

INFERIOR MYOCARDIAL INFARCTION WITH ANOMALOUS RIGHT CORONARY ARTERY

Inferior Wall Myocardial Infarction in the Setting of a High-Risk Anomalous Right Coronary Artery: A Case Report



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INTRODUCTION

The incidence of coronary artery anomalies is approximately 1% in individuals undergoing cardiac catheterization.¹ Although the majority of cases are benign and discovered incidentally, they are implicated in up to one third of sudden cardiac deaths.² Anomalous origin of the left coronary artery or right coronary artery (RCA) from the contralateral sinus of Valsalva with an intramural origin or interarterial course may cause ischemia, malignant arrhythmias, and sudden cardiac death.³ We present a case of an inferior wall myocardial infarction in a patient with an anomalous RCA arising from the left coronary cusp.

CASE PRESENTATION

A 43-year-old man with a history of long-standing tobacco use developed crescendo angina and diaphoresis that woke him from sleep. His symptoms progressed to loss of consciousness, and a family member notified emergency medical services. He was transported to our hospital, where his presenting blood pressure was 70/30 mm Hg, and the physical examination was significant for an S3 gallop. Electrocardiography showed 2-mm ST-segment elevation in leads II, III, and aVF and ST-segment depression in leads V₁, V₂, and V₃ compatible with an acute inferoposterior ST-segment elevation myocardial infarction (Figure 1). Laboratory studies showed a troponin T level of 3.1 ng/mL. Emergent coronary angiography revealed nonobstructive disease in the left coronary artery system (Video 1). The RCA could not be engaged despite significant effort and trial of multiple of diagnostic catheters (Table 1) by two experienced operators (Video 2). Aortography was performed and showed no clear origin of the RCA (Video 3). An infusion of the glycoprotein IIb/IIIa inhibitor tirofiban was initiated. Intravenous dopamine was used for inotropic and hemodynamic support, and an intra-aortic balloon pump was placed via the left common femoral artery to augment coronary perfusion. He was admitted to the coronary care unit for further management.

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The patient underwent an expedited diagnostic evaluation for a suspected anomalous origin of the RCA.

Urgent coronary computed tomographic angiography (CCTA) revealed an anomalous RCA that originated from the left sinus of Valsalva (Figure 2A) with an intramural course within the aortic wall as well as an interarterial course between the aorta and pulmonary artery (Figure 2B). There was significant vascular calcification in the mid-RCA, which contributed to blooming artifact, limiting assessment for obstructive atherosclerotic plaque. However, the vessel was patent, without evidence of thrombotic occlusion, suggesting spontaneous reperfusion.

Transthoracic echocardiography showed normal left and right ventricular size and function, with hypokinesis of the basal inferoseptal and basal to mid inferior walls. The etiology of the patient's myocardial infarction was felt to be secondary to plaque rupture and thrombotic occlusion of the mid-RCA rather than a physiologic consequence of an anomalous origin of the RCA. The patient was stabilized with medical therapy, and because of suspected severe atherosclerotic disease of the midvessel and the high-risk anatomic features of the RCA, surgical revascularization was recommended to prevent recurrent ischemic events. Because of presumed obstructive coronary artery disease in the midvessel, the patient underwent coronary artery bypass graft surgery with a pedicle right internal mammary artery graft to the RCA with an anastomotic site distal to the calcific lesion. Intraoperative transesophageal echocardiography demonstrated an intramural origin of the RCA from the left sinus of Valsalva with an interarterial course and systolic compression between the aortic root and pulmonary artery (Figure 3, Video 4). Gross operative findings showed a long intramural course of the proximal RCA (Figure 2D), which correlated with the findings on CCTA, as illustrated in a three-dimensional reconstruction (Figure 2C). His postoperative course was unremarkable, and he remains stable at 6 months of follow-up.

