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TWO-DIMENSIONAL TRANSTHORACIC ECHOCARDIOGRAM (TTE) AND DOPPLER IMAGING IN HYPERTROPHIC CARDIOMYOPATHY (HCM) WITH MID-CAVITY SYSTOLIC OBSTRUCTION

Dr. Virendra C. Patil*

Associate Professor, Department of Medicine, Krishna Institute of Medical Sciences Deemed University (KIMSDU), Dhebewadi Road, Karad Dist: Satara, Maharashtra - 415110, India.

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***Correspondence for Author**
Dr. Virendra C. Patil
Associate Professor, Department of Medicine, Krishna Institute of Medical Sciences Deemed University (KIMSDU), Dhebewadi Road, Karad Dist: Satara, Maharashtra - 415110, India.

ABSTRACT

Systolic obstruction of the left ventricular outflow tract (LVOT), either at rest or during provocation, is a frequent finding in patients with hypertrophic cardiomyopathy (HCM). Left ventricular mid-cavity obstruction is uncommon. Intraventricular pressure gradients in association with either mid-cavity of apical hypertrophy are reported only rarely. We describe a trasthorasic 2-dimensional echocardiogram and Doppler findings of 48 year male patient with hypertrophic cardiomyopathy (HCM) with evidence of systolic left ventricular mid-cavity obstruction with significant gradient.

KEYWORDS: Left ventricular outflow tract (LVOT), hypertrophic cardiomyopathy, echocardiogram.

INTRODUCTION

Hypertrophic cardiomyopathy (HC) is a hereditary myocardial disorder, associated with mutations of sarcomeric proteins. In the past, HCM was considered a rare disorder associated with a poor prognosis but more recent population screening studies suggest that it is actually common (1:500 individuals).[1] Definition: Hypertrophic cardiomyopathy (HCM) is diagnosed on the basis of left ventricular hypertrophy (LVH) for which there is insufficient explanation (e.g. mild hypertension or mild aortic stenosis with marked hypertrophy).[2] Pathophysiology: The most characteristic morphological abnormality in hypertrophic cardiomyopathy (HCM) is the excessive hypertrophy of and no dilated left ventricle with absence of other cardiac or systemic diseases that could produce left ventricular hypertrophy. Often, it is asymmetric in nature, with a

preference for ventricular septum and occurs either in sporadic or familial forms. From the genetics point of view the disease is highly variable with respect to the specific gene mutation and degree of penetration.[3] Haemodynamics: Twenty five percent of cases of HC there is associated obstruction to left ventricular (LV) outflow (LVOT). Obstruction can occur at several locations within the ventricle, depending on the distribution of hypertrophy, including at the mitral valve level with systolic anterior motion (SAM) of the mitral valve, at the mid-ventricle; or within the cardiac apex. In about 5% of patients a mid-ventricular obstruction (MVO) is be observed. These patients are often symptomatic from hemodynamic causes and are also prone to symptomatic and even lethal ventricular arrhythmias. MVO may be associated with hypertrophy of the papillary muscle(s) and apical LV aneurysm. MVO usually presents as local obstruction but can be associated with SAM and obstruction at both the mid-cavity and outflow levels.[1] Symptomatology: There is no correlation between the severity of the disease and symptomatology. Often, many patients with HCM are lack symptoms for a long period of time and frequently, the disease is detected in case of presence murmur or arrhythmia. Classically, symptoms include pulmonary congestion, fatigue, palpitations, chest pain, syncope and congestive heart failure. The clinical manifestation of the classic form of HCM result from systolic dysfunction and dynamic left ventricular outflow tract obstruction, diastolic dysfunction, mitral regurgitation, a high prevalence of arrhythmias and sudden cardiac death. A number of patho-physiological components and process are identified: systolic dysfunction and left ventricular outflow tract obstruction, diastolic dysfunction, coronary artery abnormalities, leading to myocardial ischemia, mitral regurgitation, arrhythmias and sudden cardiac death.[3] Echocardiography is an invaluable tool in the diagnosis and follow-up of patients with HCM. It is essential when assessing a patient with a potential diagnosis of hypertrophic cardiomyopathy to carry out a systematic study of cardiac structure and function.[2] Diagnosis: M-mode and 2-D echocardiography are the primary screening and evaluation of HCM, whereas Doppler imaging can fully delineate the entire spectrum of hemodynamic abnormalities. An atypical form of hypertrophic cardiomyopathy is presented with mid-cavity obstruction of left ventricle and fast increasing intra-cavity pressure. This form involves selective hypertrophy and obstruction at the mid-left ventricular level.[3, 4] Echocardiographic assessment requires a comprehensive assessment in several imaging planes with careful attention to correct beam alignment in order to minimize errors in the measurement of maximum pressure gradient, LVOT obstruction, LV wall thickness and appropriate segment identification of hypertrophy with an unusual distribution.[2] Diagnostic criteria: Although HCM is typically characterized by asymmetric

