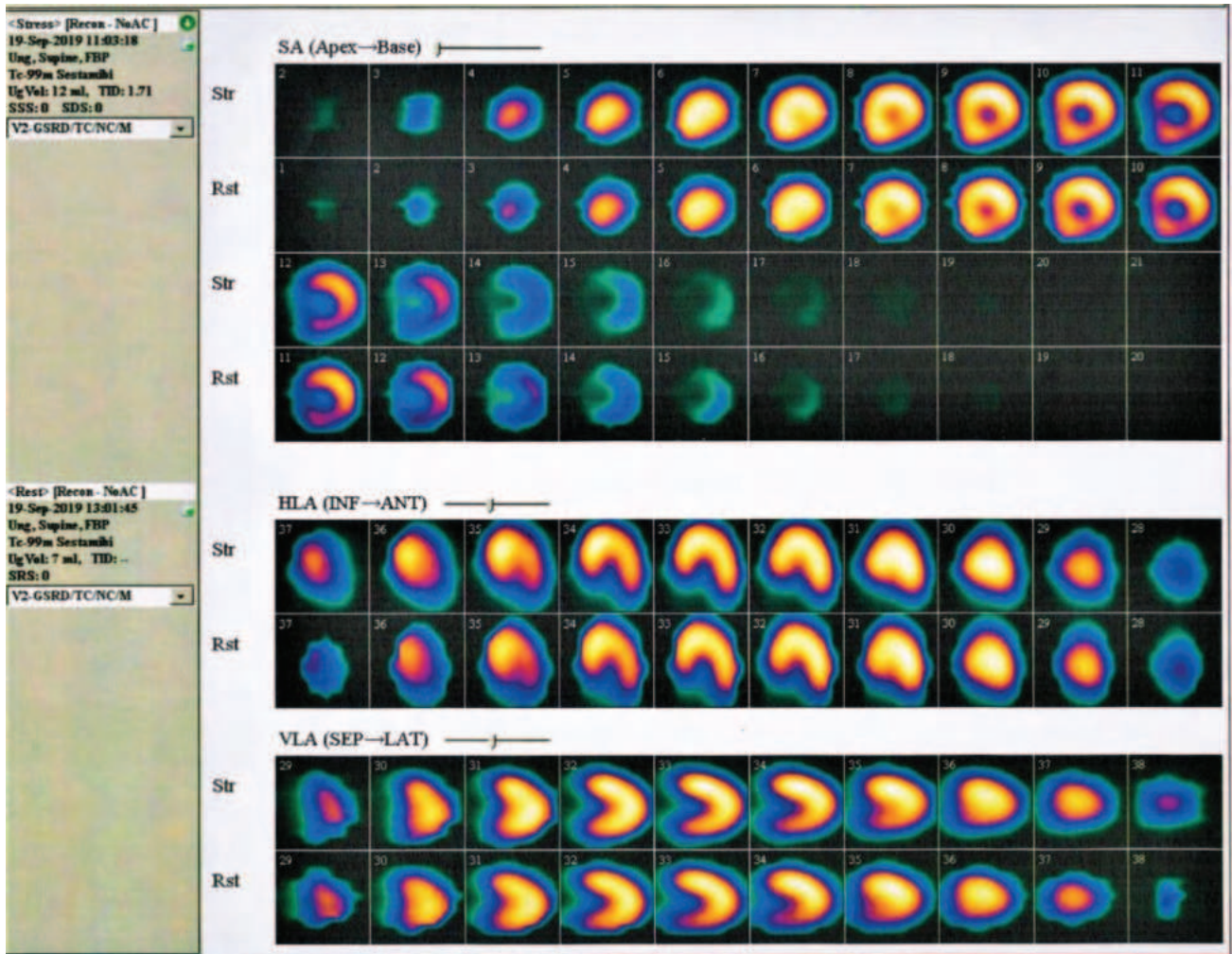


Figure 2 : Myocardial perfusion scan image of the same patient with hypertrophied apex

