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Case Report

Apical Hypertrophic Cardiomyopathy: A Special Entity.

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ABSTRACT:

Introduction: Apical Hypertrophic Cardiomyopathy (AHCM) is a unique variant with distinct clinical presentation, genetics, treatment, complications and outcome. **Case:** A 52 year non-hypertensive Asian male presented with exertional shortness of breath for two years without chest pain, palpitation or syncope. Apex beat was heaving. Electrocardiogram revealed non q wave deep symmetrical T wave inversion in anterolateral leads and echocardiography demonstrated hypertrophied apical septum. Coronary angiogram showed normal coronaries with typical "Ace of Spade" configuration during ventriculography. **Conclusion:** Characterization of various forms of hypertrophic cardiomyopathy is essential for management purpose as apical hypertrophic cardiomyopathy usually have benign course.

Key words: Apical Hypertrophic Cardiomyopathy, Hypertrophic Cardiomyopathy

INTRODUCTION:

First described by Sakamoto et al, Apical Hypertrophic Cardiomyopathy (AHCM) is an infrequent variant localized to apex of the heart. [1] Yamaguchi et al. subsequently portrayed its typical end systolic left ventriculogram pattern resembling "Ace of Spade". Hence it is also known as Yamaguchi Disease.[2] In Japanese population, it encompasses about 15% of all cases of hypertrophic cardiomyopathy,[1,2] however in non Japanese population it is only 1-3%.[3,4] It has dissimilar genetics, age distribution, clinical course, and complications compared to other forms of hypertrophic cardiomyopathy.[3,5,6] These attributes make AHCM an unique entity of discussion.

CASE REPORT:

A 52 years diabetic gentleman with a body mass index of 24.5kg/m^2, non-smoker, non-hypertensive presented to cardiac OPD with complaint of occasional shortness of breath on prolonged exertion for two years without orthopnoea or paroxysmal nocturnal dyspnoea. He denied any history of fever, cough, weight loss, chest tightness, noisy breathing, chest pain, palpitation, dizziness, excessive fatigability, pre-syncope, syncope or sleep related disorders. There was no past history of chest problem, cardiac disease, thyroid illness, neurological or hematological disorder. There was no family history of cardiac diseases. His diabetes had been on good control with anti-diabetes medications for last six years. Blood Pressure: 120/80 mm Hg, Heart Rate: 82/min regular, saturation: 100% in ambient air. There was no pallor or edema. Apex beat was localized at left 5th intercostal space 9cm from mid-sternal line and was heaving.

Other systemic examinations were unremarkable. Hematology, glycated hemoglobin, renal function test with electrolytes, creatinine phosphokinase (CPK-MB), troponin I and thyroid profile were normal. Chest X-ray was unremarkable. Electrocardiography (ECG) showed non-q wave deep symmetrical T wave inversion in leads I, II, avl, V2-V6 with giant T wave inversion (1.0 mv) in

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leads V3 and V4 (Fig. 1).

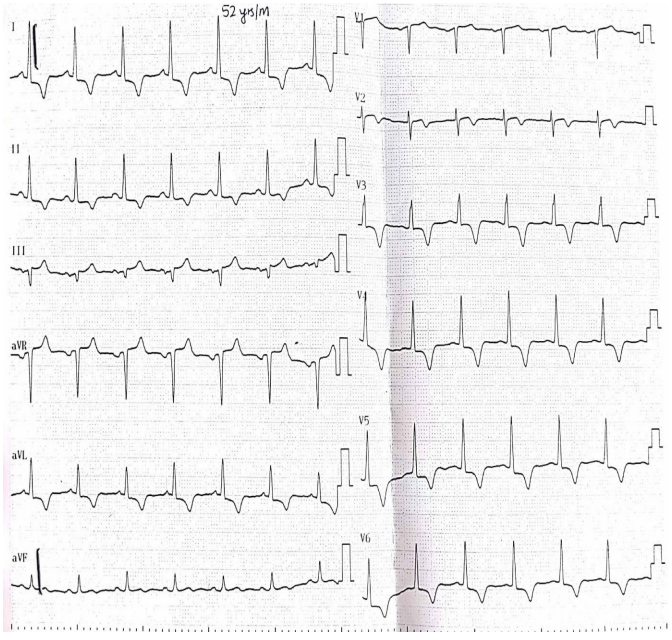

Fig.1. ECG showing giant T wave inversion in precordial leads.