septal hypertrophy (ASH), almost any myocardial segment may be involved. The following 2-dimensional echocardiographic criteria are used to aid diagnosis: [unexplained maximal wall thickness >15 mm in any myocardial segment, or septal/posterior wall thickness ratio >1.3 in normotensive patients, or septal/posterior wall thickness ratio >1.5 in hypertensive patients.[2] Systolic anterior motion (SAM)] Haemodynamics of SAM and LVOT obstruction: The haemodynamic consequences of SAM include prolongation of ejection time and a reduction in stroke volume. Coaptation of the mitral leaflets may be disrupted resulting in mitral regurgitation. The presence of SAM is documented using M-mode echocardiography and is characterized by mid-systolic notching of the aortic valve and contact of the anterior mitral valve leaflet/chordae with the septum. The severity of SAM can be inferred from the duration of leaflet/chordal contact with the septum, being mild if contact occurs for <10% of systole, and severe if >30% of systole. The presence of resting obstruction, defined as a peak LVOT gradient >30 mmHg, has prognostic significance in HCM as a predictor of the risk of sudden cardiac death (SCD) and progression to heart failure. LVOTO arises due to narrowing of the LVOT by septal hypertrophy, anterior displacement of the mitral apparatus, and SAM.[2]

Summary of echocardiogram and Doppler report: Left ventricular systolic function was normal with LVEF of 66%. There was significant severe LV hypertrophy [IVSd:16.0 mm; LVPWd: 18.6 mm; LV mass: 311 gm] There was grade –III diastolic dysfunction (Pseudo-normalisation pattern)by PW and TDI. There was significant systolic pressure gradient at LV mid-cavity level [3.61m/s; 52.1 mmHg. Both the atria were mildly dilated with mild mitral regurgitation and mild tricuspid regurgitation. There was no SAM at rest. There was no resting regional wall motion abnormality. IAS and IVS were intact. Great arteries and IVC was normal. There was no pericardial effusion, no vegetation and no clot. All above findings favor diagnosis of hypertrophic cardiomyopathy with left ventricular mid-cavity obstruction with gradient of 52.1 mmHg with mild pulmonary artery hypertension (PAP: 37.2 mmHg.) [2-D-echocardiogram and Doppler study done by Siemens accuson X-300 machine with 3-5 MHz probe, as per standard guidelines][4] [Table no.1, figure no.1 and 2].

