JACC: CASE REPORTS VOL. 4, NO. 9, 2022

CASE REPORT

ADVANCED

HEART CARE TEAM/MULTIDISCIPLINARY TEAM LIVE

Acute Perimyocarditis Unmasking Anomalous Coronary Artery

Is Treating the Inflammation Enough? Does Surgery Fix All?

Mariyam Sheidu, MD,[a] Xiaoxiao Qian, MD,[b] Hooman Bakhshi, MD,[b] Raghav Gattani, MD,[b] Araba Ofosu-Somuah, MD,[b] Abbas (Reza) Emaminia, MD[b]

ABSTRACT

We report the case of a patient with anomalous right coronary artery (RCA) unmasked by acute perimyocarditis who continued to have ischemic symptoms despite total resolution of perimyocarditis and required surgical intervention of the anomalous RCA. This case was further complicated by ventricular arrhythmia after surgical repair. Collaboration among different cardiac specialists was essential in this case. (**Level of Difficulty: Advanced.**) (J Am Coll Cardiol Case Rep 2022;4:507–511) © 2022 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

HISTORY OF PRESENTATION

An 18-year-old previously healthy man presented with acute chest pain for 2 days. He reported sore throat, dry cough, and low-grade fever 2 weeks previously. His vital signs were as follows: temperature 36.3 °C, blood pressure 97/48 mm Hg, heart rate 67 beats/min, and oxygen saturation 98% on room air. His physical examination results were unremarkable. His laboratory determinations were significant for troponin 2.74 ng/mL (normal 0.00-0.09 ng/mL) and C-reactive protein 2.8 mg/dL (normal 0.0-0.8 mg/dL). An electrocardiogram (ECG) revealed diffuse ST-segment elevation (**Figure 1**), suggesting pericarditis. Shortly after presentation, the patient experienced hypotension, and repeat troponin trended up to 32.33 ng/mL. Urgent coronary artery angiography was performed to rule out acute coronary syndrome, and it showed no obstructive coronary artery disease (CAD) but an anomalous origin of RCA from the left coronary sinus (**Figure 2,** Video 1). Transthoracic echocardiography demonstrated mild inferior wall hypokinesis and left ventricular ejection fraction of 50% (Video 2). Coronary CT angiography again demonstrated the anomalous RCA (**Figure 3**).

LEARNING OBJECTIVES

- To keep in mind that acute perimyocarditis can unmask asymptomatic anomalous coronary artery.
- To be familiar with surgical indications for anomalous coronary artery.
- To be familiar with postoperative complications in patients undergoing surgical repair of anomalous coronary artery.

QUESTION 1: WHAT IS THE DIFFERENTIAL DIAGNOSIS AT THIS STAGE?

Given the viral prodrome symptoms, acute chest pain, elevated troponin, diffuse ST-segment

From the [a]Department of Internal Medicine, Inova Heart and Vascular Institute, Falls Church, Virginia, USA; and the [b]Department of Cardiology, Inova Heart and Vascular Institute, Falls Church, Virginia, USA.

The authors attest they are in compliance with human studies committees and animal welfare regulations of the authors' institutions and Food and Drug Administration guidelines, including patient consent where appropriate. For more information, visit the Author Center.

Manuscript received December 27, 2021; revised manuscript received February 14, 2022, accepted March 7, 2022.

ISSN 2666-0849 https://doi.org/10.1016/j.jaccas.2022.03.008

ABBREVIATIONS AND ACRONYMS

ACA = anomalous coronary arteries

CAD = coronary artery disease

ECG = electrocardiogram

ICD = implantable cardioverter defibrillator

RCA = right coronary artery

SCD = sudden cardiac death

VT = ventricular tachycardia

elevation on ECG, and the absence of obstructive coronary artery disease, acute perimyocarditis was highly suspected. Other differential diagnoses included chest pain caused by the anomalous RCA.

QUESTION 2: WHAT INVESTIGATIONS ARE NEEDED TO CLARIFY THE DIAGNOSIS?

Cardiac magnetic resonance showed myocardial edema within the midmyocardium of the lateral wall. Late gadolinium enhancement mapping revealed delayed enhancement within the lateral and inferolateral walls along with pericardial thickening (**Figure 4**), suggesting acute perimyocarditis.

QUESTION 3: WHAT IS THE MANAGEMENT PLAN?

