

CASE REPORT - ISOLATED APICAL HYPERTROPHIC CARDIOMYOPATHYMonica A¹, Prabhu G², Balasubramaniyan S³¹Postgraduate, Department of General Medicine, Rajah Muthiah Medical College and Hospital.²Reader, Department of General Medicine, Rajah Muthiah Medical College and Hospital.³HOD and Chief, Department of General Medicine, Rajah Muthiah Medical College and Hospital.**ABSTRACT**

A rare form of hypertrophic cardiomyopathy predominantly involving the left ventricular apex is called as Apical Hypertrophic Cardiomyopathy (AHCM). This variant, more commonly seen in Japanese populations had been detected in a middle-aged South-Indian man by 2D echocardiography.

KEYWORDS

Cardiomyopathy, Apical Hypertrophy.

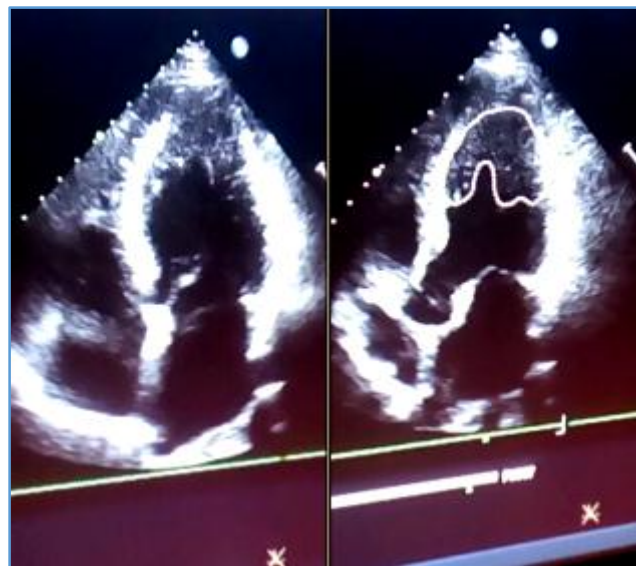
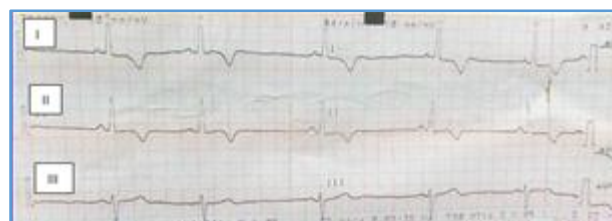
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INTRODUCTION

Apical Hypertrophic Cardiomyopathy (AHCM) is a rare form of Hypertrophic Cardiomyopathy (HCM), which usually involves the apex of the left ventricle and rarely involves the right ventricular apex or both.¹ This relatively rare variant of HCM, first described in Japan, constituted 13% to 25% of all cases of HCM in Japan²; however, it is seen much less often in non-Japanese populations.³

CASE REPORT

A 55-year-old South-Indian male, a chronic smoker with a history of systemic hypertension presented with complaints of an acute onset chest pain which was left-sided, intermittent, dull aching with no radiation. History of associated profuse sweating present. He had no complaints of dyspnoea, palpitations, syncope or haemoptysis. There was no family history of sudden cardiac death, congestive heart failure or cardiomyopathy. On examination his blood pressure was 160/100 mmHg, heart rate was 78 bpm with no heart murmur or any signs of congestive heart failure. Other systemic examination was unremarkable. A 12-lead electrocardiogram (ECG) showed left ventricular hypertrophy and inverted T-waves in I, II, aVL, aVF, V2-V6 (Figure 2). The cardiac enzymes and chest x-ray were normal. A Transthoracic Echocardiogram (TTE) showed apical hypertrophy with adequate LV systolic function and no evidence of regional wall motion abnormality (Figure 1).

**Fig. 1**

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Corresponding Author:

Monica A,

234, Lal Bahadur Sastry Street,

Pondicherry-605001.

E-mail: drmonicaanand5@gmail.com

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Fig. 2

Patient was treated with antiplatelets and antihypertensive drugs. Further hospital stay was uneventful.

