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A Rare form of Hypertrophic Cardiomyopathy with Mid-cavity Obstruction

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Abstract Hypertrophic cardiomyopathy (HCM) is a genetic disorder of cardiac myocytes with variable penetrance and heterogenous phenotypic expression leading to different morphologies of left ventricular (LV) hypertrophy which are usually associated with dynamic left ventricular outflow tract (LVOT) obstruction. Up to 2 percent of cases of HCM have a morphology of mid-cavity or mid-ventricular obstruction in which LV cavity is divided into two cavities by apposition of the hypertrophied septum and free wall, and is many times associated with pressure gradients between both cavities usually in the range of 50-70 mmHg but as high as 110 mmHg has been reported (3). We present a case of this rare form of mid-cavity obstruction with an exceptionally high intraventricular pressure gradient of 154 mmHg and apical aneurysm formation.

Keywords: *Hypertrophic cardiomyopathy, left ventricular outflow tract obstruction, intraventricular pressure gradient*

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1. Introduction

Hypertrophic cardiomyopathy (HCM) is a genetic disorder of cardiac myocytes with variable penetrance and heterogenous phenotypic expression leading to different morphologies of left ventricular (LV) hypertrophy [1]. Up to 2 percent of cases of HCM have a morphology of mid-cavity or mid-ventricular obstruction in which LV cavity is divided into two cavities by apposition of the hypertrophied septum and free wall, and is many times associated with pressure gradients between both cavities usually in the range of 50-70 mmHg but as high as 110 mmHg has been reported [3]. We present a case of this rare form of mid-cavity obstruction with an exceptionally high intraventricular pressure gradient of 154 mmHg and apical aneurysm formation.

2. Case Presentation

A 59 years old man with a history of cardiomyopathy and automatic implantable cardiac defibrillator (AICD) placement 22 years prior, presented to the emergency room (ER) with a complaint of intermittent episodes of anginal chest pains at rest. For the past several months, he had not been taking any of his home medications. Patient also reported his AICD going off at least 3 times a few weeks ago but he did not seek any medical attention. In the emergency department, the patient's heart rate was 75 beats per minute and blood pressure was 124/79 mmHg. His EKG showed sinus rhythm with occasional premature ventricular complexes, septal Q waves and marked T wave inversions in the majority of limb and chest leads (Figure 1). His first Troponin level was elevated at 0.08 ng/ml (normal < 0.05 ng/ml). Serum electrolytes, complete blood count and lipid panel were all within normal limits. Device interrogation showed a total of 19 episodes of AV dissociation for which 1 shock was delivered, and later anti tachyarrhythmia pacing (ATP) was successful in converting rhythm into sinus rhythm. A 2D echocardiogram showed LV ejection fraction of 80%, and severe LV hypertrophy in mid-cavity area, consistent with hypertrophic cardiomyopathy with mid-cavity obstruction (Figure 2, Figure 3 & Figure 4). A cardiac catheterization was done which showed non-occlusive coronary artery disease. Left ventricular pressure in the main proximal chamber was 126/70 mmHg and 280/31 mmHg in the apical portion of the left ventricle with a gradient of 154 mmHg across the mid cavity obstruction (Figure 5, Figure 6 & Figure 7). Left ventricular end diastolic pressure in the main basal portion of the left ventricle was 13 mmHg while it was 45 mmHg in the apical portion of the left ventricle. It appeared that when the basal part of the left ventricle was contracting, the blood was not only going towards the aorta, but it was also going towards the apical chamber with varying pressures through a narrow tunneled connection between basal and apical chambers, making apical chamber to

appear ballooned during systole. Patient was placed on a low dose beta blocker which he tolerated well and remained symptom free. He was discharged in stable condition with a follow up appointment in a tertiary care hospital for cardiac surgery evaluation for left ventricular myomectomy.

