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Case Report

Hypertrophic Cardiomyopathy Unmasked: The Hidden Danger of Left Ventricular Apical Aneurysm through Coronary Embolization

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Abstract: Hypertrophic Cardiomyopathy (HCM) is a genetic heart disease characterized by left ventricular (LV) hypertrophy, with outcomes ranging from asymptomatic to sudden cardiac death (SCD). Accurate diagnosis and risk stratification are essential. This case report discusses a 52-year-old female with family history of sudden deaths, presenting with acute coronary syndrome symptoms. Investigations revealed atrial fibrillation, ST-elevation, and a thrombus in the left anterior descending artery (LAD). Echocardiography and cardiac CT confirmed a left ventricular apical aneurysm, multiple apical thrombus, and hypertrophic septal wall with systolic LV dysfunction. The report highlights the risk of thromboembolic events and SCD in patients with LV apical aneurysms. Meta-analyses and guidelines emphasize the importance of integrating risk markers like LV apical aneurysms and late gadolinium enhancement into clinical assessments. Given the patient's family history, aneurysm, and LV dysfunction, primary SCD prevention with an implantable cardioverter-defibrillator (ICD) was deemed suitable. HCM patients with LV apical aneurysms represent a high-risk phenotype requiring comprehensive management, including SCD and stroke prevention.

Keywords: Hypertrophic Cardiomyopathy, Sudden Cardiac Death, Acute Coronary Syndrome, Apical Aneurysm.

INTRODUCTION

Hypertrophic cardiomyopathy (HCM) is a common genetic heart disease with a prevalence of 1 in 500 individuals [1], characterized by left ventricular hypertrophy and associated with increased cardiovascular morbidity and mortality; it's been related to more than 1400 mutations mostly sarcomeric, resulting in a large spectrum of phenotype with different clinical outcome; ranging from asymptomatic patient to heart failure, thromboembolic events and even sudden cardiac death (SCD). Hence the importance of accurate risk stratification and recognition of HCM phenotypes that are clinically significant.

The diagnosis of HCM in adults is based on the presence of left ventricular (LV) wall thickness (WT) of 15 mm or greater on one or more LV myocardial segments and not explained solely by abnormal cardiac loading conditions; a 13 mm threshold is used for familial screening in first degree relative. Apical aneurysm defined as a discrete thin-walled dyskinetic or akinetic segment of the most distal portion of the LV chamber; independent of size, studies have shown that this subset experienced increased risk of adverse outcomes [2].

CASE REPORT

A 52 old female with no significant personal history and a family history of sudden deaths (mother; sister and brother); presented in a clinical scenario of acute coronary syndrome.

The patient complained of an acute prolonged retrosternal pain irradiating to the left arm; 12 leads EKG at H10 of pain onset showed atrial fibrillation rhythm and ST elevation on V1 to V3 with inverted T waves in apico-latéral and inferior leads) figure 1; cardiac troponins I was at 8.6 ng/ml (*400 normal values).

An emergent coronary angiogram was performed that showed a large thrombus on the descending coronary artery (LAD) with a TIMI 3 flow, and smooth coronary arteries without disease, and thromboembolic origin was strongly suggested and the patient was set on anticoagulant therapy.

2D echocardiography was performed showing thickened septal wall of 15 mm, an apical insertion of mitral pillar without mitral systolic motion, and bi-atrial dilation with AP diameter of left atria over 50 mm.

