

CASE REPORT

Apical hypertrophic cardiomyopathy: Is it a conundrum for Gated SPECT MPI?

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Abstract

Apical hypertrophic cardiomyopathy, AHCM, is common in Japanese and Chinese populations but uncommon in western population and in IndoPak subcontinent. AHCM patients tend to have milder symptoms; however, in up to one-third cases, myocardial infarction, left ventricular aneurysm formation, atrial and ventricular arrhythmias and strokes may develop. Echocardiography and cardiac MRI are the good modalities for diagnosis. Gated SPECT myocardial perfusion imaging is a good alternative for diagnosis due to well defined scan patterns in AHC. A case of AHCM is presented and the subject is briefly reviewed.

***Key words**: apical hypertrophic cardiomyopathy, cardiac perfusion scan, hypertrophic cardiomyopathy*

Introduction

Hypertrophic cardiomyopathy (HCM) is an important clinical entity characterized by an excessive myocardial mass with resultant small ventricular cavity. The left ventricle is predominantly involved with the process initiating in the septum in most patients with associated frequent appearance of asymmetrical septal hypertrophy. More recently, genetic transmission and mutations have been identified and are likely to contribute to its etiology and hence screening of family members is recommended.

Apical HCM (AHCM) is a rare variant characterized by predominant involvement of the apex and varying degrees of extension basally [1]. Although uncommon in the Western world [2], AHCM is relatively common in Japanese and Chinese populations, accounting for up to 40% in these areas [1].

Typical electrocardiographic and angiographic criteria features of AHCM include giant negative T waves on the electrocardiogram (ECG), an "ace-of-spades" configuration of the left ventricle at end-diastole and absence of an intraventricular pressure gradient on contrast ventriculography.

Patients can present with symptoms of angina and dyspnea with a predominant exertional component. AHCM patients tend to have milder symptoms; however, in up to one-thirds of the cases, myocardial infarction, left

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ventricular aneurysm formation, atrial and ventricular arrhythmias and strokes may develop [2]. Atrial arrhythmias (due to atrial dilation) and serious ventricular arrhythmias have been described in patients with AHCM. Patients with ventricular arrhythmias are at a high risk for sudden cardiac death (SCD) and should have further electrophysiological evaluation.

In current clinical practice, the diagnosis of AHCM is usually made by echocardiograph. However, the apical and the lateral forms of HCM may be missed, particularly when contrast is not used [3] or as a result of a poor acoustic window. Cardiac MRI (CMR) is found to be a powerful tool in finding out "missing spade" in such situations [1]. Gated myocardial perfusion single photon emission computerized tomography (GSPECT) is frequently indicated in patients with markedly abnormal ECG with or without symptoms. AHCM is diagnosed on GSPECT by abnormally increased tracer uptake over apex at rest (spade shape deformity), solar polar map pattern at rest with or without perfusion defects (both reversible and fixed) [4-6]. There is only one report of post-stress

