

Apical Hypertrophic Cardiomyopathy in Pakistan: Electrocardiographic, Echocardiographic and Myocardial Scintigraphic Features

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Abstract

Apical hypertrophic cardiomyopathy is a recently recognised entity, with typical electrocardiographic, echocardiographic and myocardial scintigraphic features. Thallium imaging is more sensitive than echocardiography, but electrocardiogram appears to be an important clue to its diagnosis which shows changes in the mid precordial leads in most of the patients (JPMA 45:117,1995).

Introduction

Hypertrophic cardiomyopathy (HC) is a subset of cardiomyopathies, whose morphologic features have been described in many reports^{1,2} and in HC further subgroups have been recognised depending upon the site of hypertrophy, which may have diagnostic and prognostic implications^{3,4}. Apical hypertrophy of the left ventricle has been proposed in Japanese literature⁵ and there are isolated case reports from South Africa, Asia and America. Apical hypertrophic cardiomyopathy was first reported in a group of Pakistani patients in 1985⁶ and its clinical and angiographic features have been described. Different non-invasive and invasive diagnostic modalities are used to evaluate myocardial hypertrophy: echocardiography, myocardial scintigraphy, gated cardiac blood pool scanning, X-ray computed tomography and contrast left ventriculography. Echocardiography is the most widely used method, but often it is inadequate or technically poor in quality, in patients with chronic obstructive lung disease, obesity and other conditions. Also apex and lower ventricular septum are difficult to evaluate echocardiographically even by 2-D sector scanning. Myocardial scintigraphy with thallium-201 has the advantage of visualising the entire LV myocardium in all patients. This paper describes three Pakistani patients with apical hypertrophic cardiomyopathy with distinctive electrocardiograms and echocardiographic features diagnosed by conventional thallium-201 planar imaging.

Case 1

A 45-years old male was admitted to the CCU with a history of atypical chest pain and fainting triggered during strenuous activity and abnormal findings on electrocardiogram. The patient was in excellent health until the day of admission. Serial cardiac enzymes were not elevated and electrocardiogram did not change. The only risk factor was obesity. There is a strong family history of diabetes mellitus and ischaemic heart disease. On physical examination there was mild hypertension and an audible S4. Inderal 40mg three times daily and Moduretic one tablet daily were prescribed. Electrocardiogram showed abnormal changes (tall R waves and giant negative T waves) in mid precordial leads. R wave was highest (42 mm) in V4 and T wave was deepest in V4 (16 mm). QRS axis was normal and the highest R wave was noted in limb lead II. ST was depressed 2 mm in V2, V3 and V4 and septal q was absent (Figure 1).