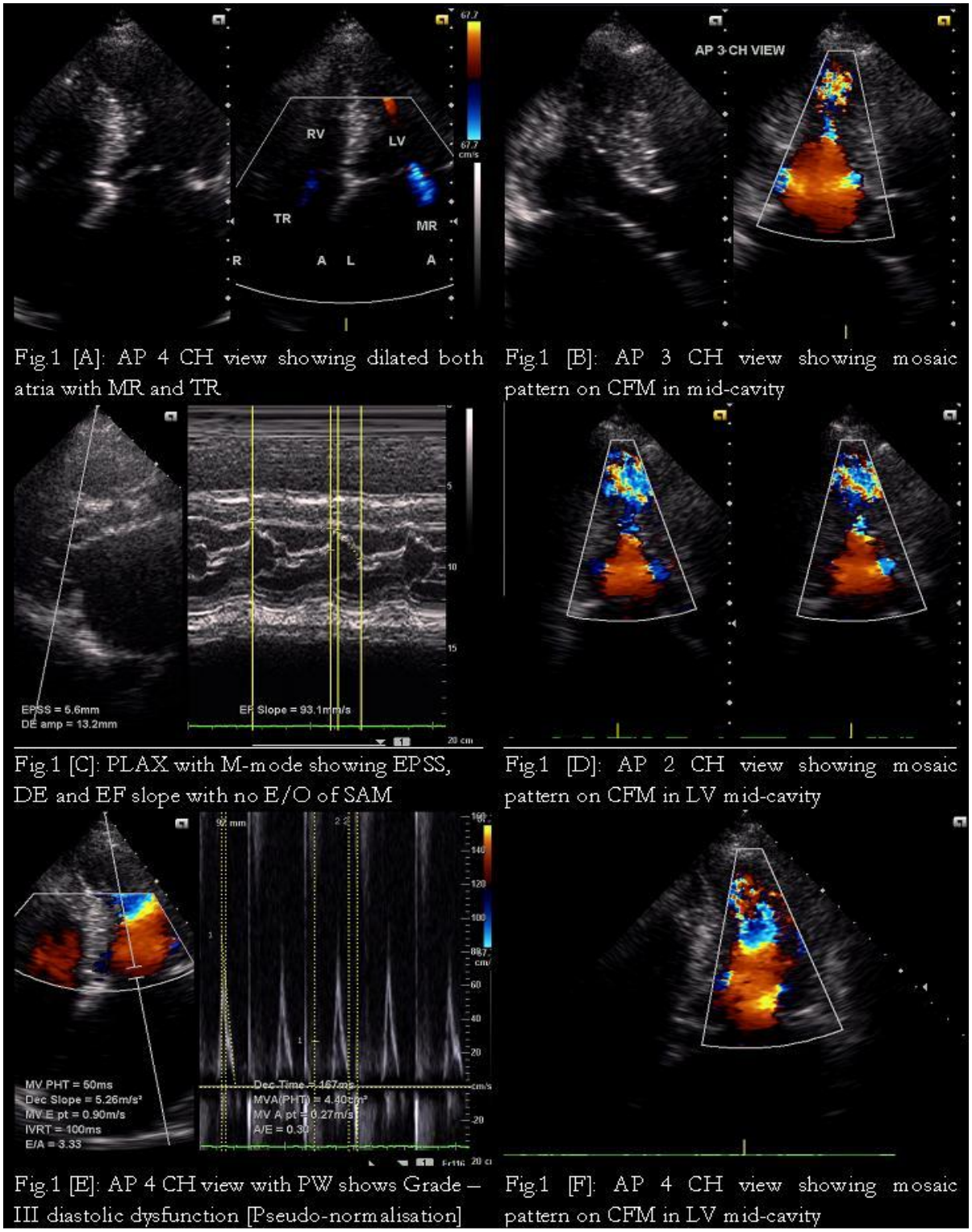

Fig.1 [A]: AP 4 CH view showing dilated both atria with MR and TR

Fig.1 [B]: AP 3 CH view showing mosaic pattern on CFM in mid-cavity

Fig.1 [C]: PLAX with M-mode showing EPSS, DE and EF slope with no E/O of SAM

Fig.1 [D]: AP 2 CH view showing mosaic pattern on CFM in LV mid-cavity

Fig.1 [E]: AP 4 CH view with PW shows Grade – III diastolic dysfunction [Pseudo-normalisation]

Fig.1 [F]: AP 4 CH view showing mosaic pattern on CFM in LV mid-cavity

Figure No. 1 [A]: AP 4 CH view showing dilated both atria with MR and TR, Fig.1 [B]: AP 3 CH view showing mosaic pattern on CFM in mid-cavity, Fig.1 [C]: PLAX with M-mode showing EPSS, DE and EF slope with no E/O of SAM, Fig.1 [D]: AP 2 CH view showing mosaic pattern on CFM in LV mid-cavity, Fig.1 [E]: AP 4 CH view with PW shows Grade –III diastolic dysfunction [Pseudo-normalisation], Fig.1 [F]: AP 4 CH view showing mosaic pattern on CFM in LV mid-cavity.