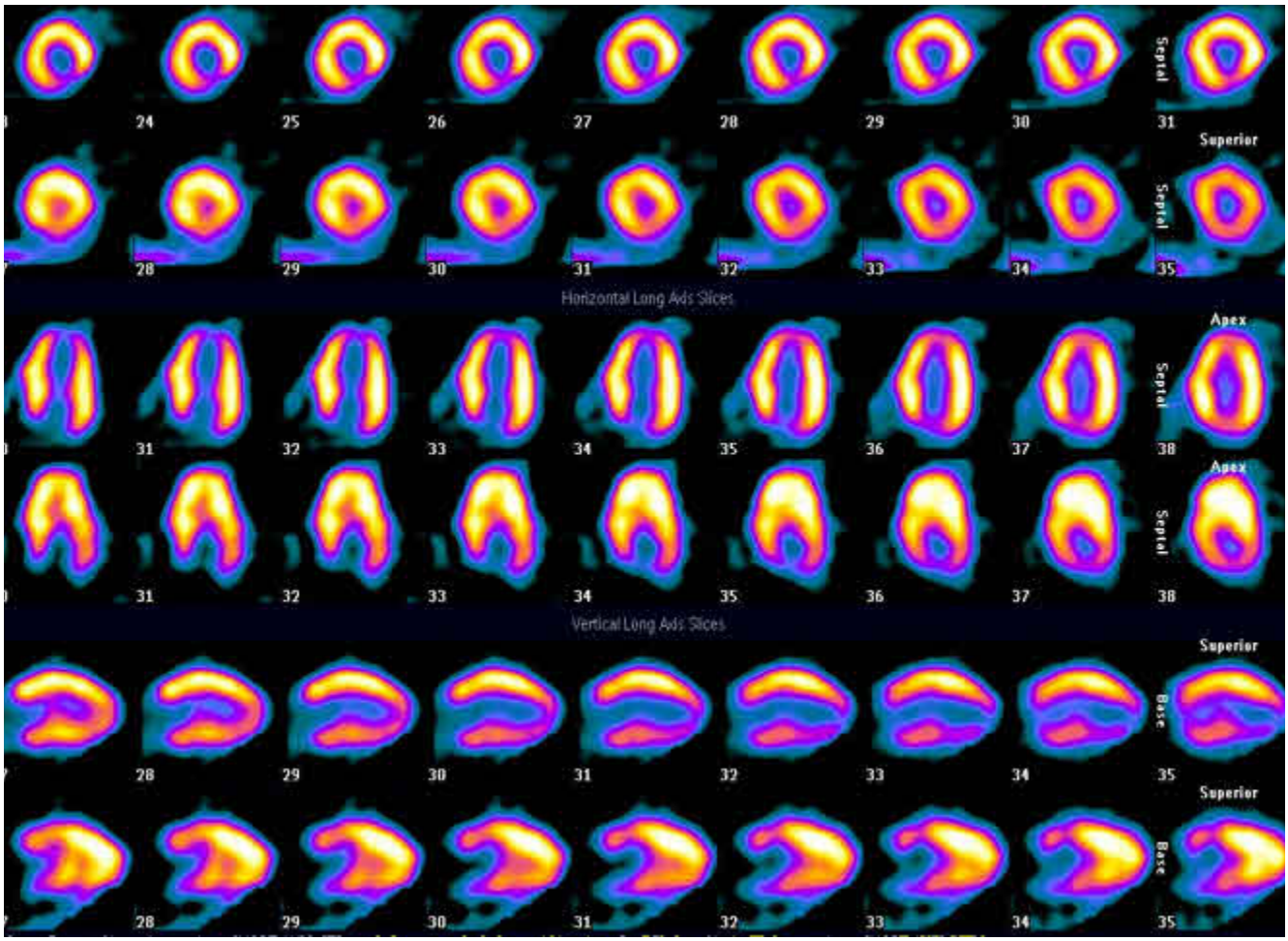

Figure 1 *Stress-Rest Gated Myocardial Perfusion scan revealed a medium size inducible ischemia of high intensity involving apex and distal inferolateral wall with evidence of transient ischaemic dilatation. Resting VLA and HLA images show "ace-of-spade" pattern of the LV cavity*

