Journal of Community Hospital Internal Medicine Perspectives

Volume 14 | Issue 2 Article 11

2024

Multiple Anomalous Coronary Arteries with Right Sinus of Valsalva Origin: A Case Report

Abraheim AL-Nasseri
Internal Medicine, HCA Florida Citrus Hospital, Inverness, Florida, USA, abe.alnasseri@gmail.com

Nathaniel Leavitt
Internal Medicine, HCA Florida Citrus Hospital, Inverness, Florida, USA

Usman Kazi
Internal Medicine, HCA Florida Citrus Hospital, Inverness, Florida, USA

Aakash Patel
Departments of Cardiology, HCA Florida Citrus Hospital, Inverness, Florida, USA

Hari Kannam
Departments of Cardiology, HCA Florida Citrus Hospital, Inverness, Florida, USA

Recommended Citation

AL-Nasseri, Abraheim; Leavitt, Nathaniel; Kazi, Usman; Patel, Aakash; and Kannam, Hari (2024) "Multiple Anomalous Coronary Arteries with Right Sinus of Valsalva Origin: A Case Report," *Journal of Community Hospital Internal Medicine Perspectives*: Vol. 14: Iss. 2, Article 11.
DOI: 10.55729/2000-9666.1314
Available at: https://scholarlycommons.gbmc.org/jchimp/vol14/iss2/11

CASE REPORT

Multiple Anomalous Coronary Arteries With Right Sinus of Valsalva Origin: A Case Report

Abraheim AL-Nasseri [a,*], Nathaniel Leavitt [a], Usman Kazi [a], Aakash Patel [b], Hari Kannam [b]

[a] Internal Medicine, HCA Florida Citrus Hospital, Inverness, FL, USA
[b] Departments of Cardiology, HCA Florida Citrus Hospital, Inverness, FL, USA

Abstract

Coronary artery anomalies (CAA) define a wide array of congenital abnormalities that stem from the origin, course, and distribution of coronary arteries. CAAs can lead to severe complications such as arrhythmias, myocardial ischemia, and even sudden cardiac death. We describe the case of a 58-year-old female who presented to the emergency department with chest discomfort and shortness of breath and received a workup for acute coronary syndrome. She underwent a cardiac catheterization, which incidentally found an anomalous left anterior descending artery with a right sinus of Valsalva origin, an absent left circumflex coronary artery, and a dominant right coronary artery of unusually large caliber and distribution. There were no identified atherosclerotic plaques. This anomalous configuration of the coronary arteries is exceptionally rare. She required medical management with daily oral acetylsalicylic acid 81 mg, atorvastatin 80 mg, twice daily metoprolol tartrate 50 mg, and hydrocodone/acetaminophen 7.5mg/325 mg oral tablet to be taken every 4 h, as needed for severe pain. Despite optimal medical management, she continued to have chronic angina. A surgical evaluation by a cardiovascular surgeon deemed her anomaly to be inoperable.

Keywords: Coronary artery anomalies, Absent left circumflex coronary artery, Dominant right coronary artery anomaly, Anomalous left anterior descending artery with a right sinus of Valsalva origin

1. Introduction

Coronary artery anomalies (CAA) are an infrequent finding in the general population, with an overall prevalence of less than 5.6 %.[1] Anomalies of the coronary arteries are classically held to be benign and asymptomatic; however, 20 % of cases can produce significant clinical symptoms such as dyspnea, palpitations, angina pectoris, syncope, and dizziness, and may lead to sudden cardiac death.[2,3] Symptomatic patients with CAA have 3 treatment options: medical/observation, coronary angioplasty with stent deployment, or surgical repair.[3] Among these anomalies, multiple phenotypes exist with variable prevalence. The phenotype of the coronary anomaly determines whether or not a patient will experience symptoms. These coronary anomaly phenotypes compromise blood flow and result in coronary artery steal. If left untreated, symptoms may develop in approximately 19 % of patients under the age of 20 and 63 % of patients over the age of 20.[12] For example, an anomalous origin of the left anterior descending artery (LAD) from the right coronary artery (RCA) or the right sinus of Valsalva is found in only 0.15 % of the population.[3] This particular anomaly can be symptomatic in some patients as the LAD becomes compressed between the pulmonary artery and ascending aorta during systole, causing angina, dyspnea, and potentially sudden cardiac death. In other instances, the LAD may pass anterior to the pulmonary artery. When this occurs, patients are less likely to exhibit sudden cardiac death; however, angina and dyspnea are still frequently reported. Very infrequently, the complete absence of a left circumflex coronary artery (LCX) may occur, with a prevalence between 0.003 % and 0.067 % and this anomaly is frequently symptomatic.[4,2] Herein we report an incidental case