DISCUSSION

Our case describes an unusual presentation of hypertrophic cardiomyopathy. Apical variant of HCM has been mainly reported in Japan with significantly much lesser prevalence in other populations.¹⁻⁴ HCM is transmitted as a Mendelian trait with an autosomal dominant pattern of inheritance. Mutations in two sarcomere genes, those encoding β -myosin heavy chain (MYH7) and myosin-binding protein C (MYBPC3) are by far the most common accounting for 70% of those successfully genotyped.⁵ The apical HCM is frequently sporadic; however, a few families have been reported with autosomal dominant inheritance.⁶

Morphologically, AHCM is divided into 3 types: pure focal, pure diffuse and mixed, of which pure focal is most common.⁷ However, in clinical practice, this sub-classification is not widely accepted and its clinical relevance is unknown. Others have divided AHCM into two groups based on whether they had isolated asymmetric apical hypertrophy (pure AHCM) or had co-existent hypertrophy of the interventricular septum (mixed AHCM).⁸ Isolated hypertrophic obstructive cardiomyopathy of the right ventricle had been reported without any involvement of the interventricular septum or left ventricle.

The mean age of presentation of AHCM is 41.4 ± 14.5 years and is most commonly seen in males.⁴ About 54% of patients with AHCM are symptomatic and the most common presenting symptom is chest pain followed by palpitations, dyspnoea and syncope.⁸ AHCM may also manifest as morbid events such as atrial fibrillation, myocardial infarction, embolic events, ventricular fibrillation and congestive heart failure. Other complications of AHCM include apical aneurysm and cardiac arrest. Physical findings of an audible/palpable fourth heart sound and a new murmur are common. Our patient was relatively asymptomatic with no family history and no physical findings.

The most frequent ECG findings are negative T-waves in the precordial leads, which are found in 93% of patients followed by LV hypertrophy in 65% of patients.⁸ Negative T-waves with a depth > 10 mm are found in 47% of patients with AHCM.⁸ "Giant T-wave inversion" (depth > 10 mm) and loss of septal Q waves should raise strong suspicion of apical HCM and are found in 47% of cases. The ECG in our patient showed LV hypertrophy and negative T-waves.

Echocardiography has been universally accepted as the initial imaging modality in investigation of patients with apical HCM.⁸ The diagnostic criteria for apical hypertrophic cardiomyopathy included demonstration of asymmetric hypertrophy, confined predominantly to the apex with an

apical wall thickness ≥ 15 mm and a ratio of maximal apical to posterior wall thickness ≥ 1.5 , based on echocardiography.⁹ In this case, asymmetric apical wall is 26 mm and its posterior wall is 4.2 mm and a ratio of maximal apical to posterior wall thickness 6.19, satisfying the diagnosis of apical left ventricular hypertrophic cardiomyopathy. The degree of thickness increases from base to apex resulting in a markedly diminished apical cavity and a spade-shaped left ventricular cavity suggesting apical left ventricular hypertrophic cardiomyopathy in the apical 4 chamber view of two-dimensional echocardiographic image of this patient. Although, the initial diagnostic test for AHCM is most commonly TTE, the best diagnostic tool is considered to be cardiac MRI.

ACHM may mimic other conditions including apical cardiac tumours, LV apical thrombus, isolated ventricular non-compaction, Endomyocardial Fibrosis (EMF) and coronary artery disease. Chest pain in a patient with ACHM can be mistaken for ischaemia from coronary artery disease.¹⁰

The course of apical HCM is relatively benign. The overall mortality rate is 10.5%. Some apical HCM patients may develop life-threatening complications such as sudden cardiac death,¹⁰ which is more commonly seen in asymmetric septal hypertrophy than apical HCM having syncopal episodes, arrhythmias, ventricular wall thickness > 30 mm or a family history of sudden death may benefit from ICD (Implantable Cardioverter Defibrillator) placement. Beta adrenergic blocking agents are the first line therapy for symptomatic patients.¹¹ Since our patient was asymptomatic, he was managed conservatively with advice for regular followup.

CONCLUSION

Isolated apical left ventricular hypertrophic cardiomyopathy had been detected in a 55-year-old South-Indian male who is a hypertensive and chronic smoker; 2D echocardiography confirmed the rare finding of isolated hypertrophy of the left ventricular apex.

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