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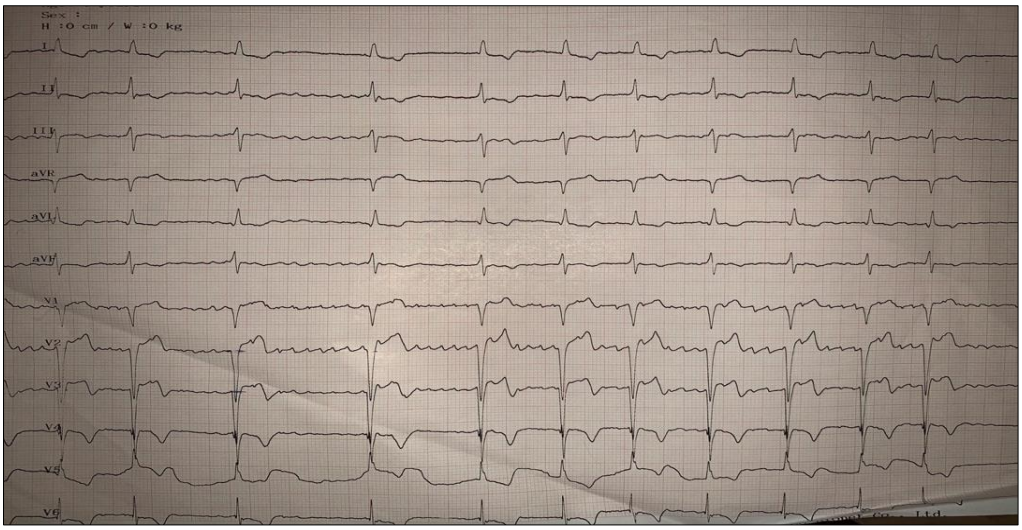

Fig. 1: 12 leads EKG

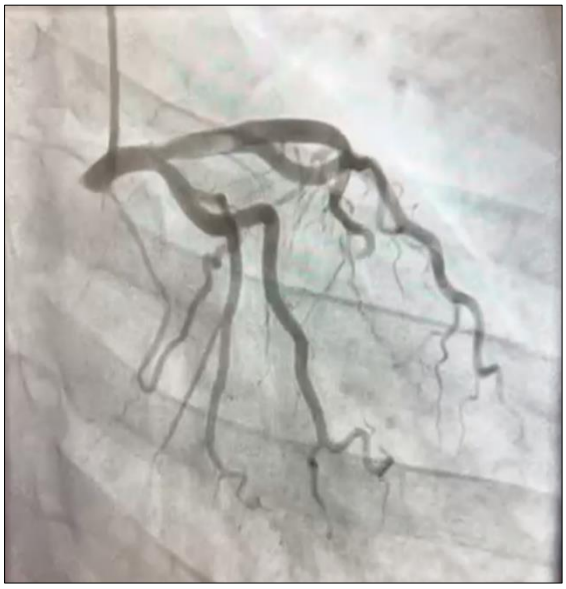

Fig. 2: Caudal view of croronary angiogram showing a large thrombus on the proximal LAD

A proper evaluation of the apex has objectified a thinned akinetic apical wall with multiple thrombus; we noted the presence of wall motion abnormalities and LV systolic dysfunction with a 40 % ejection fraction. (figure3), the doppler investigation showed no LV outflow tract acceleration even with Valsalva maneuver.

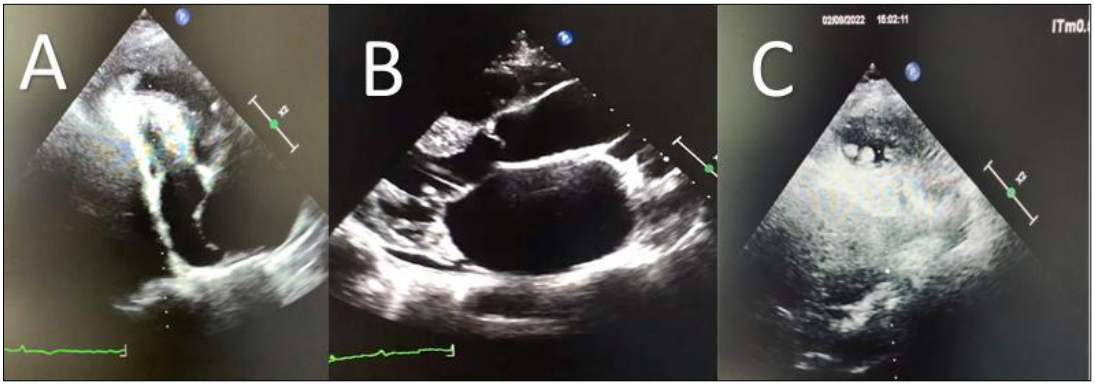

Fig. 3: 2D echocardiography views; A apical four chamber view showing apical aneurysme with thrombus and an hypertrophic septum, bi-atrial dilataion; B: asymetric anteroseptal hypertrophy and abnormalities of insertion of papillary muscle on mitral valve; C: apical multiples thrombis

Transesophageal echocardiography was performed and no clot was found in the atrial appendage.

