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The Single Cusp of Life: A Case Report

Farid Khan[1*], Amit Bansal[1], Laurene Tuider[2], Michail Vitellas[1], Sreekanth Kondareddy[1]

[1]UHS Heart & Vascular Institute, Johnson City, NY, USA
[2]Internal Medicine, UHS Wilson Medical Center, Johnson City, NY, USA
Email: *drfaridkhan@hotmail.com, *farid.khan@nyuhs.org, Sreekanth.Kondareddy@nyuhs.org, Amit.bansal@nyuhs.org, Michail.Vitellas@nyuhs.org, Laurene.tuider@nyuhs.org

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Abstract

Background: Anomalous coronary artery (CAA) is a rare condition occurring in less than 1% of individuals. The most common form consists of the left circumflex artery (LCx) arising from the right coronary sinus. These vascular anomalies have been associated with an increased risk of sudden cardiac death. We present a rare case of an anomalous coronary artery in a patient with all three coronary arteries arising from a shared ostium. **Case presentation:** A 67-year-old transgender Caucasian male with medical history of dyslipidemia and hypertension presented for ischemic workup due to occasional chest pain prior to undergoing male to female gender reassignment surgery. A Regadenosan stress test with SPECT myocardial perfusion imaging revealed moderate sized, moderate intensity perfusion defect reversible in inferolateral wall and fixed in inferior wall. Coronary angiography revealed anomalous origin of the coronary arteries, with all three major coronary arteries arising from a shared, single, ostium originating from the right coronary cusp. Coronary computed tomographic angiography (CCTA) showed a "malignant course" of LAD running between the pulmonary artery and aorta. A conservative management was pursued in view of mild symptoms with close follow ups. **Conclusion:** It is important to evaluate all coronary artery anomalies (CAA) for a "malignant" course due to its associated risk for various cardiac events including sudden cardiac death. Surgical management is indicated in high-risk patients with malignant courses. However, asymptomatic patients and those with mild symptoms with or without malignant course can be followed closely.

Keywords

Coronary Artery Anomaly, Malignant Course, Angiography, Sudden Cardiac Death

1. Introduction

Coronary artery anomalies (CAA) result from developmental and/or congenital

malformation of the coronary arteries during fetal development. These can lead to anomalous location of the coronary vessels and/or ostium [1]. Most common coronary anomalies include abnormal origin, structure or course of the coronary arteries. These abnormalities have been detected in less than 1% on autopsy and 0.27% to 1.6% amongst patients undergoing coronary angiography [2] [3]. There exist numerous variations of CAA, the most common being the left circumflex artery (LCx) arising from the right coronary sinus [4].

CAA are often asymptomatic and are incidentally discovered. However, common symptoms include syncope, anginal chest pain, and sudden cardiac death often related to exercise [4] [5]. High risk "malignant" CAA refers to the anomalous coronary artery when it traverses between the aorta and pulmonary artery. This critical location of the coronary artery predisposes to numerous cardiac events including arrhythmias, myocardial ischemia, and sudden cardiac death [1]. As such, it is crucial to assess and timely diagnose anomalous coronary arteries particularly for the malignant course.

We present an uncommon case of anomalous coronary arteries found incidentally in a patient who was evaluated for chest pain, pertinent workup and recommendations.

2. Case Presentation

A 67-year-old Caucasian transgender male was evaluated for occasional chest pain prior to gender reconstructive surgery. Their chest pain was intermittent and localized over lower sternal region with no association with physical activity. There were no accompanying palpitations, syncope or dyspnea. Their medical history was remarkable for hypertension, dyslipidemia, and former tobacco smoking. Their chronic medications consisted of simvastatin, aspirin, and spironolactone. They were on estradiol as part of gender change therapy. Family history was remarkable for coronary artery disease in the mother.

On physical examination, they were afebrile and not in acute distress. Their blood pressure was 139/82mmHg, heart rate 76 beats/minute and normal respiratory rate as well saturations. Cardiovascular examination revealed a normal and regular heart rate, normal heart sounds with no jugular venous distension or extremity edema. Lungs were clear to auscultation with nontender chest wall.