CASE 2

A 56-year-old woman, normotensive, non-diabetic, and dyslipidemic, habituated to betel nuts and jarda and family history of IHD, presented with occasional chest pain. She had a previous history of heart attack five years ago. Her ECG showed non-specific ST-T wave abnormality. However, echocardiography reported preserved ejection fraction (63%), grade 2 diastolic dysfunction (mild restrictive pattern), moderately dilated left atrium (63 ml volume), mildly elevated pulmonary artery systolic pressure (35 mm Hg), and left ventricular hypertrophy. Angiography was not done. The patient underwent a single-day stress-relief MPI. Rest and stress imaging demonstrated unusual morphology, with prominent apex. Her myocardial perfusion study was normal, with increased radiotracer uptake in the apex and associated dyskinesia. Then we sent the patient to review the echocardiography, which

revealed a small left ventricular cavity with apical hypertrophy. (Thickness: 16 mm)

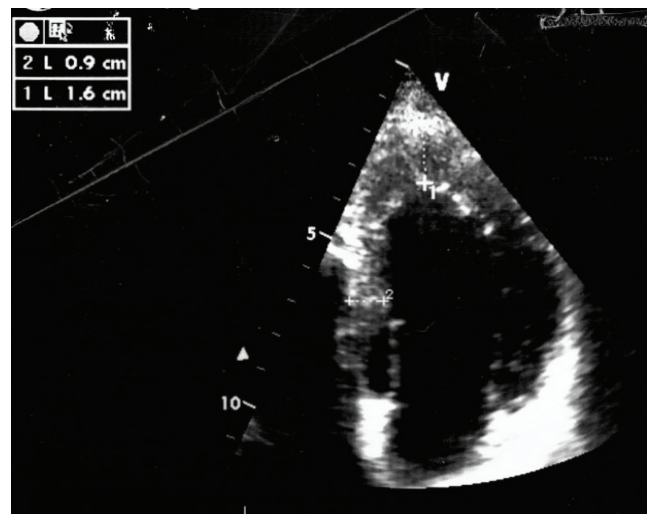


Figure 3: Echocardiography image of a 56-year-old woman showing thickened apex (1.6cm)

