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A Rare Variant of an Anomalous Origin of the Left Coronary Artery, the Great Masquerader of Angina?

Case Report

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Abstract

Anomalous aortic origin of the coronary arteries (AAOCA) is a rare set of anatomical variants that can present with symptoms such as angina and dyspnea on exertion with complications including acute coronary syndrome and sudden cardiac death. This case demonstrates a patient presenting with classic anginal symptoms with an incidental finding of one of the rarest variants and its complexity in cannulation: a left main coronary artery arising from the non-coronary cusp. Despite insignificant left coronary disease on coronary angiography, we had a high index of suspicion for underlying coronary vasospasm as a major contributor to her recurrent symptoms. AAOCA is an anatomical finding that warrants clinician awareness and close follow-up with recommendation for surgery if symptomatic.

keywords: Anomalous coronary artery; Percutaneous coronary intervention; Angina; Sudden Cardiac death

Learning Objectives

a. AAOCA is a rare anatomical variant with significant risk for potentially fatal complications including sudden cardiac death (SCD), myocardial infarction, and stable angina.

b. In patients with AAOCA, a high index of suspicion for vasospasm should be present when there is a recurrence of anginal symptoms despite complete revascularization and guideline directed medical therapy.

c. Symptomatic AAOCA patients should be referred for surgical repair given the risk of potentially fatal sequalae

Background

Anomalous aortic origin of the coronary arteries (AAOCA) is a rare set of anatomical anomalies with a prevalence of 0.31% during invasive and noninvasive diagnostic testing. It is often found incidentally during cardiac testing including transthoracic echo, coronary angiogram, MRA, and invasive coronary angiography [1]. AAOCA is further classified based upon the artery and by course subtypes such as interarterial, subpulmonic (intraconal or intraseptal), pre-pulmonic, retroaortic, or retrocardiac [1]. 50% of patients with AAOCA are symptomatic (i.e., angina, dyspnea on exertion) with over 31% having sudden cardiac death and 8% with myocardial infarction [1]. We report a case of a symptomatic patient presenting with an ultrarare subvariant of AAOCA: a left main coronary artery arising from the non-coronary cusp with an interarterial course. A great deal of technical skill is required to engage the left coronary artery in these patients with AAOCA of the noncoronary cusp as described. This case serves to highlight the complexity and multimodality approach in assessing patients with suspected AAOCA. Further, close monitoring of symptoms is warranted given the high risk of morbidity and mortality with recommendation for surgery if symptomatic

Case Presentation

An 86-year-old female presented to her PCP with complaints of worsening periods of shortness of breath, chest discomfort on exertion and palpitations on exertion. She was referred to the outpatient

clinic for ischemic workup and evaluation. Physical exam revealed an elderly woman, in no acute distress; vital signs were stable with a blood pressure 120/70mm Hg, pulse 61 beats/min, and resting oxygen saturation of 94% on room air. No jugular venous distention was noted however 1+ bilateral lower extremity pitting edema was appreciated on exam. Cardiac auscultation over the right chest revealed a III/VI systolic ejection murmur. Pulmonary exam was unremarkable, as noted to have symmetric excursion of her chest with inspiration and breath sounds throughout the lung fields with no crackles, wheezing, or rhonchi. The patient's medical history was notable for hypertension, hyperlipidemia, paroxysmal atrial fibrillation, heart failure with preserved ejection fraction, and noninsulin dependent diabetes mellitus. When she presented to our outpatient clinic, she was on amiodarone 200mg once a day, metformin 1000mg twice a day, metoprolol tartrate 50mg two times a day, sitagliptin 100mg once a day, atorvastatin 80mg once a day, and warfarin 5mg once a day.

Investigations

A resting electrocardiogram demonstrated normal sinus rhythm with left ventricular hypertrophy; nonspecific ST segment changes with inferior wall perfusion defects were noted on pharmacological stress testing. Transthoracic echocardiogram revealed moderate aortic stenosis with a peak gradient of 20mmHg and mild mitral stenosis with grade 3 diastolic dysfunction. The patient was referred for a right and left cardiac catheterization for pre-TAVR and ischemic workup. Right heart catheterizations findings revealed pulmonary pressures of 60/30mmHg with a cardiac output by thermodilution of 4L/min. Gradient across the AV was found to be consistent with the echocardiogram at 20mmHg significant for moderate aortic stenosis. On diagnostic catheterization, attempts at cannulating the left main system were met with difficulty despite numerous attempts using an FL4, FL3.5, CLS3 guide, JL3, AL 0.75, AL 1, and AR 1 all failed. The right coronary artery was easily visualized with an FR4 guide catheter and revealed a 90% stenotic mid RCA lesion. The patient was taken off the table for further non-invasive investigation. A Cardiac CT angiography was subsequently performed (Figures 1A and 1B), which revealed a retrocardiac left coronary artery arising from the noncoronary cusp. With a better appreciation of the left coronary artery anatomy, a second diagnostic catheterization was performed the following day. Initially, a pigtail was placed in the non-coronary cusp and a non-selective angiogram was performed Figure 2A). Subsequently, the LM was selectively cannulized utilizing a left internal mammary artery (LIMA) guide catheter. The LIMA guide catheter was pushed into the noncoronary cusp leaflet for structural support and then curved upward to selectively cannulate the left main coronary artery which had a superior take-off (Figure 2B). The left coronary system revealed insignificant disease on angiography as well as intravascular ultrasound (IVUS). The RCA was successfully treated with a drug eluting stent (DES). Although the transthoracic echocardiography demonstrated severe aortic stenosis, thermodilution data and utilization of the Gorlin's formula demonstrated moderate AS.