DISCUSSION

Anomalous RCA originating from an inappropriate sinus of Valsalva is a rare entity and an uncommon cause of acute myocardial infarction. Although the true prevalence is unknown, a recently reported weighted estimate from prior studies was 0.23% of the population, with a range from 0.02% to 0.79%.⁴ The incidence of adult patients presenting with acute coronary syndromes who are found to have anomalous coronary artery origins is also unknown. A single-center registry in Italy reported an incidence of anomalous coronary arteries in 0.4% of patients who presented with ST-segment elevation myocardial infarction.⁵ A large registry from the Cleveland Clinic before the era of percutaneous coronary intervention showed an overall incidence of 1.3% for all patients who underwent coronary angiography.⁶ Several studies have shown an association of an anomalous RCA from

VIDEO HIGHLIGHTS

Video 1: Coronary angiography of left coronary artery. Selective coronary angiography of the left main coronary artery shows a patent left anterior descending coronary artery with moderate stenosis in the midvessel and a patent left circumflex coronary artery.

Video 2: Coronary angiography of RCA. The ostium of the RCA could not be localized.

Video 3: Aortography was performed with a pigtail catheter and showed no definitive origin to the RCA.

Video 4: Transesophageal echocardiography, midesophageal short-axis view, demonstrating systolic compression of the proximal RCA between the aorta and the pulmonary artery.

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an inappropriate sinus of Valsalva with sudden cardiac death, with particularly high risk attributed to an intramural course or an intra-aortic course, both of which were observed in our patient.^{7,8} Furthermore, systolic compression of the intramural segment of the RCA, which was demonstrated by transesophageal echocardiography in our patient, has been associated with chest pain and syncope.⁹ Despite the association with sudden cardiac death, many patients remain asymptomatic, and the mortality rate may be less than 1%.⁹ Regardless of the presence of high-risk features such as an intramural origin and interarterial course, an anomalous origin of the RCA is frequently a benign finding, and patients are diagnosed either incidentally or with stable symptoms.¹⁰

An anomalous coronary artery origin may be diagnosed at the time of cardiac catheterization because of an inability to engage the coronary ostium with standard preshaped diagnostic coronary

artery catheters. In patients who are acutely ill and present with ST-segment elevation myocardial infarction, there is heightened urgency to restore coronary perfusion to the affected myocardium, but this may not be possible if the coronary artery cannot be engaged with a catheter. There is significant heterogeneity in the management of these patients, and patients may undergo surgical revascularization, percutaneous coronary intervention, thrombolysis, or medical management.^{11,12} Surgical management may include unroofing of the ostial RCA with or without reimplantation of the coronary or coronary artery bypass graft surgery.¹³ Our patient was stabilized with glycoprotein IIb/IIIa infusion, dopamine as a vasopressor, and an intra-aortic balloon pump. CCTA showed a severe calcified atherosclerosis in the midvessel as well as an anomalous RCA with high-risk features, and the multidisciplinary recommendation was for urgent surgical revascularization with a right internal mammary artery-to-RCA bypass graft to prevent recurrent myocardial ischemia.

Often, the diagnosis and characterization of coronary artery anomalies requires the use of cross-sectional imaging. In patients who are hemodynamically unstable, imaging may need to be performed at the bedside with transthoracic or transesophageal echocardiography, or the patient may need to be stabilized before performing imaging. Transthoracic echocardiography is among the first diagnostic strategies, but identification of coronary ostia may be challenging because of body habitus and limited imaging planes to adequately assess coronary artery origins. Detailed investigation of coronary course (intramural, interarterial, retroaortic, acute angle takeoff) may be possible with transesophageal echocardiography, as demonstrated in this case (Figure 3, Video 4).¹⁴ CCTA and cardiac magnetic resonance imaging have Class I indications to characterize anomalous coronary arteries in stable patients.¹⁵ The superior resolution of CCTA yields precise and comprehensive assessment of the orifice configuration, intramural course, intraluminal plaque, and distal vessel course, which favors its utility over cardiac magnetic resonance imaging. As such, CCTA is the modality of choice at many institutions. In patients who are acutely ill, CCTA may have an additional advantage over