Holter monitor showed stage 2 ventricular hyperexcitability with no major events.

A cardiac CT scan was performed to visualize the right coronary artery that was large with no evidence of stenoses and the disappearance of LAD and aneurysm thrombus.

An MRI was later performed that confirmed the diagnosis and showed no extensive LGE.

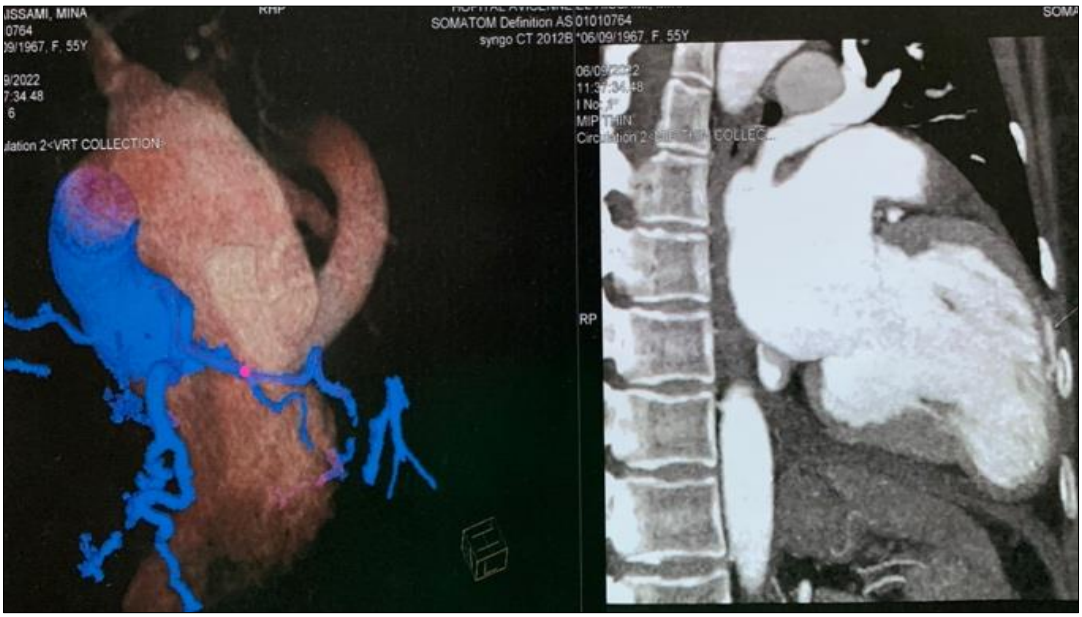

Fig. 4: Cardiac CT scan showing on the left coronary arteries and on the right large anerysm

DISCUSSION

Morphological expression of HCM is particularly heterogeneous because of the wide variety of sarcomere gene mutations associated with this disease.

In some cases, the diagnosis of HCM can be challenging especially when anterolateral or apical wall are involved or the occurrence of wall thinning along with depressed LV ejection fraction in end stage form of HCM. Also in adult patients carrying causative gene mutation associated with HCM may present with lesser degrees of hypertrophy (13-14mm) in such cases the diagnosis requires integration of family history, clinical and multimodality imaging [3].

Patients with left ventricular apical aneurysm are an underestimated subgroup within the broad spectrum of HCM. The prevalence of left ventricular apical aneurysm in the general HCM population is considered low (2%), although this may be an underestimate because 2D echocardiography may be unreliable in detecting small aneurysms (compared to cardiac magnetic resonance MRI) [4].

This case shows a late revelation of the disease by a thromboembolic event , the scenario of coronary embolization was the most likely because of absence of cardiovascular risk factors and the large thrombus with normal and large coronary arteries, a 2D echocardiography was performed to identify a potential cause of embolism and the diagnosis of HCM was suspected by the asymetrical hypertrophy aspect of the anteroseptal wall mesured at 15 mm with abnormality of mitral valve and also family history of sudden cardiac death , the apical anuryisme was carefully looked for and the diagnosis was successfully done by echocardiography , later MRI was done not only for confirmation but to evaluate the degree of myocardial fibrosis and to measure the extent of ate gadolinium enhancement LGE.