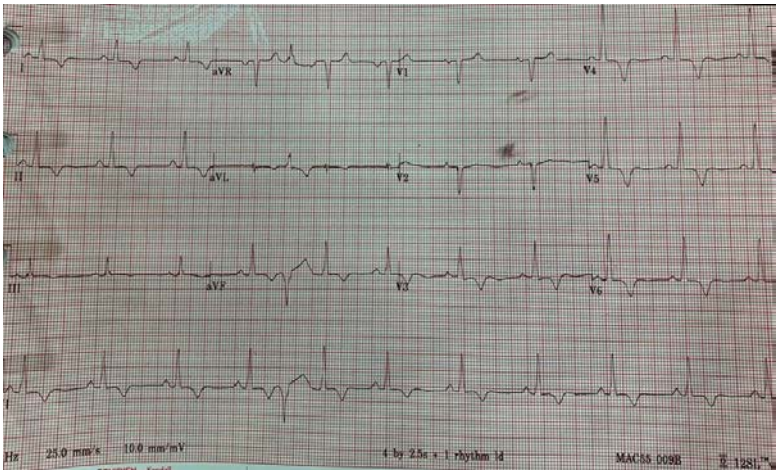

Figure 1. EKG showing deep T wave inversions predominantly in several chest and limb leads

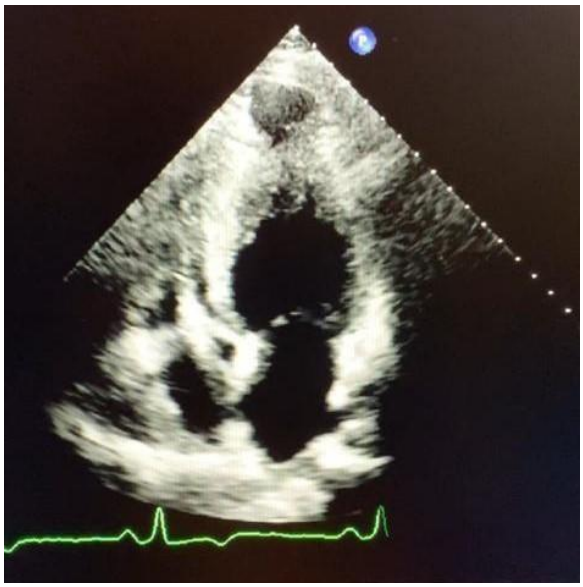

Figure 2. 2 D echocardiogram. Apical 4 view chamber showing hypertrophied mid LV with mid-cavity narrowing

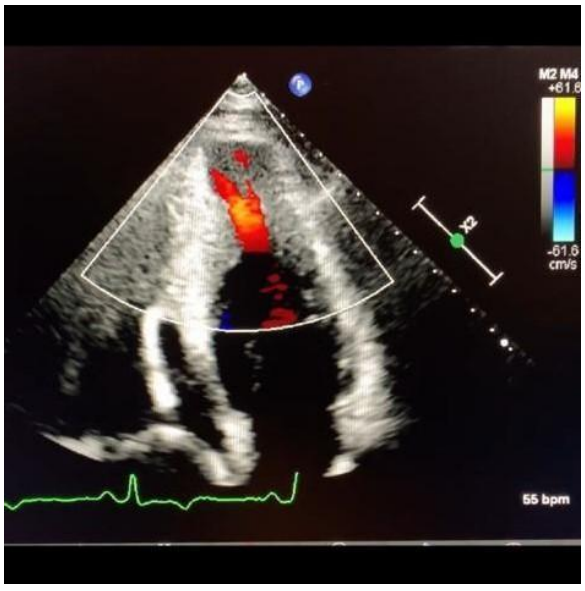

Figure 3. 2 D Echocardiogram with color doppler showing a jet of blood flow from proximal portion of LV to apical of LV