Received 2 October 2023; revised 5 December 2023; accepted 2 January 2024.
Available online 4 March 2024

* Corresponding author.
E-mail address: abe.alnasseri@gmail.com (A. AL-Nasseri).

https://doi.org/10.55729/2000-9666.1314

in which our patient was found to have a very rare congenital coronary anomaly of an absent LCX, anomalous LAD origin from the right sinus of Valsalva, and a dominant RCA supplying the majority of the patient's myocardium.

2. Case description

Our patient was a 58-year-old female with a past medical history of hypertension and hyperlipidemia, who presented to the emergency department complaining of a 3-day history of insidious chest heaviness and shortness of breath on exertion and at rest. She described her chest pain as left-sided with radiation to the left shoulder and an 8/10 in intensity. The pain was not relieved by sublingual nitroglycerin or aspirin. Her vital signs and chest x-ray were unremarkable. An electrocardiogram (EKG) revealed sinus tachycardia with nonspecific T-wave abnormalities and no evidence of acute cardiopulmonary disease. Her troponin level was 0.114 ng/mL on admission and trended down to 0.098 ng/mL 6 h later (normal <0.010 ng/mL). The patient had been taking an oral tablet of amlodipine 5 mg daily for hypertension and an oral tablet of atorvastatin 80 mg daily for hyperlipidemia. She had experienced intermittent chest pain on exertion throughout the past year; however, she stated that her chest pain on this admission was worse and present at rest. A stress EKG performed two months prior to admission revealed sinus rhythm with no perfusion defects. A transthoracic echocardiogram taken at the same time demonstrated a left ventricular ejection fraction of 50–55 % without systolic and/or diastolic dysfunction.

The thrombolysis in myocardial infarction risk score was calculated at 3, based on acetylsalicylic acid use within the past 7 days, having two anginal episodes within 24 h, and having elevated serum cardiac markers (e.g., Troponin I). Therefore, the medical team opted for coronary angiography, which revealed an anomalous LAD with right sinus of Valsalva origin, a left coronary cusp (LCC) without an orifice, and a dominant RCA with an unusually large caliber and distribution. The distribution of the RCA was so extensive that it involved the LCX territory (Figs. 1 and 2). The coronary angiography did not identify any coronary artery stenosis and a computer tomography angiography of the coronary arteries was recommended for a more effective evaluation. The computer tomography angiography demonstrated the LAD originating from the right sinus of Valsalva 1.5 mm inferomedial to the RCA and passing anterior to the pulmonary artery to supply the left ventricle. Given

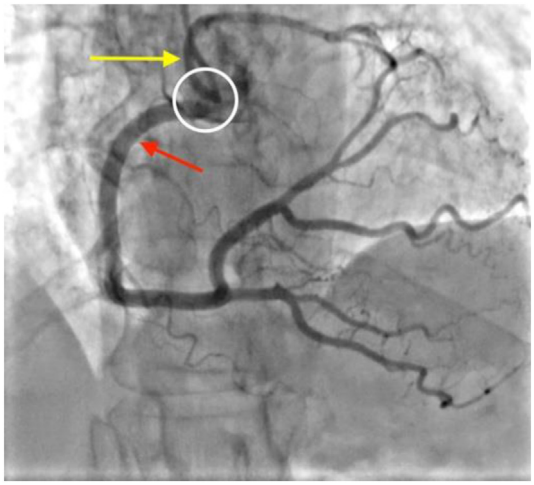

Fig. 1. CA showing right sinus of Valsalva white circle) with RCA (red arrow) supplying LCX territory and anomalous LAD (yellow arrow).

