

Review Article



Triple T pattern on ECG and Apical Hypertrophic Cardiomyopathy

Md. Fakhurul Islam Khaled¹, Md. Azharul Islam², Md. Abu Salim³, Md. Mukhlesur Rahman⁴,
Mostashirul Haque⁵, MSI Tipu Chawdhury⁶, Mohammed Emran⁷,
Md. Walidur Rahman⁸, Dipal Krishna Adhikary⁹.

Abstract

T wave on ECG is the positive deflection after QRS complex which reflect the ventricular repolarization. The amplitude of T wave is 5mm in limb lead and 10mm in chest leads. T wave is upright in all leads except aVR and VI, but it may be inverted in VI-V3 in pediatric age group. Triple T pattern is the negative T waves in inferior leads, anterior leads and on aVR. Triple T pattern is a very common finding in Apical hypertrophic cardiomyopathy (AHCM). Apical hypertrophic cardiomyopathy is a rare form of cardiomyopathy that affects LV apex and rarely RV apex or both. Patients with AHCM has a wide range of presentations, ranging from asymptomatic to palpitation, nonspecific chest discomfort, chest pain etc. It does not present with features which are common in other type of obstructive hypertrophic cardiomyopathy like presyncope, syncope, and it has less chance of sudden cardiac death. First clue of diagnosis of AHCM is widespread negative T wave in ECG. Although negative T-waves may be found in chest leads in 93% of cases. Cardiovascular magnetic resonance (CMR) is the best diagnostic tool. Proper transthoracic echocardiographic evaluation demonstrates apical wall thickness >15 mm and a ratio of maximal apical to posterior wall thickness >1.5 mm. Beta-blockers are mainstay of treatment whereas implantable cardioverter defibrillator (ICD) is recommended for high risk cases. As it is a genetic disease genetic counseling and periodic follow up is required.

Keywords: Triple T pattern, Apical hypertrophic cardiomyopathy (AHCM).

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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 Patient with AHCM may be asymptomatic and they can be suspected with wide spread T wave changes in ECS specially triple T pattern that is deep T wave inversion in anterior leads ,inferior leads and aVR. AHCM constitutes 15% of all the HCM patients in Japan, whereas in USA the prevalence was only 3%.²

Defining the conditions

1. Triple T pattern: Inverted T wave in anterior leads , inferior leads and in lead aVR .
2. Apical hypertrophic cardiomyopathy (AHCM) : The diagnostic criteria for AHCM included demonstration of asymmetric LV hypertrophy, confined predominantly to the LV apex, with an apical wall thickness >15 mm and a ratio of maximal apical to posterior wall thickness >1.5 mm, based on an echocardiogram or magnetic resonance imaging (MRI).³

1. Assistant Professor, Department of Cardiology, Bangabandhu Sheikh Mujib Medical University, Dhaka, Bangladesh.
2. Resident, Department of Cardiology, Bangabandhu Sheikh Mujib Medical University, Dhaka, Bangladesh.
3. Associate professor, Department of Cardiology, Bangabandhu Sheikh Mujib Medical University, Dhaka, Bangladesh.
4. Associate professor, Department of Cardiology, Bangabandhu Sheikh Mujib Medical University, Dhaka, Bangladesh.
5. Assistant Professor, Department of Cardiology, Bangabandhu Sheikh Mujib Medical University, Dhaka, Bangladesh.
6. Medical Officer, Department of Cardiology, Cox's Bazar Medical College & Hospital, Cox's Bazar, Bangladesh.
7. Assistant Professor, Department of Physical Medicine and Rehabilitation, KhwajaYunus Ali Medical College and Hospital, Sirajganj, Bangladesh.
8. Resident, Department of Cardiology, Bangabandhu Sheikh Mujib Medical University, Dhaka, Bangladesh.
9. Associate professor, Department of Cardiology, Bangabandhu Sheikh Mujib Medical University, Dhaka, Bangladesh.

Correspondent: Dr. Md. Fakhurul Islam Khaled, Assistant Professor, Department of Cardiology, University Cardiac Center, Bangabandhu Sheikh Mujib Medical University, Dhaka. Mobile: +8801817517505, E-mail: drfikhaled@gmail.com

Morphological variants

1. T wave : T wave may be flat , tall ,biphasic, camel hump or inverted. Deep symmetrical T wave inversion in anterior lateral and lead aVR highly specific for AHCM. Although Deep symmetrical T wave inversion indicate ischemia but it is rarer in anterior inferior and lead aVRaltogether.

