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Research paper

Contrast Study and Speckle Tracking Echocardiography for Assessment of Left Ventricular Function in Apical Hypertrophic Cardiomyopathy

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ABSTRACT

Apical hypertrophic cardiomyopathy (AHCM) is a rare phenotypic variant of hypertrophic cardiomyopathy (HCM), most commonly seen in Asian men (Yamaguchi syndrome). It is characterized by hypertrophy, predominantly affecting the cardiac apex, with an "ace of spades"-shaped left ventricular (LV) cavity best seen on the 4-chamber view of a transthoracic echocardiogram (TTE). However, TTE can be falsely negative in 30% of AHCM cases, largely due to difficulties in delineating endocardial border. The diagnostic criteria for apical cardiac hypertrophy are: 1) asymmetric LV hypertrophy - predominantly at the apex of the ventricle; 2) LV wall thickness of 15 mm or more during diastole; and 3) apical to posterior wall thickness ratio of 1.5 or more determined by 2-dimensional echocardiography or cardiac magnetic resonance imaging. Here, we are presenting a case of elderly male who visited our cardiac OPD for a check up due to atypical chest pain, cough and mild fever for 5 days. His resting ECG showed left ventricular hypertrophy with deep T wave inversion in precordial leads V_2-V_6, consistent with AHCM. An exhaustive transthoracic echocardiography (TTE) with contrast study (CS) and speckle tracking imaging by 4Dimensional XStrain speckle tracking echocardiography (STE) was performed to provide a clinching diagnosis.

1. Introduction

Hypertrophic cardiomyopathy, an inherited heterogeneous cardiac condition with diverse phenotypic expression, is a common cause of sudden cardiac death in young adults. Apical hypertrophic cardiomyopathy (AHCM), a phenotypic variant of hypertrophic cardiomyopathy, was first described in Japan in 1976 by Sakamoto et al. (1976) (Figures 1- 4).

AHCM is characterized by apical hypertrophy, an ace-of-spades configuration of the left ventricular cavity in end diastole, and giant negative precordial T waves. In AHCM, hypertrophy is localized to the left ventricular apex with or without midsegment or basal involvement and with or without apical aneurysm (Jan et al., 2016) (Figure 5).

In AHCM, there is typically no LV outflow tract obstruction from systolic anterior motion of the anterior mitral valve leaflet. AHCM can exist with or without midventricular obstruction and cavity obliteration (MVOCO) (Hughes et al., 2020).

Apical hypertrophic cardiomyopathy is distributed world-wide, affecting more men than women. It is typically diagnosed in midlife (Eriksson et al., 2002; Sakamoto et al., 1986). However, it can be diagnosed late in life and can carry a normal life expectancy. We report a case of AHCM presenting to us at the age of 76 years.

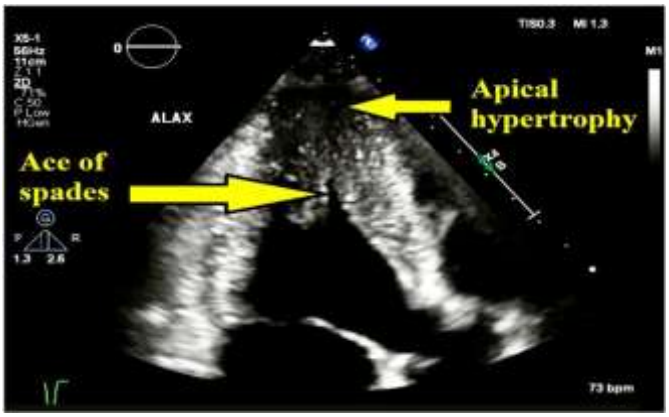


Fig. 1 Transthoracic echocardiography. Apical 3CH view delineates the typical features of AHCM with "ace of spades" like LV cavity

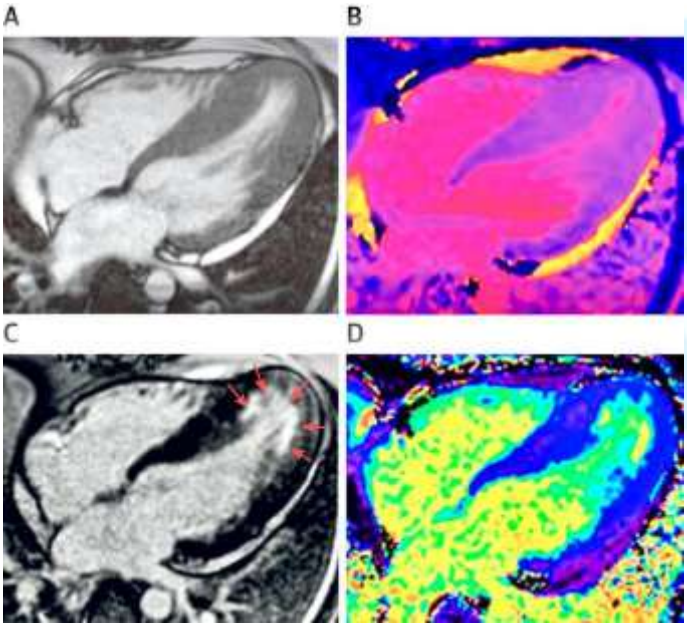


Fig. 2 Cardiac MR images of AHCM, (A) Four-chamber cine; (B) native myocardial T1 mapping by MOLLI; (C) LGE; (D) ECV. There is subendocardial patchy LGE in the mid-to-apical lateral wall and mid-to-apical septum including the true apex (red arrows). Native myocardial T1 and ECV values are increased in the mid-to-apical lateral wall (~1,130 ms and 49%, respectively, when normal native myocardial T1 range is 970 to 1,050 ms and normal ECV is 26% to 28% by MOLLI. AHCM = apical hypertrophic cardiomyopathy; ECV = extracellular volume fraction; LGE = late gadolinium enhancement; MOLLI = modified Look-locker inversion recovery sequence.

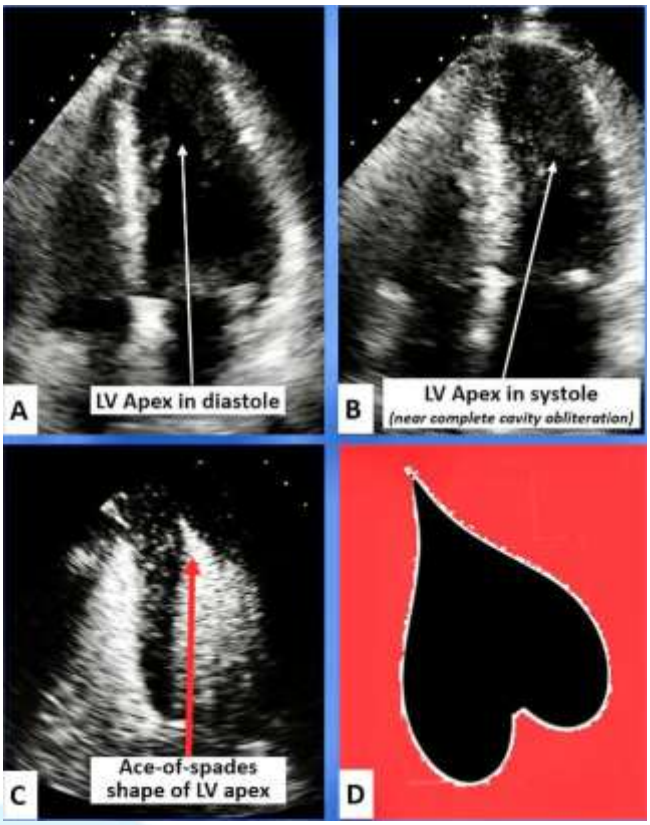


Fig. 3 Contrast echocardiography. The figure above shows 2-D echocardiographic images in apical hypertrophic cardiomyopathy, also referred to as Yamaguchi syndrome. Figure A shows the thickening of the mid to apical wall segments of the left ventricle (LV) in diastole, while figure B shows the near complete obliteration of the LV apical cavity in systole due to the hypertrophy. Figure C demonstrates to characteristic Ace-of-spades appearance of the LV using contrast, as depicted in the cartoon in figure D.

