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A Missed Malignant Right Coronary Artery Anomaly Detected Post-Cardiac Event in an Adult Patient

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Authors' contributions

This work was carried out in collaboration between all authors. Author AS designed and wrote the manuscript. Authors TRL and SKAJ managed the literature searches and critically reviewed the manuscript. All authors read and approved the final manuscript.

Case Study

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ABSTRACT

Aims: We present this case to raise awareness of this unusual presentation of a malignant anomalous right coronary artery arising from the left coronary cusp in a patient older than 50 years. We describe a useful imaging modality and discuss therapy.
Presentation of the Case: We report a case of a 63-year-old male with an interarterial coursing right coronary artery arising from the left coronary cusp with a history of a mechanical aortic valve replacement. The patient presented to our emergency department after being resuscitated from a cardiac arrest and later had a normal coronary angiogram. High suspicion of his right coronary artery angulation, he underwent a computed tomography with angiogram that revealed his anomalous course and anatomy.
Discussion: We discuss the importance of coronary artery anomaly detection in the young at risk population (athletics), pathophysiology, diagnostic modalities, and treatment recommendations. Surgical revascularization has been advocated in malignant coronary anomalies, however, the lack of large randomized clinical trials for patients older than 50-year-old left this topic controversial. Medical therapy augmented with implanted cardioverter defibrillator (ICD) was utilized in this case. Medical management could be considered in centers that don't have surgical experience or in adult patients who are a

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poor surgical candidate because of other comorbidities or life expectancy.
Conclusion: Computed tomography with angiogram seems to be the best noninvasive modality to delineate coronary course and anatomy. There is a need for randomized clinical trials to determine the best management of anomalies arising from opposite sinus with an interarterial course in adults >50-year-old.

Keywords: Anomalous right coronary artery; implantable cardioverter defibrillator; CT-coronary angiogram.

ABBREVIATIONS

CAAs: Coronary artery anomalies; RCA: Right coronary artery; SCD: Sudden Cardiac death LAD: Left anterior decending artery; LCX: Left circumflex artery; LMCA: left main coronary artery; ACC/AHA: American College of Cardiology/American Heart Association; CT: Computed tomography; MRCA: Magnetic resonance coronary angiography; ECG: Electrocardiography; ICD: implanted cardioverter defibrillator.

1. INTRODUCTION

Coronary arteries anomalies (CAAs) occur in less than 1% of the general population [1]. Anomalies of the right coronary artery (RCA) are particularly rare with an estimated prevalence of 0.026% as reported by Alexander et al in a review of almost 19 thousand autopsies [2]. Kaku et al. corroborated this estimate in a series of 17,731 angiograms, detecting RCA anomalies in 44 patients (0.25%) [3]. CAAs can be classified into the following four major categories: 1) Anomalies of course and origin of the artery. 2) Anomalies of intrinsic coronary artery anatomy. 3) Anomalies of coronary termination (which include fistulas). 4) Anomalous anastomotic vessel [4].

Anomalous RCA originating from the left sinus were considerably less common than other types of coronary anomalies [1,4]. Fortunately, the majority of RCA anomalies are clinically benign and thus remain clinically silent [5]. Coronary anomalies are of clinical importance when they predispose to coronary artery compression which can compromise myocardial blood flow and lead to myocardial ischemia.

Herein we present a case of an anomalous RCAs. Our case presents with sudden cardiac arrest, the patient underwent cardiac catheterization for possible acute myocardial infarction resulting in arrhythmia. Coronary artery angiogram revealed no occlusive coronary disease. A computed tomographic (CT)-angiogram revealed the anomalous RCA.