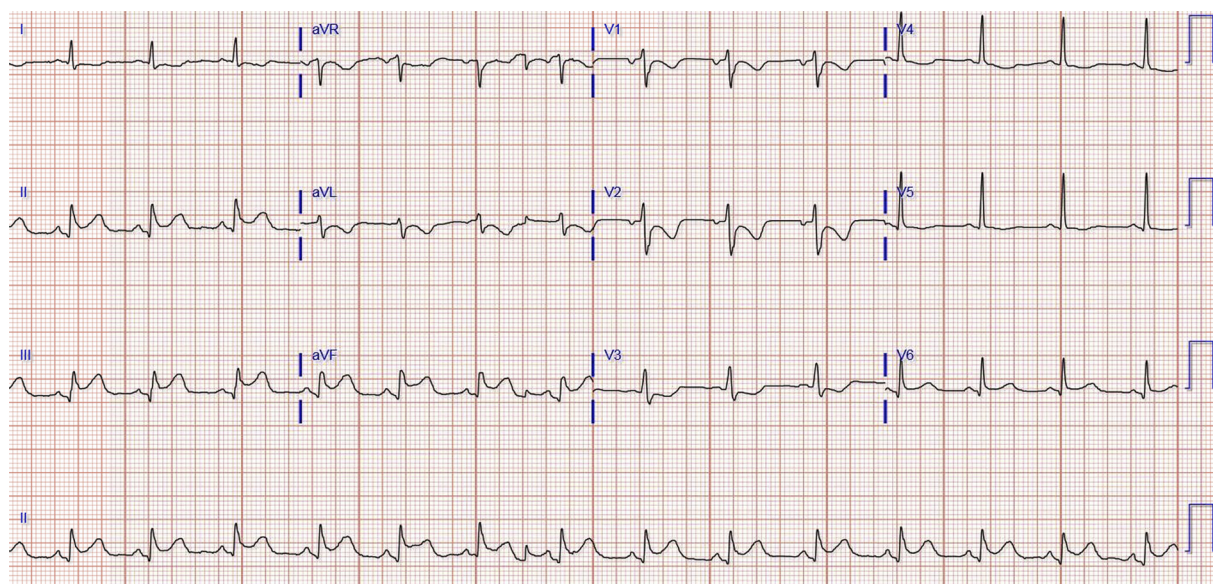


Figure 1 Twelve-lead electrocardiogram. Presenting 12-lead electrocardiography showed 2-mm ST-segment elevation in leads II, III, and aVF as well as reciprocal ST-segment depression in the lateral leads consistent with myocardial infarction in the territory supplied by the RCA.

Table 1 Timeline of initial presentation and cardiac catheterization

Time	Description
1:40 PM	Presentation to the emergency department, aspirin 324 mg
1:50 PM	Loaded with ticagrelor 180 mg
2:04 PM	Arrival to cardiac catheterization laboratory
2:15 PM	Arterial access
2:17 PM	Left coronary angiography
2:19 PM	Judkins JR4 guiding catheter inserted, unsuccessful
2:28 PM	No Torque guiding catheter inserted, unsuccessful
2:33 PM	AR 2 Launcher guiding catheter inserted, unsuccessful
2:37 PM	Pigtail catheter: aortography
2:42 PM	AL 1 Launcher guiding catheter, unsuccessful
3:03 PM	Tirofiban (glycoprotein IIb/IIIa inhibitor) initiated
3:05 PM	Blood pressure 70/53 mm Hg; dopamine initiated
3:29 PM	MB 1 Launcher guiding catheter, unsuccessful
3:33 PM	Pigtail catheter: repeat aortography
3:36 PM	AL 2 Launcher guiding catheter, unsuccessful
3:44 PM	Intra-aortic balloon pump inserted
4:00 PM	Admitted to coronary care unit