Differential Diagnosis

Stable angina, severe aortic stenosis, congestive heart failure, pulmonary hypertension, pulmonary embolism

Outcome and Follow Up

Following successful placement of a drug eluting stent in the RCA, the patient was discharged home on post procedural day 1, reporting no symptoms at her two-week and three-month outpatient follow up appointments. Her symptoms were carefully monitored given her anomalous left coronary system. Eight months later, her symptoms and evidence of ischemia returned prompting further evaluation. She was referred for surgical evaluation, however the patient deferred further workup and interventions.

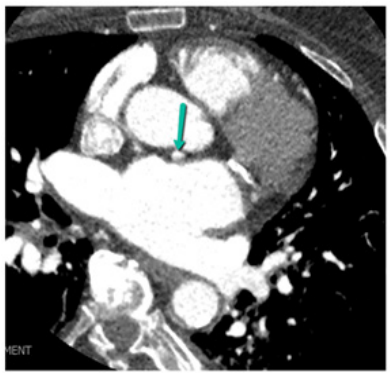

Figure 1: Cardiac CT angiography demonstrating the left main coronary artery originating from the noncoronary cusp in a retroaortic fashion (arrow).

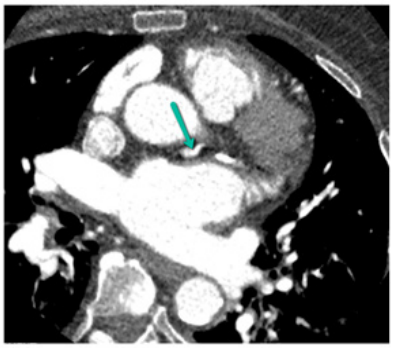

Figure 1B: Left main coronary artery continuing to traverse along the aortopulmonary window.

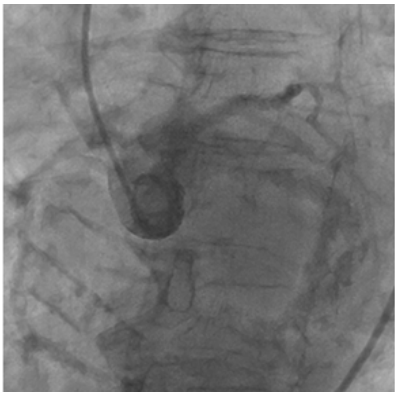

Figure 2A: Non-selective RAO caudal angiogram of the aortic cusps with visualization of the left main coronary artery arising from the posterior noncoronary cusp.

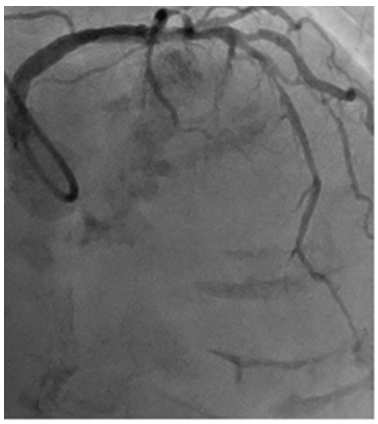

Figure 2B: Selective RAO Caudal angiogram of the left main coronary artery using a LIMA guide catheter with the non-coronary cusp serving as support.

Discussion

In summary, this patient presented with symptoms of classic stable angina secondary to significant disease in the right coronary artery with return of her symptoms several months later. Interestingly, there was difficulty in engading the left coronary system reuqiring Coronary CTA which revealed an ultra rare anatomical finding: an anomalous left main coronary artery arising from the non-coronary cusp (AAOCA). The question arose of whether the patient's presentation was attributed to the disease of her RCA and potential vasospasm of the anomalous left coronary system? Further, does this patient warrant future monitoring of her left coronary system? The return of the patient's symptoms months later raised our clinical suspicion of coronary vasospasm of the left main coronary artery. Our thought process was supported by documented cases of patients presenting with angina and vasospasm at rest secondary to AAOCA [2]. No structured guidelines are present regarding diagnostic monitoring for this ultra-rare and high-risk subset of patients.

Diagnostic workup for AAOCA should include CT Angiography of the chest or Cardiac MRA, a Class 1 recommendation noted by ACC. As seen in the described CT images of our case report, CT angiography provides valuable information to the operator when cannulating the left main coronary artery [1]. Treatment for symptomatic patients involves surgical repair with reported symptomatic relief in 97% of patients [1].

Conflicts of Interest

This manuscript has not been published in whole or in part, nor has it been considered for publication elsewhere. The authors do not have any conflict of interests to declare, and all authors had access to the case's data and equal contribution.

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