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Case Report

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Intra-Atrial Right Coronary Artery and Anomalous Origin of Left Circumflex Artery Found Concurrently

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Abstract

Coronary artery anomalies (CAAs) are known to be anatomical aberrations in the origin and structure. Due to the diverse anatomical variants, surgeons and angiographers have struggled when faced with patients who have CAA. To frame the complicated issues surrounding CAA, we present a case of a young patient found to have two CAAs, concurrently on coronary computed tomography angiography (CCTA), while in the emergency room being evaluated for chest pain. Patient was medically observed without any invasive procedure since he was deemed to have low cardiovascular risk. Subsequently, literature on prevalence, as well as high risk findings are reviewed. Further studies to evaluate pharmacological, angiographic, and surgical interventions may have additional benefit for both patients and practitioners. Our aim is to help shed the light on difficulties cardiologists are facing during angiography. Additionally, our paper offers some guidance for how to evaluate and follow patients with similar findings into the future.

Keywords: Coronary artery anomalies; Coronary angiography; Chest pain; Coronary artery disease; Congenital

Introduction

Coronary artery anomalies (CAAs) are known to be anatomical aberrations in the origin, structure, and course of the epicardial arteries. Classically, CAAs are rare and are incidentally found either during post-mortem autopsy or open-heart surgery. In recent years, due to the increased usage of advanced cardiac imaging, the incidence of CAA is increasing. More patients are being diagnosed with CAA, either as a cause of their symptoms or incidentally. While studies estimate CAA incidence to be about 1% of the general population, many patients may live asymptomatic lives with these anomalies and never be diagnosed [1, 2]. Studies estimate about 80% of CAAs to be benign, with about 20% causing symptoms [2].

Over the years, surgeons and angiographers alike have struggled when faced with patients who have CAA [3]. CAAs have been considered as possible causes for dyspnea, angina, and syncope [2]. Serious complications have been documented to include myocardial infarction (MI), congestive heart failure, cardiac arrhythmias, or sudden death [2, 4]. Focus has also been given to CAA as a cause of sudden death in the young [5] after hypertrophic cardiomyopathy and idiopathic left ventricular hypertrophy [6].

To frame these complicated issues surrounding CAA, we present the case of a young patient found to have two CAAs, concurrently on coronary computed tomography angiography (CCTA), while in the emergency room being evaluated for chest pain.

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Case Report

Investigations

A 30-year-old healthy male presented to the emergency room for intermittent sharp left-sided chest pain that radiated to his left arm. Patient reported that the pain started 1 week prior to presentation, and it was brought on by exercise and relieved with rest. He denied any past medical history or taking any medications. Additionally, he denied the use of cigarettes, alcohol, or illicit drug. Family history was irrelevant for any cardiac disease. Vital signs on admission were remarkable for blood pressure of 180/104 mm Hg, pulse of 71 beats per minute, and respiratory rate of 16 breath per minute. Physical exam was unremarkable. Electrocardiogram (ECG) on admission showed normal sinus rhythm with no significant ST-T wave abnormalities. Troponin T on admission were negative on two separate occasions. All other laboratory findings were within normal limits. CCTA was ordered as for initial noninvasive evaluation of patient's chest pain.

Diagnosis

A CCTA was done for further evaluation, showing a short

segment of intra-atrial course in distal right coronary artery (RCA). Incidental findings on CCTA also revealed that origin of the left circumflex artery emerged from the right coronary cusp and followed a retro-aortic course without evidence of compression. Additionally, there was a focal shallow myocardial bridging in the mid left anterior descending artery (LAD). For a better assessment of any possible blood flow compromise at effort, the patient underwent nuclear stress test that came back negative for a reversible defect.

Treatment

Patient was found to be in the coronary artery disease reporting and data system (CAD-RADS) group 1 with minimal nonobstructive coronary artery disease. He was observed for 24 h with no episodes of chest pain or abnormal events on telemetry monitors. He was treated medically with low-dose aspirin and pain management.

Follow-up and outcomes

The patient was discharged home with instructions to return to the emergency department if symptoms recur and to follow up with cardiology as outpatient. The patient did not require any further intervention later.

Discussion

Many challenges exist in defining anomalous coronary arteries as there are expected variations to normal anatomy. Since the development of detailed and noninvasive imaging techniques, more efforts have been launched into describing and categorizing these anomalies with precise anatomy description. As Angelini had first described and other authors seemingly agree, an "anomaly is observed in less than 1% of the general population" and anything more is considered a typical variation on a normal distribution [5]. However, there is no strict classification system in defining these anomalies. Given the increasing propensity for noninvasive imaging, such as CCTA, our definitions of an anomaly may change as more abnormalities are found. CCTA was shown to be a high-quality study for diagnosing CAA [7].

Most common anomaly is the origin of left circumflex artery from RCA or right coronary cusp, as seen in our patient. According to the study of angiographies by Yuksel et al, which was performed on approximately 16,000 patients, 0.29% were found to have anomalous coronary arteries. Of those patients, 58% were found to have the origin of left circumflex from RCA (17 patients) or right sinus of Valsalva (11 patients). The study of Yuksel et al is a single-center study, in which two cardiologist retrospectively reviewed angiograms over a 10-year period to determine the prevalence of different coronary anomalies [8]. While angiographies are an excellent diagnostic tool and remain gold standard in determining coronary artery disease state, it is still considered an invasive test and carries many risks including but not limited to bleeding, radiation risk, contrast reaction, arrhythmia, and infection.