With such ECG findings, he underwent treadmill test multiple of times in past and all of them were negative and was advised for coronary angiography. Echocardiography revealed hypertrophic apical one-third of interventricular septum (17mm), normal aortic gradient, absent left ventricular outflow gradient, absence of intraventricular/apical mass with no regional/global wall motion abnormality (Fig. 2).

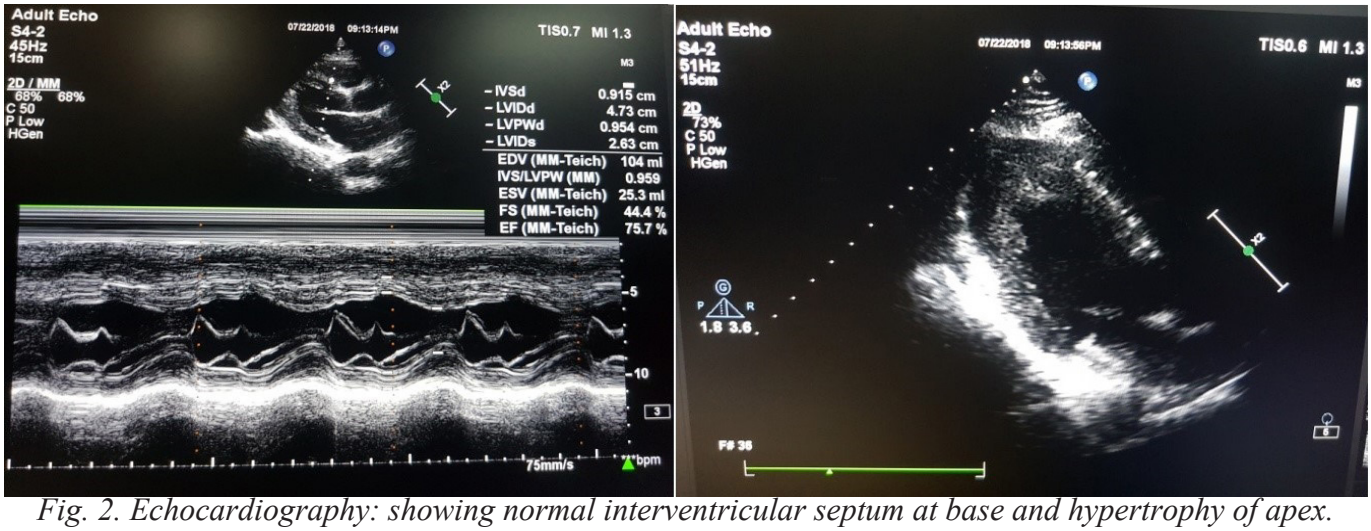

Fig. 2. Echocardiography: showing normal interventricular septum at base and hypertrophy of apex.

Coronary angiography showed normal epicardial coronary arteries with typical "Ace of Spades" configuration of left ventricle at end systole during ventriculography (Fig. 3). He was put on beta-blocker, advised for family screening including regular follow up examination.

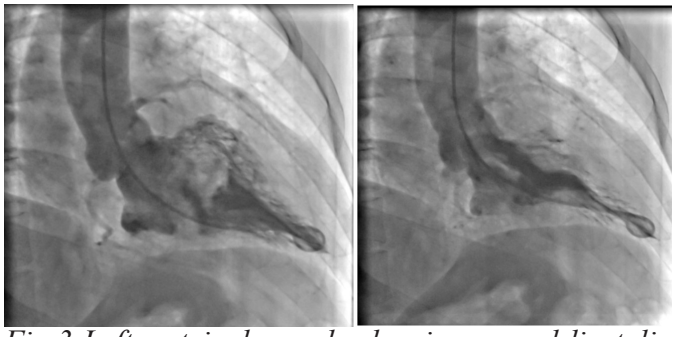

Fig. 3. Left ventriculography showing normal diastolic flow and end systolic ace of spade configuration.