2. CASE REPORT

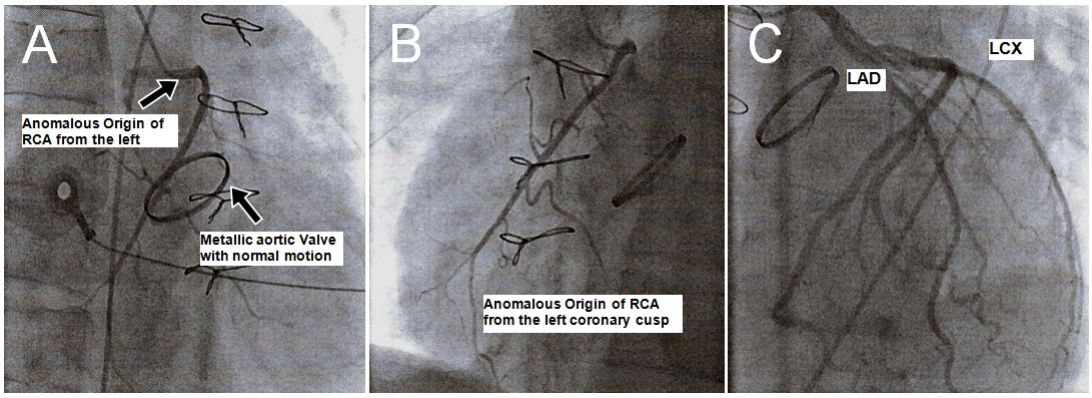
A 63-year-old Caucasian male suffered a cardiac arrest while awake in his bedroom. His wife heard him fall, he was found unconscious, emergency medical services were contacted and chest compressions were begun. First responders successfully resuscitated him with unsynchronized cardio version, epinephrine, and amioderone. His airway was maintained with a combitube and he was transferred to the hospital. Upon arrival to the emergency department he was pale, cool, and diaphoretic. Initial vital signs included blood pressure of 92/71 mmHg, heart rate 100 beats/minute, and temperature was 91.6 degrees Fahrenheit. Pupils were equal, round, and reactive to light. His cardiac exam revealed a regular

tachycardia, S1 and S2 present, and no murmurs, gallops, or rubs heard. The rest of the chest exam demonstrated normal bronchial sounds, with no wheezing, rhonchi, or rales. The remainder of the exam was unremarkable. His wife was able to provide his medical history which was only significant for an aortic mechanical valve replacement in 2009. He underwent aortic valve replacement with a mechanical valve secondary to congenital bicuspid valve resulting in aortic stenosis.

Initial laboratory data demonstrated cardiac troponin T of <0.02 ng/ml (normal reference<0.02 ng/ml), total CK was 117 unit/L, and pro-BNP 713 pg/ml. Initial electrocardiography (ECG) in the hospital showed minor ST-segment elevation in the inferior leads (leads II, III, and aVF) and ST-segment depression in the lateral leads. He was resuscitated with IV fluid, IV amiodarone continuous infusion, 2 grams of magnesium IV, and rectal aspirin. He was endotracheally intubated and mechanically ventilated to support respiration. He was emergently transferred to the cardiac catheterization laboratory for possible percutaneous coronary intervention. Coronary angiography identified a RCA arising from the left coronary cusp superior to the origin of the left main coronary artery (LMCA). The RCA coursed anterior to the aorta (Fig. 1). No significant arterial stenosis or atherosclerotic occlusion was identified. A mechanical aortic valve was visualized with overlaying sternotomy wires from a previous valve replacement surgery (Fig. 1).

The anomalous RCA was further characterized with a computed tomography scan with angiography (CT-angiogram). The CT-angiogram confirmed a cephalic origin of the RCA 8 mm from the origin of the LMCA with a course traveling between the aorta and pulmonary artery (interarterial) (Fig. 2 and Fig. 3).

With EKG evidence of ischemia and an interarterial coursing RCA, the patient was thus classified a class I according to the American college of Cardiology/American heart association (ACC/AHA) guidelines. Surgery for re-implantation of the anomalous RCA was discussed however, this procedure was felt to be technically difficult as the patient had undergone prior heart surgery, thus he elected medical management. He was then placed on beta blockers with a rigorous flow up schedule. Prior to discharge he received an implantable cardioverter defibrillator (ICD) for secondary prevention of arrhythmias and SCD.