Less than a year later, another study was published by Opolski et al who performed approximately 9,000 CCTAs at a single center over a 3-year period; and these images were reviewed retrospectively for coronary anomalies. The results were similar to the study of Yuksel et al in that the most common anomaly is left circumflex artery with right-sided origin (0.37%), and then secondly, RCA arising from left-sided origin (0.23%) [8]. Due to the ability to reconstruct a three-dimensional image of the heart from performing CCTA, this study had additional information on the course of these anomalous arteries instead of the origin site alone as seen on angiographies. The intra-atrial course of RCA was found in 0.15% of all patients with CCTA; and in most of these cases, the RCA traverses through the inferolateral portion of right atrium. The study also found that the intra-atrial course of RCA is most often associated with anomalous left circumflex artery with right-sided origin and an intramuscular course of the LAD [9]. Our patient had all three characteristics. While coronary anomalies independently are uncommon, it is even more rare to find multiple anomalies in one patient. The clinical course and symptom management remains a mystery in patients with multiple coronary anomalies due to the lack of compiled data on adverse cardiac-related events and overall prognosis. In a single-center retrospective study done to evaluate CAA in Turkish populations, high take-off coronary was the most common anomaly at 0.77% [7].

Some high-risk anomalies that are strongly associated with sudden cardiac death are in the class of anomalous coronary artery from the opposite sinus (ACAOS). When the right main or left main coronary artery arise from the contralateral sinus, the risk of compression, stenosis and arterial hypoplasia is more pronounced [5]. These patients can have more serious complications such as MI or sudden cardiac death from ventricular arrhythmias. In 2004, Eckart et al [10] published a study reviewing autopsies of sudden death in young adults recruited for intense military training over a 25-year period. This study was significant in that it found that 50% of deaths were identified to be cardiac in origin. Of this cardiac-related death, 33% were due to ACAOS which was found on autopsy. In these patients that expired due to ACAOS (21 patients), it was suspected that half of them had symptoms such as chest pain, dyspnea, or syncope but may not have been forthright due to fear of disqualification from the military training [10]. In comparison, only 8% of sudden cardiac deaths were due to hypertrophic cardiomyopathy, but this can be accounted for the pre-screening testing the military recruits undergo for hypertrophic cardiomyopathy characteristics via baseline ECG and obtaining family history.

Given the aforementioned studies detailing the incidence of CAA, which are summarized in Table 1 [7-10], these anatomical aberrations are an under-recognized cause of cardiac morbidity and mortality. The risk of sudden cardiac death due to coronary artery anomalies is highly unpredictable and heavily dependent on many factors, most importantly the aberrant anatomy and course of the coronary arteries. Currently, the treatment is based on expert opinion, type of CAA, and clinical signs and symptoms. For asymptomatic patients, the expert consensus of practitioners is to manage conservatively and allow these patients to participate in competitive athletic

Table 1. Prevalence Data for Coronary Artery Anomalies With Various Imaging Studies

Author, year	Modality	Total number	Prevalence of total coronary anomalies/(percentage)	Most common anomaly	Prevalence of most common anomaly/(percentage of anomalies)
Yuskel et al, 2013 [8]	Angiography	16,573	48/(0.003%)	Left circumflex from right sinus Valsalva	28/(58.3%)
Opolski et al, 2014 [9]	Coronary CTA	9,284	76/(0.008%)	Left circumflex from right-sided origin	21/(27.6%)
Yesilyurt et al, 2017 [7]	Coronary CTA	2,973	79/(2.65%)	High take-off coronary artery	23/(0.77%)
Eckart et al, 2011 [10]	Autopsies	Total recruits n = 6,290,000	21/(0.0003%)	Left coronary artery from right sinus of Valsalva	21/(100%)
		Cardiac-related deaths n = 64	21 (33%)		

CTA: computed tomography angiography.

sports with informed consent. For patients experiencing clinical signs and symptoms of ischemia with a high-risk anomaly, many practitioners recommend limiting exertional activity and consider surgical intervention. Some of the surgical interventions described are unroofing, ostioplasty, ostial translocation, and bypass grafting [11]. A major limitation in current research and expert consensus is our inability to isolate and appropriately manage the asymptomatic patients with high-risk CAA that have sudden cardiac deaths without any preceding signs or symptoms of ischemia. Similarly, we are presenting a sole case of CCA that we rarely encounter in practice which makes it hard for us to conduct whether a retrospective or even a randomized clinical trial for a better understanding and risk stratification of such disease.

Learning points

CCAs are sparsely described in literature because of the rarity of this entity, which makes it hard for cardiologist to classify these patients based on their anatomical variants. For now, it is important to classify these patients based on their symptomatology as well as other risk factors, in conjunction with their anatomical variant. With the advancement of noninvasive cardiac imaging, we might be able to identify more anomalies and maybe a scoring system for us to better manage these patients. Despite the advancements in the field of CAA, there is a long road ahead in order to understand the full impact of CAA. For cardiologists and internists alike to properly understand the natural history and physiology of CAA we call for large multi-center prospective studies to be conducted. These studies should evaluate the natural history of CAA, helping practitioners to determine which anomalies are of highest risk for future cardiac events. Finally, we also call for cardiology societies to consider releasing guidelines about who and when to screen for CAA.

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Conflict of Interest

The authors declare that there is no conflict of interest.

Informed Consent

Informed consent was obtained.

Author Contributions

All authors have made contributions to writing this case report. Michael Cinelli, Hamfreth S. Rahming and Marc Assaad helped drafting the manuscript. Peter C. Olson and Michael Cinelli are responsible for the concept and editing. Jonathan Spagnola and James C. Lafferty are responsible for the editing and supervision.

Data Availability

The authors declare that data supporting the findings of this study are available within the article.

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