

Anomalous Origins of All Three Coronary Arteries from Separate Ostia within the Right Aortic Cusp: A Case Report and Review of the Literature

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Abstract Coronary artery anomalies are congenital defects which are found incidentally or after cardiac events. While these are rare abnormalities with the majority of patients remain asymptomatic and largely undiagnosed, it remains to be a major cause of sudden cardiac death (SCD). Anomalous origin of left coronary artery (ALCA) from the opposite right aortic sinus is extremely rare with less than 100 cases reported to-date. These patients are at increased risk for significant cardiac events, including SCD. In this report, we present a 48-year-old man with hypertension and marijuana use who was admitted initially with multi-lobar pneumonia and acute kidney injury, developed respiratory failure and sustained ST elevation myocardial infarction (STEMI). Coronary angiography demonstrated anomalous origin of all three main coronary arteries arising from right aortic sinus. In this report we also discuss the genesis of this rare and potentially fatal congenital abnormality and we highlight the diagnostic and management strategies available to-date.

Keywords: *anomalous origin of coronary arteries, anomalous left coronary artery, right coronary artery, st- elevation myocardial infarction*

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1. Introduction

Anomalous origin of coronary arteries (ACA) constitute a group of morphological abnormalities occurring congenitally. These are relatively uncommon and depending on the type of anatomic variant, the clinical manifestations range from being asymptomatic to typical chest pain and sudden cardiac death [1]. The incidence of ACA has been found to be between 0.3 % to 1.3% from retrospective angiographic studies [2,3,4]. The subtypes vary based on origin, location, intrinsic anatomy and termination [1]. Anomalous origin of a coronary artery from an opposite sinus (ACAOS) is a subtype where either of the three coronary arteries differ in their normal anatomy and arise from opposite aortic sinuses. These have an incidence of less than 0.1% [3]. Of which the left coronary arteries arising from the right aortic sinus known as anomalous left coronary artery(ALCA) is much rarer and poses higher risk. Here in, we report a case of distal right coronary artery (RCA) ST elevation myocardial

infarction (STEMI) in which ACAOS was incidentally noted. And, all the three main coronary arteries (Left anterior descending, Left circumflex and right coronary artery) arose from separate ostia within the right aortic sinus.

2. Case Report

A 48 year-old male with a known history of hypertension and marijuana use presented with low-grade fevers, non-productive cough, dyspnea, pleuritic chest pain and diarrhea of 3 days duration. Patient was initially found with leukocytosis of 12.3 cells/mm³, lactic acidosis, transaminitis, elevated creatinine to 6.3 significantly increased from baseline. A chest X-ray demonstrated multi-lobar pneumonia and serology returned positive for Influenza. Patient was initially hemodynamically stable in the emergency room and was admitted to the medical floors for further management. However, oxygen saturation began to drop necessitating supplemental oxygen therapy and further investigation.

Venous doppler ultrasound of lower extremities revealed acute thrombosis of left soleal and peroneal veins. Patient was started on full dose anticoagulation for a lower extremity thrombosis. Subsequent ventilation perfusion scan showed intermediate probability (20-80%) of pulmonary embolism. A sputum culture growth revealed methicillin-resistant staphylococcus aureus bacteria. Patient was initiated with oseltamivir, vancomycin and piperacillin-tazobactam. Upon experiencing worsening respiratory distress, the patient was transferred to the intensive care unit where he was intubated and placed on cardiac monitor.

While being monitored in the intensive care unit, telemetry showed ST elevations. 12 lead EKG revealed ST elevations in the inferior leads (II, III, aVF) as well as V5-V6 with reciprocal ST depression in leads I and aVL. STEMI alert was immediately activated. Troponin I drawn prior to cardiac catheterization was >200 ng/mL.

Subsequent transthoracic echocardiogram revealed mild systolic left ventricular dysfunction (ejection fraction - 45%) with mild to moderate inferior and posterior wall hypokinesia.