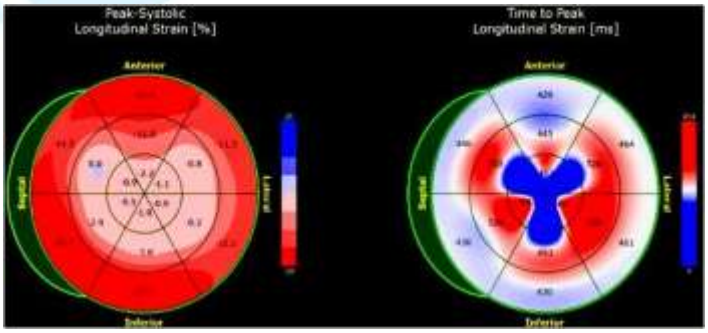


Fig. 4 Global longitudinal strain imaging. Parametric maps (bull's-eye displays) of myocardial global longitudinal peak systolic strain (left) and time to peak longitudinal strain (right) demonstrate the characteristic blueberry-on-top patterns.

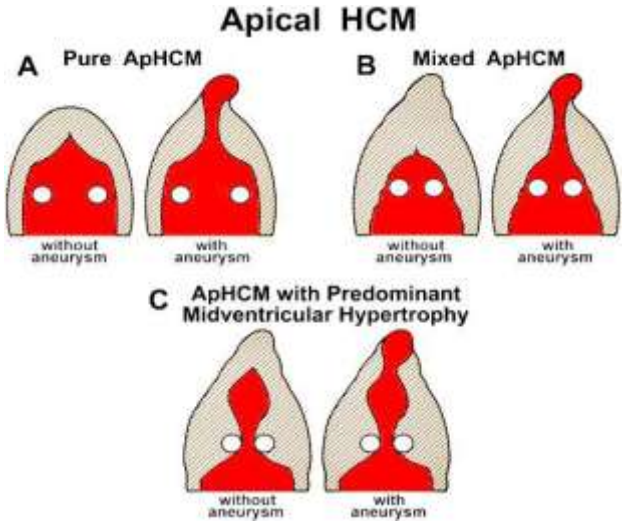


Fig. 5 AHCM presenting with apical aneurysm. (A) Pure apical hypertrophic cardiomyopathy with and without aneurysm; (B) Mixed apical hypertrophic cardiomyopathy with and without aneurysm; (C) Apical hypertrophic cardiomyopathy accompanied by mid ventricular hypertrophy- with and without aneurysm

2. Case Report

A 76 year elderly male presented to our cardiology OPD for a check up due to atypical chest pain, cough and mild fever since last 5 days. The patient denied any history of cardiovascular risk factors (smoking, tobacco chewing, hypertension, diabetes, dyslipidemia).

On clinical examination, the patient was healthy looking and normally built (Figure 6). The patient's weight was 73 kg, height was 156 cm, pulse rate was 69/min, blood pressure was 130/80 mmHg, respiratory rate was 16/min and SPO2 was 98% at room air. All the peripheral pulses were normally palpable without any radio-femoral delay. Cardiovascular and systemic examination were normal.

Fig. 6 Facial appearance of the elderly gentleman

Xray chest (PA) view (Figure 7) showed a normal cardiac size with a non-homogeneous opacity in the lower zone of right lung, suggestive of pneumonitis patch.

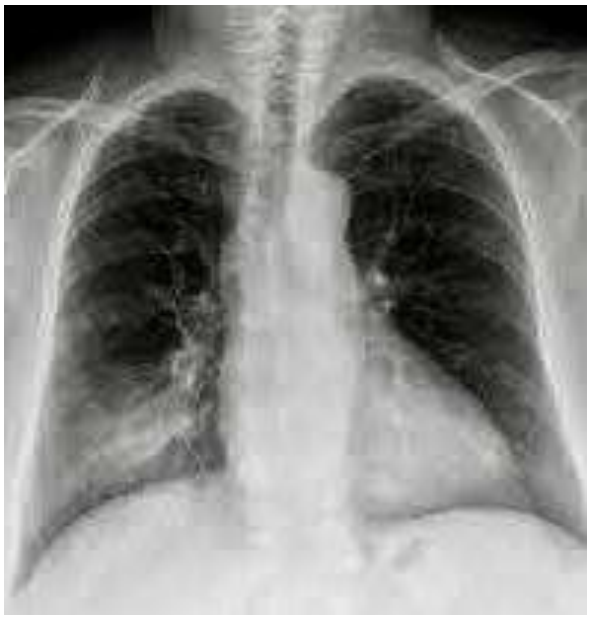


Fig. 7 X-ray chest (PA view). The cardiac size is normal with normal pulmonary blood flow. A non homogeneous opacity is visualized in the lower zone of right lung suggestive of pneumonitis patch

The resting ECG (Figure 8) revealed classical features of LVH with deep T wave inversion in precordial leads V_2-V_6, consistent with apical hypertrophic cardiomyopathy.

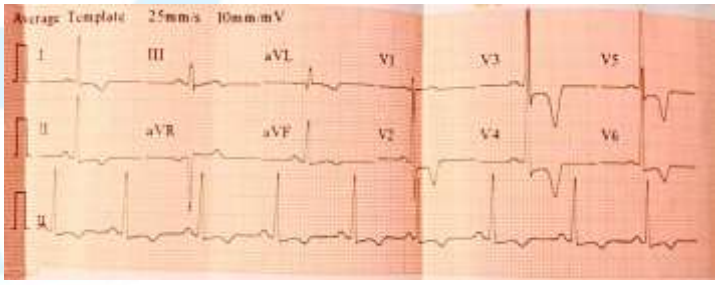


Fig. 8 Resting ECG. Classical ECG features of apical hypertrophic cardiomyopathy is elucidated accompanied by left ventricular hypertrophy. The conspicuous deep T wave inversion in the precordial leads V2- V6 is striking. There is normal sinus rhythm with a ventricular rate of 70/min and normal QRS axis

2.1 Transthoracic Echocardiography

All echocardiography evaluations were performed by the author, using My Lab X7 4D XStrain echocardiography machine, Esaote, Italy. The images were acquired using an adult probe equipped with harmonic variable frequency electronic single crystal array transducer while the subject was lying in supine and left lateral decubitus positions.

Conventional M-mode, two-dimensional and pulse wave doppler (PWD) and continuous wave doppler (CWD) echocardiography was performed in the classical subcostal, parasternal long axis (LX), parasternal short axis (SX), 4-Chamber (4CH), 5-Chamber (5CH) and suprasternal views (Figures 9-17).

2.2 M-mode Echocardiography

M-mode echocardiography of left ventricle was performed and the estimated measurements are outlined (Table 1, Figure 9).

