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Malignant Course of the Right Coronary Artery Originating from the Left Main Coronary Artery: An Odd Exit

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Abstract ***Introduction:*** Congenital coronary artery anomalies are quite uncommon with estimates ranging from 0.2% to 1.3% on coronary angiography. The rarity of these anomalies makes their diagnosis a formidable challenge. Variable arterial courses have been described. In this report, we present a case with unique arterial course, which starts as a hyperacute take-off of the right coronary artery (RCA) from the left main coronary artery (LMCA), with subsequent coursing, without external compression, between the right ventricular outflow tract and aorta. Our case is relevant to the concept of whether we should keep a reasonable index of suspicion for coronary artery anomalies during cardiac evaluations of patients undergoing non-cardiac surgeries. ***Clinical case:*** This is an asymptomatic 47-year-old African American female who presented for cardiac clearance for renal transplantation. She had a past medical history of hypertension, bronchial asthma, and former smoking of 10 years (quitting 15 years prior to presentation). She also has end-stage renal disease on hemodialysis. Cardiac workup revealed left ventricular hypertrophy on EKG, multi-chamber dilation seen on echo, and anomalous RCA course seen on CT coronary angiography. Cardiac catheterization revealed non-obstructive coronary artery disease of the LCX and RCA. After consulting with cardiothoracic surgery, conservative medical management was decided based on the patient's risk stratification. She was advised to have close monitoring of her condition. ***Discussion:*** Coronary artery anomalies represent the second most common cause of sudden cardiac death in young athletes. At this time, the prevalence of right coronary artery (RCA) take off from the left coronary sinus occurs at a percentage of 0.019% to 0.49%. The RCA origination from the left main coronary artery (LMCA) accounts for only 0.65% of these anomalies. Our patient had high-risk anatomy consisting of a hyper-angulated take-off of the RCA from the LMCA as well as course between the pulmonary artery and right ventricular outflow tract (RVOT). CT coronary angiography is the most useful imaging modality that characterizes coronary artery anomalies. Although this patient exhibited no signs or symptoms of cardiorespiratory compromise, she warranted a full cardiac workup preoperatively that incidentally revealed a coronary anomaly. Recognition of this disease is critical for timely prevention of potential complications as well as discussion of goals of care. Guidelines for medical versus surgical management are available, but the management strategy should be individualized, with the highest consideration given to risk-benefit analysis.

Keywords: *congenital coronary artery anomalies, malignant course of right coronary artery*

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1. Introduction

In patients undergoing coronary angiography, the incidence of coronary anomalies (CA) is estimated to range from 0.2% to 1.3%. [1] After hypertrophic cardiomyopathy, coronary artery anomalies are the second most common cause of sudden cardiac deaths among young athletes. [2,3] At this time, the prevalence of right coronary artery (RCA) take off from the left coronary sinus occurs at a percentage of 0.019% to 0.49%. [4] The RCA origination from the left main coronary artery (LMCA) accounts for only 0.65% of these anomalies. [5] Although CA anomalies are rare, they can exist in many configurations, with each case being unique with respect to origin, continuation, and termination of a major coronary artery. An example is the anomalous origin of

the LMCA or RCA from the aorta, which then courses between the aorta and pulmonary trunk. [3] This course is malignant due to the risk of arterial compression, which can lead to life-threatening consequences in young adults such as ischemic heart disease and sudden cardiac death. Because of the clinical significance of these anomalies, it is important to identify them as a measure of primary prevention. Identification of this pathology is possible with the use of ECG-gated multidetector CT coronary angiography, which provides accurate and non-invasive detection. [6] This imaging modality also provides visualization of surrounding vessels in relation to the anomalous artery. [7,8] Use of this imaging technique facilitates risk stratification of patients.

We present the case of a middle-aged African American female with modifiable risk factors for atherosclerotic cardiovascular disease, who was found to have a malignant RCA course. This report will discuss the clinical significance of the take-off, its origination from the LMCA, and its course between the RVOT and the aorta.

