Cardiac

Interarterial course of anomalous right coronary artery: Pathophysiology, diagnosis, and treatment

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ABSTRACT

Anomalous coronary artery from the opposite sinus is a rare congenital anomaly that can present with symptoms similar to coronary artery disease, and sudden cardiac death. Management of anomalous coronary artery from the opposite sinus varies; however, current guidelines suggest surgery in symptomatic patients.

Our patient is a middle-aged male with a history of coronary artery disease and status post coronary artery bypass graft. He presented with complaints of vague chest pain. After a positive stress test, he was sent to the catheterization suite. Diagnosis of an anomalous right coronary artery from the left coronary sinus was made. The patient underwent surgical revascularization and was awaiting follow-up with cardiology at the time of study.

A timely diagnosis of an anomalous coronary artery is critical in symptomatic patients because of the risk of sudden cardiac death, especially in patients with arteries with an interarterial course. This case demonstrates the importance of making the correct diagnosis, as appropriate surgical management can drastically improve outcomes.

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Introduction

The origin of a coronary artery from the opposite aortic sinus is a rare congenital abnormality that could potentially lead to cardiac ischemia or sudden cardiac arrest [1]. Most commonly, these anomalies involve the left circumflex artery originating from a separate ostium in the right coronary sinus [2]. Here we present a case of an anomalous right coronary artery (RCA) originating from the left coronary sinus in a middle-aged patient who presented with atypical chest pain.

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Case report

O.G. is a 43-year-old male with a history of coronary artery disease, coronary artery bypass graft, hypertension, and hyperlipidemia who presented to the emergency department with intermittent, sharp, atypical chest pain. His physical examination findings were negative and troponin was mildly elevated at 0.039 ng/mL.

After monitoring of the patient overnight with electrocardiography and serial cardiac enzymes, myocardial scintigraphy was performed, which showed signs of reversible ischemia in the inferolateral portion of the left ventricle (Fig. 1). The patient was subsequently taken to the catheterization laboratory suite and left heart catheterization with coronary angiography was performed. Imaging revealed a single left sinus of Valsalva ostium that gave rise to the left coronary artery (LCA), which divided into the left anterior descending (LAD) and left circumflex arteries. However, there was also an RCA that was originating from the proximal LCA (Fig. 2). The RCA did not demonstrate any significant stenosis, nor was there any occlusion noted in the RCA during systole or diastole. No arteries were found arising from the right coronary cusp. A subsequent...
A coronary computed tomography (CT) angiography scan was performed and demonstrated an interarterial pathway of the RCA as it took a course between the aortic root and pulmonary artery (Fig. 3). In addition, a three-dimensional CT was also performed, further confirming the anomalous origin of the RCA that was suggested by the initial imaging (Fig. 4).

Based on these images and clinical findings, the decision for surgical revascularization was made. A coronary artery bypass graft was performed with anastomosis of the right internal mammary artery with the distal RCA, followed by ligation of the proximal RCA. The patient tolerated the procedure well without complications and was taken to the intensive care unit. He was subsequently discharged and was awaiting postoperative follow-up with cardiology at the time of study.

Discussion

Anomalous coronary artery from the opposite sinus (ACAOS) is a rare phenomenon with varying prevalence rates, depending on the detection method used. In P. Angelini’s group, the estimated total prevalence rate was found to be 1.07% when using coronary angiography for detection. This prevalence rate was further divided into 2 groups, with 0.92% demonstrating an RCA from the left sinus, and 0.15% demonstrating an anomalous LCA from the right sinus [3]. In a separate study where echocardiography was used for diagnosis, the prevalence rate of ACAOS was much lower at 0.17% [4]. Despite these differences in prevalence rates, one thing is clear: ACAOS is associated with the risk of sudden cardiac death (SCD) and as such should be carefully considered in symptomatic patients [5].

Clinical symptoms of ACAOS can present very similarly to symptoms of coronary artery disease, especially if the anomalous artery takes an interarterial course [6]. This can be seen in our particular case, where the patient was diagnosed with right-sided ACAOS and presented with atypical chest pain on exertion. The interarterial course associated with a right-sided ACAOS can be further classified as taking either a high or a low interarterial course. A high interarterial course is when the anomalous RCA travels between the aorta and pulmonary artery, and a low interarterial course is when the anomalous RCA travels between the aorta and right ventricular outflow tract. These two subtypes demonstrate a significant difference in clinical presentation, according to a study by H. Lee et al., with 43% of patients in the high group presenting with typical anginal chest pain, compared with only 6% in the low group. The high group also had a higher prevalence of major adverse cardiac events (MACEs), which includes cardiac death, nonfatal myocardial infarction, unstable angina, and surgical treatment, with 28% patients experiencing MACEs versus only 6% in the low group [6]. Essentially, when an individual undergoes strenuous activity, the increased cardiac output results in a greater expansion of the aorta and pulmonary artery. When there is a high interarterial course of the right-sided ACAOS, as seen with our patient, it is possible that the enlarged vessels mechanically compresses the RCA, effectively acting as an exercise-dependent form of stenosis.

In addition to determining the course of an anomalous coronary artery, it is also important to differentiate between different types of coronary artery anomalies to better stratify patient risk. Although this report outlines a unique case of a symptomatic right-sided ACAOS, it is generally considered to be more benign than other coronary artery anomalies, such
as a left-sided ACAOS. The difference in mortality between the 2 types of ACAOS has been described in past autopsy reports, with a suggested mortality rate of 57% for left-sided ACAOS and 25% for right-sided ACAOS [7]. Other coronary artery anomalies, including those that involve the LAD artery, were also discussed in these autopsy reports. In patients with an anomalous LCA or LAD originating from the pulmonary trunk, 38% of all deaths (14 of 37) were due to SCD, but in patients with both arteries originating from the pulmonary trunk, 100% of all deaths (3 of 3) were due to SCD [7].