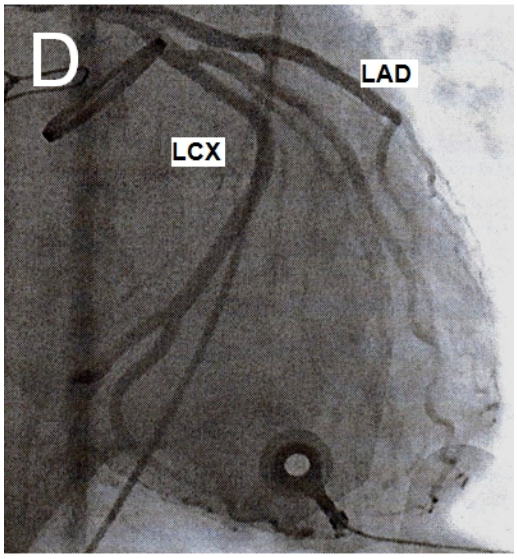


Fig. 1. Coronary angiogram of our patient

RCA arising from the left coronary cusp appears to run anteriorly. Metallic aortic valve with a normal motion and previous sternotomy (A). RCA from a right anterior orientation (B). Dominating Left main artery system with two branches from Left anterior orientation (C). Right anterior orientation of the Left anterior descending and left circumflex arteries (D). (RCA) right coronary artery. (LAD) Left anterior descending. (LCX) Left circumflex.

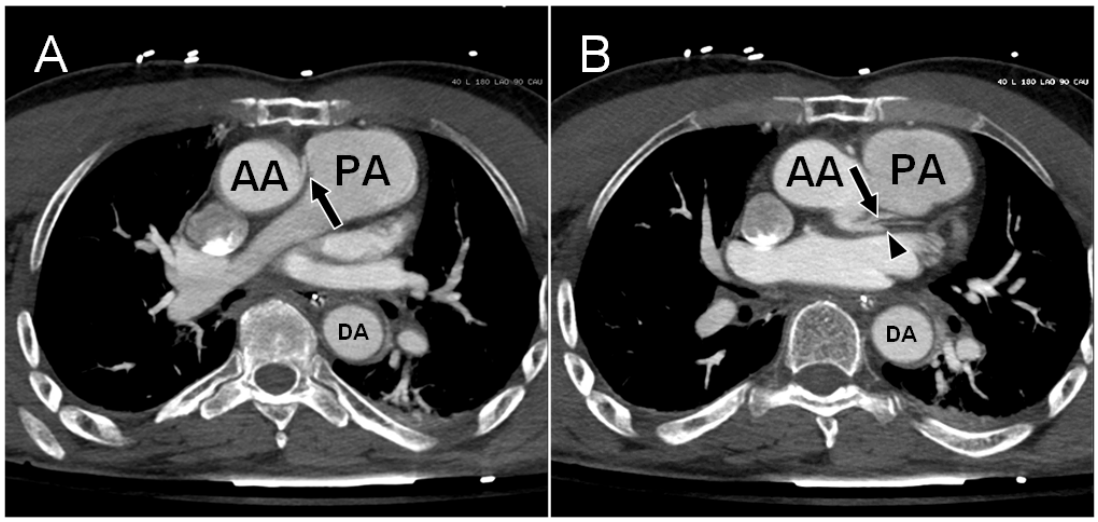


Fig. 2. Thoracic CT scan from the first case

T4-T5 CT scan with arrow pointing to the anomalous RCA arising from the left cusp of the Aorta with a malignant course that is interarterial in between the aorta and pulmonary artery (A). T4-T5 CT scan showing the bifurcating of the left dominating system into Left anterior descending artery (arrow) and left circumflex artery (arrow head) (B). (AA) Ascending aorta. (PA) Pulmonary artery. (DA) Descending aorta. (RCA) right coronary artery