2. Case Presentation

A 47 year-old African American female with a past medical history of hypertension, bronchial asthma, former smoker for 10 years (quitting 15 years prior) and end stage renal disease on hemodialysis, presented to our institution to be evaluated for kidney transplantation. She denied shortness of breath on exertion, chest pain, palpitations, or dizziness. She described her exercise tolerance as 3-4 blocks limited by fatigue. Physical examination revealed a well developed female, not in any distress. She was afebrile with a blood pressure of 138/76 and a heart rate of 71. Her electrocardiogram demonstrated characters of left ventricular hypertrophy, nonspecific T wave abnormalities, and a prolonged QTc of 534 (Figure 1). Transthoracic echocardiography revealed an ejection fraction of 55% to 60% with left ventricular wall thickness markedly increased to 16.2 mm, mild systolic and diastolic flattening of the ventricular septum consistent with right ventricular volume and pressure overload. Furthermore, there was mild to moderate dilatation of the right ventricle, left atrium and right atrium with moderate to severe tricuspid regurgitation. Her pulmonary artery systolic pressure was estimated at approximately 40 mmHg plus right atrial pressure with a small pericardial effusion (Figure 2). She was sent for an outpatient dobutamine stress echocardiogram, primarily for risk stratification before renal transplantation. The test was terminated as the patient did not achieve the target heart rate. However, pulmonary systolic pressure was increased to 60-65 mmHg plus right atrial pressure with leftward displacement of the interatrial septum consistent with increased right atrial pressure. She was referred for CT coronary angiography which revealed a malignant interarterial course of her right coronary artery (RCA) originating from the proximal part of the left main coronary artery (LMCA). The artery traversed between the aorta and right ventricular outflow tract with secondary ostial and proximal narrowing. The LMCA trifurcated with presence of a ramus intermedius. There was evidence of pulmonary arterial hypertension with enlarged pulmonary arteries, right-sided chambers, and thickening of the free wall of both ventricles (Figure 3, Figure 4). She went for cardiac catheterization for better evaluation, which revealed non obstructive coronary artery disease at the distal left circumflex artery (LCX) and proximal RCA (Figure 5). Cardiothoracic surgery was consulted to evaluate the treatment for RCA course and conservative medical management was decided.