Initial workup including electrocardiogram (EKG) showed normal sinus rhythm, rate 70/min and no ST segment changes suggestive of ischemia (**Figure 1**). Laboratory workup including complete blood count, metabolic panel and cardiac enzymes including troponin were within normal limits. Transthoracic echocardiogram (TTE) revealed normal left ventricular systolic and diastolic functions without wall motion abnormality. However, regadenosan stress test with single photon emission computed tomographic (SPECT) myocardial perfusion imaging revealed a moderate sized, moderate intensity perfusion defect reversible in the inferolateral wall and fixed in the inferior wall. Due to the abnormal stress test, the patient underwent an invasive coronary angiography that revealed only a mild non-obstructive coronary artery disease; however, an anomalous origin of

coronary arteries was observed. All three major coronary arteries were noted to arise from a shared, single, ostium originating from the right coronary cusp (RCC) (Figure 2). Subsequently, coronary computed tomographic angiography (CCTA) was performed to evaluate for vascular anatomy and particularly for "malignant course" of the coronary arteries. CCTA confirmed the right coronary artery (RCA), the left anterior descending artery (LAD), and LCx originating from the RCC. Additionally, the LAD had a malignant course running between the pulmonary artery and ascending aorta (Figure 3). No obvious left main artery was seen.

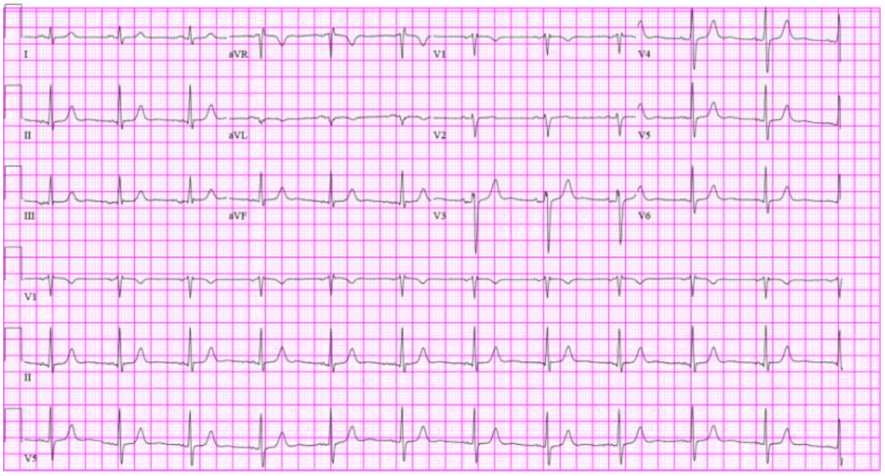

Figure 1. 12-lead EKG revealing normal sinus rhythm with no ST segment elevation or depression.