Whereas colchicine plus nonsteroidal anti-inflammatory drugs constitute the treatment of choice for pericarditis, limited data are available regarding the treatment for perimyocarditis. One small study demonstrated good response to colchicine in patients with myocarditis.[1] Our patient was treated with ibuprofen 800 mg three times a day until the resolution of chest pain and colchicine 0.6 mg twice daily for 3 months.

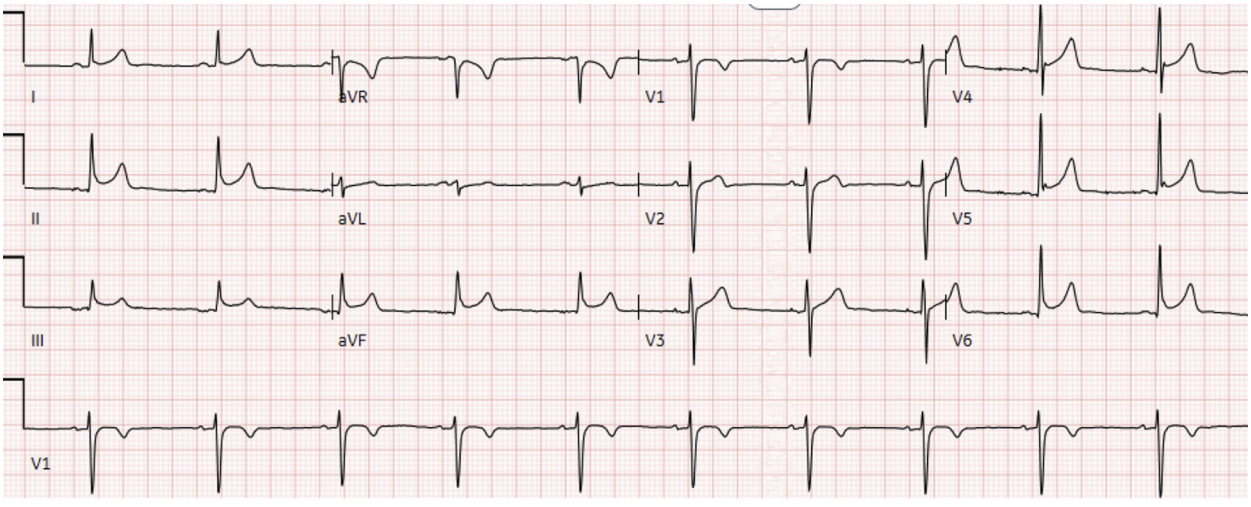
Anomalous coronary arteries (ACA) constitute a group of rare congenital vascular malformations, and the prevalence is estimated to be 1% to 1.2%.[2] Anomalous RCA originating from the left coronary sinus has an incidence of approximately 0.92%. ACAs are traditionally classified as malignant and benign variants, with malignant variants defined by an interarterial course between the aorta and the main pulmonary artery. Malignant anomalous RCA has an incidence of 0.03% to 0.17%.[3] Most ACA are incidental findings and do not cause symptoms. Less frequently, ACA may cause symptoms related to decreased perfusion to the myocardium, such as chest pain, shortness of breath, heart failure symptoms, or even sudden cardiac death (SCD). The presenting symptoms of ACAs are nonspecific and can be confused with other common cardiac diseases such as acute coronary syndrome, congestive heart failure, myocarditis, and arrhythmia. Given that this patient was completely asymptomatic before the acute perimyocarditis, no surgical intervention was planned because this was most likely an incidental finding.

The patient completed 3 months of colchicine; however, he continued to have exertional chest pain.

QUESTION 4: WHAT ARE THE DIFFERENTIAL DIAGNOSES OF PATIENT'S CONTINUED CHEST PAIN? WHAT INVESTIGATIONS COULD BE DONE?

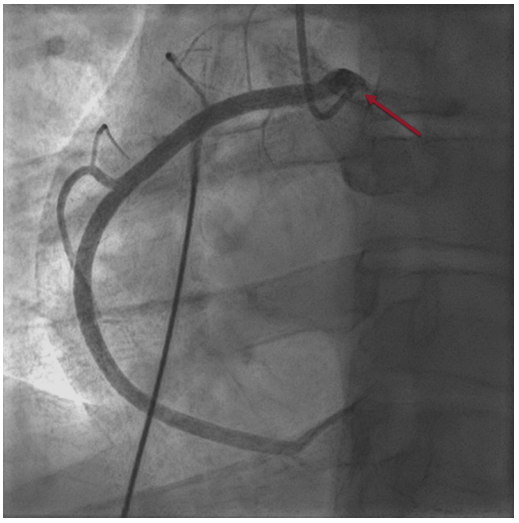
The differential diagnoses of continued chest pain included unresolved perimyocarditis, ischemia due to anomalous RCA, and noncardiac causes. Repeat cardiac magnetic resonance showed completely resolved perimyocarditis (Supplemental Figure 1). Exercise myocardial perfusion imaging showed a small, moderate-severity, and predominantly reversible inferolateral perfusion defect suggesting ischemia (**Figure 5**).