Diagnosis of ACAOS is usually incidental as most patients are asymptomatic. In those who do present with symptoms, the most common ones are exertional syncope, chest pain, or palpitations [8]. Coronary computed tomography angiography (CCTA) is a well-known imaging modality that has been shown to be effective in diagnosing ACAOS and excluding other coronary anomalies with high accuracy [9], as well as being an accurate technique for distinguishing patients at high risk for adverse events [10]. Ashrafpoor et al. demonstrated in their study that there are specific CT-derived anatomical criteria that are associated with an increased risk for MACEs such as unstable angina and myocardial infarction [10]. The patient in our case first underwent a Lexiscan stress test, which was positive for reversible ischemia in the inferolateral portion of the left ventricle. This prompted the need for a coronary angiography and CCTA, which clearly demonstrated the anomalous origin of the RCA as it left the opposite sinus and took an interarterial course between the aorta and pulmonary artery. We were also able to generate a three-dimensional rendering of the anomalous RCA from our CT images, which further helped us decide the best treatment for our patient.

Because of the increased risk of SCD, it is important to carefully consider treatment options in these patients. Symptomatic ACAOS patients have 3 treatment options: medical treatment, coronary angioplasty with stenting, and surgical repair, which includes bypass surgery, reimplantation of the anomalous artery, unroofing of intramural segments of the anomalous artery, or osteoplasty [11]. The American College of Cardiology and American Heart Association 2008 guidelines recommend surgery in patients with clinical adverse events or evidence of ischemia; in patients with neither of those criteria, surgery is not the clear-cut choice. In our case, the patient was symptomatic and had a positive stress test that demonstrated reversible myocardial ischemia. Thus, the decision was made to perform surgery. Our patient underwent a right internal mammary artery to RCA bypass graft with ligation of the proximal RCA to prevent competing flow, which has been shown to be of some benefit [12]. The patient tolerated the procedure well with no complications. He was awaiting follow-up appointment with cardiology for further assessment of symptoms.

In conclusion, although ACAOS is fairly rare, the potential risk of SCD and other adverse complications make accurate diagnosis and treatment of this condition crucial to maximizing patient outcomes. This case is a good representation of what the current literature recommends in terms of appropriate workup and treatment and serves to add to our growing knowledge of ACAOS and its management.

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References

[1] Ogden JA. Congenital anomalies of the coronary arteries. Am J Cardiol 1970;25(4):474–9. <http://www.ncbi.nlm.nih.gov/pubmed/5438244>.
[2] Click RL, Holmes DR, Vlietstra RE, Kosinski AS, Kronmal RA. Anomalous coronary arteries: location, degree of atherosclerosis and effect on survival—a report from the Coronary Artery Surgery Study. J Am Coll Cardiol 1989;13(3):531–7. doi:10.1016/0735-1097(89)90588-3.
[3] Angelini P, Villason S, Chan AV, Diez G. Coronary artery anomalies: a comprehensive approach. Philadelphia (PA): Lippincott Williams & Wilkins; 1999.
[4] Frommelt PC, Frommelt MA, Tweddell JS, Jaquiss RDB. Prospective echocardiographic diagnosis and surgical repair of anomalous origin of a coronary artery from the opposite sinus with an interarterial course. J Am Coll Cardiol 2003;42(1):148–54. doi:10.1016/S0735-1075(03)00503-5.
[5] Penalver JM, Mosca RS, Weitz D, Phoon CK. Anomalous aortic origin of coronary arteries from the opposite sinus: a critical appraisal of risk. BMC Cardiovasc Disord 2012;12:83. doi:10.1186/1471-2261-12-83.
[6] Lee H, Hong Y, Kim H, Lee J, Hur J. Anomalous origin of the right coronary artery from the left coronary sinus with an interarterial course: subtypes and clinical importance. Radiology 2012;262(1):101–8. doi:10.1148/radiol.11110823/-/DC1.
[7] Taylor AJ, Rogan KM, Virmani R. Sudden cardiac death associated with isolated congenital coronary artery anomalies. J Am Coll Cardiol 1992;20(3):640–7. doi:10.1016/0735-1097(92)90019-J.
[8] Basso C, Maron BJ, Corrado D. Clinical profile of congenital coronary artery anomalies with origin from the wrong aortic sinus leading to sudden death in young competitive athletes. J Am Coll Cardiol 2000;35(6):1493–501. doi:10.1016/S0735-1075(00)00566-0.
[9] Angelini P, Velasco JA, Flam F. Coronary anomalies: incidence, pathophysiology, and clinical relevance. Circulation 2002;105(20):2449–54. doi:10.1161/01.CIR.0000016175.49835.57.
[10] Ashrafpoor G, Danchin N, Houyel L, Ramadan R, Belli E, Paul JF. Anatomical criteria of malignancy by computed tomography angiography in patients with anomalous coronary arteries with an interarterial course. Eur Radiol 2014;25(3):760–6. doi:10.1007/s00330-014-3454-9.
[11] Angelini P. Coronary artery anomalies: an entity in search of an identity. Circulation 2007;115(10):1296–305. doi:10.1161/CIRCULATIONAHA.106.618082.
[12] Kirklin JW, Barrat-Boyes BG. Cardiac surgery. 3rd ed. New York (NY): Wiley; 2003.