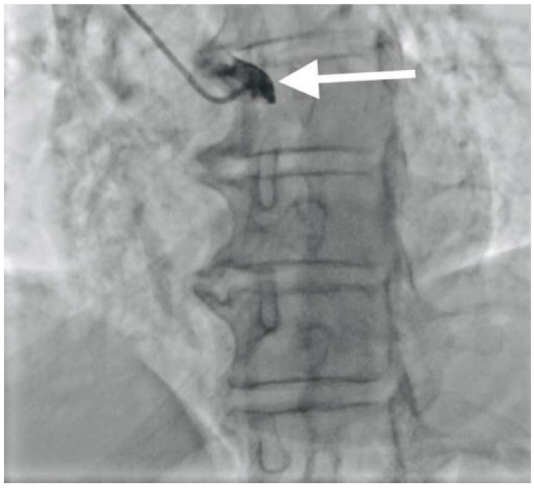

Fig. 2. CA showing the LCC (white arrow) without an orifice.

the absence of the LCX, its territory was supplied by a large dominant RCA with a posterolateral branch supplying the LCX territory along the lateral cardiac border (Fig. 3). Her total and right coronary artery calcium score was 20.

Following a diagnostic workup of her condition, the patient was educated regarding the findings, and a referral to a higher level of care was recommended for further treatment. A lipid panel and an atherosclerotic vascular disease (ASCVD) risk score were obtained upon admission to assess the patient's risk for major ASCVD, myocardial infarction, and ischemic stroke. The lipid panel revealed an LDL of 198 mg/dl, and the ASCVD risk score was calculated to be 15.1 %. Consequently, the patient was initiated on daily oral atorvastatin 80 mg and acetylsalicylic acid 81 mg, respectively. She was also started on oral

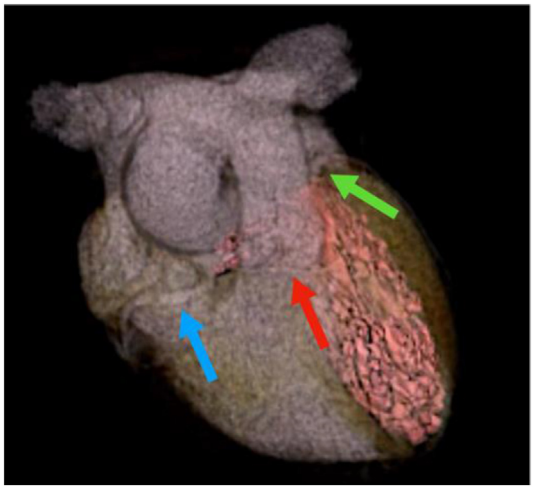

Fig. 3. (far right): CT heart 3-dimensional coronary with calcium revealed the LAD originating from the right sinus of Valsalva, 1.5 mm inferomedial to the RCA, passing anterior to the pulmonary artery (red arrow) to supply the left ventricle. Additionally, the LCX was absent, and a large dominant RCA (blue arrow) and a posterolateral branch supplied LCX territory along the lateral cardiac border (green arrow).

isosorbide dinitrate 2.5 mg twice daily and twice daily metoprolol tartrate 50 mg for angina.

At discharge, she continued to experience left-sided chest pain and pain management was consulted for optimum pain control and to establish care. She was prescribed hydrocodone/acetaminophen 7.5mg/325 mg oral tablet to be taken every 4 h, as needed for severe pain. She was also referred to cardiothoracic surgery at a tertiary care facility for surgical evaluation. A follow-up phone call was issued approximately three months after discharge. She stated that her pain was the same despite continuing all the medications that were prescribed at discharge. After evaluation for surgical correction, her anomaly was deemed to be inoperable. As a result of her severe pain and inoperable condition, she opted into palliative care.