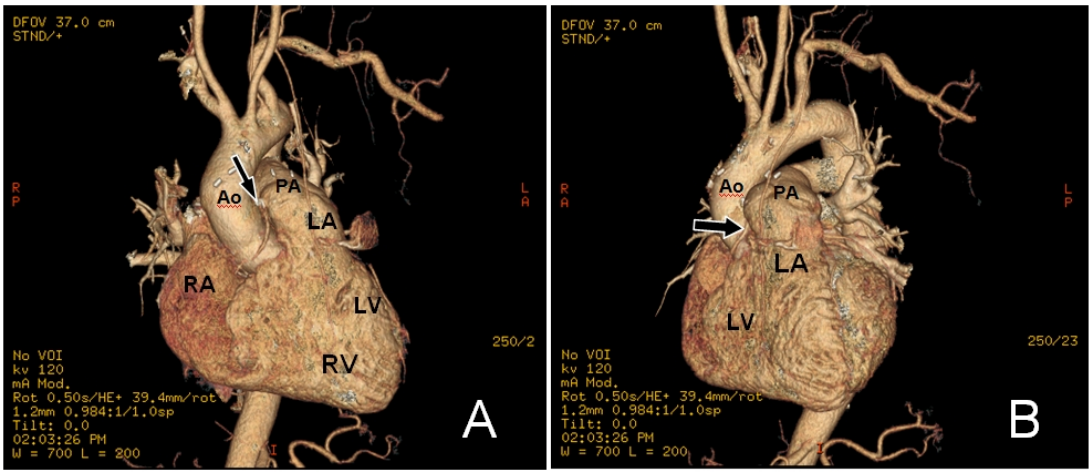


Fig 3. 3D reconstructed CT angiogram of the first case

Arrow pointing to the malignant course of the (A). Lateral projection of the heart with arrow pointing to the RCA (B). (Ao) Aorta. (PA) Pulmonary artery. (RA) Right atrium. (RV) Right ventricle. (LA) Left atrium. (LV) Left ventricle. (RCA) right coronary artery.

3. DISCUSSION

The patient's anomaly was detected at age of 63, which was undetected after undergoing a prior aortic valve replacement surgery. Although patients carrying RCA anomalies were not reported to have a specific age at presentation, all coronary anomalies (especially arising from an opposite sinus and having an interarterial course) are an important etiology in young patients who suffer sudden cardiac death (SCD) [6]. 19% of all SCD reported in patients <35 years of age were attributed to CAAs [7,8]. In asymptomatic individuals these anomalies are only detectable by imaging that can visualize the arterial origin and course (i.e. CT or magnetic resonance (MR) angiography) [1]. Guidelines and recommendations for congenital coronary anomalies of ectopic arterial origin remain controversial for patients older than 50 year-old because of the absence of large randomized clinical trials (with large number of patients) rather than small nonrandomized or observational studies [9]

Often coronary anomalies are clinically silent, if symptoms are present they are typically related to myocardial ischemia and include angina pectoris, myocardial infarction, and heart failure. If myocardial blood flow is compromised, patients become at risk for SCD, which can be the initial manifestation [8]. Other symptoms include palpitations, syncope, and ventricular arrhythmias [10].

There are several important anatomical features that predispose to ischemia in patients with CAAs originating from the opposite sinus. These malignant features include a course traveling between the aorta and pulmonary artery and the angulation at the coronary artery origin. The CT-angiogram done on the patient revealed a RCA with a cephalic take off and interarterial course (Fig. 2 and Fig. 3). The interarterial course carries the highest risk of ischemia, as progressive exertion increases cardiac output which results in dilatation of both the pulmonary artery and aorta [11]. This dilatation of the great vessels causes direct compression, compromising coronary blood flow. Arteries with sharp either acute or obtuse angles of origin are also at risk for collapse leading to subsequent ischemia [12,13].

Anomalies of intrinsic coronary artery anatomy my also result in ischemic disease and include vascular wall hypoplasia. This class of anomalies frequently results in arterial vasospasm and is usually difficult to diagnose. These anomalies predispose patients to additional complications (such as thrombosis and vasospasm) in addition to mechanical compression. It is noteworthy that patients with exertional symptoms due to CAAs were more likely to have an anomaly with an interarterial course or a compromised take off angle. If symptoms are not related to exertion, then the mechanism is more likely to be vasospasm [13,14].

