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ADVANCED

CASE REPORT: CLINICAL CASE

Multimodality Assessment of Anomalous Aortic Origin of the Right Coronary Artery Presenting With Cardiac Arrest

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ABSTRACT

Anomalous aortic origin of coronary artery (AAOCA) can range from benign anatomic variants to those presenting with sudden cardiac arrest. This unique case of right AAOCA demonstrates detailed anatomic findings from cardiac computed tomography and the effects of transient acute coronary ischemia by cardiac magnetic resonance. (**Level of Difficulty: Advanced.**) (J Am Coll Cardiol Case Rep 2020;2:2120–3) © 2020 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

A 14-year-old patient had a witnessed sudden cardiac arrest while running with friends. He lost consciousness and collapsed without any chest pain, dyspnea, or dizziness. Cardiopulmonary resuscitation was started immediately by a bystander and continued until emergency medical services (EMS) arrived. The EMS team placed an automated external defibrillator, which verified ventricular fibrillation. Two defibrillation shocks resulted in sinus tachycardia. Time from EMS arrival to return of spontaneous circulation was 5 to 10 min, a period in which the patient was unconscious. The patient arrived to the emergency department in an agitated state with minimal responsiveness with a blood pressure of 146/109 mm Hg. He was then intubated and sedated.

LEARNING OBJECTIVES

- To review CCTA anatomic details that may predict higher risk of coronary event in AAOCA, RCA from left sinus of Valsalva.
- To review CMR parametric imaging techniques that define myocardial edema in the right coronary distribution.

MEDICAL HISTORY

The patient was a previously healthy teenager who participated in competitive sports without cardiac symptoms. He had no recent illness, personal or family history of congenital heart disease, sudden cardiac death (SCD), early myocardial infarction, rhythm disorders, or need for electrophysiology devices.

DIFFERENTIAL DIAGNOSIS

Myocarditis, channelopathies including long QTc and catecholaminergic ventricular tachycardia; myopathies including hypertrophic cardiomyopathy or

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The authors attest they are in compliance with human studies committees and animal welfare regulations of the authors' institutions and Food and Drug Administration guidelines, including patient consent where appropriate. For more information, visit the *JACC: Case Reports* author instructions page.

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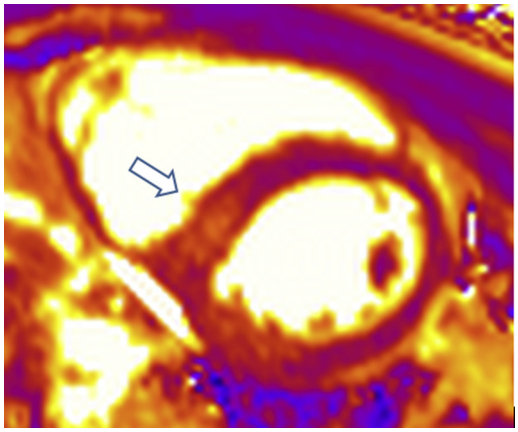
FIGURE 1 Edema in the RCA Distribution



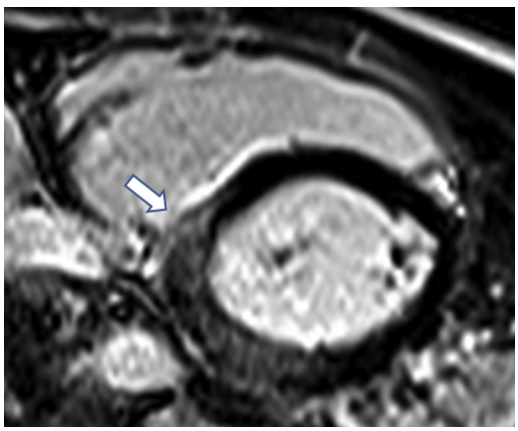
Cardiac magnetic resonance image showing increased T2 consistent with edema in the right coronary artery distribution (basal and mid inferoseptum).

arrhythmogenic right ventricular cardiomyopathy; toxin ingestion, anomalous coronary artery.