3. Discussion

Coronary artery anomalies affect about 5.6 % of the general population.[1] Approximately 19 % of all sudden death causes in young athletes are related to congenital coronary anomalies, making it the second leading cause of sudden cardiac death after hypertrophic cardiomyopathy.[3,6] A retrospective cohort study designed by Eckar and coworkers using demographic and autopsy data from the Department of Defense Recruit Mortality Registry, discovered that cardiac abnormalities were the leading identifiable cause of sudden death among military recruits.[7] Noninvasive diagnostic tests such as two-dimensional echocardiography and electrocardiography are not sensitive and/or specific enough to detect all the important coronary artery anomalies. To more accurately identify the various coronary artery anomalies, imaging such as coronary arteriography or MRI is required. Since the incidence of CAA is only 5.6 %, and the majority of cases are benign, implementing these modalities for screening of CAA is impractical as it would be neither cost-effective nor universally available.[8]

Various clinical findings associated with CAA will help to risk-stratify patients.[5] High-risk CAA is considered to lead to ventricular tachyarrhythmias and associated sudden cardiac death. These high-risk findings include the LAD originating from the right sinus of Valsalva and passing posterior to the pulmonary artery, and the RCA originating from the left aortic sinus and passing posterior to the pulmonary artery.[7] As cardiac contractility increases during exertion, the coronary artery passing between the pulmonary artery and aorta experiences external compression during systole leading to impinged blood flow and resulting in exertional angina, lightheadedness and/or syncope, and potentially sudden cardiac death. However, in some cases, these symptoms may be entirely absent prior to sudden cardiac death. Our patient was found to have a LAD originating from the right sinus of Valsalva, which then passed anteriorly (not posteriorly) to the pulmonary artery, sparing it from systolic contraction, and thus, her heart from experiencing sudden cardiac death. It remains unclear why our patient's symptoms did not develop until nearly the age of 60. We speculate that her age, psychosocial stresses, and/or calcification of her dominant RCA may have led to the onset of her symptoms. We chose to undergo a cardiac catheterization during her hospitalization due to her elevated thrombosis in myocardial infarction score and a coronary CT angiography was used to further delineate the origin, course, and termination of her coronary anomalies. CT angiography allows for a comprehensive evaluation of coronary artery anatomy and associated coronary lesions to help further guide accurate treatment and management.[13]

While this patient was also found to have an absent LCX, accommodated by the posterolateral branch from the dominant RCA, such absence has previously been shown to be benign but can cause angina-like symptoms (particularly with exertion).[9] The most widely accepted explanation for these symptoms is based on the coronary steal principle. That being said, CAA should be maintained as a differential during the workup for exertional and/or non-exertional angina with no obvious source of

atherosclerotic coronary artery disease.[10,11] To date, there has been no convincing evidence to suggest that an absent LCX can cause sudden cardiac death. At this juncture, there are no established guidelines for the treatment of CAA. The mainstay management for patients with CAA is focused on decreasing symptom severity using medical therapy, coronary angioplasty (with or without stent deployment), and surgery.

4. Conclusion

This case details an anomalous coronary artery configuration that masqueraded symptomatically as acute coronary syndrome. However, the actual origins of the patient's symptoms were revealed after catheterization, showing anomalous coronaries and the absence of atherosclerotic disease. She was managed with daily oral acetylsalicylic acid 81 mg, atorvastatin 80 mg, twice daily metoprolol tartrate 50 mg, and hydrocodone/acetaminophen 7.5mg/325 mg oral tablet to be taken every 4 h, as needed for severe pain. Despite optimal medical management, she continued to have chronic angina. A surgical evaluation by a cardiovascular surgeon deemed her anomaly to be inoperable. These congenital abnormalities must be maintained as differentials when working up patients with exertional angina, especially in young patients who are at risk of sudden cardiac death.