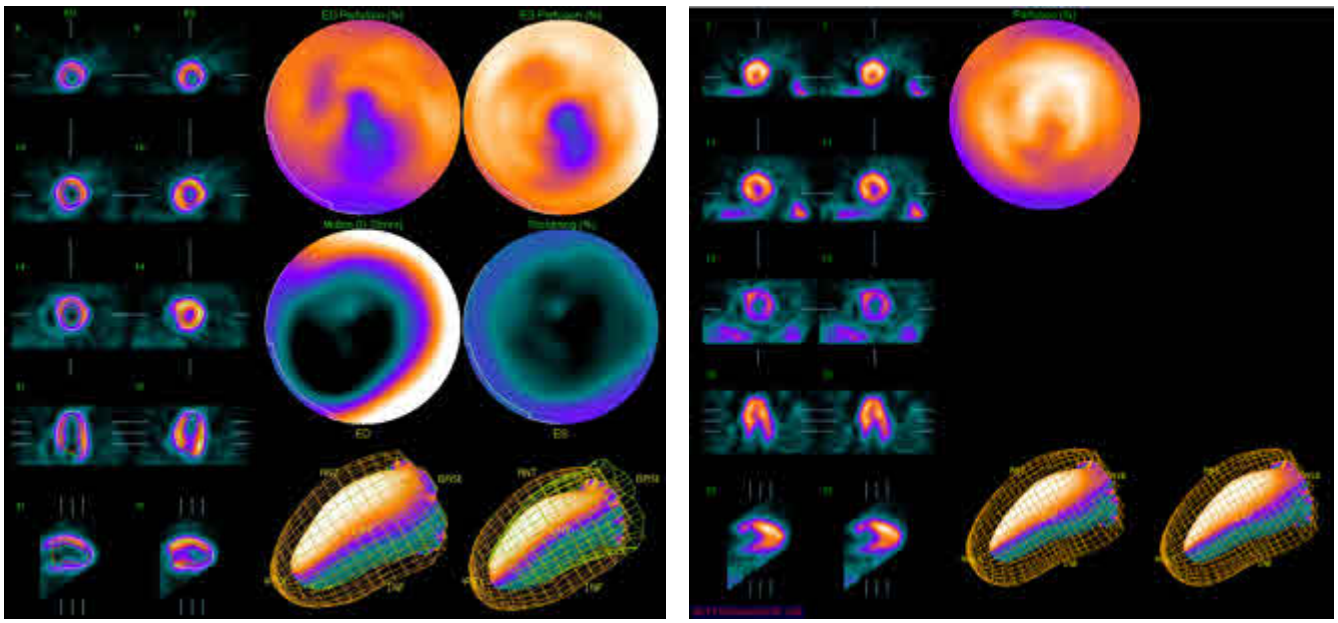

Figure 2 *Rest-stress polar maps. Stress map (a) shows a severe perfusion defect involving apex and inferior wall. Resting map (b) shows solar polar pattern with reduced slightly reduced count over inferior segments in resting polar map due to incomplete recovery of stress induced ischemia at time of resting injection. Stress map (a) shows a severe perfusion defect involving apex and inferior wall*

stunning and transient ischemic dilatation (TID) in a patient with AHCM [7].

We present an interesting case of AHCM referred to the Nuclear Cardiology Department of Karachi Institute of Heart Diseases (KIHD) whose GSPECT study showed the complete spectrum of findings including a spade-shape deformity and solar polar pattern on resting images, severe reversible perfusion defect, abnormal wall-motion and TID. A subsequent coronary angioghram showed normal epicardial coronaries.

Case History

A 51-year-old male presented with a history of progressive exertional dyspnoea for the last six months. The patient was hypertensive, though well controlled on medications, and an ex-smoker. The patient did not have diabetes nor was there a family history for coronary artery disease (CAD). His past medical history was unremarkable. Laboratory tests including a complete blood picture and lipid profile were normal. Chest x-ray and transthoracic 2D echocardiogram were also normal. However, the resting ECG revealed RBBB with inverted T waves in the precordial leads. The patient was referred to the Nuclear Cardiology Department of KIHD for myocardial perfusion imaging (MPI).