INVESTIGATIONS

A limited transthoracic echocardiogram performed on arrival to the emergency department showed moderately depressed biventricular systolic function, no left ventricular hypertrophy, and probable normal origins of the coronary arteries by 2-dimensional imaging. The electrocardiogram (ECG) showed nonspecific ST- and T-wave changes and normal QTc. After 5 days it showed T-wave inversion in inferior leads. Troponin was mildly increased at 0.22 ng/ml. Viral and toxicology screening was negative. Subsequent genetic testing for cardiomyopathy showed no known pathological variants. A detailed repeat echo was suspicious for anomalous aortic origin of the right coronary artery (RCA) from the left coronary sinus. The patient was referred for cross-sectional imaging to further clarify etiology of cardiac arrest and to help determine need for defibrillator placement.

FIGURE 2 Late Gadolinium Enhancement in the RCA Distribution



Cardiac magnetic resonance image showing abnormal late gadolinium enhancement in the right coronary artery distribution (basal and mid inferoseptum).

Cardiac magnetic resonance (CMR) was performed in a 1.5-T scanner using standard breath-hold cine imaging for function, parametric imaging, and late gadolinium imaging for tissue characterization. Cine imaging showed basal and mid inferoseptal hypokinesis and mildly decreased right and left ventricular systolic function. Parametric imaging revealed elevated T1 of 1,135 ± 59 ms (normal reference value: 950 to 1,050 ms) and elevated T2 of 68 ± 3.5 ms (normal reference value: ≤55 ms) in the basal and mid inferoseptum (Figure 1). Late gadolinium imaging was significant for low-intensity enhancement of the basal and mid inferoseptum (Figure 2). Calculated extracellular volume was 36 ± 2.5% (normal reference value: 25.3 ± 2.5%) and was also elevated in the basal and mid inferoseptum (Figure 3). Single-shot 3-dimensional post contrast of aorta showed possible anomalous RCA. The wall motion abnormality, parametric imaging abnormalities, and late gadolinium enhancement correlated with ischemia in the RCA distribution.

A coronary computed tomography angiogram (CCTA) was performed for detailed coronary artery imaging. CCTA confirmed anomalous aortic origin of coronary artery (AAOCA) with the right coronary artery arising from the ascending aorta above the left sinus of Valsalva. Anatomic details included a right dominant coronary system. Proximal RCA course was significant for ostial narrowing, acute angulation from the aortic wall, and intramural and interarterial course (Figures 4A and 4B).

MANAGEMENT

Ten days after cardiac arrest, the patient underwent surgical unroofing of the RCA with re-suspension of the aortic valve commissure. Operative course and recovery were uneventful. A defibrillator was not

ABBREVIATIONS AND ACRONYMS

AAOCA = anomalous aortic origin of coronary artery

CCTA = coronary computed tomography angiogram

CMR = cardiac magnetic resonance

ECG = electrocardiogram

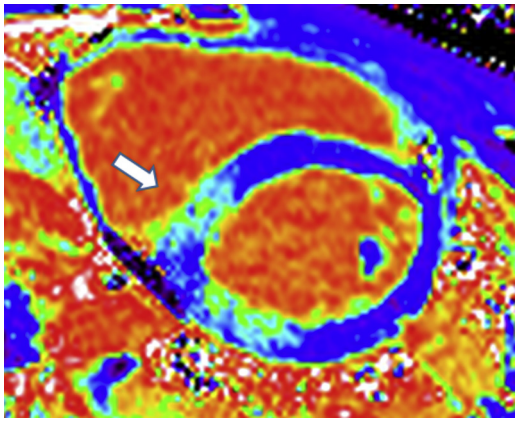
Echo = echocardiogram

EMS = emergency medical services

RCA = right coronary artery

SCD = sudden cardiac death

FIGURE 3 Extracellular Volume in the RCA Distribution



Cardiac magnetic resonance image showing increased extracellular volume (36%) in the right coronary artery distribution (basal and mid inferoseptum).

placed due to the findings of edema from acute transient ischemia in the RCA distribution associated with AAOCA as etiology for his cardiac arrest.