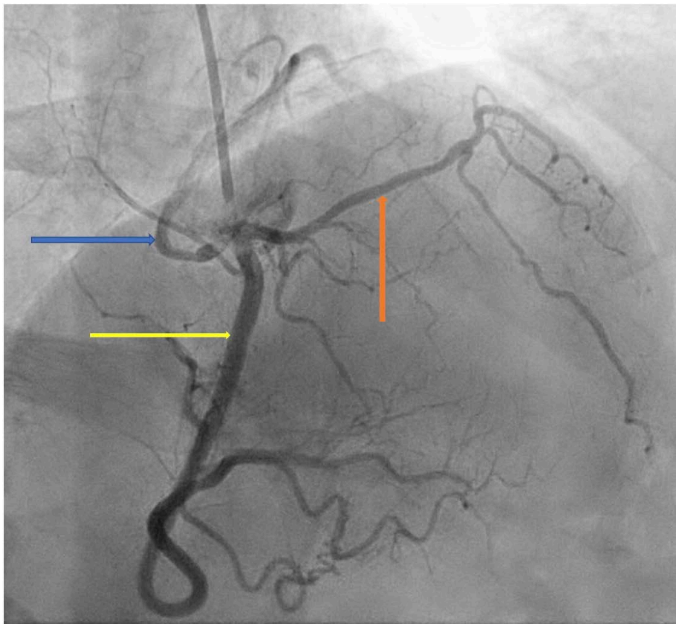

Figure 2. In this right-anterior-oblique (RAO) angle 27 and cranial angle 33, coronary angiogram showing all three major coronary arteries arising from a shared, single ostium in the right coronary cup. Right coronary artery (yellow arrow), circumflex artery (orange arrow), and anterior descending artery (blue arrow).

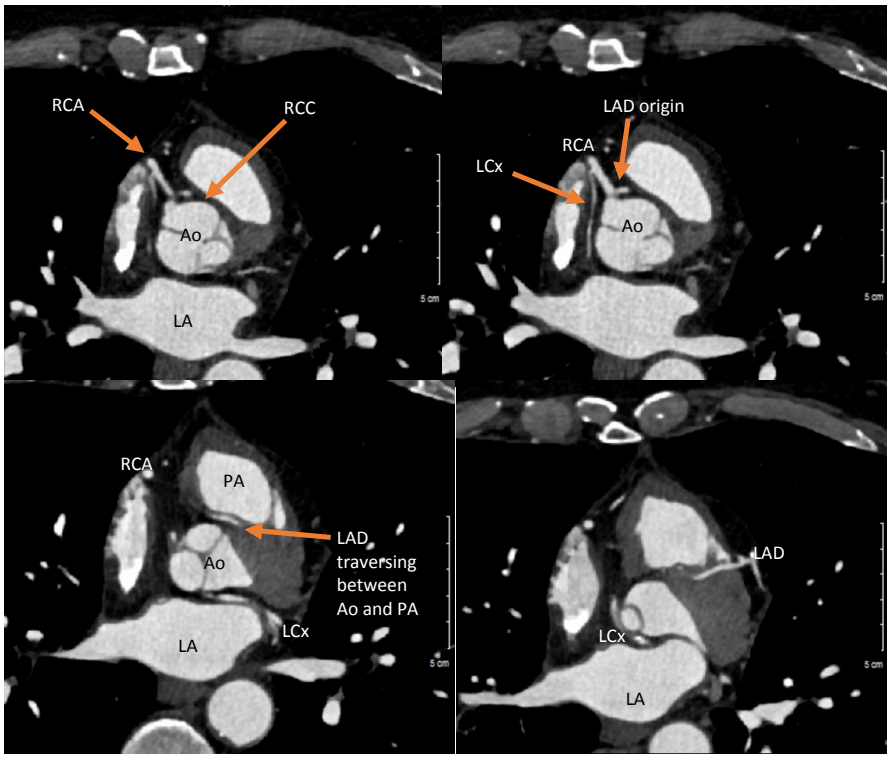

Figure 3. CT coronary arteries showing right coronary artery (RCA), left anterior descending (LAD) artery and left circumflex (LCx) artery having anomalous origin arising from right coronary cusp (RCC) and shared ostium. LCx has retro-aortic course while LAD traverses between aorta (Ao) and pulmonary artery (PA) before reaching interventricular groove. LA: Left atrium

Although the course of the LAD was "malignant", due to the patient's mild symptoms and nonobstructive atherosclerotic disease decision was made to pursue conservative management with the plan of close follow ups in the cardiology office.

3. Discussion

Our patient was evaluated for occasional chest pain prior to gender reconstructive surgery. An abnormal myocardial perfusion stress test, and personal as well as family history of cardiovascular risk factors for CAD prompted for invasive ischemic workup. Subsequent coronary angiogram and CCTA revealed anomalous origin of the coronary arteries, with all three major coronary arteries arising from a shared, single, ostium that originated from RCC. Interestingly, no left main artery was seen. CCTA revealed a malignant course of LAD traversing between pulmonary artery and ascending aorta.