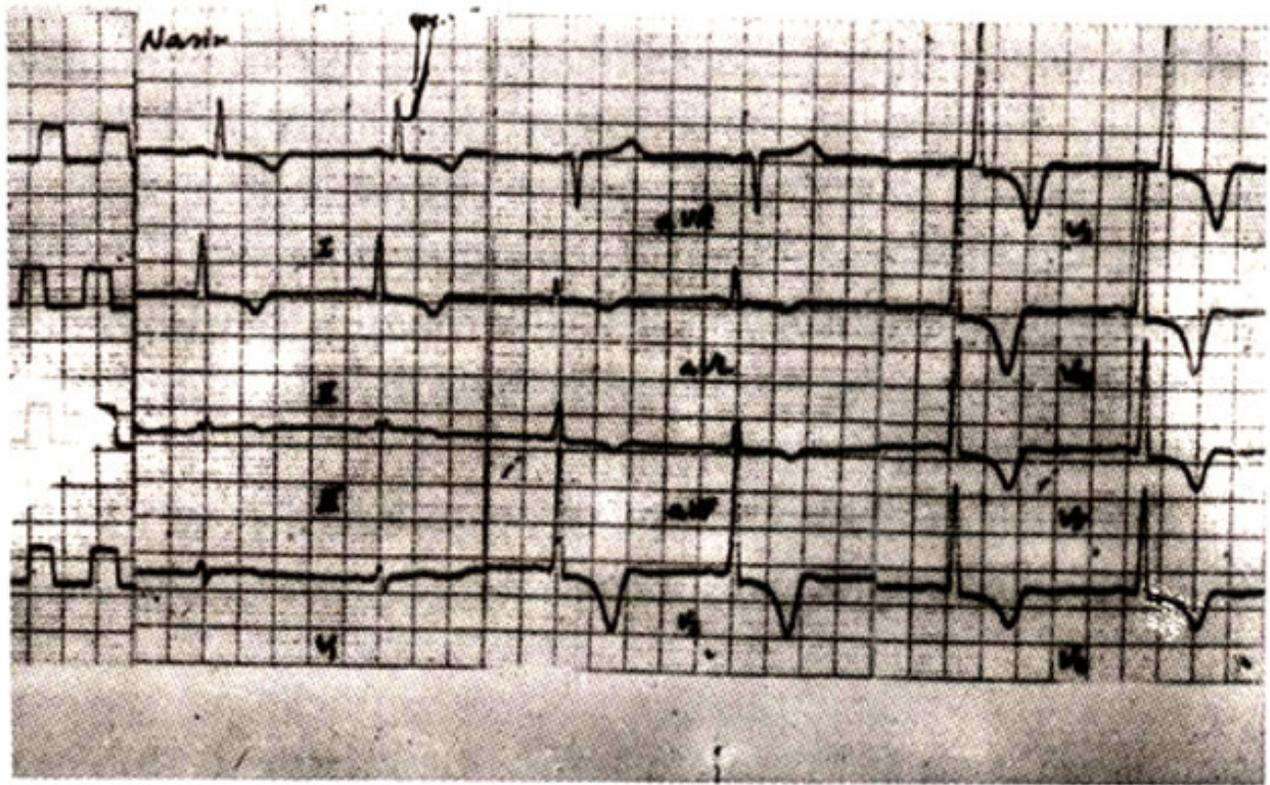


Figure 1. Twelve lead electrocardiogram from patient No.1 taken on half standard shows left ventricular enlargement with giant negative T-waves. R-wave is of maximum amplitude in lead V4 in which depth of T-wave is also maximum.

Graded exercise test was performed on a bicycle ergometer with 25-W increments every 3 minutes. Patient was able to exercise for 14 minutes and the test was terminated due to fatigue. After 12 minutes of exercise patient felt slight pinching in the chest. There were no ST or T wave changes seen at peak exercise, or during the recovery period. The morning dose of Inderal on the day of exercise was omitted. 2-D echocardiogram showed lower septal and lower free wall hypertrophy and narrowing of LV cavity towards the apex (Figure 2).