The diagnosis of CAAs is complicated given the low incidence in the general population and symptoms that mimic much more common pathology [1]. Additionally the sensitivity and specificity of diagnostic tests used to visualize CAAs are, for the most part, not known. Therefore when CAAs are suspected (by evidence of ischemia or SCD and nonocclusive atherosclerotic disease in our case) several imaging modalities were utilized. Coronary anatomy and blood flow can be delineated through many modalities including echocardiograms with Doppler interrogation (e.g. to examine the anatomy of aortic cusp and direction of blood flow), invasive angiography or CT/MR angiography [15,16]. Certain tests are more reliable than others; CT-angiography appears to be more reliable over echocardiography. Coronary angiography demonstrated a detection rate for CAAs of 1.07% compared to echocardiography detection rate of 0.17% in an adult population [1]. CT also has the added ability to delineate the origin of the anomalous artery and view its course better than coronary angiography and echocardiogram. One study was able to detect a 6.6% of CAAs in a 242 patients using a 16-slice CT [17]. Another study was able to visualize CAAs and their proximal course in patients using magnetic resonance coronary angiography (MRCA), proving another useful technique [18]. CT angiography appears to be the best available imaging modality in detecting CAAs noninvasively, however the evidence base supporting this recommendation remains relatively weak [9].

Diagnosis of CAAs and delineation of a malignant anatomy will then require prevention of long term complications after acute treatment. The ACC/AHA guidelines for the congenital coronary anomalies of ectopic arterial origin advocates surgical correction in adult patients presenting with SCD or symptoms of CAAs [9]. There are many surgical modalities including but not limited to revascularization. Revascularization may include translocation of the coronary artery, ostioplasty, or bypass grafting. Our patient elected against surgical intervention as we also did, and thus was treated medically with beta blockers. Medical therapy using beta-blockers have been advocated for and practiced in Japan where surgical therapy of CAA is not common [3,5]. Augmenting medical treatment, we also placed and ICD to prevent any future morbidity and mortality. Additionally implantation of an ICD has been shown to improve survival after an episode of SCD. The Antiarrhythmics Versus Implantable Defibrillators (AVID) trial compared ICD implantation with drug therapy using amiodarone. The ICD treated group had a relative reduction in mortality by 39% in one year and a reduced relative risk of ventricular fibrillation or tachycardia (specifically with EF 40%) by 33%, showing an overall survival greater than the amiodarone only treatment group [19]. The Cardiac Arrest Study Hamburg (CASH) randomized 288 patients to ICD, amiodarone, or metoprolol for secondary prevention of SCD due to ventricular arrhythmias. The CASH study showed that ICD implantation reduced mortality by 23% (although nonsignificant according to the study) compared to drug therapy over 5 years [20]. Although both AVID and CASH trials suggest benefits of ICD for secondary prevention of SCD due to ventricular arrhythmias, these trials do not specifically address patients who suffer SCD due to CAAs. There remains a lack of studies that evaluate ICD's versus medical antiarrhythmic therapy in the secondary prevention of SCD specifically in CAA patients. Had our patient undergone

surgical revascularization his risk of ventricular arrhythmias and SCD would have diminished and his benefit from an ICD implantation becomes unclear.

4. CONCLUSION

Malignant and symptomatic CAAs lead to myocardial ischemia and long-term heart damage through small ischemic events and/or ventricular arrhythmias prompting life-long intervention to prevent SCD. This increased risk of mortality is more specifically worrisome in athletic young population at risk and should prompt cardiologists not to rely solely on one screening or diagnostic test, but to utilize multiple tests when CAA suspicion is high using CT-angiograms to delineate the coronary course after stress testing. However, patients older than 50 year-old with coronary anomalies especially those arising from opposite aortic sinus and has interarterial course prompt revascularization surgery per ACC/AHH guidelines. This management remains controversial in the adult population, as no large randomized clinical trials have addressed these patients. Medical treatment with beta blockers is a possible alternative especially in centers without experience performing revascularization surgery or when patient comorbidities or life expectancy preclude surgery. Pharmacological antiarrhythmics for sustained ventricular arrhythmia or ICD management (a better alternative) when patients undergo medical therapy for their CAAs are both valid therapeutic considerations as a secondary prevention of SCD.

CONSENT

All authors declare that written informed consent was obtained from the patient (or other approved parties) for publication of this case report and accompanying images.

ETHICAL APPROVAL

Not applicable.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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