Anomalous coronary arteries are infrequently observed in a small subset of the population, and are often incidentally discovered. CAA are uncommon with an overall prevalence of around 1% based on the studies involving coronary angiography and autopsy [2] [3] [6]. Anomalous LCx and LAD originating from RCC carry a known prevalence of 0.7% and 0.15% respectively. However, all

three coronary arteries originating from a single and shared cusp is extremely rare.

A retrospective report by Eckart *et al.* [7] reviewed 6.3 million cases of sudden death in military recruits ages 18 - 35 to assess for sudden cardiac death over the course of 25 years. The study found that of the 126 nontraumatic deaths, cardiac abnormalities accounted for 64 deaths (51%), of which 39 (61%) were attributed to coronary artery abnormalities and about half of these patients had prodromal symptoms. CAA was present in 21 of the 64 recruits and these recruits were found to have LCx arising from the right anterior sinus of Valsalva with a malignant course between the aorta and pulmonary artery. Yuksel *et al.* [8] reported CAA in 0.29% (48 of 16,573) patients noted on retrospective analysis of the coronary angiograms at their center. LCx originating from RCC or right sinus of Valsalva was noted to be the most common coronary anomaly (28 of 48 patients).

Two major categories of CAA considered as high-risk include coronary artery travelling between the aorta and pulmonary trunk, and the left main coronary artery originating from the RCC. The sudden death in patients with a malignant coronary anomaly is close to 80% [9]. Therefore, it becomes critical to recognize their course with respect to the surrounding structures. CAA with acute-angle take off, and ostial abnormalities such as slit-like orifice and beak-shaped orifice are other high-risk categories [3] [10].

Our case has several unique aspects. All major coronary arteries originated from RCC sharing a single ostium; a rarely observed coronary anomaly. Evidence of LAD traversing between the aorta and pulmonary trunk establishes its high-risk or malignant category. A recent report observed all 3 coronary arteries with shared single ostium arising from RCC; however, the course was non-malignant, and patient was treated conservatively [3]. Another report noted all major vessels sharing a single ostium that originated from the right coronary sinus [11].

Our patient had occasional chest pain, rather atypical, and had only nonobstructive CAD as noted on cardiac catheterization. He had no history of syncope, arrhythmias or cardiomyopathy. Therefore, a conservative approach was considered with the plan of regular outpatient follow-ups.

Although, there are several observational and descriptive studies on CAA, there is a lack of guidelines for screening patients for this condition, particularly for high-risk anomalies. Asymptomatic patients can become symptomatic and at times the initial presentation may be a sudden death. In most cases, CAA is diagnosed incidentally particularly on cardiac catheterization; however, if it does reveal coronary anomaly, a CCTA should follow to further define its anatomy and severity [12]. CCTA is crucial for delineating the travel course of the artery including malignant course. It also identifies geometric orientation and evaluation of the origin from a particular ostium or cusp. Identifying these is vital for devising treatment strategy in symptomatic patients.

Management of asymptomatic patients varies widely and is not well defined [4]. At this point in time, there is nonexistence of a standardized approach and/or guideline for such patients. Often, these patients are managed conservatively with close monitoring while surgical intervention is opted in symptomatic patients with arrhythmia and/or ischemia.

Although our report has significant inherent limitations by design, we reason that prompt workup including CCTA should be considered to evaluate for high-risk coronary anomalies in patients with suspicion of CAA, irrespective of symptoms. We reason that our experience will add to the slowly growing literature on this subject.

4. Conclusion

We recommend a prompt evaluation of coronary artery anomalies for "malignant" or high-risk course by performing CCTA imaging study. In these patients, invasive coronary angiography may be considered to evaluate for ischemic coronary disease due to underlying high risk of sudden cardiac death. Asymptomatic patients can be closely followed with treatment options explored beforehand.

Informed Consent

The case was reviewed by the Institutional Review Board and informed consent was obtained from the patient.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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