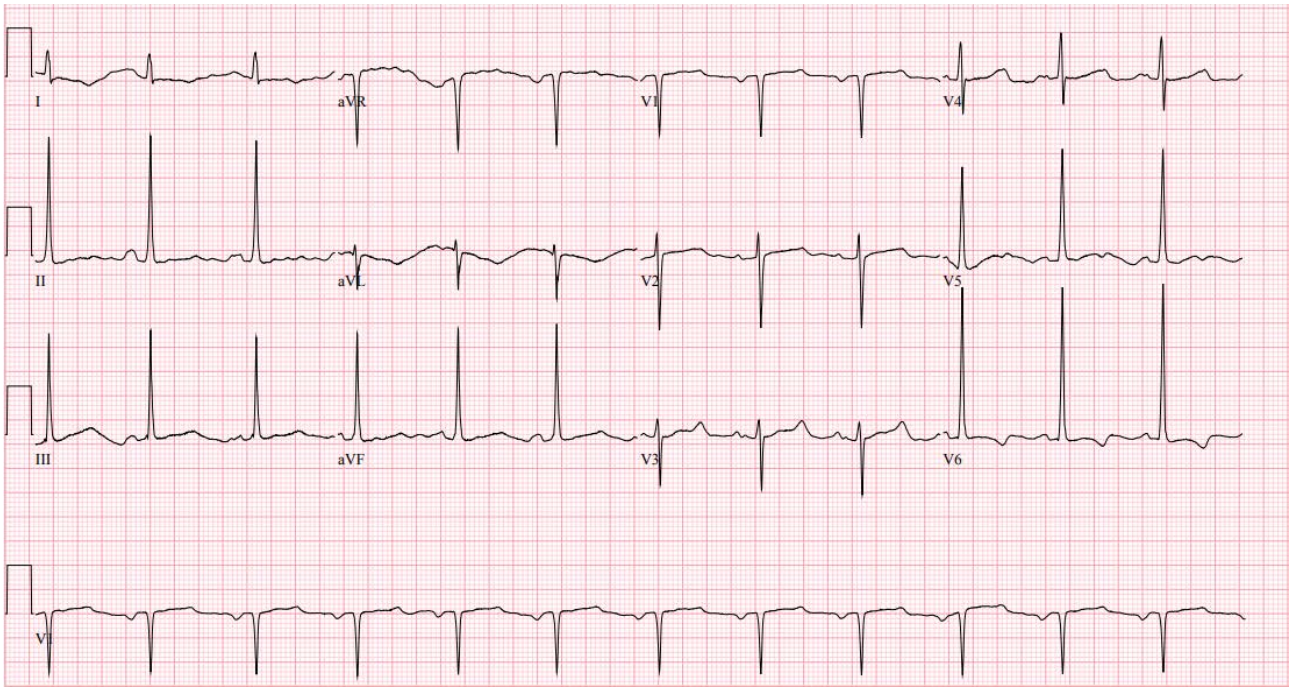

Figure 1. EKG of the patient showing left ventricular hypertrophy and prolonged QTc 534

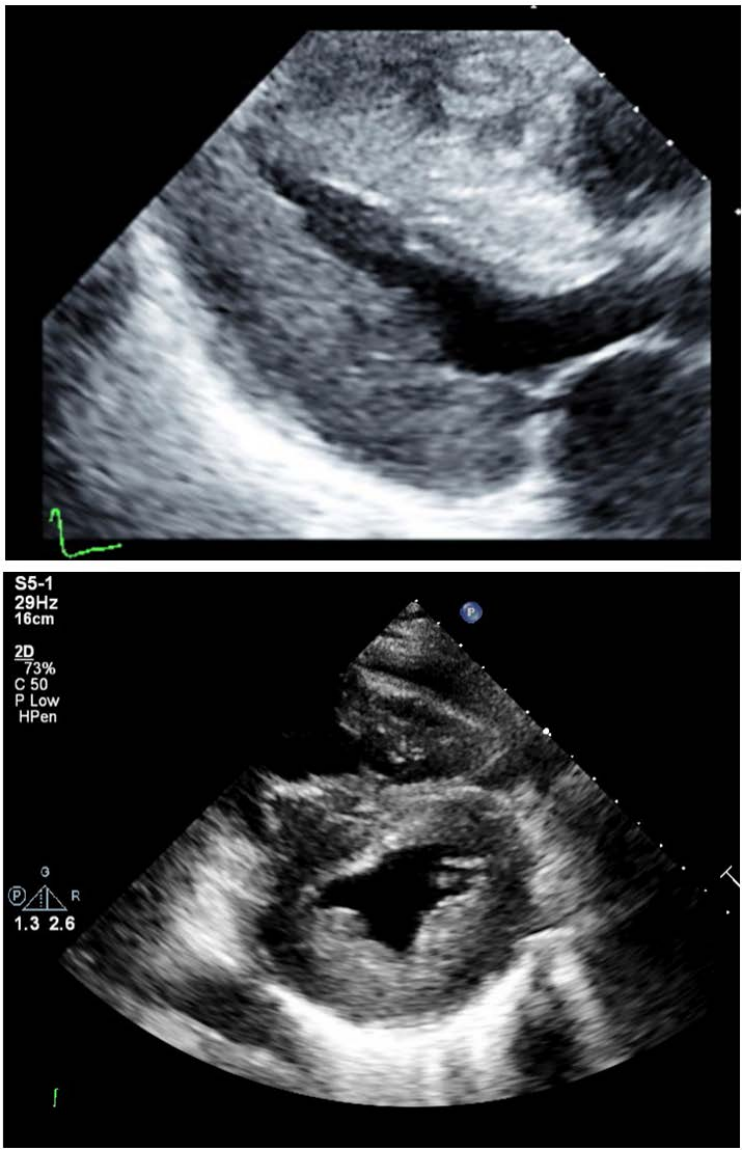

Figure 2. Transthoracic echocardiography revealed severe left ventricular hypertrophy