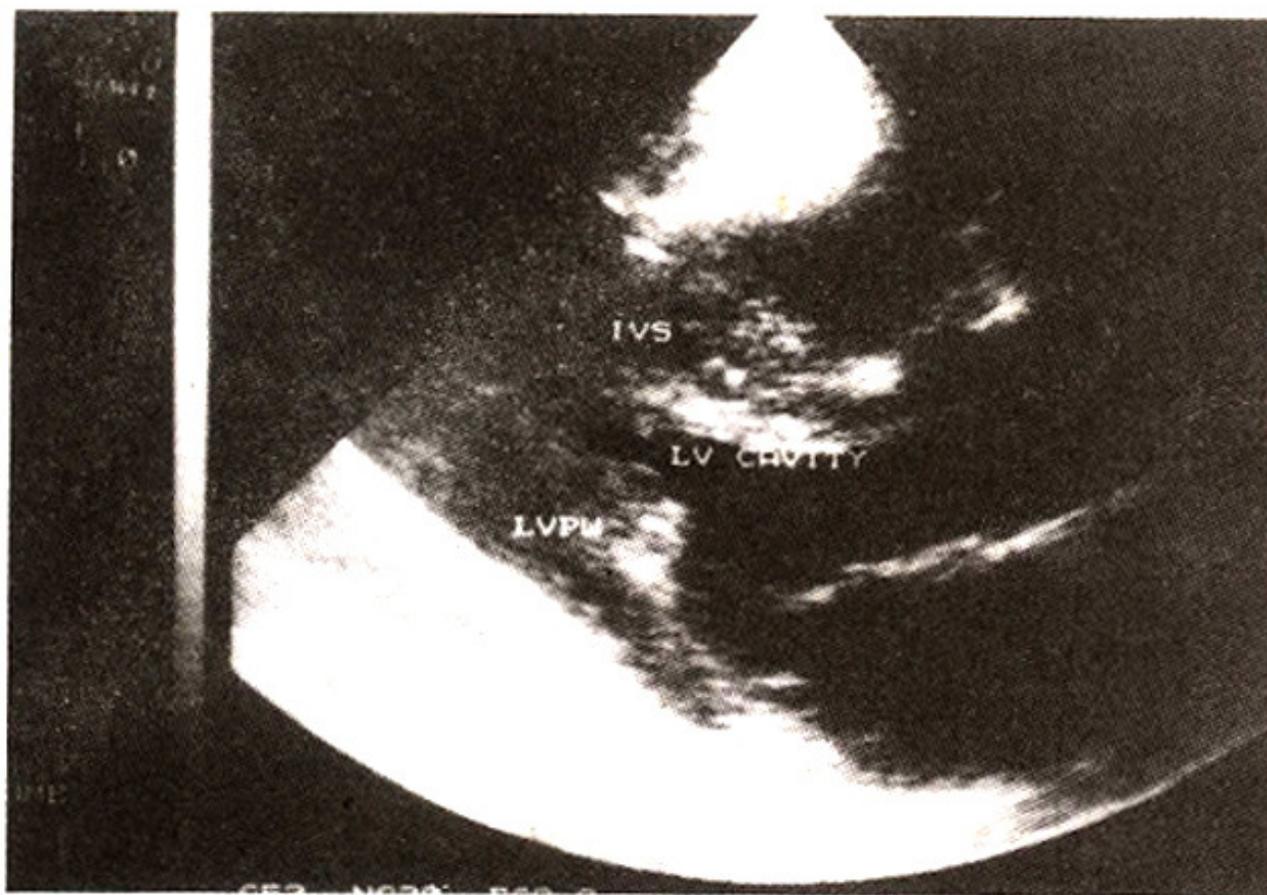


Figure 2. Two dimensional echocardiogram. Parasternal longaxis, showing typical features of apical hypertrophic cardiomyopathy. Abbreviation: LV=left ventricle; IVS= interventricular septum; LVPW= left ventricular posterior wall.

A diagnosis of apical hypertrophic cardiomyopathy was made on these findings as previously described⁵. Resting thallium images were obtained in anterior, left anterior oblique 45° and left anterior oblique 70° projections after intravenous injection of 2 mCi of thallium-201. In the thallium images marked thickening of the apex was clearly visualised. No perfusion defects or areas of relative hypoperfusion were seen (Figure 3).



Figure 3. Left anterior oblique 45° thallium image of patient No. 1 showing thickened apex.

Case 2

A 55 year old male presented to the Casualty with a history of discomfort in left shoulder non-radiating in nature and not associated with sweating. The only cardiac finding was a short grade 1/6 systolic ejection murmur at the left sternal border. Electrocardiogram showed normal QRS axis with limb lead II showing highest R wave. Tall R waves and giant negative T waves were seen in midprecordial leads. R wave was tallest in V4 (56 mm) and T wave was deepest in V4 (24mm). Twaves were also inverted in limb leads, in aVL and aVF. Graded exercise was not performed in this case. Patient was evaluated echocardiographically using 2-D sector scanner in the left recumbent position. Lower septal hypertrophy, lower free wall hypertrophy and significant narrowing of the apical LV cavity was seen. Resting thallium images were obtained as in the previous case. Marked thickening of the apex was clearly seen and none of the images showed any perfusion defect or areas of relative hypoperfusion.

Case 3

A 48 year old male developed palpitation when he was given intravenous fluids after having had diarrhoea for a couple of days. He later presented to the Cardiology department with sweating, palpitation and breathlessness on exertion. Examination of the cardiovascular system revealed a short 2/6 systolic murmur at the left sternal border. As in the previous two cases tall R waves and giant negative T waves were seen in the midprecordial leads, marked thickening of the apex. Echocardiographic features were consistent with apical hypertrophic cardiomyopathy (Figure 4).