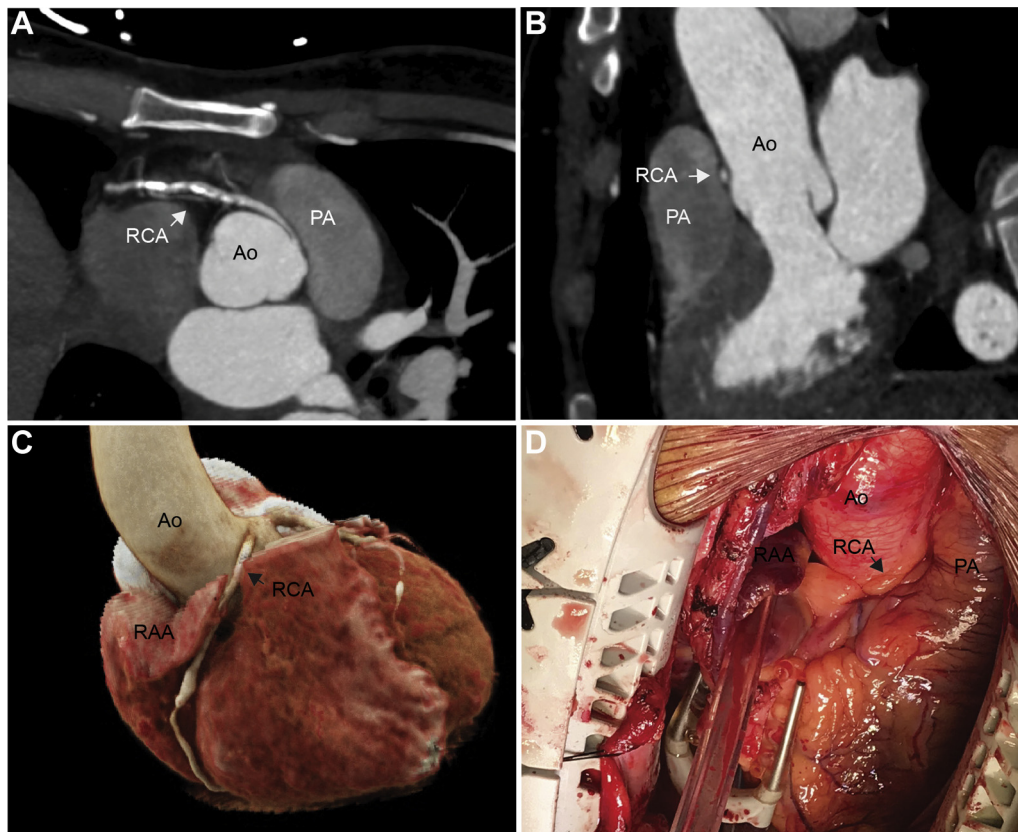


Figure 2 Retrospective gated CCTA, multiplanar reconstruction, showing anomalous RCA (arrow) from the left cusp with an acute angle takeoff and slitlike orifice (A). Modified left ventricular outflow tract plane revealed an intramural and interarterial course with vessel compression in the systolic phase (static image captured at 40% phase of the cardiac cycle) (B, arrow). Three-dimensional reconstruction with exclusion of the pulmonary trunk confirmed an anomalous origin (C, arrows), which correlated with operative findings that showed an intramural course (D, arrows). Ao, Aorta; PA, pulmonary artery; RAA, right atrial appendage.

