Recurrent sudden cardiac death secondary to anomalous right coronary artery: Insights into prevalence and management

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Abstract

A 32-year-old woman presented after ventricular fibrillation arrest requiring three defibrillations. The episode coincided with an upper respiratory infection and physical exertion. Eight years prior, she survived another cardiac arrest of unknown cause during childbirth. This time, imaging revealed an anomalous right coronary artery connecting to the left coronary cusp, with a small, slit-like osteal orifice coursing between the aorta and pulmonary artery. Surgical exploration revealed an intramural segment of the right coronary artery, which was surgically unroofed with improvement in cardiac function. An implantable cardioverter-defibrillator was implanted for secondary prevention of sudden cardiac death. Surgery is recommended for malignant anomalous coronary arteries, with a very low risk of recurrence of arrhythmia and sudden cardiac death after surgery. However, with growing evidence for persistent risk of arrhythmia and sudden cardiac death even after surgical correction of the anomalous coronary arteries, more experts choose to take secondary prevention measures as a component of initial management.

Introduction

Anomalous coronary arteries (ACAs) are defined as coronary arteries with an abnormal connection and/or course. The prevalence of these anomalies is estimated to be less than 1% in the general population.[1] The vast majority of these anomalies are benign; however, a few are defined as malignant due to their propensity to cause symptoms and sudden cardiac death (SCD). Many cases of anomalous coronary arteries are discovered on autopsy, but some patients are diagnosed after symptoms present. ACAs are particularly prevalent in young athletes, where they have been associated with 17% of exercise-related deaths.[2] There are two main mechanisms by which ACAs cause symptoms. In the first, acute exertion results in significant ischemia and malignant arrhythmia secondary to one or more of the following: (1) compression of coronary arteries between the aorta and pulmonary artery (i.e. interarterial course). This compression may either directly decrease blood flow or cause secondary vasospasm from the mechanical stimulation.[3] (2) Ostial occlusion during physical exertion (slit-like orifice or acute-angle takeoff). (3) Proximal segmental occlusion due to an intramural course in the aortic wall.[4,5] This is a particularly malignant feature due to hypoplasia of the intramural segment and susceptibility to lateral compression by the aorta during times of increased cardiac output.[1] Alternatively, chronic ischemia and fibrosis can develop in the ACA distribution resulting in malignant arrhythmia.[4] This occurs due to chronic hypoperfusion leading to tissue injury. Herein, we describe a rare case of recurrent sudden cardiac death in a

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young woman with a unique constellation of coronary artery anomalies.

Case presentation

A 32-year-old woman presented after a witnessed collapse at home while carrying her 6-year-old child. Her family initiated resuscitation efforts after noticing her "gasping and snoring." On initial assessment by paramedics, she was in ventricular fibrillation (VF) with subsequent return of spontaneous circulation after three shocks were administered. In the days leading up to the event, the patient complained of viral symptoms, including body aches, fever, runny nose, and cough.

In the emergency department, she was tachycardic to 110, with sustained blood pressures in the 130 s/90 s mmHg. She developed recurrent episodes of ectopy and pulselessness, requiring three additional defibrillations prior to stabilization. Physical examination on arrival was notable for intubated and sedated state, rapid and regular heart rhythm without murmurs, rubs or gallops, and coarse crackles throughout all lung fields.

The patient's history includes untreated hypertension and prior SCD during the delivery of her first child 8 years prior. The patient had no history of alcohol, tobacco, or other substance use. She had no history of major illnesses or infections and no recent travel, sick contacts, exposures to pets, or occupational exposures.

The differential diagnosis of the patient's VF was broad and included hypertrophic cardiomyopathy, acute myocardial infarction, underlying channelopathies such as long or short QT syndromes and Brugada syndrome, dilated cardiomyopathy related to viral myocarditis, alcohol abuse, or idiopathic. We also considered coronary artery spasm or anomalous coronary arteries.

Viral serologic testing identified rhinovirus, and a urine toxicology screen was negative. An electrocardiogram (ECG) noted sinus tachycardia with ST depressions in anterolateral leads. ST elevations were absent on standard and posterior ECG. There was no evidence of prolonged or shortened QT or other electrical signs of channelopathies. A computed tomography (CT) angiogram of the chest identified a malignant anomalous right coronary artery (RCA) with an interarterial course between the aorta and main pulmonary artery and no evidence of pulmonary embolus. A transthoracic echocardiogram demonstrated a mild dilation of the left ventricle with moderately reduced systolic function and global hypokinesis. Subsequent coronary artery angiography revealed an ACA of the RCA connected to the left coronary cusp, consistent with the CT findings, with right-dominant circulation. The left anterior descending and left circumflex arteries were angiographically normal. As a result, the patient underwent cardiac magnetic resonance imaging (MRI), which showed no evidence of acute myocarditis or infarction.