Left heart cardiac catheterization with coronary angiography was performed with a right radial approach. Coronary angiography revealed anomalous origins of the left circumflex (LCX) and left anterior descending (LAD) arteries arising from the right coronary cusp (RCC) (Figure 1B & Figure 1C), with 99% thrombotic occlusion of the distal right coronary artery was identified as the culprit lesion (Figure 1D). Percutaneous coronary intervention with a 4 x 30 mm drug-eluting stent was performed. Post STEMI hospital course was complicated by progressive respiratory failure requiring tracheostomy and ventilator-associated pneumonia. Patient was eventually discharged on appropriate medical therapy to long-term rehabilitation center.

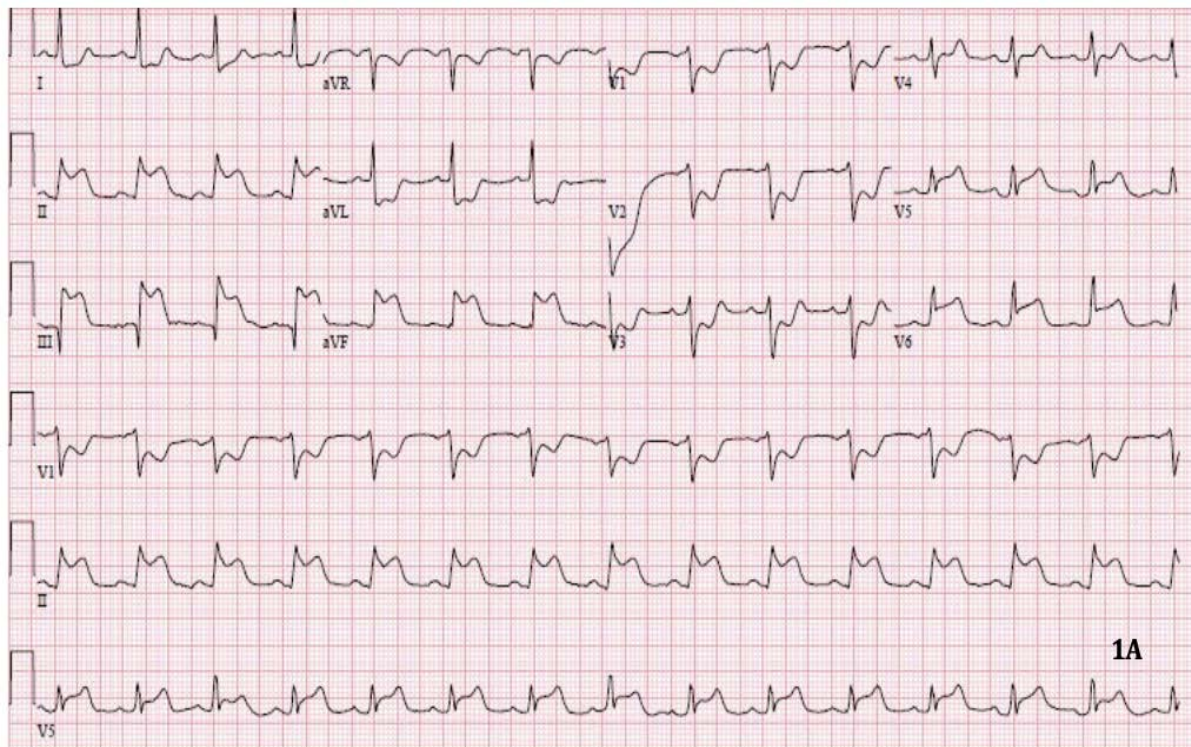


Fig. 1A: ST segment elevation in leads II, III, aVF, V5 and V6. **Fig. 1B:** Anomalous origin of the left anterior descending artery arising from the right coronary cusp. **Fig. 1C:** Anomalous origin of the left circumflex artery arising from the right coronary cusp. **Fig. 1D:** Thrombotic occlusion of the distal right coronary artery