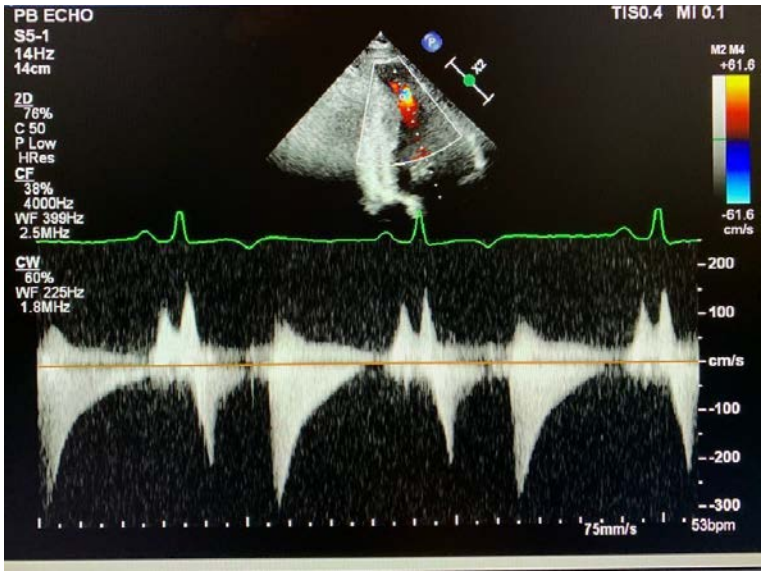

Figure 4. 2 D Echocardiogram with M mode showing spike like projections consistent with intraventricular pressure gradient

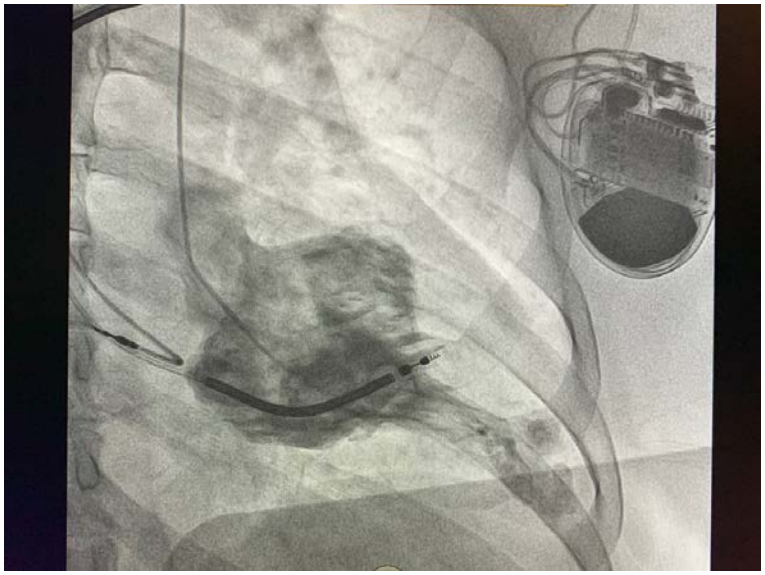

Figure 5. Cardiac catheterization with left ventriculogram showing basal and apical chambers of LV communicating through narrow tunnel like mid cavity of left ventricle

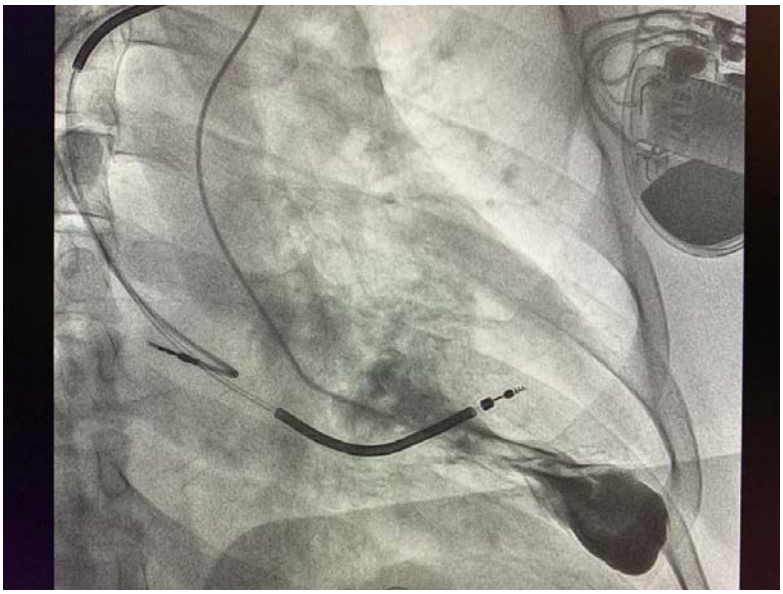

Figure 6. Cardiac catheterization with left ventriculogram showing contrast in apical chamber of LV, distal to mid-cavity obstruction