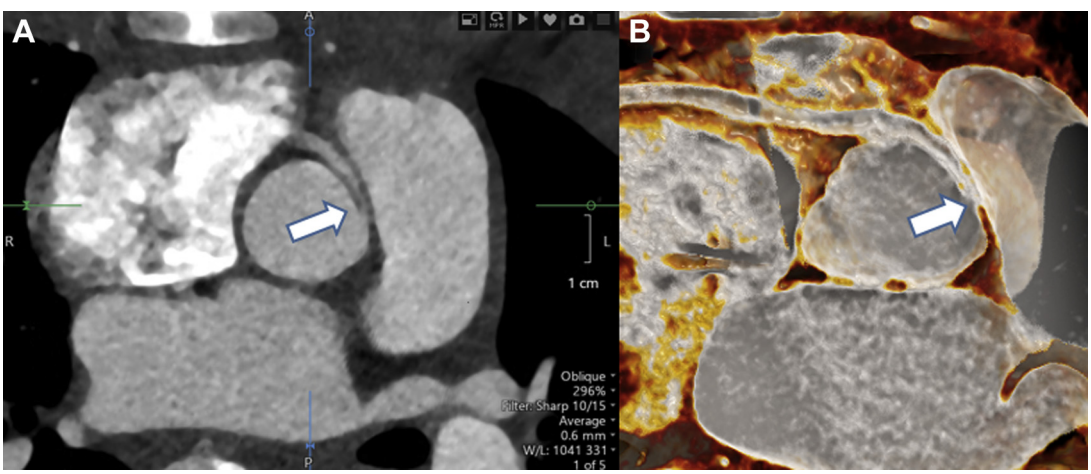
DISCUSSION

Coronary arterial anomalies range from benign anatomic variants to those presenting with SCD (1). AAOCA from the inappropriate sinus of Valsalva is common, with a prevalence estimated in 0.7% (2). The right variant of AAOCA is much more common than left AAOCA (2), but is rarely associated with SCD and management is controversial in asymptomatic patients (3).

Imaging characteristics of right AAOCA that may predict increased risk of adverse events include slit-like orifice, acute angle of origin, interarterial course, length of intramural course, and right dominant coronary artery system (1). Activity-related sudden death in AAOCA is thought to be secondary to arrhythmia triggered by myocardial ischemia from coronary compression of the interarterial or intramural segment in the setting of increased cardiac output (4). The available data suggest that myocardial ischemia is intermittent. A negative maximal-effort stress test does not exclude a potentially lethal coronary anomaly, and sudden death can be the first symptom in patients participating in high-intensity athletics (2,5). Coronary anatomic features of right AAOCA that may allow differentiation of a benign anomaly from more malignant anatomic variants are important to identify (4). If this were possible, surgical correction would be recommended in those at highest risk of adverse event while sparing those with benign lesions from the risks of surgery.

In older patients, repetitive ischemic episodes may result in patchy myocardial fibrosis creating an electrically unstable myocardial substrate with risk for arrhythmia (5). This is corroborated by autopsy

FIGURE 4 High-Risk Characteristics of AAOCA in CCTA



Coronary computed tomography angiogram images (**A** = 2 dimensional, **B** = 3 dimensional) showing origin of the right coronary artery from the ascending aorta just above the left sinotubular junction on a short axis image. Note the coronary ostial narrowing, acute angulation from the aortic wall, and intramural/interarterial proximal coronary course.

studies of adult patients who died because of AAOCA, where myocardial fibrosis is a common finding, suggesting that ischemia preceded the terminal event (6).

There are no prior case reports of right AAOCA presenting with cardiac arrest with high risk imaging features defined by CCTA and correlated to myocardial edema in the RCA distribution by CMR in the acute setting. The 2 imaging modalities are complementary in this case to define the anatomic details of the coronary anomaly and the physiologic consequence.

FOLLOW-UP

The patient did remarkably well after surgical intervention without cardiac symptoms and/or neurological sequelae. Post cardiac surgery, his echo showed normal biventricular systolic function without wall motion abnormalities. Color flow was documented in the proximal coronary arteries and there was mild aortic insufficiency. ECG remained normal. He will undergo repeat CMR and stress testing before clearance for strenuous activity.

CONCLUSIONS

CCTA and CMR are complementary imaging modalities that allow anatomic and physiologic correlation in the setting right AAOCA presenting with resuscitated sudden cardiac arrest.

AUTHOR DISCLOSURES

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KEY WORDS cardiac magnetic resonance, computed tomography, congenital heart defect, coronary vessel anomaly