2. Morphologically AHCM is divided into 3 types: pure focal, pure diffuse and mixed, of which pure focal is most common.⁴ In other way AHCM is divided 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).³

Genetic basis of AHCM

AHCM is frequently sporadic; though the pattern of autosomal dominant inheritance is reported in few families.⁵ A sarcoma gene mutation (E101K mutation in the alpha-cardiac actin gene) has been identified in these families. A family history is more common in patients with asymmetric septal hypertrophy than with AHCM.⁶

Presentation of AHCM

The mean age of presentation of AHCM is 41.4 ± 14.5 years and is most commonly seen in males.³ About 46% patient of AHCM patient are asymptomatic , who may be suspected with triple T pattern of ECG changes and rest 54% of patients with AHCM are symptomatic and the most common presenting symptom is chest pain, followed by palpitations, dyspnea and syncope.³ AHCM may also present with complications like atrial fibrillation, myocardial infarction, embolic events, ventricular fibrillation, congestive heart failure, apical aneurysm and cardiac arrest.³

Diagnostic work up for AHCM

The diagnostic work up includes 12 lead ECG, 2D M mode and Color Doppler echocardiography, Contrast echocardiography, Transesophageal echocardiography (TEE), Cardiovascular magnetic resonance (CMR). In ECG most common finding is negative T-waves in the precordial leads which are found in 93% of patients, followed by LV hypertrophy in 65% of patients.³ But negative T wave may also found in inferior leads and lead aVR. Negative T-waves with a depth > 10 mm are found in 47% of patients with AHCM.³ TTE shows hypertrophy of the LV apex and is the initial diagnostic tool for AHCM. Patients with AHCM usually show less-frequent LV outflow tract obstruction and accompanied by a higher frequency of apical aneurysm than non-AHCM.⁶ The parameters of diastolic dysfunction like E/e' value and the LA volume index (LAVI) are usually lower in AHCM patients.⁶ When the baseline images are suboptimal, a contrast echocardiogram is useful in establishing the diagnosis.⁷ On contrast ventriculography AHCM shows a distinctive LV "spade-like" configuration.³ On single photon emission computed tomography (SPECT) myocardial perfusion imaging findings of resting "solar polar" map pattern and reduced flow reserve of the apex are the characteristics of AHCM.⁸ Multislice spiral computed tomography can also be used to diagnose AHCM; besides diagnosis it provides

information on cardiac anatomy, function and coronary arteries.⁹ Cardiac MRI is also a valuable tool for diagnosing patients with inconclusive echocardiography and SPECT findings.¹⁰ Although the initial diagnostic test for AHCM is most commonly TTE, the best diagnostic tool is considered to be cardiac MRI.

Differentials for AHCM

AHCM may mimic other conditions like apical cardiac tumors¹¹, LV apical thrombus¹², isolated ventricular non-compaction,¹³ endomyocardial fibrosis (EMF)¹⁴ and coronary artery disease.¹⁵ Chest pain in a patient with AHCM can be misdiagnosed for ischemia from coronary artery disease.¹⁵ Frequently these patients undergo a nuclear scan or coronary angiography for abnormal ECG.¹⁶ The majority of the patients with AHCM who suffer myocardial infarction have an apical infarct, and in these patients' wall motion abnormalities varies from apical aneurysm to apical hypokinesis.³ Some patients may have asymptomatic apical infarction.³ Hence, in clinical practice, an apical aneurysm may sometimes be seen with AHCM in asymptomatic patients. An echocardiogram with contrast can be used to differentiate AHCM from a LV apical mass (thrombus or tumor).¹² Isolated ventricular non-compaction may be differentiated from AHCM by high resolution images of the heart obtained by cardiac MRI.¹³ A LV angiogram shows apical obliteration during both systole and diastole in EMF, whereas in AHCM apical obliteration occurs only in systole and also there is an absence of significant ventricular hypertrophy in EMF patients.¹⁴

Management

Management of ApHCM is focused on symptom control, genetic testing, and prevention of adverse cardiovascular sequelae. In symptomatic patients with AHCM, verapamil, beta-blockers and antiarrhythmic agents are used.³ Verapamil and beta-blockers are found to be beneficial in improving the symptoms in AHCM patients.^{17,18} Amiodarone and procainamide are used in the treatment of atrial fibrillation and ventricular arrhythmias.¹⁹ An implantable cardioverter defibrillator (ICD) is recommended for high risk HCM patients with (1) previous cardiac arrest or sustained episodes of ventricular tachycardia; (2) syncope; (3) a family history of sudden death; or (4) episodes of non-sustained ventricular tachycardia on serial Holter monitoring.²⁰ ICD is also recommended in AHCM patients with cardiac arrest and non-sustained ventricular tachycardia.¹⁸

Prognosis

The prognosis of AHCM is thought to be relatively benign. The overall mortality rate of AHCM patients was 10.5% and cardiovascular mortality was 1.9% after a follow-up of 13.6 ± 8.3 years.³ Sudden death and cardiovascular events occur more commonly in patients with asymmetric septal hypertrophy than in those with AHCM.²¹ A large LV end diastolic dimension may predict cardiac events in AHCM patients.²² As AHCM patients may develop sudden life-threatening complications, so close follow-up of these patients is recommended.³

Conclusion

As AHCM is a rare entity, and it has wide range of presentations, even it may be asymptomatic. Triple T type ECG changes may strike for further investigations. Multimodality imaging should be used for detection of AHCM although CMR is gold standard. Symptomatic patients should be treated and Family screening should be considered. Periodic lifelong follow-up seems indicated for even initially asymptomatic patients with AHCM.

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