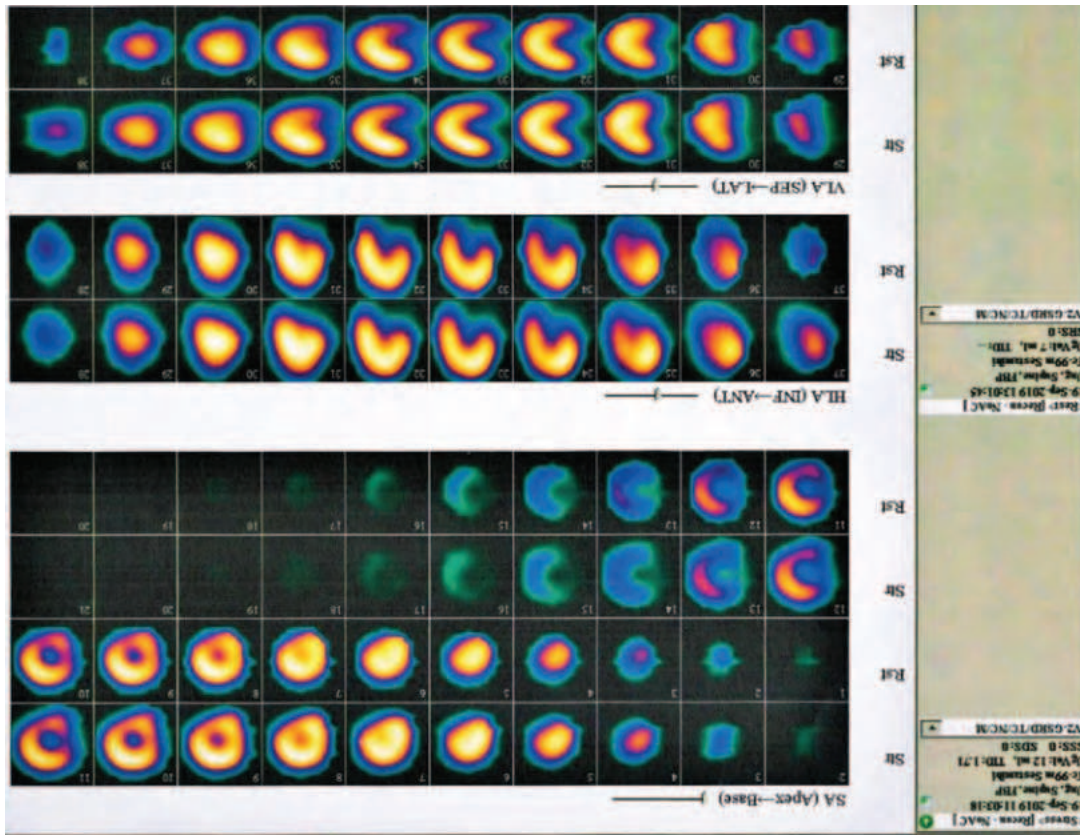


Figure 4: Myocardial perfusion scan image of same patient showing apical hypertrophy

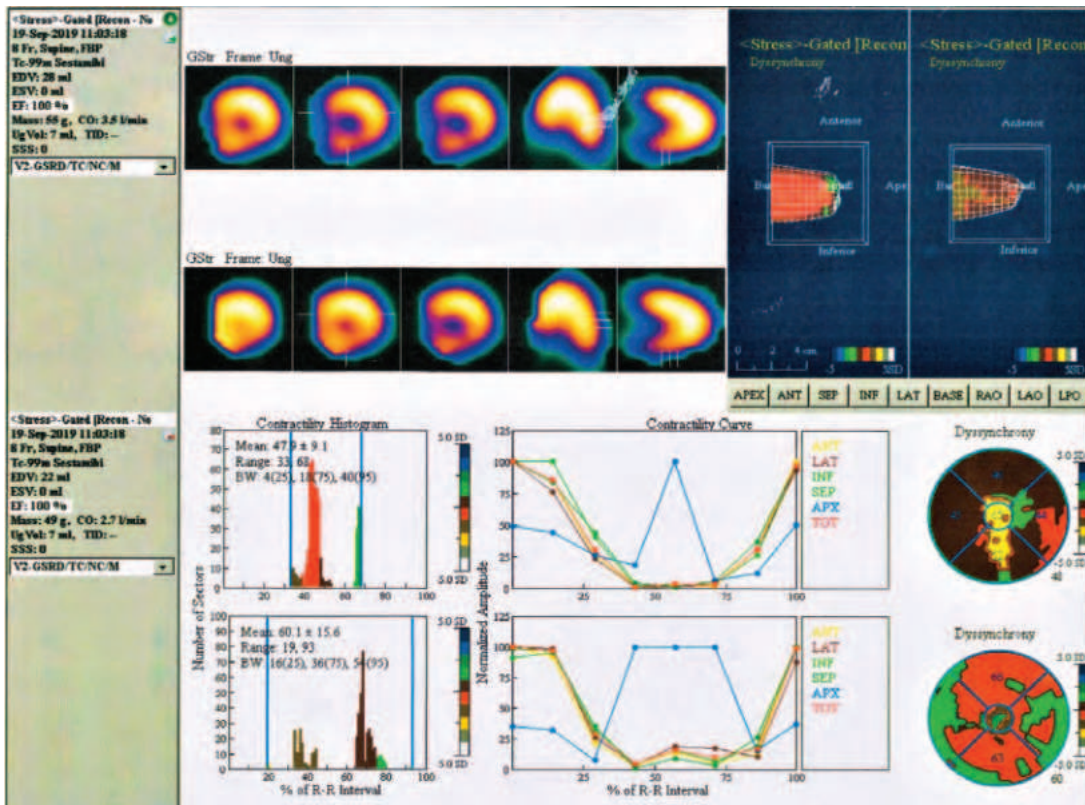


Figure 5 : Myocardial perfusion scan image showing myocardial contractility curve of same patient with dyskinetic apex and normally contracted other walls

CASE 3

A 52-year-old woman, hypertensive, diabetic, and dyslipidemic with a family history of IHD, presented with occasional central chest pain on exertion. Her echocardiography showed non-specific findings with a preserved ejection fraction (59%). The patient underwent single-day stress rest (MPI). Rest and stress imaging

demonstrated unusual morphology, with the apex appearing prominent. Her myocardial perfusion study was normal, with increased radiotracer uptake in the apex and associated dyskinesia. Our suspicion was again aroused, and we sent this patient to our cardiology department for a repeat echo. They found the apical thickness to be higher than usual (thickness: 20 mm).