Treadmill stress was performed according to Bruce protocol, but discontinued after four minutes only, when the patient developed severe dysponea together with a hypertensive response. No significant ECG changes were noted during the stress or in recovery phase. The patient achieved 76% target heart rate at 5.8 METS. One minute before termination of stress, the patient was injected with 370 MBq of 99mTc-Methoxy IsoButyl Isonitrile (MIBI) and 20 minutes postinjection, gated single photon emission computerized tomography (GSPECT) was performed using a dual-head gamma camera (Cardio MD, Philips) fitted with high resolution collimators. Commercially available software (Astonish® and Autoquan®)

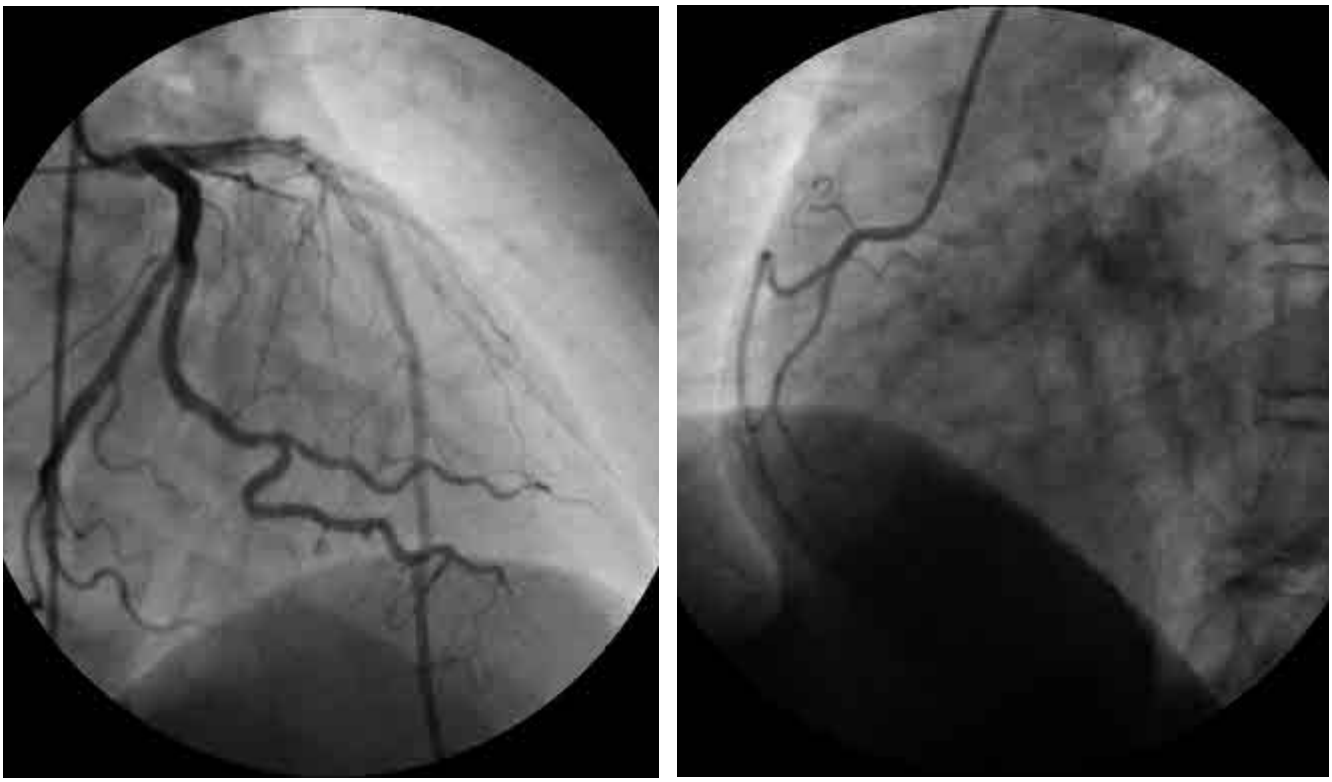

Figure 3 *Coronary angiography revealed normal epicardial coronary vessels*

were used to reconstruct the short axis (SA), vertical and horizontal long axis slices (VLA, HLA), estimation of left ventricular ejection fraction (EF), end diastolic and end systolic volumes (EDV, ESV) and wall motion score using 17 segment model. Two hours later a resting GSPECT scan was performed 20 minutes following injection of 1110 MBq 99mTc-MIBI.