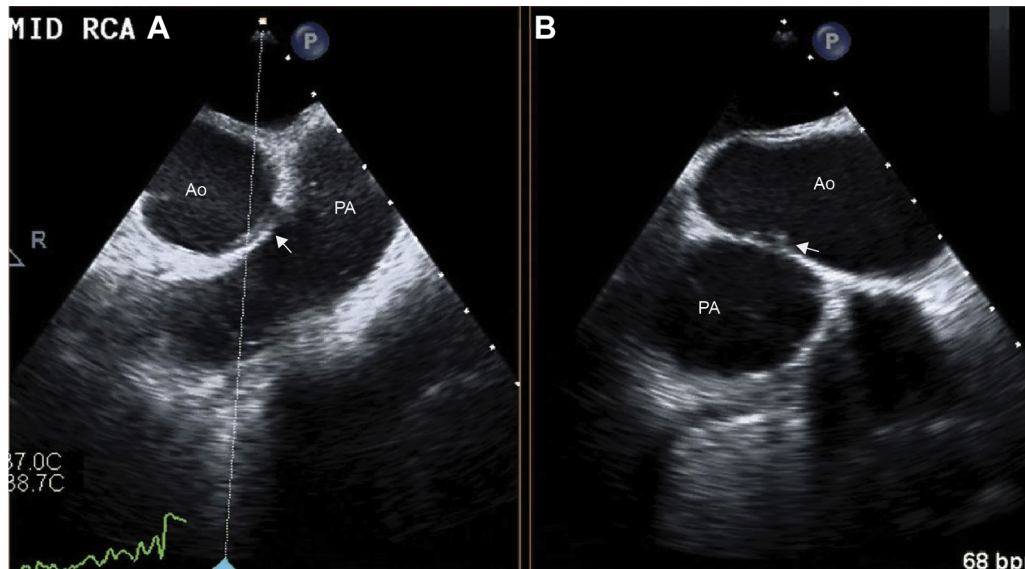


Figure 3 Transesophageal echocardiography, midesophageal short-axis view, showing an anomalous RCA arising from the left sinus of Valsalva and with an intramural and interarterial course (A). An “X-plane” cross-sectional view through the aortic root demonstrates an intramural origin within the aortic wall (B). Ao, Aorta; PA, pulmonary artery.

cardiac magnetic resonance imaging because of its shorter acquisition time. However, CCTA may be of limited value in patients with highly calcified coronary arteries, because of mischaracterization of the arterial lumen due to imaging artifacts in the presence of calcium. Additionally, CCTA requires iodinated contrast medium, which may be contraindicated in patients with renal dysfunction.

The mechanism of ischemia or myocardial infarction in patients with anomalous coronary arteries may be related to primary plaque rupture, arterial dissection in the setting of endothelial dysfunction, extrinsic compression of the vessel (particularly in cases with an interarterial course), slitlike ostium, or acute angle takeoff of the proximal vessel.^{1,16} In many patients with anomalous origin of the RCA who present with angina and dyspnea, there is evidence of myocardial ischemia on exercise stress testing with myocardial perfusion imaging.¹⁰ In our patient there was evidence of severe atherosclerotic coronary artery disease in the mid-RCA (Figure 3A) as well as extrinsic compression of the ostial RCA (intramural within the aortic wall). The culprit for the patient’s presentation was likely a combination of anatomic obstructive coronary artery disease and baseline physiologic reduction in coronary blood flow due to ostial compression. Furthermore, in patients with an anomalous RCA with an interarterial course, the proximal segment may also be compressed by the great vessels, particularly with increasing aortic dimensions with age and pulsatile expansion during exercise or exertion.^{2,3,17} In a postmortem study of 242 patients with coronary anomalies, all patients with sudden cardiac death who had origins of the left coronary artery or RCA from the contralateral sinus had presence of an acute angle takeoff, intramural segment (within the aortic wall), or an interarterial course.⁷

CONCLUSION

Anomalous coronary arteries are a risk factor for sudden cardiac death, and an origin from an inappropriate sinus of Valsalva with an intramural origin and interarterial course may be particularly ominous. However, most patients remain asymptomatic or present

with stable symptoms, and coronary artery anomalies are typically discovered in the routine evaluation of angina or as incidental findings. Rarely, patients may present with acute myocardial infarction. This may be a particularly challenging situation for management in the catheterization laboratory, and there is no consensus for diagnostic assessment or revascularization in the acute setting. On the basis of the American College of Cardiology/American Heart Association guideline for adults with congenital heart disease, surgical revascularization is recommended for anomalous left coronary artery with an interarterial course irrespective of symptoms (Class I). In instances of anomalous RCA, surgical revascularization is recommended when ischemia is demonstrated.¹⁵ Thus, the management approach should be based on anatomic features, clinical presentation, and institutional expertise.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at <https://doi.org/10.1016/j.case.2019.02.003>.

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