DISCUSSION:

AHCM has two morphological forms: a) Pure Apical form: hypertrophy (>15mm) confined to left ventricular apex below the papillary muscle, and b) Distal Dominant form: apical hypertrophy extends to the interventricular septum without basal septal hypertrophy. Transition of AHCM to and from other forms of hypertrophic cardiomyopathy has not yet been documented.[7]

Though genetic association is rare, some AHCM have familial association with autosomal dominant inheritance involving sarcomere gene mutation (cardiac actin Glu10Lys).[3,6,8,9] Patient may be asymptomatic or present with atypical chest pain, angina, palpitation, dyspnoea, pre/syncope or cerebrovascular accident.[3,6,10] They may have hypertension, forceful apical beat with or without widely split second heart sound and or fourth heart sound.[6] Essential for diagnosis is typical ECG showing non-q deep (giant) T wave inversion, high QRS voltage over the precordial leads, intra ventricular conduction abnormalities or arrhythmias in the form of atrial fibrillation, supra ventricular tachycardia, non sustained ventricular tachycardia and even ventricular fibrillation[1,2,3,6] and can occur in following entities: a) Adams-

Stokes attacks associated with complete heart block, b) ischemic heart disease, c) bradycardia, d) right ventricular hypertrophy and right bundle branch block, e) metabolic disturbances, f) changes during coronary angiography and g) cerebral disturbance.[11] This giant T wave inversion is resultant of altered repolarisation changes of hypertrophied apical musculature.[1] Echocardiography should also be performed with high degree of suspicion. Echocardiographic diagnostic criteria includes: a) asymmetric LV hypertrophy (LVH) predominantly at the apex of the ventricle; b) LV wall thickness of 15mm or more during diastole; and c) apical to posterior wall thickness ratio of 1.5 or more determined by two dimensional echocardiography.[3] Use of intravenous echocardiographic contrast agents can be of help if apical endocardium is not visualized properly.[12] Echocardiography carries sensitivity of 91% for diagnosing AHCM.[3] Angiography demonstrates no obstructive lesion. Hemodynamic studies display absence of pressure gradient in left ventricular outflow tract and complete systolic obliteration of the apex with relative sparing of ventricular cavity at base during left ventriculography forming the typical "Ace of Spade" configuration.[2] Cardiac Magnetic Resonance Imaging (CMRI) is decisive when there is suspicion of AHCM by ECG and echocardiographic feature is inconclusive or technically challenging. CMRI has the advantage of being less operator dependant, multi-planar detection and excellent soft tissue contrast.[13]

Mimickers of AHCM and their diagnostic tools are listed below in the table.[14]

Disease	Diagnostic tool to establish diagnosis of AHCM
Coronary artery disease	Echocardiogram/coronary angiogram and LVG
Left ventricular apical tumors	Echocardiogram with contrast/CCT/CMRI
Left ventricular apical thrombus	Echocardiogram with contrast/CCT/CMRI
Isolated ventricular non-compaction	CMRI/CCT
Endomyocardial fibrosis	LVG/CMRI

AHCM: Apical hypertrophic cardiomyopathy; CMRI: Cardiac magnetic resonance imaging; LVG: Left ventriculography; CCT: Cardiac computed tomography.

Treatment of symptomatic patient constitutes beta blocker, verapamil and anti-arrhythmic agents. Amiodarone and procainamide are used for atrial fibrillation and ventricular arryhthmias.[3,15,16,17] Implantable Cardioverter Defibrillator (ICD) is recommended in patient who have survived a cardiac arrest due to ventricular tachycardia, ventricular fibrillation or who have spontaneous sustained ventricular tachycardia causing syncope or hemodynamic compromise.[18]

Long term follow up studies depict AHCM as benign entity with 15 year survival of 95% and annual cardiovascular mortality of 0.1%- 0.8%.[3,19] Predictors for cardiovascular comorbidity[3] includes a) age at presentation < 41 years, b) left atrial enlargement and c) New York Heart Association functional class II or more at baseline and that for cardiovascular mortality[18] includes a) LV outflow obstruction, b) atrial fibrillation and c) female gender. Almost one third of patients (30%) can develop serious cardiovascular complication of atrial fibrillation and myocardial infarction.[3] Hence periodic lifelong follow up should be initiated even for asymptomatic patient.[20]

CONCLUSION:

Coronary artery disease, aortic stenosis and hypertension should be ruled out in every case presenting with such symptoms and electrocardiographic and/or echocardiographic presentation before designating a diagnosis of hypertrophic cardiomyopathy. Moreover, specifying the types of hypertrophic cardiomyopathy is important as apical hypertrophic cardiomyopathy carries benign prognosis compared to other variants.

Conflict of Interest:

The authors declare that no competing interest exits.

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