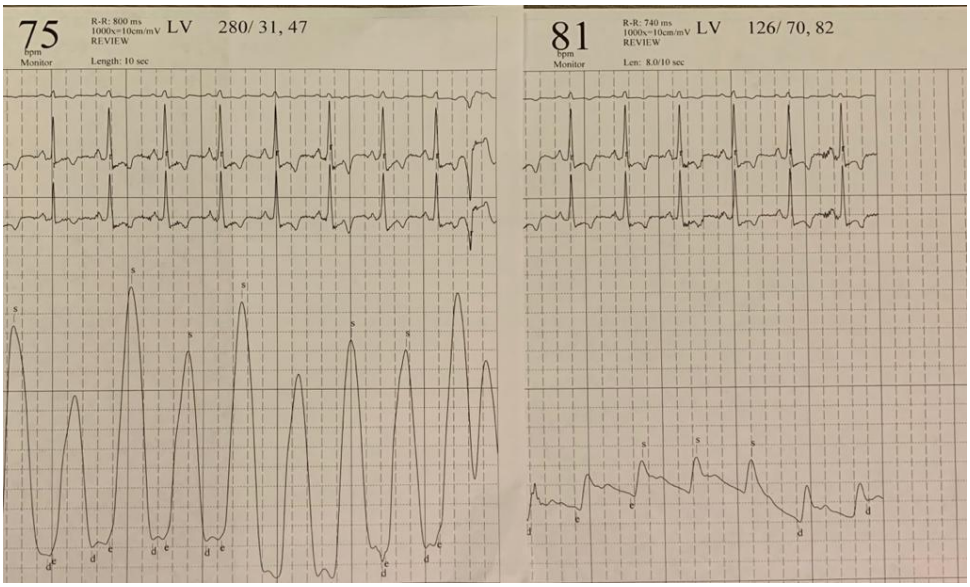

Figure 7. Cardiac catheterization with left ventricular pressure waveforms in apical chamber of LV of Left side showing pressure of 281/31 mmHg and proximal portion of LV showing BP of 126/70 mmHg; intraventricular pressure gradient of 154 mmHg

