Case Reports

Multimodal Imaging after Sudden Cardiac Arrest in an 18-Year-Old Athlete

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We report the case of a previously healthy 18-year-old male athlete who twice presented with sudden cardiac arrest. Our use of electrocardiography, echocardiography, cardiac magnetic resonance, coronary angiography, coronary computed tomographic angiography, and nuclear stress testing enabled the diagnoses of apical hypertrophic cardiomyopathy and anomalous origin of the right coronary artery. We discuss the patient's treatment and note the useful role of multiple cardiovascular imaging methods in cases of sudden cardiac arrest. ***(Tex Heart Inst J 2015;42(6):548-51)***

Key words: *Arrhythmias, cardiac/etiology/therapy; cardiomyopathy, hypertrophic/complications/diagnosis/pathology; coronary vessel anomalies/diagnosis/surgery; death, sudden, cardiac/etiology/prevention & control; defibrillators, implantable; diagnostic imaging/utilization; gadolinium/diagnostic use; risk factors; sports medicine/standards; treatment outcome*

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Sudden cardiac death (SCD) in a young athlete is considered to be a paradoxical tragedy, because athletes are presumed to be the healthiest individuals in the community. In the current period of technological advances, the collaborative use of diverse imaging tools potentially improves our understanding of sudden cardiac arrest (SCA) and SCD. Hypertrophic cardiomyopathy (HCM) and coronary anomalies are the chief causes of SCD in young athletes. We present the case of a patient with both clinical entities who presented with SCA, and we discuss the value of multiple imaging methods in revealing the pathophysiology and designing the treatment plan.

Case Report

In April 2013, a presumably healthy 18-year-old black man, a varsity athlete, collapsed while playing basketball. Emergency medical technicians arrived within 10 minutes and found him to be in ventricular fibrillation (VF). Spontaneous circulation returned after defibrillation, and the patient was intubated in the field. His social and family histories yielded nothing of note; however, it was learned that he had never undergone a complete, elective electrocardiogram (ECG). At the emergency department, initial physical examination revealed a young man of height 6 ft 1 in, weight 207 lb, and a body mass index of 27. Cardiac auscultation revealed no murmurs. Urine toxicology results were notable for tetrahydrocannabinol. The patient's electrolyte levels and serum toxin screening results were normal. His cardiac troponin level peaked at 0.32 μg/L.

Thirty minutes after the SCA, an ECG showed sinus rhythm, left ventricular (LV) hypertrophy with deep T-wave inversions in the precordial leads, and ST-segment elevation in the anterior leads (Fig. 1). One hour after the SCA, a bedside echocardiogram revealed prominent thickening of the LV apex and a preserved LV ejection fraction without wall-motion abnormalities (Fig. 2). Within 2 hours of the SCA, the patient was sent to the cardiac catheterization laboratory for investigation of obstructive coronary artery disease. A ventriculogram showed the classic "ace of spades" configuration seen in apical hypertrophy (Fig. 3). No coronary atherosclerotic lesions were identified; however, the right coronary artery (RCA) was noted to originate from the left coronary cusp.

These findings were confirmed on a coronary computed tomographic angiogram (CTA) on day 4 of the patient's hospitalization. The RCA arose from the left coronary sinus of Valsalva with a severely narrowed ostium, and it took a "malignant" course between the main pulmonary artery and the aorta (Fig. 4). On day 5, cardiac magnetic resonance images revealed patchy, near-circumferential, apical late gadolinium enhancement (LGE), caused by fibrosis that was consistent with apical-variant HCM (Fig. 5).

The patient recovered uneventfully and had no neurologic deficit. On day 7, he underwent a nuclear exercise stress test, which revealed a reversible defect of moderate size and intensity in the inferolateral and lateral wall (Fig. 6). The patient exercised on the treadmill for 10.3 min (11.6 metabolic equivalents, functional class I). Appropriate blood pressure response was noted, and 86% of age-predicted heart rate was achieved without symptoms. The ST-segment response during exercise was nondiagnostic.