The GSPECT scan images revealed a medium sized inducible ischaemia of high intensity involving the apex and distal inferolateral wall, there was evidence of apical hypokinesia and TID of the LV cavity was also evident (Figure 1). Resting images showed the typical "ace-of-spade" pattern due to enhanced count density over hypertrophied apex and a solar polar map with slightly reduced count over inferior segments due to incomplete recovery of stress-induced ischaemia at time of resting injection (Figure 2). GSPECT showed a post-stress EF 39%, EDV 113 ml and ESV 68 ml.

In view of the patient's symptoms and the abnormal GSPECT findings, coronary angiography was undertaken, which revealed normal epicardial coronary arteries (Figure 3). The left ventriculogram revealed obliteration of distal LV cavity during systole with hypertrophied LV wall (Figure 4). A diagnosis of AHCM was established and conservative management was advised with significant improvement in the patient's symptoms.

Discussion

AHCM is a fairly uncommon type of hypertrophic cardiomyopathy in this part of world. Its salient features are LV hypertrophy predominantly involving the apex with variable extension basally, giant inverted T waves (>10 mm) in mid precordial leads and spade-shaped appearance of the LV at end-diastole on echocardiogram, cardiac MRI and the rest myocardial perfusion SPECT scan.

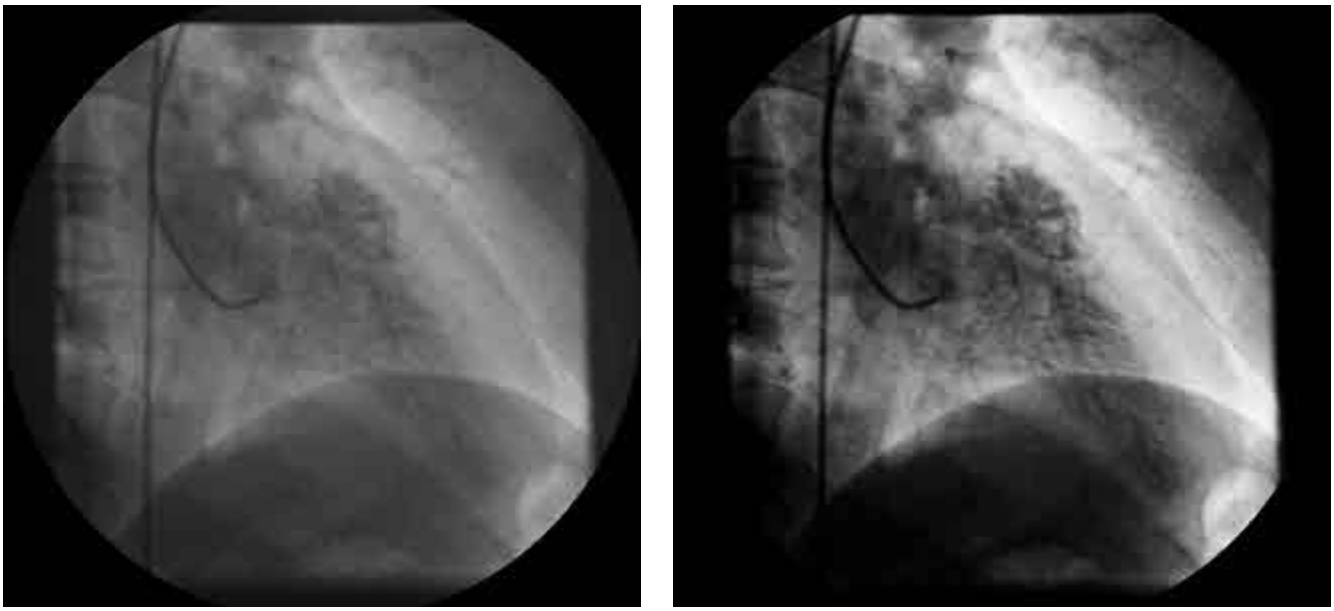

Figure 4 *Left Ventriculogram reveals obliterated distal left ventricular cavity and hypertrophied LV wall with the left image un-enhanced and right enhanced to optimise visualisation of the ventricular wall and cavity*