In addition to routine postarrest care, the patient received amiodarone and metoprolol to decrease the risk of recurrent VF. She underwent targeted temperature management and remained intubated and sedated for 17 days with evidence of status epilepticus on electroencephalogram and delayed neurologic improvement. CT coronary angiography identified a small, slit-like osteal orifice of the RCA connecting to the left sinus of Valsalva followed by an acute-angle takeoff (Figure 1). The RCA then takes a long interarterial course between the aorta and pulmonary artery (Figure 2). Given the concern that the ACA was a culprit for the SCD, the patient underwent surgical repair. It revealed a 12-mm intramural segment of the RCA for which she underwent surgical unroofing.[6] Her preoperative transesophageal echocardiogram showed moderate to severe mitral regurgitation with a left ventricular ejection fraction (LVEF) of 40%, while postoperative ultrasound showed improved LVEF (>60%) and only mild mitral regurgitation.

Following surgical unroofing of this patient's anomalous RCA, she was discharged with a wearable defibrillator with plans to undergo implantable cardioverter-defibrillator (ICD) placement for secondary prevention of SCD. This was done as there was uncertainty about whether the patient's SCD was entirely attributable to mechanical obstruction of an anomalous RCA.[7,8] Potentially, chronic ischemia from years of hypoperfusion in the distribution of the RCA left the patient at increased risk of malignant arrhythmia and recurrent SCD. However, there was no evidence of infarction or fibrosis on cardiac MRI. Over the following few months, she continued to recover neurologically to independence.

Discussion

This case is different from a previously described case of malignant ACA of the RCA in which the interarterial segment of RCA in our patient contains a long intramural portion that exposes the artery to an additional risk of hemodynamic compromise.[9] Another unique aspect of this case is the patient's previous SCD during a difficult labor 8 years prior in a resource-poor setting, without further investigation of etiology. However, we suspect that physiologic changes related to labor including an increase in blood volume and cardiac workload precipitated tachyarrhythmia and/or hypoperfusion resulting in cardiac arrest. Alternatively, her initial episode may have been a result of peripartum cardiomyopathy resulting in VF arrest. Interestingly, she gave birth 2 years later without complications. Despite this history, her most recent SCD occurred during a seemingly mild viral illness while lifting her child suggesting a heterogenous triggering mechanism. Triggers for this episode likely included exertion from lifting her child, tachycardia from viral illness, and untreated hypertension superimposed on baseline increasing the risk for SCD. This abnormal anatomy results in secondary, episodic ischemia during exertion when physiologic dilation of ventricles stretches the RCA and the proximal RCA becomes compressed against the aortic wall.[10] This compression may either directly decrease blood flow or (less likely) cause secondary vasospasm from
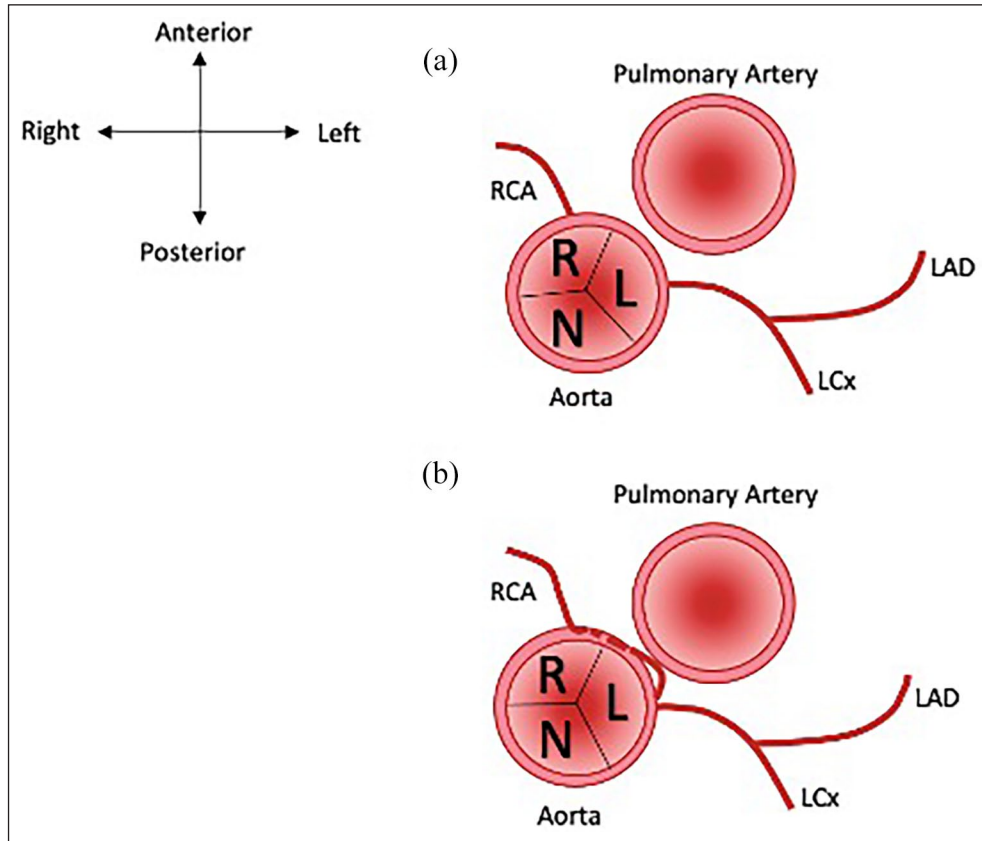
Figure 1. Normal coronary artery connection and course contrasted with anomalous RCA. (a) Normal coronary artery connection and course. (b) Anomalous RCA with slit-like orifice, interaterial, and intramural course.
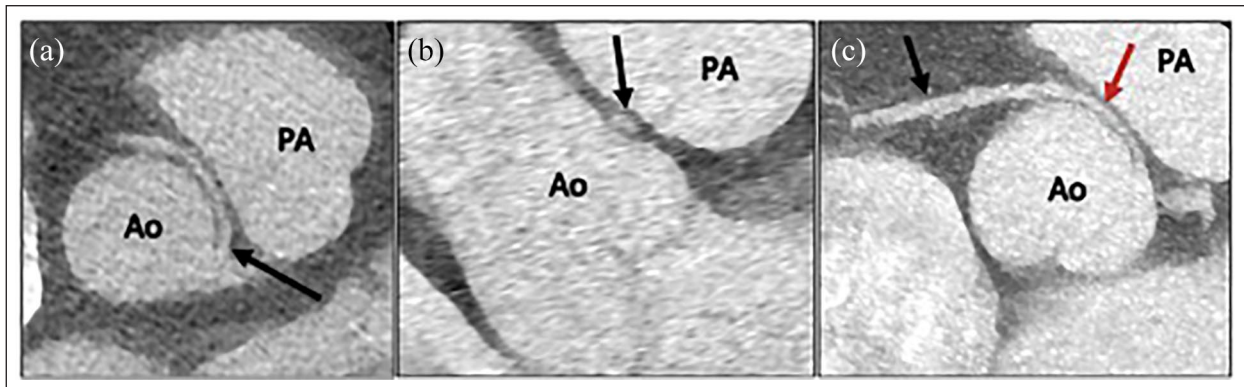LAD: left anterior descending artery; LCx: left circumflex coronary artery; RCA: right coronary artery.