FIGURE 1 Electrocardiogram



ST-segment elevation of leads I, II, III, AVF, as well as V_4-V_6.

FIGURE 2 Coronary Angiography

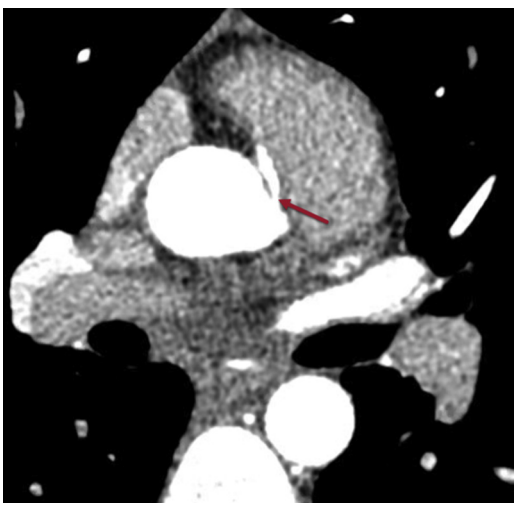


Right coronary artery originates from left coronary sinus **(red arrow)**.

QUESTIONS 5: WHAT SHOULD BE THE MANAGEMENT PLAN FOR ISCHEMIA CAUSED BY ANOMALOUS RCA?

Given the persistent chest pain and reversible perfusion defect on myocardial perfusion imaging, the patient underwent surgical translocation of the main pulmonary artery toward the left pulmonary artery to relieve impingement of the RCA by the aorta and the main pulmonary artery. A pediatric cardioplegia system was used for the surgery, and the total ischemia time was 108 minutes.

FIGURE 3 Coronary Artery Computed Tomography Angiography



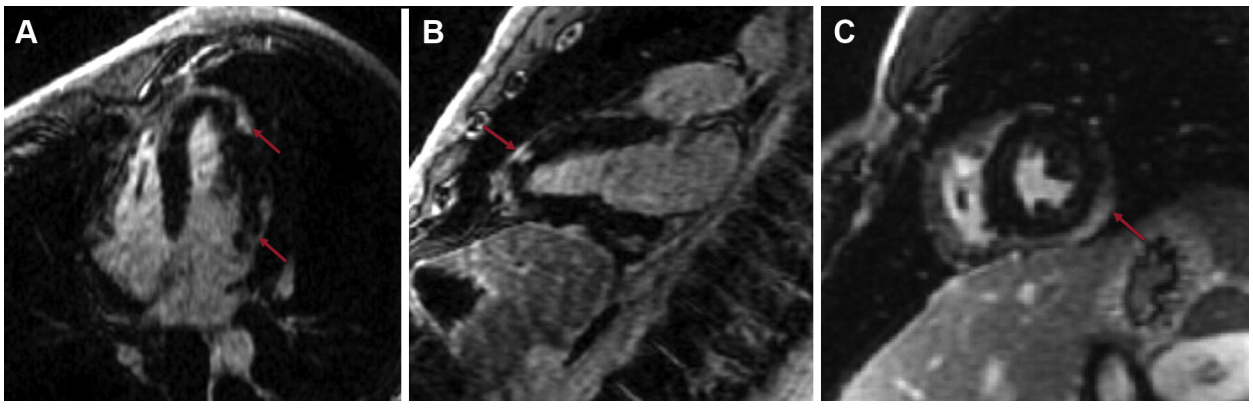
Right coronary artery originates from left coronary sinus with an interarterial course between aortic root and main pulmonary artery **(red arrow)**.

The patient was otherwise healthy before this admission; the possible explanation for his clinical picture was that the anomalous RCA was unmasked by the myocardial inflammation and edema from acute perimyocarditis. This case highlights that in patients with recent acute perimyocarditis, invasive or noninvasive coronary angiography should be considered to rule out CAD or possible ACA if chest pain continues after the acute phase of inflammation.