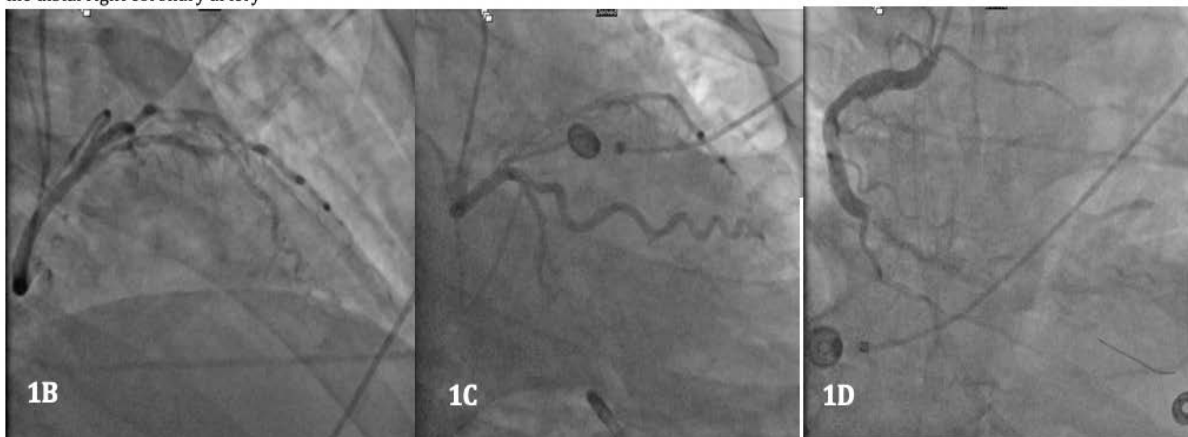


Figure 1.

3. Discussion

Amongst the coronary artery anomalies, ACAOS is of prime focus in our case. The incidence of all three coronary arteries (LAD,LCX,RCA) arising from the same right aortic sinus is very rare, about 0.008% [4]. A recent systematic review in 2006 recorded 34 cases [5]. Since then, a few cases of similar nature have been reported, but the overall incidence could be delineated to less than 100. ACA has been reported to be the most common cause of sudden cardiac death(SCD), identified from a retrospective review of autopsies among military recruits. Specifically, anomalous origin of left coronary artery from right sinus (ALCA) was the only ACA associated with increased risk for mortality [6]. In our case, we incidentally found ALCA while treating for a distal RCA STEMI. It is important to be aware of ACA in younger individuals especially, as myocardial infarction could be caused by both ACA and acute coronary syndrome (ACS), but by different pathophysiology with varied treatment approaches.

During fetal development, the epicardial coronary arteries develop out of the coronary buds located on the aortic sinuses. The truncus arteriosus gives rises to the aorta and pulmonary trunk through the conotruncal separation. The coronary buds are located such that their division would be in conjunction with the formation of right and left sinuses of Valsalva. The progenitor cells migrate and contribute to angiogenesis. Based on the aorto-pulmonary rotation, the coronary arteries are matched to their sinuses of origin [7]. Abnormalities in development of coronary buds, angiogenesis or rotation could postulate as reasonable etiologies for development of ACA. In a normal cardiac anatomy, the aortic root has three sinuses and cusps. The right coronary artery (RCA) develops from the right sinus of Valsalva(RSV) and the left main coronary artery(LMCA) from the Left sinus of Valsalva(LSV). The LMCA further bifurcates into Left anterior descending artery (LAD), left circumflex artery (LCX). Malposition of these arteries are referred to as ACA.

ACA can be classified into various types based on anatomical variations like origin, course and termination. [1,7]. Around 80% of these anomalies are benign and asymptomatic. The more perilous types include coronary artery origin from the pulmonary artery, ACAOS and single coronary artery [8]. The incidence of anomalous origin of RCA from left sinus is six times more common than anomalous origin of LCA from right sinus of Valsalva [6]. Further, the coronary arteries can course through 4 common routes. Depending on the anatomic relationship to aorta and pulmonary trunk, they can run (a) interatrial (between aorta and pulmonary artery), (b) retro-aortic (c) pre-pulmonic and (d) subpulmonic(septal). Knowing the course is of clinical significance as the interatrial course has a poor prognosis. Interatrial subtype can be detected up to 75% of patients with ALCA [3]. The underlying pathophysiology of interatrial course is that when the aorta dilates during physical exertion, the intussuscepted coronary artery segment becomes narrow, decreasing luminal diameter, possibly contributing to myocardial ischemia [1,7].