3. Discussion

Hypertrophic cardiomyopathy (HCM) is an autosomal dominant disorder occurring in approximately 1 out of 500 adults in the general population [2]. HCM is caused by mutations of cardiac sarcomere genes resulting in disorganized architecture and hypertrophy of cardiac myocytes with interstitial and replacement fibrosis which leads to different phenotypic expressions of cardiac muscle thickness regarding location, severity and extent of LVH [1,8]. LVH in HCM is typically asymmetrical in distribution and causes LVOT obstruction in the majority of patients especially during exertion, if not present at rest [4]. HCM is usually asymptomatic but can present with symptoms of heart failure, obstruction, thromboembolism or arrhythmias e.g., dyspnea, orthopnea, chest pain, syncope, stroke or even sudden cardiac death. LVOT obstruction in HCM is dynamic and usually arises from asymmetrical septal hypertrophy (ASH) along with the systolic anterior motion (SAM) of anterior mitral valve leaflet leading to bifid carotid upstroke with brisk rise and then decreased pressure in aorta as compared to LV cavity, an ejection systolic murmur and sometimes concomitant mitral regurgitation with eccentric jet. Sometimes a third or fourth heart sound might also be present due to increased left atrial pressures. Smaller LV cavity size tends to increase LVOT obstruction and systolic murmur which can be elicited by decreasing preload by Valsalva maneuver, standing from sitting position or nitroglycerine. On the other hand, systolic murmur tends to decrease in intensity with increasing preload by leg elevation or sitting from standing position; or by increasing afterload by hand grip. Electrocardiographic findings of HCM include non-specific repolarization abnormalities, prominent but narrow Q waves in inferolateral leads due to initial depolarization of thick myopathic septal tissue, and deeply inverted T waves in precordial leads associated with apical variants of HCM. An otherwise unexplained wall thickness of more than 15 mm anywhere in the left ventricular wall is diagnostic of HCM. Common locations of asymmetric ventricular hypertrophy in HCM are basal anterior septum, anterior free wall and posterior septum. In about one percent of patients, apical or concentric distribution of LVH is found. Other morphological variants are rare and include mid cavity hypertrophy, biventricular hypertrophy and LV wall thinning with biapical enlargement. Many patients with HCM who do not show LVOT obstruction at rest, express it during exercise and that is why exercise testing is recommended in patients with HCM who do not have LVOT obstruction at rest [5]. Cardiac magnetic resonance (CMR) imaging provides more reliable details than 2D echocardiography regarding LV wall thickness, morphology of LVH, and anatomy of mitral valve and papillary muscles which is useful to determine which septal reduction surgery better suits the patient whether alcohol septal ablation or surgical myomectomy [6]. CMR can be used to assess late gadolinium enhancement (LGE) which correlates with the degree of myocardial fibrosis and in some observational studies LGE is found to correlate with the risk of arrhythmias [7]. Cardiac catheterization is used in selected patients for purposes of evaluation for concomitant coronary artery disease, accurately measuring LVOT pressure gradients, pre-op evaluation for cardiac transplant or to perform endocardial biopsy to rule out other causes of cardiomyopathy like lysosomal storage diseases, Danon disease and amyloidosis. Routine genetic testing is not required for diagnosis of HCM however it is reserved to screen first degree relatives of probands with definite disease-causing sarcomere gene mutations; and for patients with HCM suspected to be related to other genetic diseases [9]. Medical management is recommended for symptomatic patients with HCM and primarily includes AV nodal blocking medications, beta-blockers being the first line of treatment [10]; nondihydropyridine calcium channel blockers like verapamil being the second choice which should be avoided in patients with low LV ejection fraction. Disopyramide is class 1A antiarrhythmic drug which is known to effectively decrease LVOT gradients in HCM but due to its proarrhythmogenic and QT-prolonging side effects, it is reserved for second line

therapy in combination with either beta blockers or verapamil for patients with LVOT obstruction not improving with monotherapy.

HCM with mid cavity or mid ventricular obstruction is a unique morphological type of HCM and occurs in up to 2 percent of all HCM cases in which apposition of septum and lateral wall creates 2 distinct hourglass shaped apical and basal cavities in left ventricle with an intraventricular gradient in between. Cardiac catheterization with left ventriculogram can be done to accurately measure pressures in basal and apical ventricular cavities. The mechanism of LVOT obstruction in these cases is usually not secondary to systolic anterior motion of mitral valve but instead occurs due to approximation of free wall with hypertrophied papillary muscle or less occasionally secondary to anomalous insertion of papillary muscle into mitral valve. High pressures in the apical part of LV cavity can sometimes lead to formation of apical thinning and aneurysm which can act as a nidus or ventricular arrhythmias and thromboembolism. Therefore, patients with midventricular obstruction should be considered for primary prophylactic ICD as well as prophylactic anticoagulation along with beta blockers which are the mainstay of initial treatment for symptomatic patients. Patients with persistent symptoms and gradients require surgical myomectomy.

4. Conclusion

HCM is an autosomal dominant disorder with variable phenotypic expressions leading to different morphologies of left ventricular (LV) hypertrophy. Mid cavity HCM is a unique morphological type of HCM which can lead to dynamic mid left ventricular obstruction. Our case describes an exceptionally high intraventricular pressure gradient of 154 mmHg and apical aneurysm formation in a patient with mid cavity HCM.

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