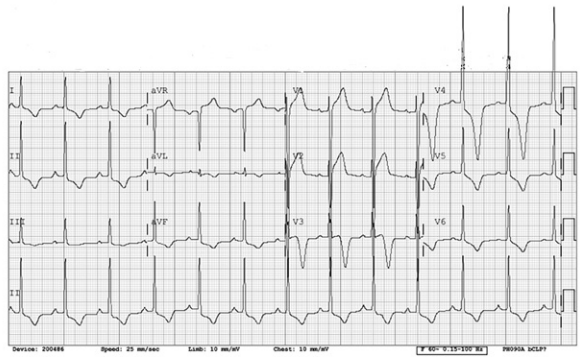

Fig. 1 *Electrocardiogram shows sinus rhythm with diffuse ST-T-wave changes (ST-segment elevation in leads V_1 and V_2 and giant negative T waves in leads V_3 through V_6), diagnostic of apical hypertrophy.*

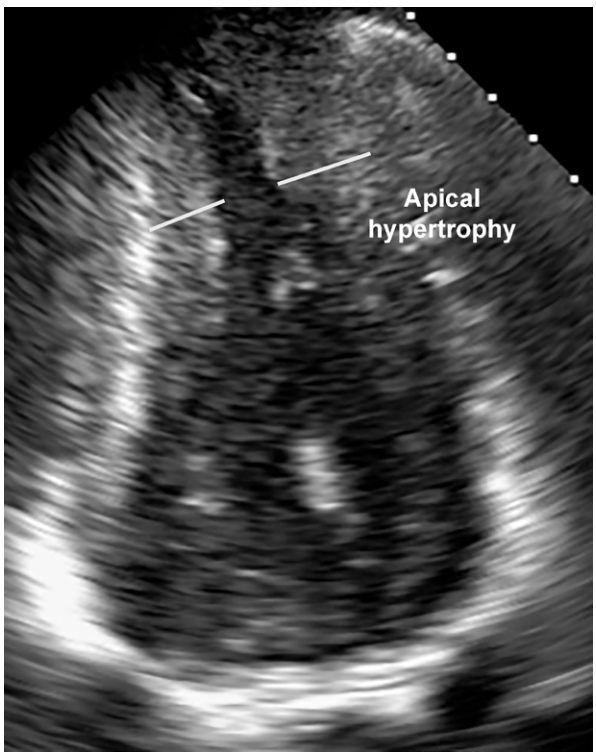

Fig. 2 *Echocardiogram shows a thickened, echo-bright apex with complete systolic collapse of the apical left ventricular cavity. The lines indicate the substantial thickness of the apical ventricular wall, consistent with apical hypertrophy.*

Before the patient's discharge from the hospital after a 9-day stay, an implantable cardioverter-defibrillator (ICD) was placed for secondary prevention. Five months after the SCA, he successfully underwent a surgical unroofing (marsupialization) of the anomalous RCA, was placed on prophylactic β-blocker therapy, and was instructed not to resume competitive sports.

However, 6 months after the surgical procedure, the patient decided to join an informal basketball game with his friends. He sustained another cardiac arrest, and emergency medical technicians found him lying on the basketball court after an ICD discharge. The ICD memory revealed VF. The patient was offered imaging to confirm the adequacy of the RCA unroofing; however, he declined further cardiac testing. As of September 2015, he had experienced no more ICD discharges, was consulting regularly with his cardiologist, and had abandoned sports activities.

Discussion

Sudden cardiac death in young athletes is rare yet tragic. It is most prevalent in athletes younger than 25 years of age. The chief causes of SCD are HCM (in 36% of cases) and coronary anomalies (in 17%).[1] Apical HCM, also called Yamaguchi syndrome, is a variant of HCM and is classically described in the Asian population. The characteristic ECG finding is giant negative T waves (the Yamaguchi sign) in the precordial leads.[2] Echocardiography is the standard tool for the diagnosis of apical hypertrophy, although the "ace of spades" appearance of the ventricular cavity on a ventriculogram is considered to be pathognomonic.[3] Despite evidence that the use of ECG as a screening tool for sports has substantially reduced SCD occurrences in Europe,[4] this issue is contro-

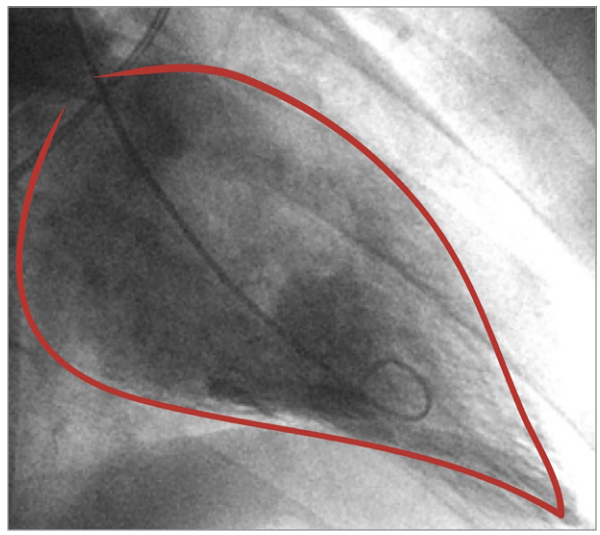

Fig. 3 *Ventriculogram at end-diastole shows the classic "ace of spades" configuration associated with apical hypertrophy.*

versial in the United States. Opponents of routine ECG screening often cite the rate of false-positive results and the undue cost without evidence of substantial benefit from such universal screening. The American Heart Association recommends against routine ECG screening.[5]