Table 1 Calculations of M-mode echocardiography

Measurements	LV
IVS d	12.7 mm
LVID d	45.8 mm
LVPW d	10.3 mm
IVS s	15.8 mm
LVID s	23.4 mm
LVPW s	18.6 mm
EF	80 %
% LVFS	49 %
LVEDV	96.3 ml
LVESV	18.9 ml
SV	77.4 ml
LV Mass	192 g

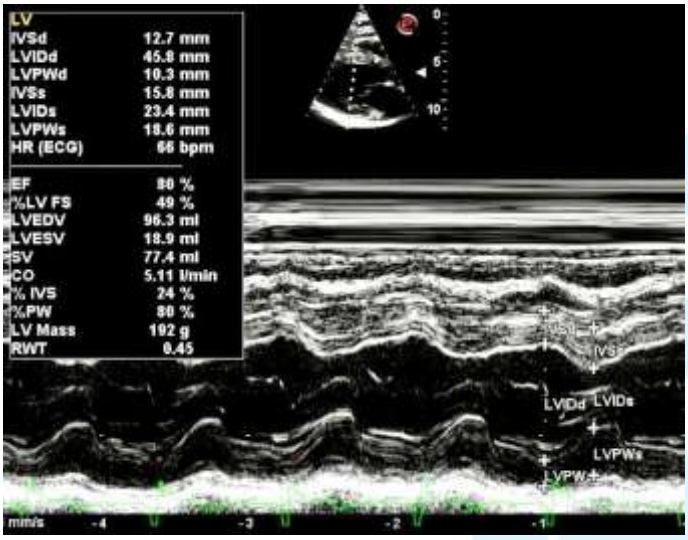


Fig. 9 M-mode measurements of left ventricle

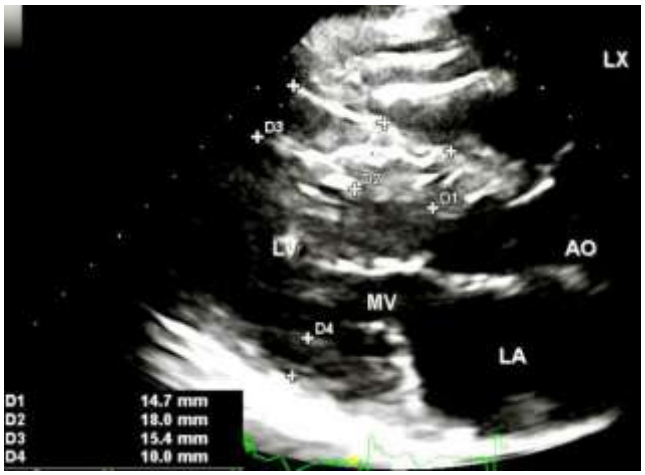
2.2.1 Summary of M-mode echocardiography

The LV was of normal size and the LVEF was 80 %. There was asymmetrical septal hypertrophy: interv-enetricular septal thickness and posterior wall thickness was 12.7 mm and 10.3 mm respectively. There was no apparent regional wall motion abnormality.

2.3 2 Dimensional transthoracic echocardiography

In the LX view, there was conspicuous presence of asymmetrical septal hypertrophy (Figure 10).

IVS thickness basal (D) 14.7 mm
Mid (D) 18.0 mm
Apical (D) 15.4 mm
LVPW thickness (D) 10.0 mm
IVS/LVPW ratio = 1.8:1



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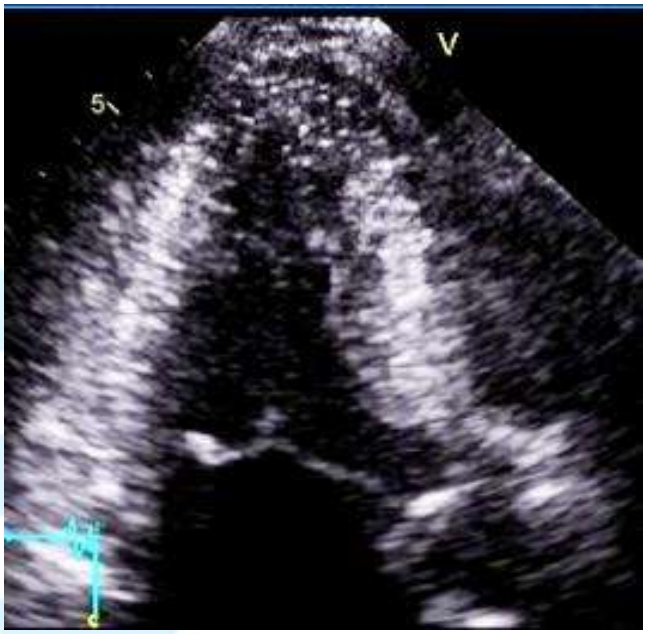

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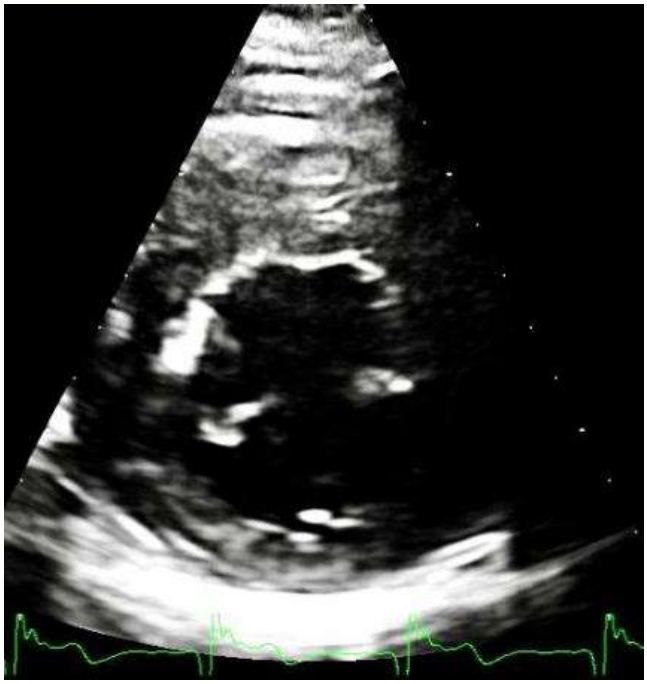

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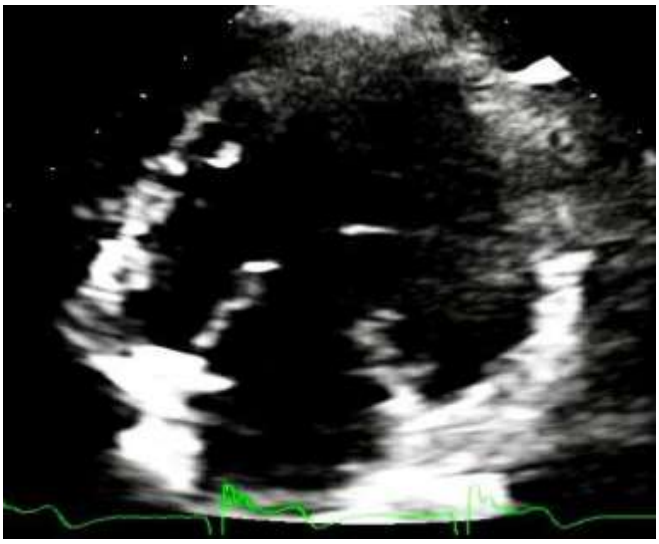


(D)

Fig. 10 2Dimensional Transthoracic Echocardiography. (A) LX view; (B) Apical 3CH view (LAX view); (C) SX view at the level of papillary muscle; (D) Apical 4CH view - large elongated AML and PML are visualized

Additionally, other important echocardiographic findings are mentioned below:

- Markedly thickened LV apex and apical lateral wall was appreciated.
- There was no SAM/LVOT obstruction/MVP.
- LVEF was normal
  M-mode - 80 %
  Simpson's biplane method - 51 % (Figure 11).
- Mitral regurgitation (mild) was present (Figure 12);
  MV was thickened, MR velocity was 4.42 m/sec
  On color flow mapping MR JET area was 1.10 sqcm; 10 % of LA area, eccentric posterior jet.
- The dimensions of LV were normal and there was no regional wall motion abnormality.
- Mild PAH was present and estimated RVSP/PAP was 37 mmHg.
- Pulse wave doppler analysis across the mitral valve and tissue doppler imaging of the base of lateral wall of LV were un-remarkable (Figure 13).