Funding

This research was supported (in whole or in part) by HCA Healthcare and/or an HCA Healthcare affiliated entity. The views expressed in this publication represent those of the author(s) and do not necessarily represent the official views of HCA Healthcare or any of its affiliated entities.

Acknowledgment

Michael G. Flynn, Ph.D., Medical Writer, provided writing and copy-editing assistance on the final version of the manuscript.

References

1. Angelini P, Villason S, Chan AV, et al. Normal and anomalous coronary arteries in humans. In: Angelini P, ed. *Coronary artery anomalies: a comprehensive approach*. Philadelphia: Lippincott Williams & Wilkins; 1999:27–150.
2. Hongsakul K, Suwannanon R. Congenital absence of left circumflex artery detected by computed tomography coronary angiography: a case report. *Case Rep Vasc Med*. 2012;2012:204657. https://doi.org/10.1155/2012/204657.
3. Angelini P. Coronary artery anomalies: an entity in search of an identity. *Circulation*. 2007;115(10):1296–1305. https://doi.org/10.1161/CIRCULATIONAHA.106.618082.
4. Yamanaka O, Hobbs RE. Coronary artery anomalies in 126,595 patients undergoing coronary arteriography. *Cathet Cardiovasc Diagn*. 1990;21(1):28–40. https://doi.org/10.1002/ccd.1810210110.
5. Villa AD, Sammut E, Nair A, Rajani R, Bonamini R, Chiribiri A. Coronary artery anomalies overview: the normal and the abnormal. *World J Radiol*. 2016;8(6):537–555. https://doi.org/10.4329/wjr.v8.i6.537.
6. Maron BJ, Thompson PD, Puffer JC, et al. Cardiovascular preparticipation screening of competitive athletes: a statement for health professionals from the sudden death clinical congenital cardiac defects Committee (cardiovascular disease in the young). *American Heart Association. Circulation.* 1996;94:850–856.
7. Eckart RE, Scoville SL, Campbell CL, et al. Sudden death in young adults: a 25-year review of autopsies in military recruits. *Ann Intern Med*. 2004 Dec 7;141(11):829–834. https://doi.org/10.7326/0003-4819-141-11-200412070-00005. PMID: 15583223.
8. Schmied C, Borjesson M. Sudden cardiac death in athletes. *J Intern Med*. 2014 Feb;275(2):93–103. https://doi.org/10.1111/joim.12184. PMID: 24350833.
9. Shaikh SSA, Deshmukh V, Patil V, Khan Z, Singla R, Bansal NO. Congenital absence of the left circumflex artery with Super-dominant right coronary artery: Extremely rare coronary anomaly. *Cardiol Res*. 2018;9(4):264–267. https://doi.org/10.14740/cr749w.
10. Pun B, Shrestha A, Karki B, Tuladhar AS. Congenital absence of left circumflex artery with right coronary dominance. *Clin Case Rep*. 2021;9:570–571. https://doi.org/10.1002/ccr3.3457.
11. Gentzler 2nd RD, Gault JH, Liedtke AJ, McCann WD, Mann RH, Hunter AS. Congenital absence of the left circumflex coronary artery in the systolic click syndrome. *Circulation*. 1975;52(3):490–496. https://doi.org/10.1161/01.cir.52.3.490.
12. Liberthson RR, Sagar K, Berkoben JP, Weintraub RM, Levine FH. Congenital coronary arteriovenous fistula. Report of 13 patients, review of the literature and delineation of management. *Circulation*. 1979;59(5):849–854. https://doi.org/10.1161/01.cir.59.5.849.
13. Popa RM, Pântea R, Macașoi P, Manea RM. A rare case of Single coronary artery: Getting to the heart of the Matter. *J Belg Radiol*. 2023;107(1):50. https://doi.org/10.5334/jbsr.3122.