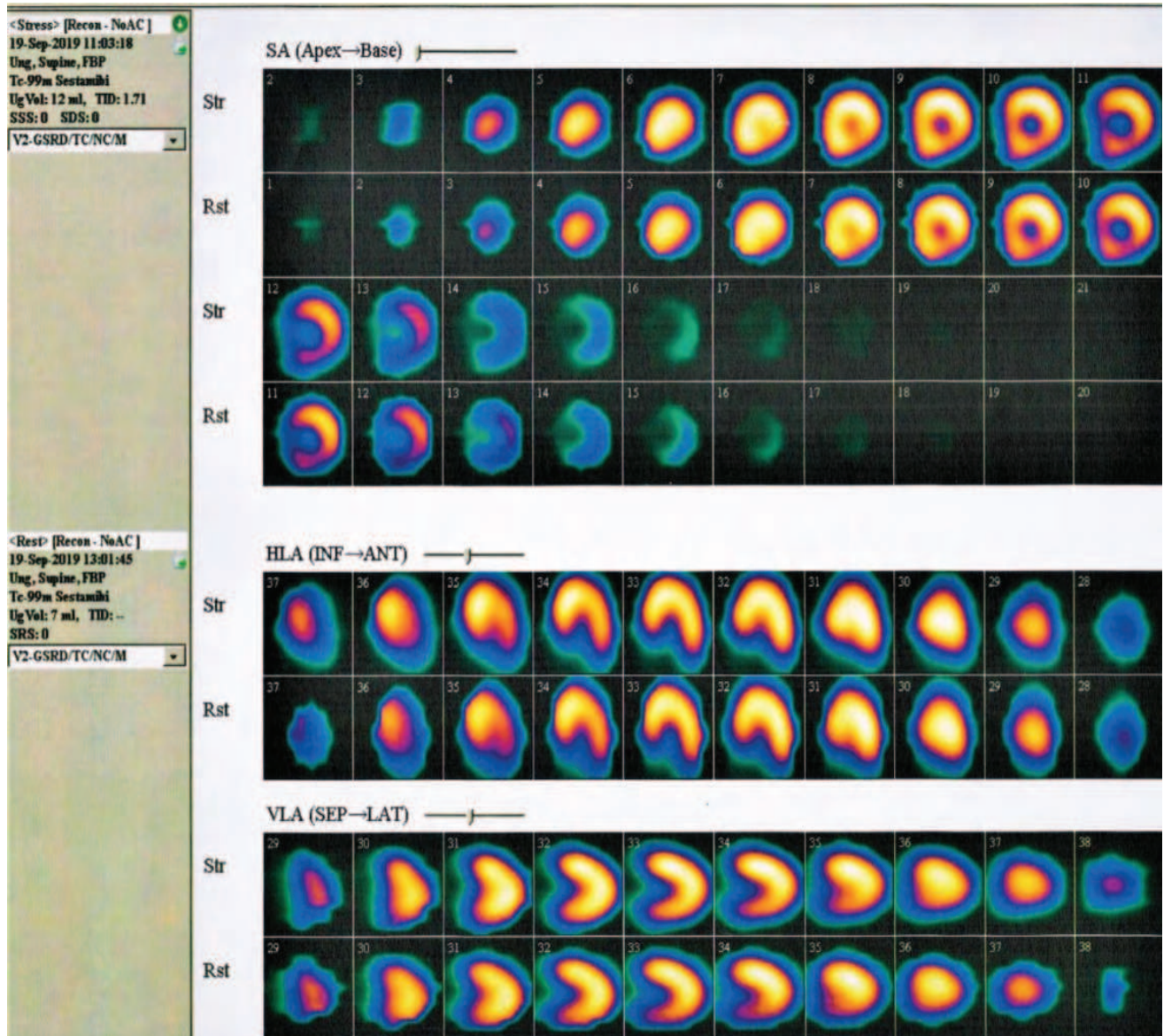


Figure 6: Myocardial perfusion scan image showing marked apical hypertrophy