LV apical aneurysm was associated in some studies with a significantly increased risk of thromboembolic events and/or apical thrombus with 6-fold for thromboembolic events [5]. A possible explanation of this finding is that the dyskinetic/akinetic aneurysm can provide an ideal structural nidus for thrombus formation.

Whether LV apical aneurysm could be a robust independent risk factor of SCD in patients with HCM has been debated for many years.

On a recent meta-analysis, the presence of a LV apical aneurysm was found to be a significant prognostic factor, conveying more than 4-fold higher risk of potentially lethal ventricular tachyarrhythmia events compared to HCM patients without apical aneurysm (fig4). These data are also consistent with the largest study in this meta-analysis in which the appropriate ICD

therapy rate for primary prevention was 4.0%/year, nearly 5-fold greater than the SCD event rate in HCM patients without LV aneurysms and similar to the rate observed in other high-risk HCM cohorts [5].

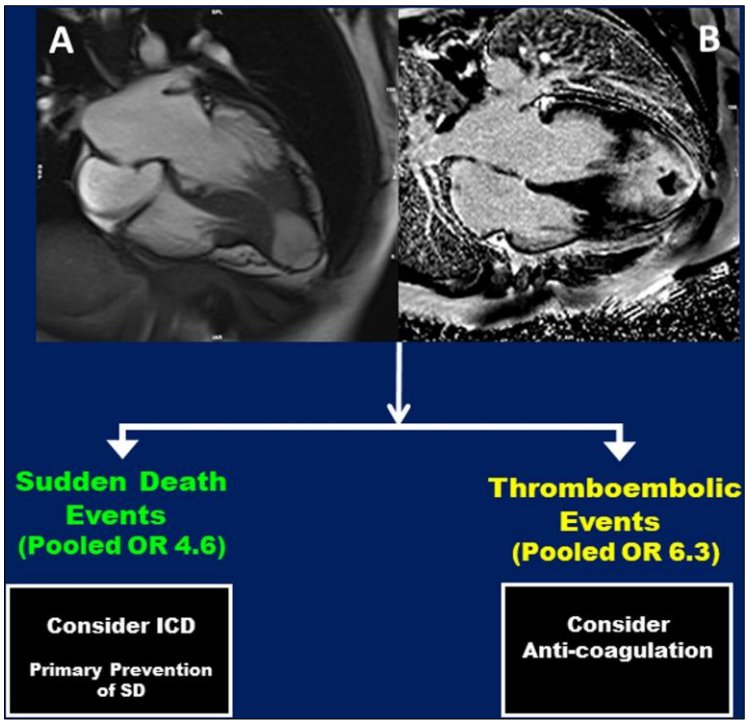

Fig.5: Clinical implications of the meta-analysis. A. End-diastolic still frame of a 4-chamber cine showing mid-ventricular hypertrophy with a large apical aneurysm. B. LGE 4ch-view showing mid-ventricular hypertrophy with patchy fibrosis and a large apical aneurysm with transmural fibrosis. Note the apical thrombus at the aneurysmal apex (arrow). ICD: Implantable cardioverter defibrillator; OR: Odds ratio; SD: Sudden death

On the other hand, the enhanced American College of Cardiology/ American Heart Association (ACC/AHA) guidelines for the diagnosis and treatment of HCM have incorporated novel high-risk cardiac magnetic resonance markers such as LV apical aneurysms, as well as extensive late gadolinium enhancement, and systolic dysfunction (ejection fraction <50%) [2].

Indeed, the high sensitivity (95% with intention to treat) associated with the ACC/AHA SCD risk marker method is in large part due to the fact that these 3 novel risk markers now account for a significant proportion (~25%) of appropriate ICD therapies among HCM patients [6].

On ESC guideline, task Force recommends that individualized ICD decisions should be based using well-established risk factors and not solely on the presence of an LV apical aneurysm.

ICD implantation is not free of risk and the patients should be wisely selected in our case the patient had 3 major risk factors an apical anurysm and LV systolic dysfunction plus family history of SCD therefor a primary prevention of SCD was reasonable even in the absence of documented ventricular arrythmia.

CONCLUSION

HCM patients with LV apical aneurysm represent a high-risk phenotype associated with substantially increased risk for adverse disease related events, including SCD events and thromboembolism. This feature of HCM impacts management, including SCD prevention with ICD and stroke prophylaxis with anticoagulation.

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