With more frequent use of diagnostic imaging to rule out CAD, there has been an increase in absolute numbers of ACA identified. Medical treatment of ACA or surgical intervention is indicated for patients with symptoms, and evidence of myocardial ischemia or ACA with a malignant course. Malignant variants have been recognized in autopsy studies to be an underlying cause of SCD in young athletes.[4] The major gap for patients with asymptomatic malignant ACA is the lack of ability to adequately risk-stratify these patients for surgery versus observation. Functional noninvasive perfusion imaging can assess potential ischemia induced by dynamic compression of malignant ACA and should be considered in this population. For patients with ischemic symptoms or evidence of inducible ischemia, like our patient, both the 2018 American College of Cardiology/American Heart Association guideline for the management of adult congenital heart disease and the American Association of Thoracic Surgery expert consensus guideline for anomalous origin of a coronary artery recommend activity restriction and surgical treatment (Class I; Level of Evidence: B).[2,5] Our patient was not initially considered for surgery because he was completely asymptomatic before this admission. In asymptomatic patients with similar ACA, the guideline states a Class IIa indication for the evaluation of inducible ischemia using an exercise stress test with additional imaging.[5] Different surgical procedures have been proposed to eliminate the potential cause of myocardial ischemia. These procedures include unroofing, pulmonary artery translocation, reimplantation, osteoplasty, and bypass grafting.[6] For this patient, pulmonary artery translocation was done without RCA unroofing because no significant intramural segment was seen intraoperatively.

Elderly patients with ACA can be difficult to treat because this population could have significant

FIGURE 4 Cardiac Magnetic Resonance



(A) 4-chamber view. **(B)** 2-chamber view. **(C)** Short-axis view demonstrating pericardial thickening **(red arrow in B)** and late gadolinium enhancement of lateral and inferolateral walls **(red arrows in A and C)**.

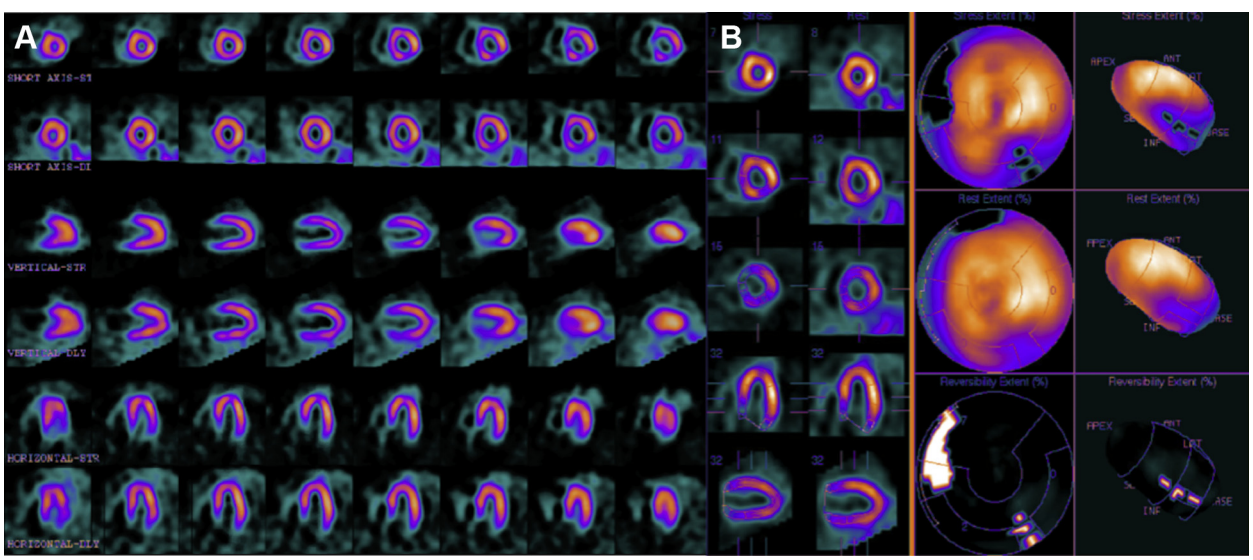
atherosclerosis and tortuosity of the vessels, making percutaneous intervention or surgical intervention of ACA challenging if become symptomatic with concurrent obstructive coronary artery disease.[7,8]

The patient's chest pain resolved after surgery, but he started to have nonsustained ventricular tachycardia (VT) postoperatively. Despite medical therapy with diltiazem 180 mg daily, he continued to have symptomatic nonsustained on loop recorder.