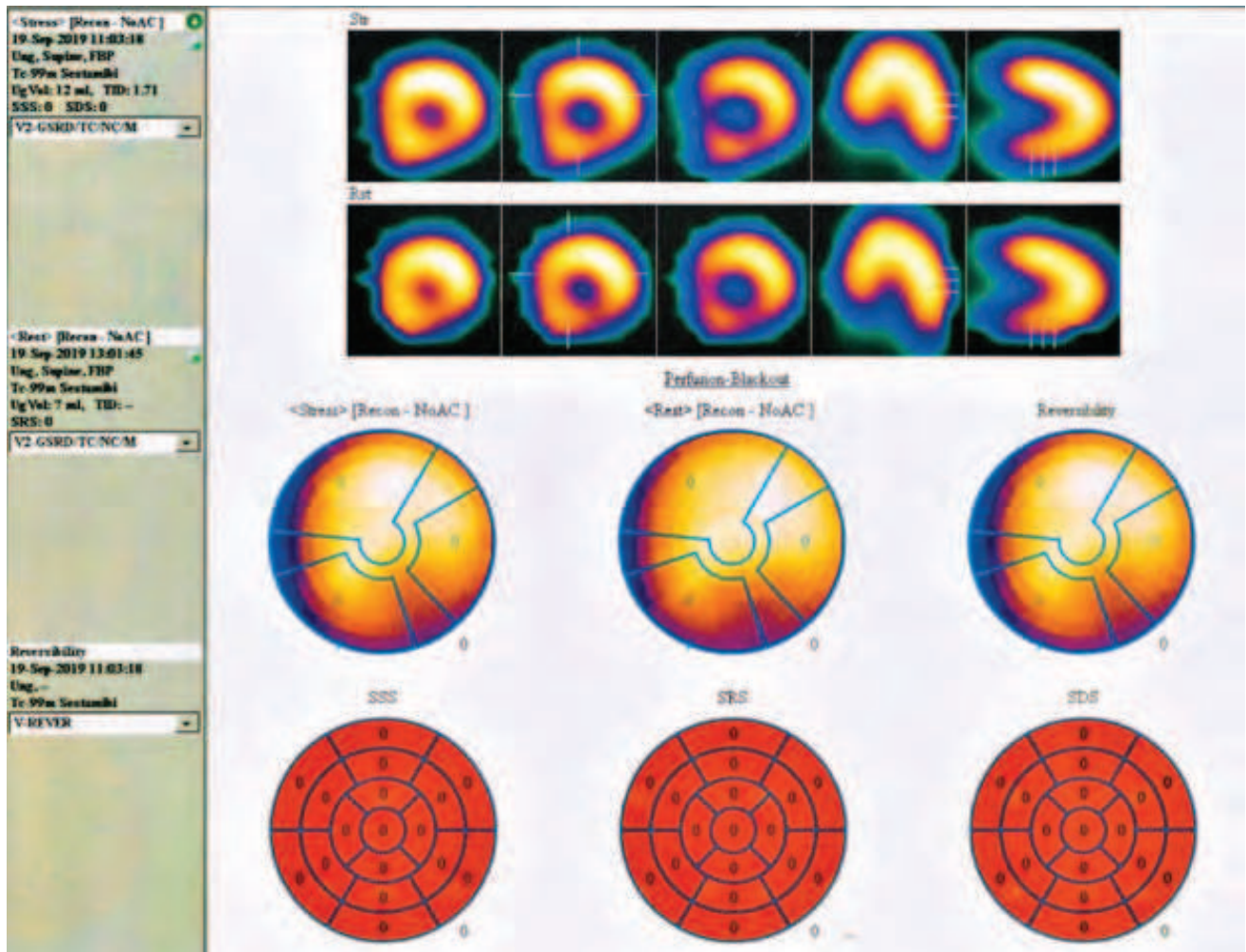


Figure 7: 17 segment perfusion scan image case 3 with normal perfusion and thickened apex