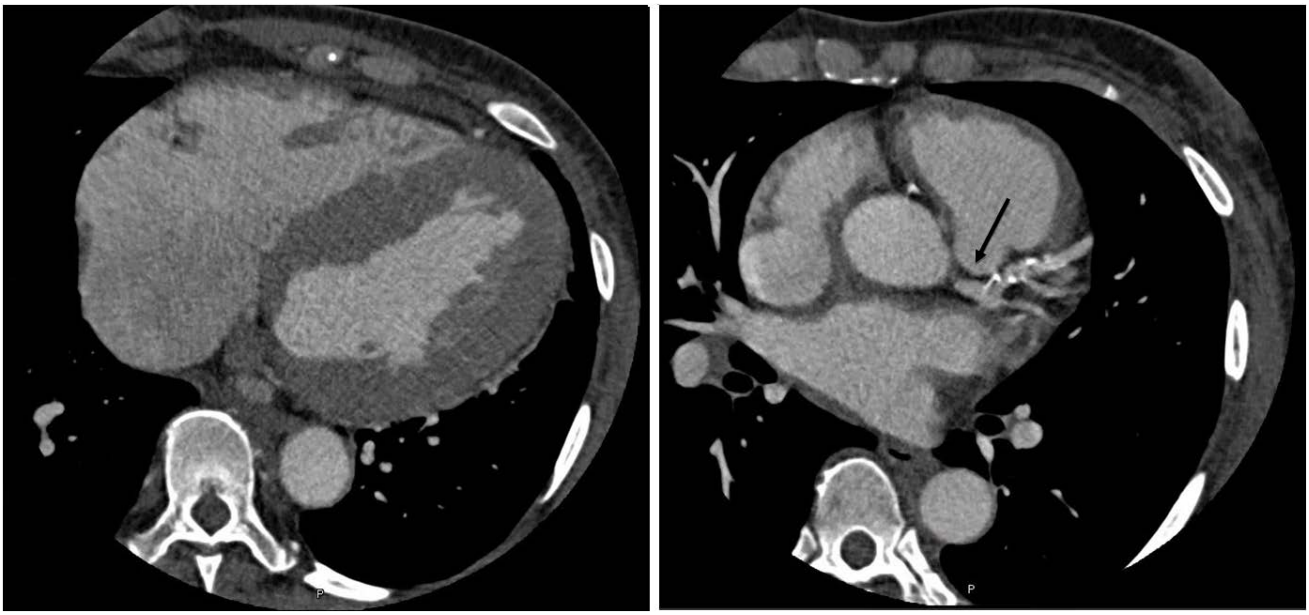

Figure 3. CT coronary angiography which revealed malignant interarterial course of right coronary artery (RCA) has originating from the proximal part left main coronary artery (LMCA) between the aorta and right ventricular outflow. LMCA demonstrated variant trifurcation: ramus intermedius. There is evidence of pulmonary arterial hypertension with enlarged pulmonary artery, right-sided chambers, and thickening of the free wall of both ventricles