Figure 2. Imaging from CT coronary angiography. (a) An oblique reformat demonstrates an acute-angle takeoff of the anomalous RCA (arrowhead) as it courses interarterially. (b) The coronal image demonstrates lateral luminal compression of the anomalous RCA (arrow). (c) This oblique maximal intensity projection again demonstrates luminal compression (red arrow), with distal restoration of the luminal shape and diameter (red arrow) as it exits the intramural, interarterial portion.
Ao: aorta; PA: pulmonary artery; RCA: right coronary artery.

the mechanical stimulation.[10] The intramural course within the tunica media of the aortic wall is particularly a malignant feature due to hypoplasia of the intramural segment and susceptibility to lateral compression by the aorta during times of increased cardiac output.[1] Of note, ST-segment depressions isolated to the anterolateral leads on the initial ECG are likely a result of acute nonlocalizing ischemia, with a notable absence of ST segment elevation or Q-waves in the inferior leads suggesting the absence of acute myocardial infarction.

In patients with malignant ACA, surgical intervention is recommended and can include unroofing and/or coronary artery bypass procedures.[11] Outcomes after surgical intervention are excellent, with a very low risk of symptom recurrence, as was seen for this patient.[12] Recent studies have highlighted a possible persistent risk of recurrent symptoms after surgical intervention for malignant ACA, with some experts recommending the addition of ICD for secondary prevention.[7,8]

In some cases where patients experience SCD in the setting of ACA, noninvasive imaging, including cardiac MRI may help with the identification of an underlying arrhythmogenic substrate presenting as scarring/fibrosis resulting from myocardial infarction, and rendering a persistent risk of recurrent arrhythmia even following surgical correction of the ACA.[13] However, in this particular case, there was no evidence of scarring on cardiac MRI, suggesting an acute ischemic episode as the trigger for this patient's arrhythmia.

Conclusion

Cases of SCD have a broad differential and imaging studies are effective in helping determine the most likely trigger. ACAs are rare, but even more so, a case of anomalous RCA with a slit-like orifice, significant proximal intramural course and interarterial course resulting in recurrent SCD has not previously been noted. These anomalies result in SCD due to ischemia and tachyarrhythmia. While surgical intervention can be technically challenging, it can effectively correct the anomaly and prevent SCD. There is an increased focus on secondary prevention in these cases with many experts recommending ICD placement.

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Ethical approval

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Informed consent

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