DISCUSSION

Apical HCM is a more common variant that occurs in Asian non-Asians, with the prevalence reported to be as high as 41% of the HCM patients in China (1) and in 15% of HCM patients in Japan (2). In non Asian population, reports suggest a prevalence between 1% and 3% of HCM patients (3-5). The pathophysiology is thought to be associated with sarcomere gene mutations (6). But a direct genetic link has not been established. Patients may present asymptomatic or symptomatic. Most patients with apical HCM are asymptomatic, and those who do develop symptoms usually present with angina, heart failure, myocardial infarction, pre-syncope, syncope, atrial fibrillation, or ventricular fibrillation. These occur usually due to diastolic dysfunction with low cardiac output (3). The typical clinical and diagnostic features of apical HCM include audible fourth heart sounds and “giant” negative T

waves on the ECG.

Spectrum of findings applicable to AHCM. The most common finding is the spade-shaped appearance of LV in resting VLA and HLA slices due to excessive tracer uptake over the hypertrophied apex (7). In all of our patients, this finding was present.

Reversible or irreversible perfusion abnormalities have been reported in 50% of patients in a few studies (7). In these patients, an inducible ischemia involving the apex, apical anteroseptal, and inferior walls with normal epicardial coronaries raises the possibility of microvascular disease in AHCM. The indication of MPI to characterize HCM is not well established (10). ECHO and CMR have both been described in the literature as the most sensitive and specific imaging modalities for the diagnosis of HCM (11).

Sometimes echocardiography fails to diagnose AHCM. Though Doppler echocardiography has become the gold standard for the diagnosis of hypertrophic cardiomyopathy, there are many pitfalls in its use. Some of these pitfalls are technical in nature, resulting from inadequate image quality, incorrect transducer angulation, and improper equipment settings.

CMR helps in the diagnosis of HCM by identifying areas of hypertrophy not well visualized by echocardiography, providing more accurate wall thickness measurements, and differentiating HCM from other causes of left ventricular (LV) hypertrophy. But in some cases where CMR is contraindicated, MPI may be a helpful tool in the diagnosis of AHCM.

Previous studies of stress MPI in patients with AHCM have diagnosed normal to reversible apical perfusion defects along with normal coronary arteries (8). The first two cases demonstrated characteristic ECG changes in left precordial leads, apical wall motion abnormalities including hypokinesis and aneurysm formation, and a “spade-like” configuration of the left ventricular cavity at end-diastole on imaging.

Increased apical tracer uptake in SPECT-MPI horizontal and vertical long axis slices that are consistent with the diagnosis of AHCM (8, 10). The apical hypertrophy produces a focal hotspot, with apparent decreased intensity in the mid and basal segments.

Myocardial SPECT imaging is a useful imaging tool in the diagnosis of apical HCM. Nuclear cardiologists should be aware of myocardial SPECT findings in order to avoid inappropriate other tests for the diagnosis of apical HCM. Sometimes significant myocardial ischemia may occur in patients with AHCM, probably secondary to the closure of the epicardial coronary vasculature during systole. But our three patients showed no perfusion defect in stress images. Stress echocardiography can be useful in correlating images in such cases. In our patients, stress echocardiography was not performed.

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.

CONCLUSION

Though earlier studies suggested AHCM as a benign clinical course, recent studies report increased morbidity and mortality. So, when we make a differential diagnosis for chest pain, structural heart disease, including apical and other variants of hypertrophic cardiomyopathy, should be kept in mind. Furthermore, when screening tests such as echocardiography cannot adequately establish the diagnosis of AHCM, then SPECT-MPI findings in association with ECG and ECHO findings can be a good diagnostic tool for the diagnosis of AHCM.

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