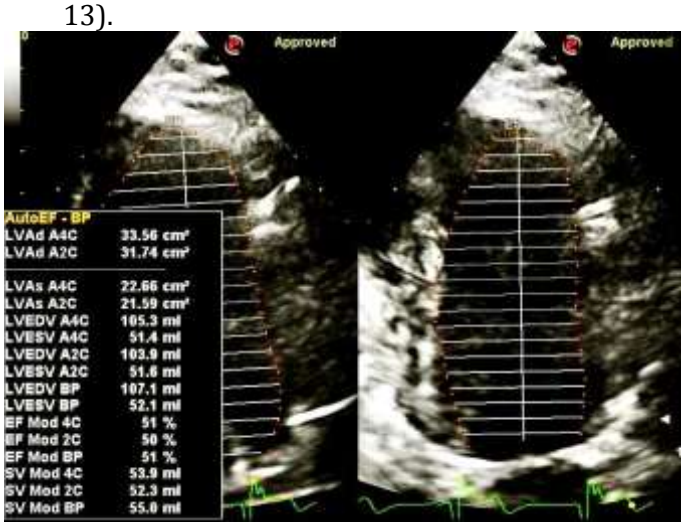


Fig. 11 Simpson's biplane method- LVEF 51 %

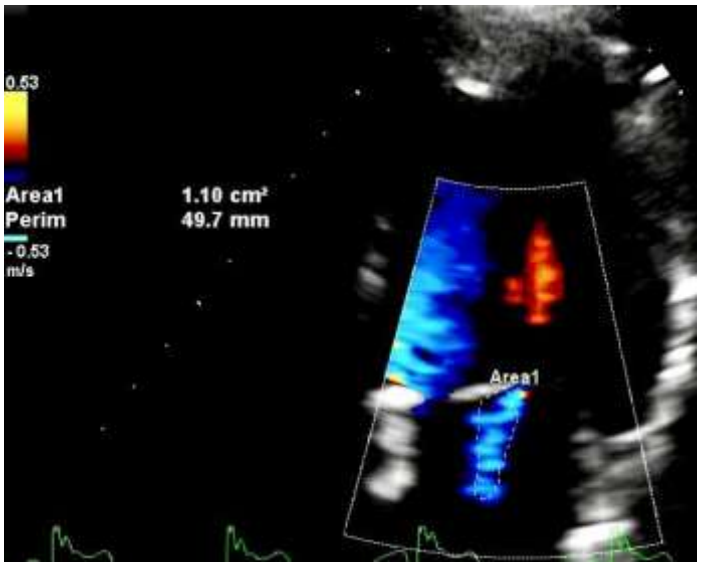
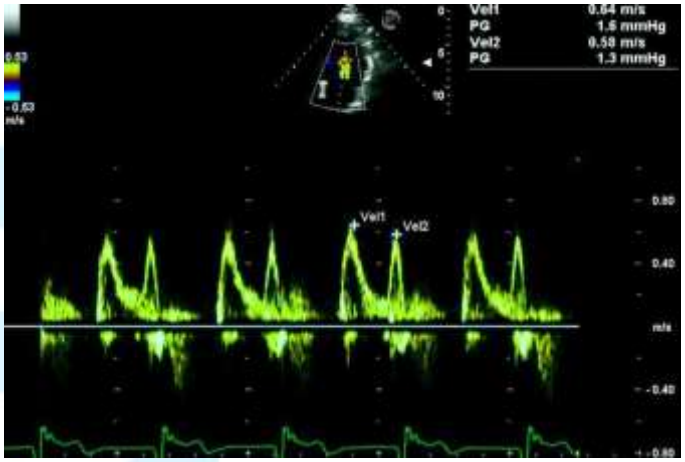


Fig. 12 Mild mitral regurgitation is identified with a jet area of 1.10 sqcm and a central jet



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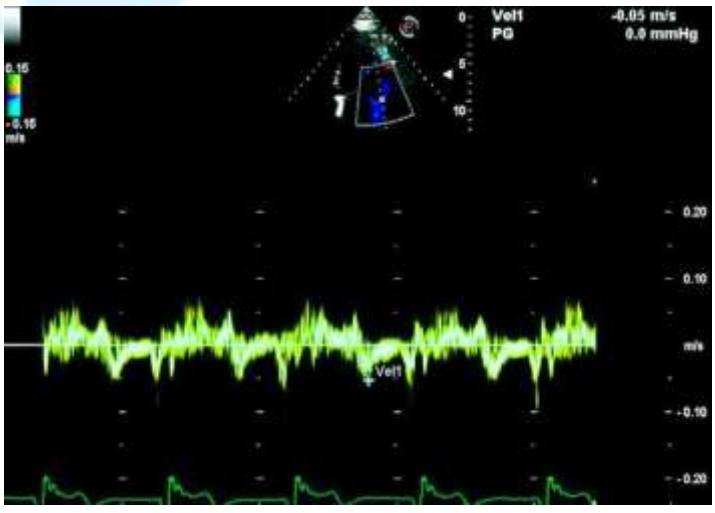


(B)

Fig. 13 (A) Pulse wave Doppler across mitral valve; (B) Tissue Doppler imaging of the base of lateral LV

2.4 4 Dimensional volumetric data

Table 2 depicts the 4 Dimensional volumetric data (Figure 14).

Table 2 4 Dimensional volumetric data

Parameters	Values
LVEDV	109.40 ml
LVESV	67.24 ml
EF	38.54 %
CO	2593.62 ml/min
Sph i d	0.56
Sph i s	0.42

EDV, end diastolic volume; ESV, end systolic volume; EF, ejection fraction; CO, cardiac output; Sph i d, sphericity index diastole; Sph i s, sphericity index systole

Interestingly, the 4Dimensional LVEF acquired from 4 Dimensional STE was 38.54%.