The mechanism of SCA in HCM is not fully known. In theory, elevated end-diastolic pressure compresses the coronary circulation and triggers ischemia. It is suspected that, during stress, myocardial blood flow is blunted because of increased oxygen demand and elevated diastolic pressure. Underlying myocyte disarray is suspected to be another mechanism of the blunted myocardial flow. The episodic ischemia leads to further death of myocytes and their consequent replacement by fibrotic tissue.[6]

The advantageously high spatial and temporal resolution of cardiac magnetic resonance enables tissue characterization with use of nonionizing radiation. In our patient, the LGE was consistent with the classic fibrosis of HCM. This abnormal tissue might serve as a substrate for reentrant tachyarrhythmia that leads to SCD. Although the correlation of the extent of LGE and the

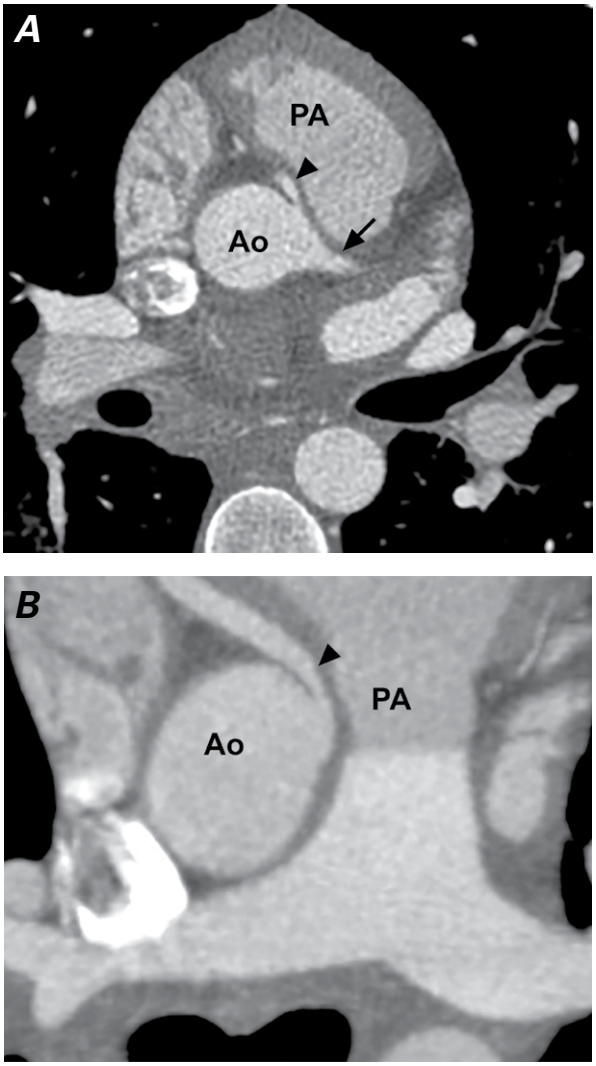

Fig. 4 *Computed tomographic coronary angiograms show* ***A****) the right coronary artery (arrowhead) and left main coronary artery (arrow) originating from the left sinus of Valsalva, and* ***B****) an abnormal origin of the right coronary artery (arrowhead) from the left sinus of Valsalva. The proximal vessel is acutely angled and narrowed from compression between the ascending aorta (Ao) and the main pulmonary artery (PA).*