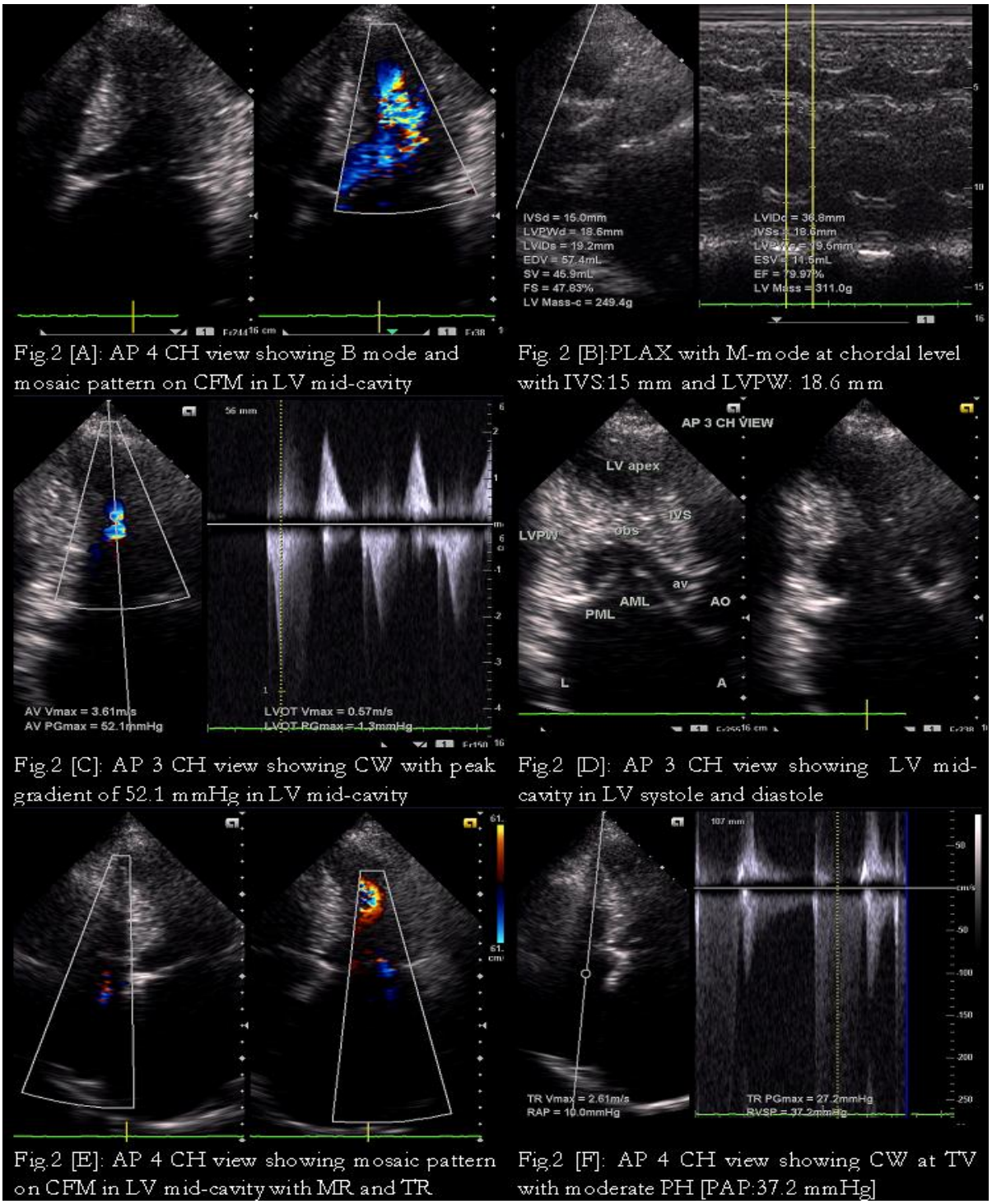

Fig.2 [A]: AP 4 CH view showing B mode and mosaic pattern on CFM in LV mid-cavity