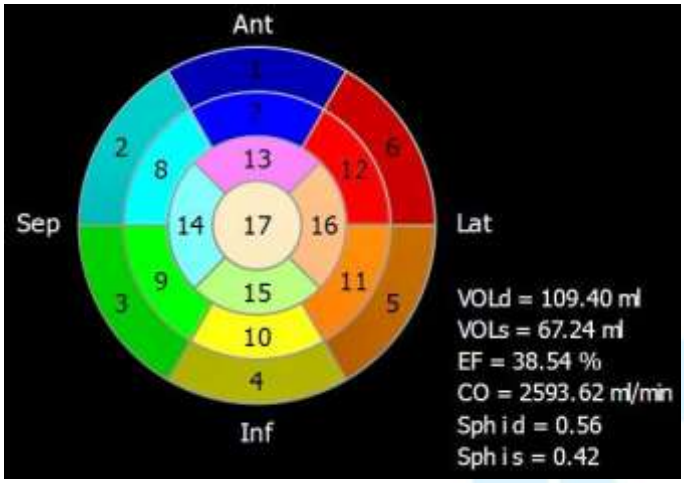


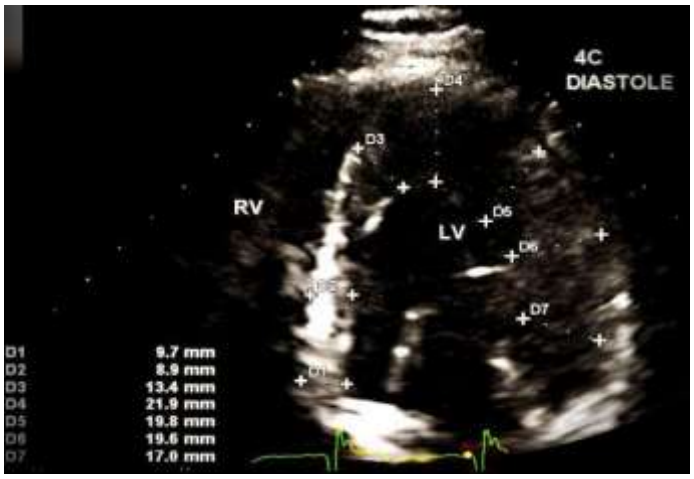
Fig. 14 4 Dimensional Volumetric data derived from 4 Dimensional XStrain Echocardiography. LVEF was 38.54 %

2.5 Contrast Study

In the normal Apical 4CH view the LV endocardial borders were fuzzy and not clearly visible (Figure 15 A). Therefore contrast study was carried out and it distinctly delineated the LV endocardial border (Figures 15 B, C). The contrast study exhibited marked thickness of LV apex and the lateral wall.

2.5.1 4 Chamber view (Diastole)

Normal Study	**Contrast Study**
Apex (D) 21.9 mm	Apex (D) 19.0 mm
Apical lateral (D) 15.4 mm	Apical lateral (D) 18.0 mm
Mid lateral (D) 19.6 mm	Mid lateral (D) 18.1 mm
Basal lateral (D) 17.0 mm	Basal lateral (D) 17.6 mm
Apical septum(D) 19.8 mm	Apical septum (D) 11.3 mm
Mid septum (D) 8.9 mm	Mid septum (D) 11.5 mm
Basal septum (D) 9.7 mm	Basal septum (D) 12.4 mm
IVS/LVPW ratio = 2.19:1	IVS/LVPW ratio = 19:1



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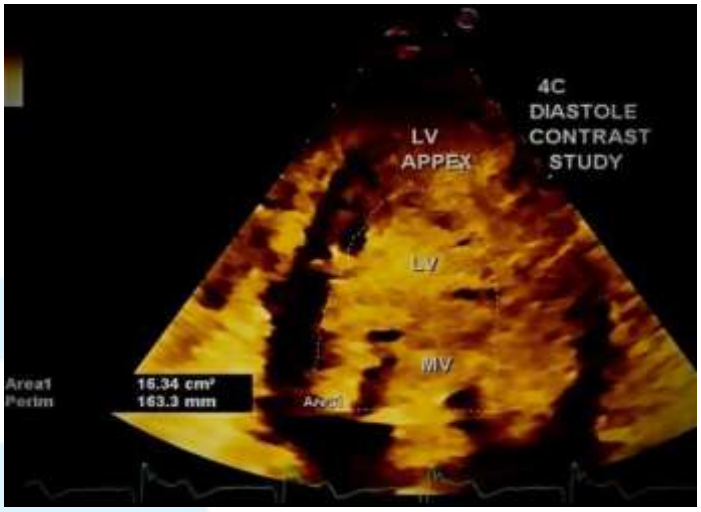

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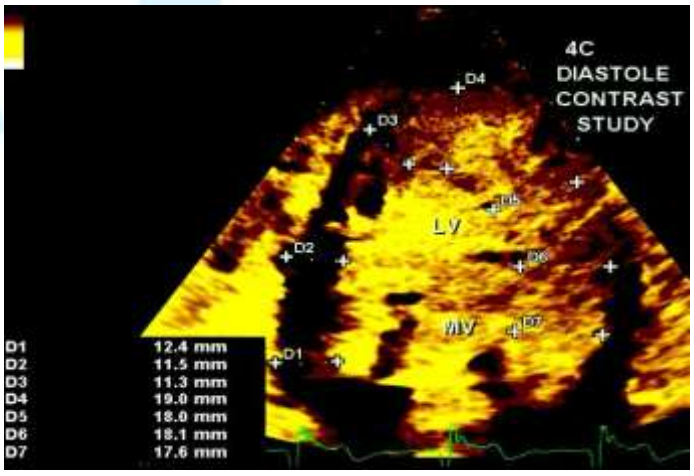


(C)

Fig. 15 Contrast Study. (A) LV apical 4CH view depicting indistinct endocardial border; (B) Contrast study depicting the distinct LV endocardial border; (C) Contrast Study elucidating the measurements performed at different locations of LV myocardial thickness in diastole

Table 3 exhibits LVEF variability with different methodologies of measurements. The 4Dimensional LVEF derived from 4DXStrain STE showed remarkable reduction of LVEF with a value of 38.54 % despite normal LVEF obtained from M-mode and Simpson's biplane method.

Table 3 LV ejection fraction variations by different methodologies

Technique	LVEF (%)
M-mode echocardiography	80%
Simpson's biplane method	51 %
4Dimensional LVEF derived from 4DXStrain echocardiography	38.54%
LVEF, left ventricular ejection fraction	

2.5.2 Speckle tracking echocardiography

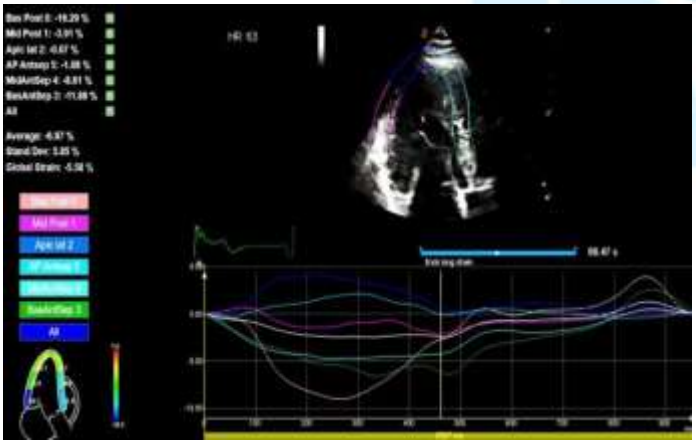
Comprehensive speckle tracking echocardiography (STE) was accomplished by 4Dimensional XStrain technique. The values obtained of various LV strain parameters are enumerated (Table 4):

Table 4 4 Dimensional XStrain echocardiography - estimated values of LV strain parameters in our index patient

LV strain Parameters	Strain (%)
Global longitudinal strain (GLS)	
AP 2C	-7.31
AP LAX (3CH)	-5.58
AP 4C	-7.97
Global strain	-6.96

AP, apical; 2C, two chamber; LAX, long axis; 4C, four chamber

There was severe reduction of global longitudinal strain (GLS) in apical 2CH, 3CH and 4CH view with a GLS value of -6.96 % (Figures 16, 17).



