Cardiomyopathy Conundrum, A case of Apical Hypertrophic Cardiomyopathy with an Apical Aneurysm

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Introduction:

Hypertrophic Cardiomyopathy (HCM) is a genetically determined disorder with a prevalence of 0.2%. HCM can present with diverse morphologies and depending on the morphology can present with cardiac abnormalities such as Diastolic Dysfunction, Left Ventricular Outflow Tract Obstruction (LVOT), Myocardial Ischemia, Arrhythmias or Mitral Regurgitation. They often present with clinical features of Chest Pain, Syncope or Near Syncope, and Dyspnea. An increasing number of patients with HCM are being identified with having Left Ventricular Apical Aneurysms which is believed to perpetuate the risk of HCM related mortalities.

Case:

We hereby report the case of a 47-year-old Male presented for follow up with complains of chronic stable angina which was relieved with Nitroglycerin. He was previously diagnosed with a right coronary artery (RCA) occlusion on cardiac catheterization with the presence of collaterals. EKG was performed which showed normal sinus rhythm (NSR), Heart rate 78, normal axis and intervals with deep T-wave inversions in the antero-septal lateral leads. Patient had also undergone a cardiac stress test which showed inferior wall ischemia which was indicative of the RCA occlusion. Upon further investigation Transthoracic Echocardiogram was performed without image enhancing agent which showed Left ventricular apical hypertrophy. However with the help of image enhancing patient was also found to have left ventricular apical aneurysm. Patient was then sent for a cardiac MRI to rule out presence of a thrombus which was ruled out and diagnosis was confirmed. Patient also underwent Ventriculogram in the past which illustrated gradient of 80mmHg between apex and mid cavity at rest which however increased to 200mmHg post PVC. However clinical significance of this is yet limited. Patient did not have family history of sudden cardiac deaths, did not show signs of non-sustained Vent Tach/Vent Fib hence it was decided to avoid placement of AICD and to monitor the patient closely with yearly Holter monitoring to detect presence of life threatening arrhythmias.

Discussion:

HCM can present with varying phenotypes. In this case we report a case of an Apical Variant of HCM with the presence of an Apical Aneurysm. Presence of an apical aneurysm increases the risk of cardiac adverse events. The junction of the aneurysm with the normal myocardium works as an arrhythmogenic substrate for the generation of malignant ventricular arrhythmias which could lead to SCD. Apical aneurysm also poses an increased risk of thrombus formation which could lead to thromboembolic events. Studies have shown HCM with LV apical aneurysm having an increased incidence adverse cardiac events such as heart failure, life threatening
V.Tach/Fib, thromboembolic events. Identification of such phenotypes is essential to help in risk stratification. Study has shown benefit of treatment interventions in such phenotypes such as AICD placement or Heart Transplant.

Conclusion:

The purpose of this case report is to highlight the importance of identifying phenotypes of HCM with apical aneurysms, their appropriate risk stratification in order to prevent adverse cardiac events with appropriate treatment interventions.