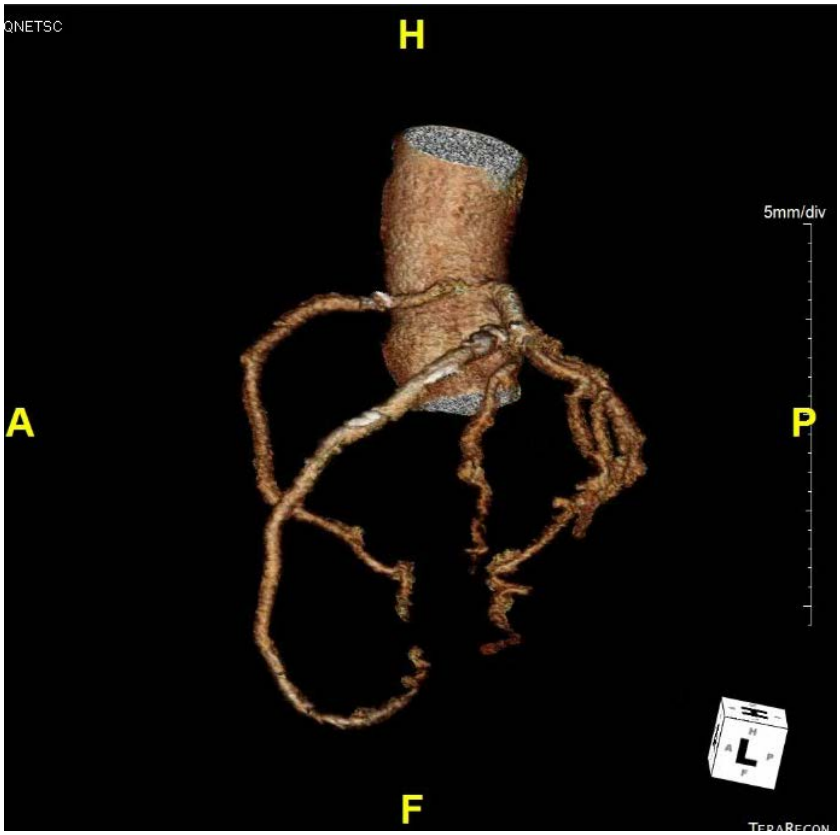

Figure 4. CT coronary angiography 3D Reconstruction of coronary vessels

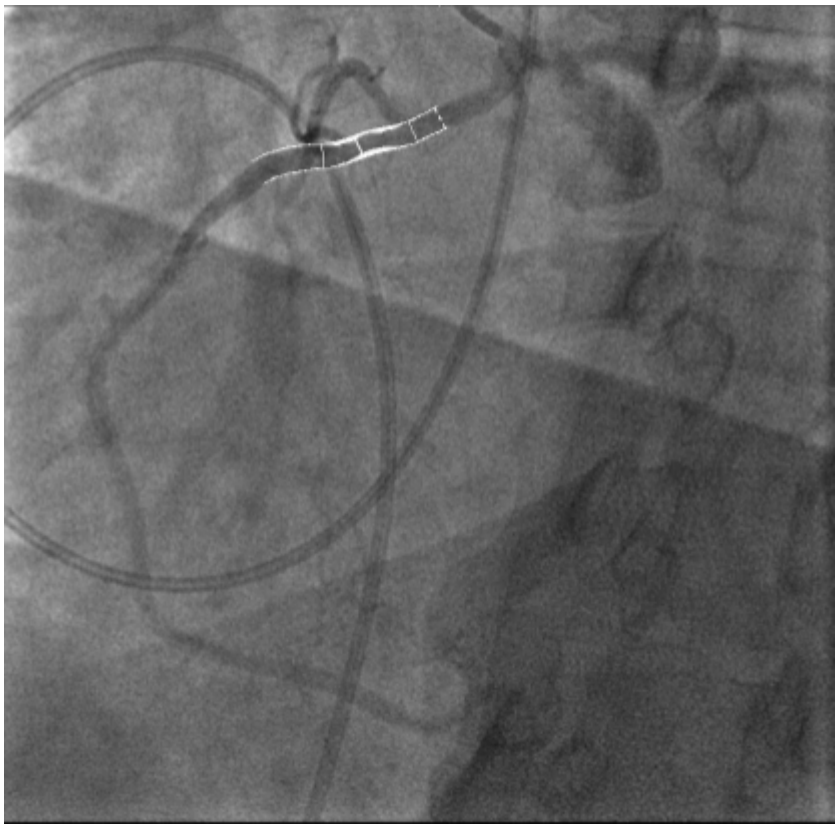

Figure 5. Cardiac catheterization revealed non obstructive coronary artery disease at proximal RCA