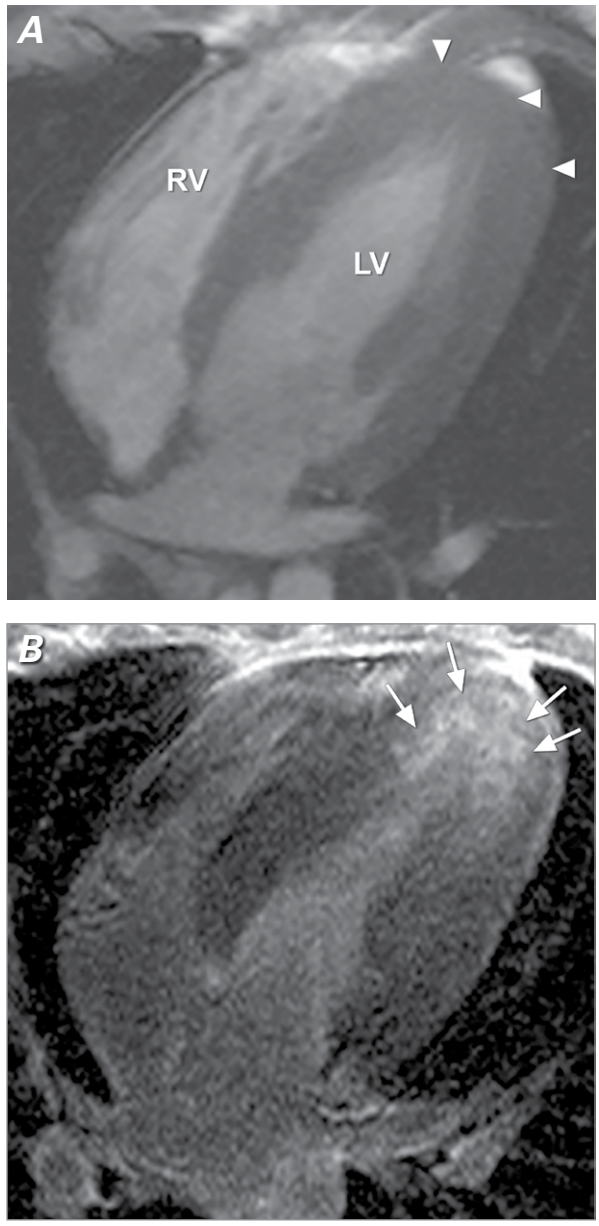

Fig. 5 *Cardiac magnetic resonance (4-chamber views).* ***A****) Bright-blood image shows abnormal apical thickening (arrowheads).* ***B****) Post-contrast image reveals bright areas of apical fibrosis (arrows) within regions of abnormal thickening. The normal myocardium has a dark signal. The findings confirm the diagnosis of apical hypertrophic cardiomyopathy.*

LV = left ventricle; RV = right ventricle

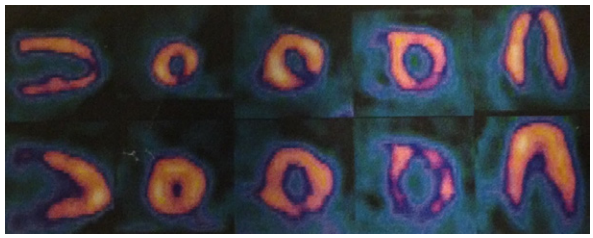

Fig. 6 *Nuclear perfusion images show inferior reversible defects during stress (upper row) and rest (lower row).*

risk of SCD is unproven, the absence of LGE is associated with a lower risk of SCD and vice-versa.[7]

Coronary anomalies account for approximately 17% of SCD cases in athletes.[1] Catheter-based coronary angiography has long been the means of identifying and characterizing coronary anomalies; however, this method is invasive and has risks and complications. In addition, anatomic complexity can preclude precise identification of the origin and course of the anomaly. Coronary CTA—with its fast 3-dimensional acquisition time, relatively high temporal resolution, and very high spatial resolution—has become the gold standard for fully characterizing coronary anomalies.[8]

The proposed mechanism of SCA and SCD in coronary anomalies is described as follows. The proximal vessel, as shown in the reconstructed image (Fig. 4B), is acutely angled and causes kinking of the coronary artery, particularly during elevated heart rates—possibly creating an acute occlusion that triggers ischemia. In addition, the acute takeoff and course of the artery give rise to a small slit-like coronary orifice that is subject to ischemia during exercise. It is suspected that catecholamine release and increased sympathetic tone during exercise contribute to a systolic expansion of the great vessels, which compresses the malignant artery and triggers ischemia.

Angelini and colleagues[9] have suggested that ischemia in RCAs originating from the left coronary sinus might be secondary to stenosis. They used intravascular ultrasound (IVUS) in their study of 63 adult patients and found strong evidence that the severity of stenosis correlated with the occurrence of ischemic symptoms.[9] The investigators further proposed that stenting would reduce not just the risk of SCD, but also angina, dyspnea, and syncope. Had we used IVUS, it might have enabled further investigation of our patient's SCA.

It was difficult to determine the chief cause of our patient's cardiac arrest. Initially, we thought that the myocyte disarray and fibrosis in HCM served as a substrate for the VF, which was probably triggered by ischemia upon compression of the RCA by both large vessels during exercise.

The nuclear stress test finding of the reversible defect in the inferior territory suggested a role of the RCA and contributed to our decision to perform the surgical unroofing. The indication for the ICD was secondary prevention of SCD.[10]

In view of our surgical correction of the patient's coronary anomaly, his 2nd SCA strongly suggests that the apical HCM was the principal mechanism underlying his cardiac events.

This case illustrates how multiple imaging methods can be used in the diagnosis, risk stratification, screening, and selection of appropriate therapy in complex cardiac conditions.

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