Fig. 2 [B]:PLAX with M-mode at chordal level with IVS:15 mm and LVPW: 18.6 mm

Fig.2 [C]: AP 3 CH view showing CW with peak gradient of 52.1 mmHg in LV mid-cavity

Fig.2 [D]: AP 3 CH view showing LV mid-cavity in LV systole and diastole

Fig.2 [E]: AP 4 CH view showing mosaic pattern on CFM in LV mid-cavity with MR and TR

Fig.2 [F]: AP 4 CH view showing CW at TV with moderate PH [PAP:37.2 mmHg]

Figure no. 2 [A]: AP 4 CH view showing B mode and mosaic pattern on CFM in LV mid-cavity, Fig. 2 [B]:PLAX with M-mode at chordal level with IVS:16 mm and LVPW: 18.6 mm, Fig.2 [C]: AP 3 CH view showing CW with peak gradient of 52.1 mmHg in LV mid-cavity, Fig.2 [D]: AP 3 CH view showing LV mid-cavity in LV systole and diastole, Fig.2 [E]: AP 4 CH view showing mosaic pattern on CFM in LV mid-cavity with MR and TR, Fig.2 [F]: AP 4 CH view showing CW at TV with moderate PH [PAP:37.2 mmHg].

Table no. 1: Measurements of B-mode, M-mode and Doppler [PW, CW and colour flow map (CFM)]

Mitral Valve [PW]					
MV E pt	0.90	m/s	MV A pt	0.27	m/s
Dec Time	167	ms	Dec Slope	5.26	m/s^2
IVRT	100	ms	MV PHT	50	ms
E/A ratio	3.33		A/E ratio	0.30	
MVA(PHT)	4.40	cm^2	LVOT diam	16.1	mm
Aortic Valve [CW]					
LV mid cavity	**3.61**	**m/s**	**LV-Mid PG max**	**52.1**	**mmHg**
LVOT Vmax	0.57	m/s	LVOT PGmax	1.3	mmHg
LVOT diam	16.1	mm	AVA(Vmax)	0.32	cm^2
LV/Teich [M-mode]					
IVSd	**16.0**	**mm**	LVIDd	36.8	mm
LVPWd	**18.6**	**mm**	IVSs	18.6	mm
LVIDs	19.2	mm	LVPWs	19.5	mm
EDV	57.4	mL	ESV	11.5	mL
SV	45.9	mL	EF	79.97	%
FS	47.83	%	**LV Mass**	**311.0**	**g**
Mitral Valve [M-mode]					
DE amp	13.2	mm	EPSS	5.6	mm
EF Slope	93.1	mm/s			
MR [mitral regurgitation]					
MR Vmax	2.43	m/s	MR PGmax	23.6	mmHg
Dt	61	ms	dP/dt	524	mmHg/s
TR [tricuspid regurgitation]					
TR Vmax [CW]	2.2	m/s	RAP	10.0	mmHg

Differential diagnosis: In cases of unexplained LVH, accurate diagnosis of the underlying cause is inherently difficult and conditions such as HCM and hypertension may coexist. No single echocardiographic parameter is ideal, and history and clinical examination play a vital role. However, several echocardiographic clues may help in differential diagnosis. Hypertensive heart disease SAM is a recognized but uncommon finding in patients with hypertensive heart disease (HHD). It is mandatory to look for coarctation of aorta and renal artery stenosis in patient with LV symmetric hypertrophy. Although LVH is common in cardiac amyloid, several other characteristic echocardiographic features may help to distinguish this condition from HCM, including thickened LV walls and interatrial septum, increased myocardial echogenicity, thickening of the valve leaflets, and the presence of a pericardial effusion. In athletes, LVH demonstrate normal or supranormal TDI velocities with LV cavity dilation whereas, in HCM patients have impaired systolic and diastolic function on TDI analysis and presence of LA enlargement and abnormal diastolic function.[2, 5]