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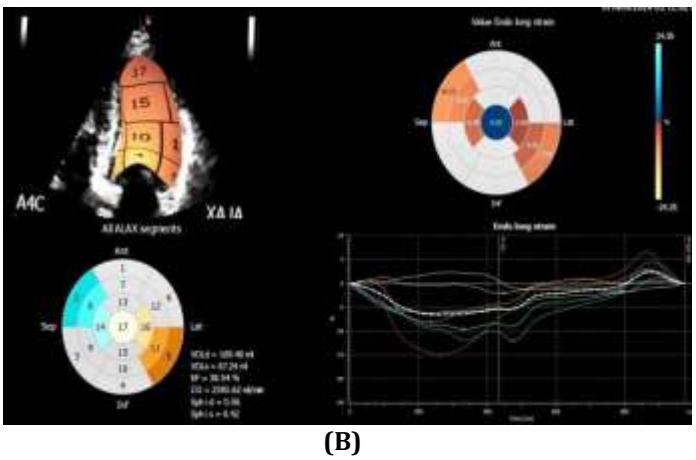

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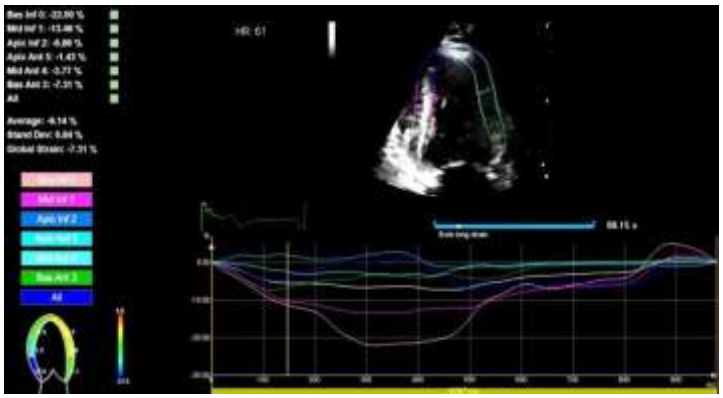

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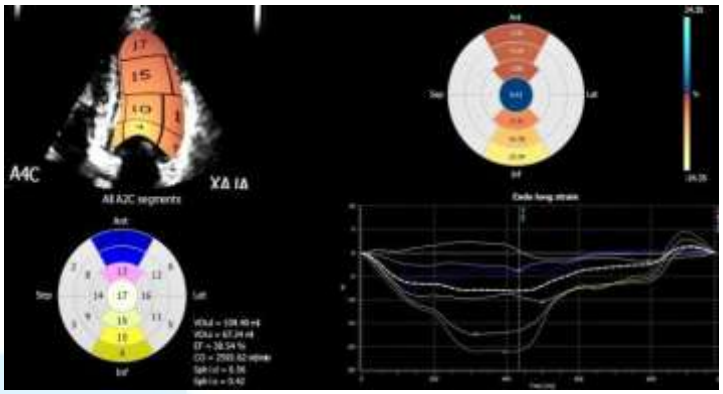

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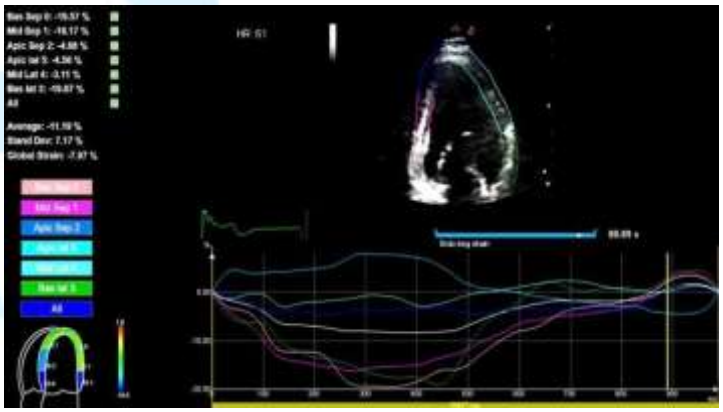

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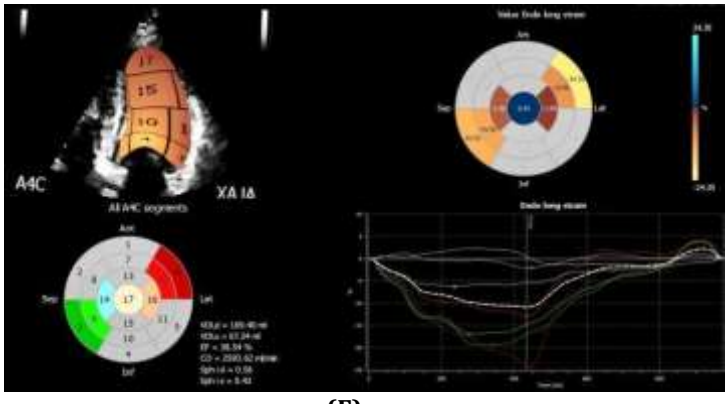
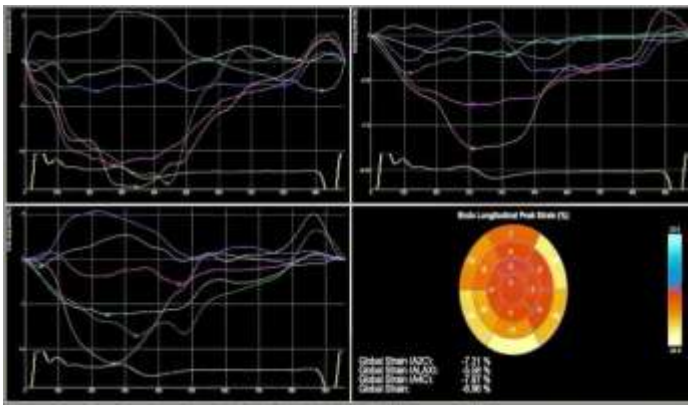
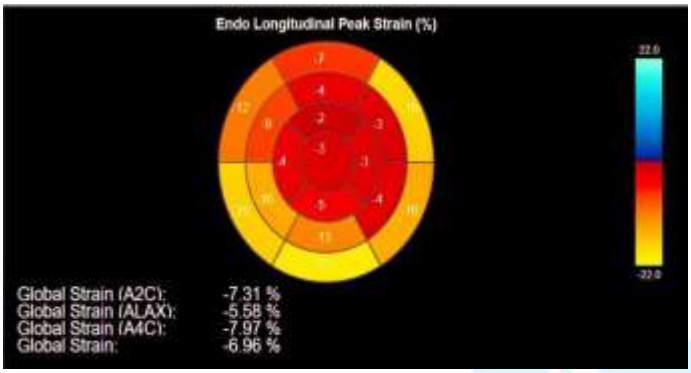


(F)

Fig. 16 Apical peak global longitudinal strain. (A) Apical LAX (3CH) GLS; (B) Apical LAX parametric maps (Bull's -eye displays) and graphs; (C) Apical 2CH GLS; (D) Apical 2CH parametric maps (Bull's -eye displays) and graphs; Apical 4CH GLS; (F) Apical 4CH parametric maps (Bull's -eye displays) and graphs



(A)



(B)

Fig. 17 (A) Global Longitudinal Strain. Parametric analysis and graphical display; (B) Bull's eye plot showing GLS values

A 17 segment model of 4D XStrain global segmental strain, provided a detailed picture of regional myocardial function, by analyzing each segment individually, all with in a 4D (three-dimensional over time) imaging framework. The values obtained are mentioned in Table 5.