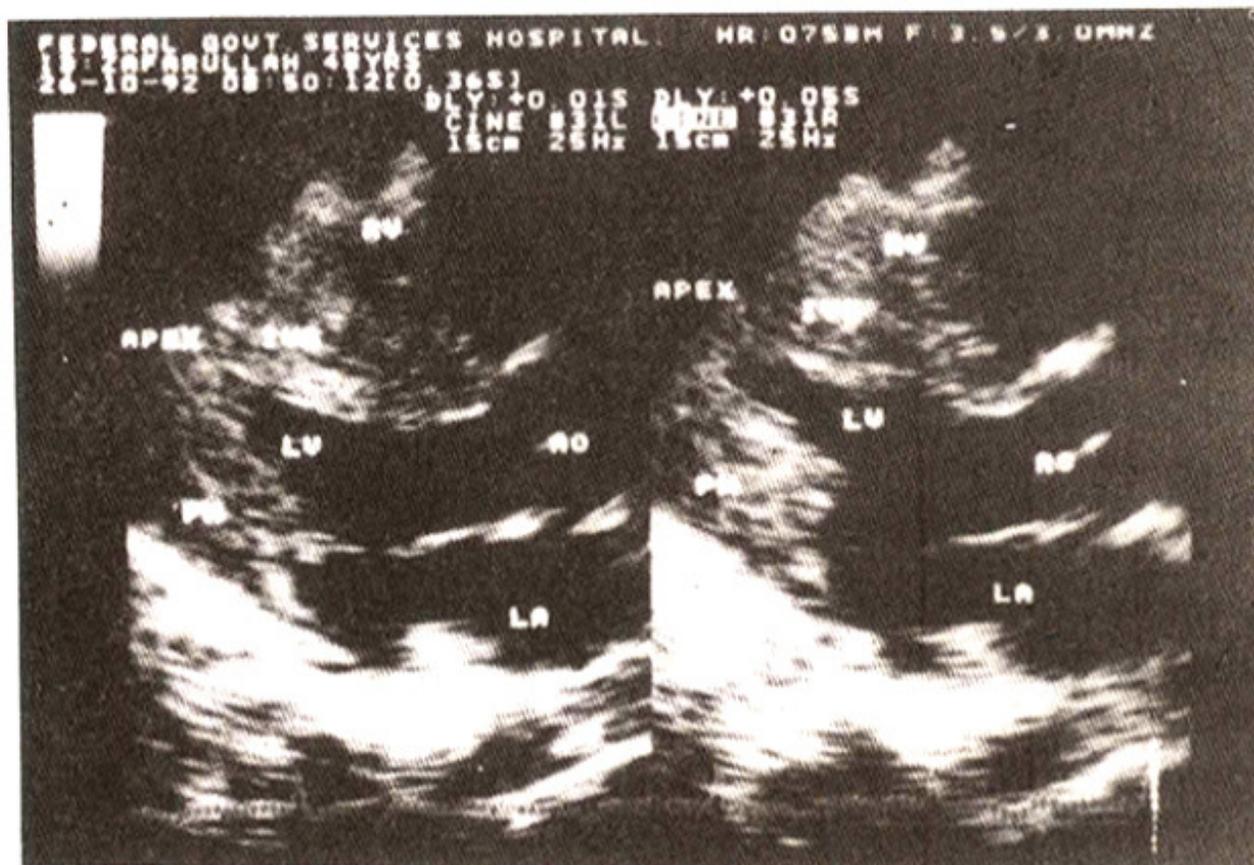


Figure 4. Two dimensional echocardiogram, parasternal long axis (Case III). RV= right ventricle; IVS= interventricular septum; LV= left ventricle; PW= posterior wall; AO= aorta; LA= left atrium .

Thallium images showed marked thickening of the apex.

Discussion

Yamaguchi et al⁵ reported giant negative T waves in a series of hypertrophic cardiomyopathy which they labelled as "apical hypertrophy". The electrocardiographic pattern satisfying the diagnostic criteria of LVH with the most remarkable changes in the midprecordial leads, Lead II, with a normal axis, is characteristic for non-obstructive hypertrophic cardiomyopathy⁷. Thallium-201 single-photon emission-computed tomography (SPECT) with a rotating gamma camera can provide useful clinical information about regional myocardial hypertrophy⁸. Kuzeshi et al⁹ showed that in APH the midprecordial leads reflected abnormal changes which were most significant in lead V4 and the that the morphologic findings in APH derived by SPECT coincided with the ECG findings. In our patients the T wave was most deeply inverted in lead V4 where the R wave was also the highest. The giant negative T waves might be secondary to the result of reversed order of repolarisation from the hypertrophic lower septum and apical area⁷. Although relative myocardial ischaemia may be another factor related to T wave change, true coronary artery disease is not the cause. The electrocardiographic abnormalities in patients with apical hypertrophy may serve as sensitive diagnostic markers for the disease. The cases in the report of Maron et al¹⁰ showed features different from the Japanese patients, probably because only the septum rather than the entire apical segment is involved. Our patients demonstrated the typical ECG changes characteristic of APH as described in earlier reports. In all three