In the majority of patients, symptoms are mild but in about one-third of the cases, the patients may experience symptoms like dyspnoea and exertional chest pain and some of the patients may get myocardial infarction and life threatening arrhythmias. The patients with exertional symptoms and resting ECG abnormalities are frequently referred for MPI to investigate the possibility of obstructive CAD.

SPECT MPI has been reported to show a wide spectrum of findings pertinent to AHCM. Most common finding is spade shape appearance of LV in resting VLA and HLA slices due to excessive tracer uptake over hypertrophied apex and "Solar Polar" pattern in polar maps[7]. In our patient, both of these findings were present (although slightly reduced count over inferior segments in resting polar map due to incomplete recovery of stress induced ischaemia at time of resting injection). Reversible or irreversible perfusion abnormalities have been reported in 50% of patients in few studies [7]. In this patient, an inducible ischaemia involving apex, apical anteroseptal and inferior wall with normal epicardial coronaries raise the possibility of micro vascular disease in AHCM. Similarly, TID with post -stress stunning draws our attention to a severe and diffuse micro vascular element resulting in compromised coronary flow reserve [5] and ischaemia. Other possible causes of myocardial ischaemia in AHCM are myocardial hypertrophy and bridging, diastolic dysfunction and LV outflow tract obstruction resulting subendocardial ischaemia [9].

Echocardiography is the most common tool used for the diagnosis of HCM in clinical practice. In this case transthoracic 2D echocardiogram could not detect AHCM (false negative). There are published reports that 2D echocardiogram may miss apical, lateral and extreme forms of HCM when contrast is not used [2, 4]. A LV wall thickness of above 30 mm have been reported as a strong risk factor for SCD, and may influence future assessments for implantable cardioverter-defibrillator (ICD) [10].

Cardiac catheterization is frequently done in patients with AHCM due to clinical suspicion and positive GSPECT findings. Obliterated apex and small basal LV cavity during systole

are the typical features on ventriculography which were seen in this case too. In majority of patients there is no intraventricular pressure gradient, but in symptomatic individuals, a pressure gradient is demonstrated between the left ventricular apex and the base. However, in this patient with severe exertional symptoms, pressure gradient was not calculated. Concomitant coronary artery disease can be found in patients with risk factors for atherosclerosis. In addition, the left anterior descending coronary artery and its septal branches may demonstrate phasic narrowing during systole (myocardial bridging) [11] in the absence of underlying obstructive lesions.

In majority of patients with AHCM, symptoms are milder and well controlled on medication. The purpose of pharmacologic therapy (beta blocker, calcium blocker, etc) is to reduce the pressure gradient across the LV outflow tract by reducing the inotropic state of the left ventricle, improving compliance of the left ventricle, and reducing diastolic dysfunction. One third patients with AHCM may have MI, apical aneurysm, stroke or arrhythmias [2]. In patients with arrhythmogenic SDC, amiodarone (Cordarone) or ICD have been found with better outcome. For patients who have limiting symptoms despite optimal medical treatment, apical myectomy can improve functional status by decreasing LV end-diastolic pressure, thus improving the effective operative compliance of the LV and increasing stroke volume. This procedure may be of value in other patients with HCM who have severe hypertrophy and small LV end-diastolic volumes [12].

AHCM although uncommon in this part of world but is an important clinical entity due to its presentation mimicking obstructive CAD and possibility of arrhythmia or SCD. Gated SPECT MPI is an important and non-invasive tool which has characteristic patterns pertinent to AHCM helping nuclear physician to diagnose this condition with high level of confidence.

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