Table 5 4 Dimensional XStrain echocardiography estimation of LV segmental endocardial longitudinal strain

Bull's eye analysis		
Endo long strain (Peak)		
Bas Ant	-7.31	%
BasAntSep	-11.88	%
Bas Sep	-19.57	%
Bas Inf	-22.00	%
Bas Post	-16.92	%
Bas lat	-19.07	%
Mid Ant	-3.77	%
MidAntSep	-8.61	%
Mid Sep	-16.17	%
Mid Inf	-13.46	%
Mid Post	-3.91	%
Mid Lat	-3.11	%
Apic Ant	-1.89	%
Apic Sep	-4.32	%
Apic Inf	-5.36	%
Apic lat	-3.47	%
Apex	-3.11	%
Global Strain (A2C)	-7.31	%
Global Strain (ALAX)	-7.97	%
Global Strain (A4C)	-5.58	%
Global Strain	-6.96	%

Red squares depict significant reduction in strain values in multiple segments of LV

2.5.3 Summary of 2 Dimensional transthoracic echocardiography

2D TTE in LX, SX Apical 4CH and Apical 3CH views demonstrated the presence of AHCM in our elderly asymptomatic patient. The thickness of LV apex was 21.9 mm and ratio of apex to posterior wall was 2.19:1. The LVEF determined by M-mode and Simpson's biplane method was normal, 80% and 51%, respectively. However, 4 Dimensional volumetric analysis acquired from 4DXStrain STE validated a markedly reduced LVEF - 38.54 %.

Due to the presence of ill-defined LV endocardial borders, we conducted a contrast study to clearly demarcate the boundaries of LV. We found LV apical thickness of 19 mm and the ratio of apex to posterior wall was 1.9:1.

Furthermore, speckle tracking imaging was executed by 4DXStrain STE technique which exhibited severe reduction of GLS and moreover, on segmental strain analysis, nearly all the segments illustrated substantial decline in strain values.

3. Discussion

Hypertrophic cardiomyopathy (HCM) is mostly an autosomal dominant disease characterized predominantly by the detection of left ventricular (LV) hypertrophy in the absence of another cardiac, systemic, or metabolic disease (Ommen et al., 2020).

3.1 Epidemiology

Hypertrophic cardiomyopathy is estimated to affect 1 out of 500 people. The AHCM has a different prevalence in cohorts of patients with HCM, which is relatively higher in Asian ethnicity. It is considered less common (8%) in Europe and North America with a majority (84%) of white race (Neubauer et al., 2019). However, AHCM may occur more frequently in Asian race, in whom it is seen in up to 40% of HCM patients (21% in China (Yin et al., 2022), 30% in Japan (Kubo et al., 2009), and 38% in Korea (Moon et al., 2011)) of patients with HCM.

AHCM is worldwide in distribution and affects males more frequently than females, with male-to-female ratios typically 1.6 to 2.8:1 (Klarich et al., 2013). Most commonly diagnosed in midlife (Neubauer et al., 2019), the early- and late-onset expressions are also known to occur.

3.2 Classification

According to the segments of LV hypertrophy, HCM can be classified into basal (also called classic HCM), midventricular, and apical (Ommen et al., 2020; Elliott et al., 2014).

Morphologically AHCM can be subclassified into three subtypes (Hughes et al., 2020): pure, mixed, and relative AHCM (Figure 18).

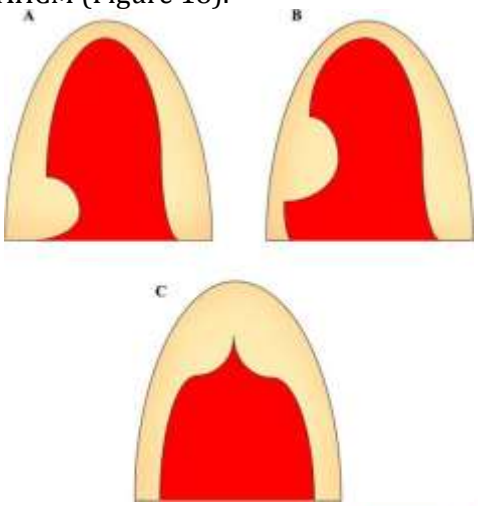


Fig. 18 Diagrammatic illustration of AHCM variants. Variants of hypertrophic cardiomyopathy. Different morphologic types of hypertrophic cardiomyopathy: A basal, B midventricular, and C apical

Pure AHCM presents with hypertrophy that is confined to the apex. Mixed AHCM displays both apical and septal hypertrophy but with the thickest apex. Finally, relative AHCM is believed to be an early AHCM phenotype. Individuals with relative AHCM do not meet conventional diagnostic criteria for AHCM but have similar imaging findings with the pure type. Relative AHCM can be diagnosed when electrocardiography shows characteristic precordial T-wave inversion and cardiac imaging techniques show apical wall thickness exceeding basal wall thickness, although failing to reach the cutoff of wall thickness ≥ 15 mm (Flett et al., 2015).

3.3 Clinical Diagnosis

Octo- and nonagenarians with AHCM are rare. An 84-year-old woman with AHCM was reported by Stöllberger et al (2015). In that case, a 12-lead ECG showed inverted T waves in the lateral precordial leads, and an echocardiogram showed predominantly apical hypertrophy. Cardiac magnetic resonance imaging confirmed the diagnosis of AHCM and showed a small, left ventricular apical aneurysm. They had no evidence of life-threatening arrhythmias before they died at 93 years of age due to noncardiac causes despite the presence of an apical aneurysm. This suggests that many patients remain undiagnosed due to the benign course of AHCM. Only 10% to 20% of patients with hypertrophic cardiomyopathy are diagnosed clinically (Maron et al., 2022), and this number is likely lower in patients with AHCM.

Støylen et al. (2003) reported the case of a 92-year-old woman with AHCM who presented with syncope and dyspnea and was found to have nonobstructive hypertrophic cardiomyopathy with postsystolic shortening leading to delayed emptying of the apex. Doppler flow showed delayed emptying of the apical chamber extending into the early filling phase.

In a study of a patient cohort at Mayo Clinic, Rochester, Minnesota, a patient with AHCM is reported who died at 92 (Klarich et al., 2013).

The differential diagnosis of AHCM should always be considered in elderly patients.

Table 6 Differential diagnosis of apical hypertrophic cardiomyopathy (Yusuf et al., 2011)

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

3.3.1 Diagnostic criteria

AHCM is described as an electrocardiographic pattern of giant negative T-waves (≥ 1 mV) together with a "spade-like configuration" of the LV cavity on the left ventriculography (Yamaguchi et al., 1979). With advances in imaging techniques, the current definition mainly relies on demonstrating LV hypertrophy predominating in the LV apex, with the apical wall thickness ≥ 15 mm and a ratio of maximal apical to posterior wall thickness ≥ 1.5, based on echocardiography or CMR (Eriksson et al., 2022).