patients 2-D sector scan showed lower septal hypertrophy, lower free wall hypertrophy and towards the apex the LV cavity was significantly narrowed. Echocardiographic techniques may not always adequately visualise this type of distal hypertrophy. A recent study⁸ in Japanese patients with APH demonstrated adequate visualisation of apical hypertrophy by 2-D echocardiography in only 6 of 10 patients. Therefore, 2-D sector scanning may not be an adequate scanning method for all types of hypertrophic cardiomyopathy. Hypertrophic cardiomyopathy has been evaluated by conventional thallium-201 myocardial imaging by Bulkely et al¹¹, who found a thickened apex in both obstructive and non-obstructive types of HC. Suzuki et al⁸ showed that a thickened apex was characteristic of non-obstructive HC with giant negative T waves but not of obstructive HC or concentric LV hypertrophy. This apparent discrepancy between conventional scintigraphy and SPECT imaging may be related to the radionuclide superimposition of conventional thallium-201 myocardial imaging. In our cases apical hypertrophy was clearly seen by the conventional thallium imaging, but the number of cases is small to make any inference (Table).

Table. Clinical data on three Pakistani patients with apical hypertrophic cardiomyopathy.

Age (Years)	45	55	48
Symptoms	Atypical chest pain dyspnea and fainting	Pain in left shoulder, giddiness on and off	Sweating, palpitation and breathlessness on exertion
Abnormalities on physical exam	Audible S4	1/6 Systolic murmur at LLSB	2/6 Systolic murmur at LLSB
Chest X-Ray	Normal	CTR: 53% Prominence of left ventricle	Normal
ECG precordial Voltage (mm)			
Highest R wave	V4 (42 mm)	V4 (56 mm)	V3 (42 mm)
Maximum depth of "giant" negative T wave	V4 (16 mm)	V4 (24 mm)	V4 (24 mm)
QTC (Sec.)	0.55 sec.	0.54 sec.	0.55 sec.
GXT	Maximal, stopped for fatigue, dyspnea no ECG changes	Not done	Not done
Echocardiography	Lower septal and lower free wall hypertrophy and concentric apical hypertrophy	Lower septal and lower free wall hypertrophy and narrowing of the LV cavity towards the apex.	Same
Thallium-201 myocardial imaging	Apical hypertrophy	Apical hypertrophy	Apical hypertrophy
Abbreviations: ECG= electrocardiogram; (QT) LLSB= Left lower sternal border R-R		GXT= graded exercise test;	QTC= corrected QT interval

Most patients with apical HC are asymptomatic and over 90% are male and usually over 40 years⁵. The most common symptom appears to be chest pain (anginal or atypical), followed by dyspnea on exertion, palpitations, dizziness and easy fatigue⁵. These patients do not show appreciable impairment of left ventricular function or symptoms of heart failure. Stress test in our patients had shown a good exercise tolerance without any ST segment changes. Abnormal cardiac performance during exercise could be an important factor of sudden death. Koga et al¹² have shown that risk of sudden death was significantly higher in patients with positive single Master's two-step test. Graded exercise test appears a good way to assess prognosis in patients with apical hypertrophic cardiomyopathy. Familial occurrence is uncommon in APH and hypertension and strenuous physical activity are probably associated risk factors. APH appears to have an excellent prognosis in contrast to

other form of hypertrophic cardiomyopathy¹². Few symptoms may be the cause of lack of case reports or the distinctive electrocardiogram is not recognised and so the diagnosis is not considered. Possibly in our patients the electrocardiographic changes evolved over a passage of time. Patients with striking electrocardiographic T wave changes may represent the final product of a hypertrophic process changing in response to unknown stimuli such as humoral factors, as suggested by Yamaguchi and colleagues⁵. As our patients did not show any evidence of perfusion defects or areas of relative hypoperfusion on thallium-201 myocardial imaging and had typical diagnostic features of APH, coronary angiography was not performed.

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