Although the exact arterial course is unclear in our case, it makes sense to assume that this could be a contributing factor. Our patient had a complicated hospital course with

respiratory distress requiring intubation and subsequent development of spontaneous pneumothorax. The dynamic lung pressure changes could have reduced the pre load, inducing a stress response with increased cardiac demand and reflex tachycardia. The aortic pulsatility during tachycardia could have led to further narrowing of arterial lumen. The patient was also a marijuana smoker which as such is a potential risk factor for acute coronary syndrome [9]. Although the ACAOS was incidentally found, these factors could have propagated the myocardial infarction.

20% of patients who develop symptoms experience exertional chest pain, palpitations, syncope, transient ischemic attack [3,5]. In a study by Yildiz et al, only 31% had cardiovascular symptoms. Eckhart et al looked at autopsy reports of military recruits and found that only 52% who experienced SCD reported symptoms [3]. So, a majority of these patients remained asymptomatic and undiagnosed. Electrocardiography shows nonspecific ST-T wave changes and varies depending on the acuity of presentation, like the case of STEMI in our patient. Malignant arrhythmias are more commonly seen in the left coronary artery originating from the pulmonary artery [10]. Transthoracic echocardiography (TTE) can be used as an initial screening test to detect ACAOS, but its sensitivity is limited compared to other imaging modalities [11]. Coronary Computed tomography angiography (CCTA) is the diagnostic imaging modality to evaluate high risk anatomic features. Cardiac magnetic resonance imaging (CMR) can be utilized to visualize ACA without radiation, but has a lower spatial resolution than CCTA. Myocardial perfusion imaging can be performed to correlate ischemic symptoms. Cardiac catheterization can be performed if noninvasive testing is non diagnostic and if there is suspected coronary artery disease [12]. Furthermore, intravascular ultrasound (IVUS) and fractional flow reserve (FFR) is increasingly being performed and these are highly sensitive to depict the exact anatomy and stenotic severity [13].

Treatment options include surgical correction, percutaneous coronary intervention(PCI) or medical and conservative management with sports restriction. Decisions should be made after careful risk stratification since abnormalities could be diagnosed incidentally. ACC guidelines stratifies patients with anomalous left coronary artery from right sinus (ALCA) as a Class I recommendation for surgical intervention if they have ischemic symptoms or develop ischemia during diagnostic testing [14]. The surgical approaches include (a) Unroofing procedure done in case of intramural segments where the walls of the segment are resected, (b) coronary or pulmonary translocation, (c) creation of neo - ostium, (d) coronary artery bypass grafting [12]. In our patient, considering the increased peri-operative risk and poor functional status, decision for surgical intervention was deferred. PCI can be performed for simultaneous acute coronary syndrome, however no study comparing the efficacy of surgery vs PCI has been done. A recent systematic review showed a 50% symptomatic reduction in surgery, but a 2.2 % persistence of residual coronary artery stenosis on repeat CT scans posing a risk for repeat ischemia [15]. For asymptomatic or incidentally discovered patients, the decision for surgery versus medical management should be individualized. There are unclear recommendations regarding medical therapy due

to paucity of data. The data on follow up recommendations is limited to some case reports and institutional guidelines as there are no prospective trials. But, vigilant monitoring of these patients at a timely interval is imperative.

4. Conclusion

Anomalous origin of the left coronary artery from right aortic sinus is a rare congenital defect with increased risk for SCD. Oftentimes, they are incidentally found, but can be symptomatic. If found to have myocardial ischemia either from ACS or concurrent coronary stenosis, emergent invasive intervention either through PCI or surgery is vital. The objective of this case with literature review was to point out the rare incidental finding which changed the course of subsequent management. With autopsy reports proving such a presentation as the most common cause of SCD, clinicians should be watchful. Over the last decade, there has been growing incidence owing to increased awareness. Routine screening is not recommended at this point. But, ACA should be considered in the differentials apart from hypertrophic cardiomyopathy in a young patient presenting with typical chest pain and no cardiovascular risk factors. Decision regarding surveillance is scarce. Hence, when such a finding is appreciated in an asymptomatic individual, a thorough history and detailed discussion regarding the prognosis and sports restrictions should be sought. The patient must further have an ischemic workup and management plans should be formulated accordingly.

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