3. Discussion

Congenital coronary anomalies are not prevalent, discovered in approximately 1% of the general population undergoing coronary angiography. [9] However, the true incidence is unknown as coronary angiography is not typically routine in these patients without evidence of ischemic heart disease. Prospective angiographic studies, specifically looking for anomalies, suggest the incidence may be as high as 5.6%. [9] Surgical revascularization is

appropriate for patients who have a malignant course of the RCA between the aorta and PA and who manifest ischemia.

What makes our case unique is the combination of the RCA origination from the LMCA, absence of ischemia, and existence of its course between the aorta and RVOT. Other cases reported similar arterial courses but with the pulmonary trunk as one of the surrounding vessels, rather than the RVOT. [3] In both situations, the path taken by the RCA is malignant, which can have serious consequences for the patient. However, our patient remained hemodynamically stable and with a multidisciplinary approach, decided to pursue close outpatient monitoring. As she was unable to achieve her target heart rate with stress echocardiography, she underwent CT angiography, which incidentally revealed the coronary anomaly. This case highlights the importance of considering diagnostic imaging in the elucidation of coronary anatomy, especially in a young female patient undergoing cardiac clearance for a major non-cardiovascular operation. What helped us to decide on conservative management was the accurate three-dimensional visualization of the RCA course as well as its surrounding structures. In other presentations of compromising coronary anomalies, reduction in the coronary blood flow could be due to compression of the coronary artery during its interarterial course, hyper-angulated takeoffs, or slit-like orifices. [12] Ischemic compromise of anomalous coronary arteries manifests as chest pain, loss of consciousness, congestive heart failure, dysrhythmias, and sudden cardiac death, none of which occurred in our case. [13] However, she remains susceptible to any of these complications because of nonsurgical management, and will require routine follow up.

One mechanism of ischemia has been proposed for this anomaly. The great vessels, which dilate during exercise, may externally compress the coronary artery at variable intervals during systole and diastole. [14,15] Hemodynamic status becomes compromised with increasing age due to natural hypertrophy of the aorta and pulmonary trunk; in our patient's case, this hypertrophy was accelerated by her malignant RCA anomaly.

On the other hand, coronary anomalies are often asymptomatic and discovered incidentally, as seen in our patient. Therefore, screening patients for coronary anomalies, regardless of whether there are cardiac risk factors, may be effective as primary and secondary prevention of cardiovascular events.

One study showed that irrespective of modifiable risk factors such as atherosclerotic cardiovascular disease (ASCVD), younger patients who had coronary anomalies were more likely to die of sudden cardiac death compared to their older counterparts. [16] Age was deemed a strong risk factor, in addition to the anomalous course, for clinical outcomes. [16] It is an acceptable Class I indication to operate on the left coronary artery from the opposite cusp that traverses between the aorta and the PA without evidence for ischemia; this is not the case with the RCA from the opposite cusp. [17] Because our case demonstrated no ischemia on cardiac workup, it was decided that the patient should not undergo surgery, which is in keeping with the ACC/AHA guidelines (class I, level B evidence). Even with conservative management for our patient, she will continue to have an elevated risk for acute coronary events compared to her peers who have benign anomalies. [18]

4. Conclusion

Anomalies of coronary arteries exist throughout the general population but are rarely associated with symptoms. They are commonly encountered during routine coronary angiography. They can be categorized into anomalies of origin, trajectory, and destination. It is essential to recognize which anomalies can cause significant clinical consequences, including sudden death. [20] The malignant course of a coronary artery is a poorly understood, infrequently diagnosed condition, which is unrelated to ASCVD. However, it has an excellent prognosis if managed appropriately. Early recognition and treatment is important to achieve favorable outcomes and improve prognosis.

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