Standard TTE may miss this entity where a contrast echocardiography may be useful but cardiac MR imaging (CMR) is best (Yamaguchi et al., 1979; Hansen and Merchant, 2007).

CMR is superior to TTE in detecting variants of HCM such as apical HCM, presence of focal hypertrophy, severe hypertrophy (>30 mm) and LV apical aneurysm. Patchy mid-wall-type delayed hyperenhancement of myocardium, on contrast-enhanced CMR, identifies fibrosis. It is associated with a higher risk of adverse LV remodelling and systolic dysfunction. Its association with ventricular tachycardia has been reported but its predictive value for sudden cardiac death is not clear (Hansen and Merchant, 2007; Maron, 2012).

3.3.2 Electrocardiography

The trademark electrocardiographic finding for AHCM is LV hypertrophy with giant T waves, which are defined as T-waves ≥ 1 mV in any electrocardiography lead. These giant T waves are dynamic and will change over time, or even disappear, as a part of the natural history of AHCM (Koga et al., 1995). The depth of T waves varies among patients, in a study of 208 AHCM patients, Yan et al. (2012) reported that 60 (28%) patients had giant T-waves, but only 11% of AHCM patients had giant T waves in a series from Mayo Clinic (Klarich et al., 2013). Although giant T-waves in the setting of LV hypertrophy are considered pathognomonic for AHCM, it is unlikely to rule out other causes of ST-T wave abnormalities, such as other types of HCM, coronary heart disease, medication effect (e.g., digoxin), and neurological diseases (e.g., subarachnoid hemorrhage).

Ambulatory 24- or 48-h electrocardiography is crucial to detect non-sustained ventricular tachycardia (NSVT), AF, and ventricular fibrillation. Eriksson et al. (2002) showed that Holter monitor recordings revealed NVST in 20 patients (23%) with AHCM, which was found to be correlated with the presence of fibrosis. Several studies have reported that the prevalence of AF was 11–17% (Yin et al., 2022; Kubo et al., 2009). LA enlargement secondary to LV diastolic dysfunction in patients with AHCM can predict subsequent AF, which is prognostically adverse (Chen et al., 2018).

3.3.3 Transthoracic echocardiography

In contemporary clinical practice, transthoracic echocardiography (TTE) is the first-line imaging modality for the evaluation of AHCM because of its noninvasiveness, wide availability, and low cost (Tower-Rader et al., 2020; Nagueh et al., 2022). A comprehensive echocardiographic examination should involve the assessment of (1) the distribution and magnitude of LV hypertrophy; (2) the presence of cavity obliteration and apical aneurysm; (3) obstruction at any level in the LV; (4) systolic and diastolic function; (5) and mitral apparatus and papillary muscle abnormalities (Tower-Rader et al., 2020; Nagueh et al., 2022; Cardim et al., 2015).

Local dysfunction involves the apical segments, manifested as hypokinesia, akinesia, or dyskinesia, especially in the presence of an apical aneurysm (Binder et al., 2011). LV twist is significantly decreased due to a reduction in apical rotation, leading to the loss of early diastolic suction and eventually diastolic dysfunction in AHCM (Chang et al., 2010). In AHCM patients with MVOCO, diastolic intraventricular gradient often exists, manifesting as a directed flow of apex to base during early diastole—the PJF phenomenon, (Sherrid et al., 2023) which is associated with a high risk of systemic embolism, perfusion abnormalities, and ventricular arrhythmias. In our patient, TTE in the apical 4CH and LX view showed thickness of LV apex and posterior wall to be 21.9 mm and 10 mm, respectively. The ratio of apical to posterior wall thickness was 2.19:1. The LVEF was normal by M-mode and Simpson's biplane method 80% and 51 %, respectively, even though there was marked reduction of LVEF (38.54 %) by 4D volumetric analysis.

3.3.4 Contrast echocardiography

Because of frequent difficulties in visualizing the apical endocardium, 2D echocardiography may result in misdiagnosis, as it was reported that echocardiography failed to diagnose AHCM in 31.7% of patients (Yan et al., 2012). Therefore, there is a need for an alternative imaging technique, and contrast echocardiography has been recommended as an alternative when 2D images are suboptimal (Senior et al., 2017). The administration of echocardiographic contrast allows the excellent visualization of LV morphology and the presence of an apical aneurysm.

In our index patient, on contrast study we identified thickness of LV apex to be 19 mm and the apical to posterior wall thickness ratio was 19:1. We did not find any LV apical wall motion abnormality or apical aneurysm.

3.3.5 Late Onset AHCM

The reason for the late and gradual onset of apical HCM compared to other HCM variants is not established (Towe et al., 2015). Compared to non-apical HCM, apical HCM seems to carry a more favourable prognosis (Itzhaki et al., 2018).

3.3.6 Speckle tracking echocardiography

Regional LV function may be assessed non-invasively by measuring strain or systolic deformation. Recently, a method derived from the two-dimensional (2-D) echocardiogram, called "speckle tracking" of 2-D strain, has been developed to measure systolic strain (Leitman 2004). In a study of Saccheri et al (2017) global longitudinal peak systolic strain (GLPSS), was studied in patients of AHCM. GLPSS curves represent the maximum myocardial longitudinal shortening during contraction in each of the 17 LV segments. In a normal subject GLPSS varies between -15% and -20% (Marwick et al., 2009). Mutations of genes that code for contractile proteins of the sarcomere are responsible for the structural and functional changes seen in patients with HCM, and cause ventricular hypertrophy, myofibrillar disarray and interstitial fibrosis. In spite of the hyperdynamic systolic function

seen by echo, 2-D strain detected a decrease in myocardial strain values (Saccheri et al., 2017).

In patients with HCM, Popović et al. (2008) have shown that 2-D strain was lower in patients whose MRI showed myocardial fibrosis than in patients without fibrosis. Moreover, in patients with apical HCM, longitudinal midwall strain identified subclinical global systolic dysfunction, with a lower intra and interobserver variability than for strain derived from colour tissue Doppler (Afonso et al., 2008).

In our index patient, similarly, despite normal LV systolic function on Simpson's biplane method there was severe reduction of GLS predominately affecting nearly all the myocardial segments.

4. Conclusion

AHCM may present in very elderly patients and may have a benign course. Further research is needed to understand why some patients have a higher risk of arrhythmias, heart failure, and SCD, while manifest with a more benign course.

Apical HCM is an under-recognized pathology with symptoms and ECG-changes mimicking coronary artery disease that can manifest gradually at a later age in a previously asymptomatic individual. Cardiac magnetic resonance imaging is useful to distinguish apical HCM from other causes of chest pain and ECG changes.

AHCM can be a challenging diagnosis. Contrast echocardiography must always be applied in cases of poor delineation of the LV apical endocardial border at baseline echocardiography. Timely detection and appropriate lifestyle intervention might reduce the severity of LV hypertrophy and minimize and delay HF related symptoms and arrhythmias. 2-D strain assessed by "speckle tracking" is a sensitive method to detect subclinical systolic LV dysfunction. We utilized an innovative technique of 4D XStrain speckle tracking echocardiography to substantiate global longitudinal strain values in our patient. The clinical application of this new finding may help in the pathophysiological interpretation of AHCM. Future studies, with more subjects, will allow assessing whether patients with greater change in strain may be at higher risk for ventricular arrhythmias, sudden death or progression to heart failure due to systolic dysfunction. Additionally, the method could help in evaluating the benefit of conventional treatment and new therapeutic strategies.

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