DISCUSSION

Echocardiography plays an important role in diagnosis, assessing hemodynamic of left ventricle, choosing treatment option and prognosis of HCM. Luckie M et al described a patient with hypertrophic cardiomyopathy with evidence of systolic mid-cavity obstruction, and with complex diastolic paradoxical flow abnormalities within the left ventricular cavity detected by colour and pulsed-wave Doppler.[6] Similarly our patient with HCM had LV hypertrophy with LV mid-cavity obstruction with pressure gradient of 52.1 mmHg with mild MR and mild pulmonary hypertension with grade-III diastolic dysfunction with preserved LV systolic function. Matsuno Y et al reported mid-ventricular obstruction in a 60 year woman with hypertrophic cardiomyopathy using a real-time two-dimensional M-mode echocardiography showed asymmetric septal hypertrophy: thickness of the end-diastolic left ventricular posterior wall was 9 mm, and that of the interventricular septum was 19 mm without systolic anterior motion (SAM) of the mitral apparatus. Doppler color flow imaging showed a mid-left ventricular narrowing in late-systole and a mosaic pattern was depicted from the mid-ventricle to the outflow tract. Continuous wave Doppler echocardiography disclosed a peak velocity of 2.0 m/sec (pressure gradient (PG) = 16 mmHg).[7] Similarly our patient had LV mid-cavity obstruction with pressure gradient of 52.1 mmHg without SAM on M-mode. Nakamura T et al studied seven patients with hypertrophic cardiomyopathy having mid-ventricular obstruction (MVO) to investigate intraventricular flow conditions. All MVO patients had "hour-glass" LV cavities during systole, resulting from either hypertrophy at the mid-ventricular level or hypertrophied papillary muscles, where systolic mosaic signals originated. Systolic peak flow velocities at the mid-ventricle ranged from 2.5 to 4.2 m/s, proving the presence of a pressure gradient between the apex and the base of the LV.[8] similarly our patient had masaic appearance at LV mid –cavity level with peak gradient of 52.1 mmHg [3.61m/s]. Georgios K et al described a case of HCM with mid-ventricular obstruction and apical aneurysm formation in 3 patients coming from a single family.[9]In contrast to their echocardiographic findings our Patient had LV mid-cavity without apical aneurysm. The MO-HCM is responsible for syncope and shows a high probability of sudden death, suggested that an aggressive prevention for sudden death should be considered in patients with MO-HCM.[10] Intracavitary LV obstruction is an important determinant of clinical outcome in hypertrophic cardiomyopathy (HCM). In a minority of patients the obstruction is at the level of the papillary muscles. Mid-cavity obstructive HCM may be associated with a distal LV aneurysm and a worse prognosis.[11] Mid-ventricular obstructive hypertrophic cardiomyopathy (MVOHC) is a rare type of cardiomyopathy. The diagnosis is

based on the presence of pressure gradient between apical and basal chamber of the ventriculum on the hemodynamic assessment.[1] Tengiz et al quoted a case of MVOHC associated with systolic anterior motion of the mitral valve and obstruction at both the mid-ventricular and outflow levels treated with successful percutaneous treatment with septal ablation.[1]

CONCLUSIONS

The presence of mid-cavity obstruction in hypertrophic cardiomyopathy may be associated with ventricular arrhythmias and systemic embolism. Echocardiography plays an important role in diagnosis, assessing hemodynamic and risk stratification, based mainly on the assessment of maximal wall thickness and the presence or absence of LVOT or mid-cavity obstruction. HCM is diagnosed on the basis of LVH for which there is no or insufficient explanation. Echocardiography is an invaluable tool in the diagnosis and follow-up of patients with HCM. The diagnosis of HCM using is challenging, and diagnosis can only be 100% reliable when a gene mutation is identified. In cases of unexplained LVH, accurate diagnosis of the underlying cause is inherently difficult and conditions such as HCM and hypertension may coexist. There is no single echocardiographic parameter which is ideal, and history and clinical examination play a vital role. We suggest a meticulous echocardiographic assessment by experienced echocardiographer to diagnose hypertrophic cardiomyopathy.

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