YOU ONLY DIE TWICE: A CASE OF SUDDEN CARDIAC ARREST IN A PATIENT WITH SEVERE APICAL HYPERTROPHIC CARDIOMYOPATHY AND A CORONARY ANOMALY

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Introduction: Apical hypertrophic cardiomyopathy (AHCM) is an uncommon variant of hypertrophic cardiomyopathy which may carry less risk of sudden cardiac arrest (SCA) than others phenotypes. A 66 y/o M who had previously suffered resuscitated cardiac arrest attributed to severe AHCM was found years later to have an anomalous origin of the right coronary artery from the left coronary sinus of valsalva, casting doubt on the presumed cause of his arrest.

Methods: A 66 year old man presented with worsening angina and exertional dyspnea. He had a history of AHCM, SCA due to ventricular fibrillation, and ICD placement. He reported progressive dyspnea on exertion. His EKG showed atrial fibrillation with right ventricular pacing. The patient underwent exercise stress transthoracic echocardiography exercise for 5 minutes (6 METS), stopping due to dyspnea. The peak intracavitary gradient was 55 mm Hg. Coronary angiography was performed, which revealed an anomalous right coronary artery (aRCA) originating from the left coronary cusp, coursing anteriorly between the pulmonary artery and the ascending aorta. He turned down surgical evaluation for bypass of the anomalous coronary artery.

Result: AHCM and anomalous coronary anatomy are both rare causes of SCA. We present a case of SCA in a man with severe AHCM later found to have a coronary anomaly. Anomalous coronaries have a reported incidence of 0.6-1.3% in autopsy series; anomalous origin of the RCA from the left coronary cusp between the great vessels is 0.026%- 0.25%. Coronary anomalies may account for less than 1% of SCA, though the true risk of SCA in a given coronary anomaly is not known. Apical HCM is generally felt to have a low risk of malignant arrhythmia, though numerous case reports and some case studies suggest an increased mortality risk.

Conclusion: Apical HCM and anomalous right coronary artery (aRCA) are both rare causes of SCA. We highlight a case of SCD where both phenotypes existed simultaneously in a patient who suffered SCA. This case highlights the need for comprehensive testing in patients with SCA when a rare potential cause is initially found.