QUESTION 6: WHAT SHOULD BE THE MANAGEMENT PLAN FOR PATIENT'S RECURRENT NONSUSTAINED VT?

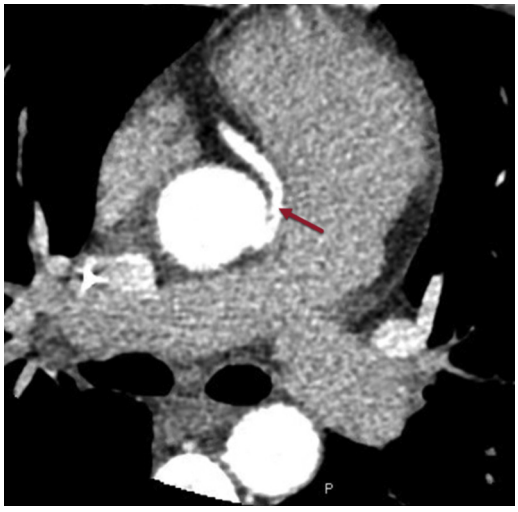
Repeat coronary CT angiography showed successful translocation of the main pulmonary artery (Figure 6). The results of additional workup, including repeat echocardiography, electrolytes, and serial ECG, were unremarkable. He eventually underwent implantation of an implantable cardioverter defibrillator (ICD).

FIGURE 5 Nuclear Medicine Myocardial Perfusion Imaging



Attenuation-corrected stress and rest myocardial perfusion imaging examination revealed **(A)** small moderate-intensity predominantly reversible perfusion defect of the mid to basal inferolateral wall of left ventricle. **(B)** Polar maps redemonstrated left ventricular reversible perfusion defect of inferolateral wall.

FIGURE 6 Repeat Coronary Artery Computed Tomography Angiography



No impingement of the right coronary artery by aortic root and main pulmonary artery **(red arrow)**.

Patients who have undergone surgical repair of anomalous ACA remain at risk. Short-term to midterm complications of surgery include aortic valve insufficiency, ischemic symptoms, ischemic change on proactive testing, arrhythmia, and as SCD.[3,8,9] Thus, the guideline recommends close followup care with serial ECGs and cardiac imaging after surgery.[5] Lifelong follow-up care is also important because the long-term outcomes from surgical repair are largely unknown. Although both perimyocarditis and anomalous RCA increase the risk of ventricular arrhythmia,[3,9] our patient had no detected ventricular arrhythmia before surgical intervention. He had short episodes of VT after surgery, and he continued to have nonsustained VT despite medical therapy with diltiazem. The incidence of ventricular arrhythmia after surgical repair of ACA could be still as high as 7%.[10] For patients with continued VT without reversible or correctable causes in whom medical therapy has failed, ICD may be considered for primary prevention.

REMAINDER OF THE CLINICAL COURSE

The patient was doing well at his 18-month follow-up visit. He continued to take diltiazem 180 mg daily, and interrogation of his ICD showed no recurrence of VT.

CONCLUSIONS

Asymptomatic ACA may become symptomatic after myocardial inflammation, and symptoms can persist. Surgical repair should be considered for patients with evidence of ischemia. Postsurgical monitoring is critical to detect or prevent short-term and long-term complications.

ACKNOWLEDGMENTS The authors thank Chris Smarte and Stella Li for their valuable contribution.

FUNDING SUPPORT AND AUTHOR DISCLOSURES

The authors have reported that they have no relationships relevant to the contents of this paper to disclose.

ADDRESS FOR CORRESPONDENCE: Dr Xiaoxiao Qian, Department of Cardiology, Inova Heart and Vascular Institute, 3300 Gallows Road, Falls Church, Virginia 22102, USA. E-mail: xiaoxiao.qian@inova.org. Twitter: qian_xiaoxiao.

REFERENCES

1. Behbahani-Nejad O, Mikolich B, Morenstern D, Mikolich JR. Myocarditis response to colchicine therapy based on cardiac MRI diagnostic criteria. *J Am Coll Cardiol*. 2021;77(18suppl1):1432.

2. Smith SC Jr, Jacobs AK, Adams CD. ACC/AHA 2008 Guidelines for the management of adults with congenital heart disease: executive summary. *J Am Coll Cardiol*. 2008;52:1890–1947.

3. Angelini P. Coronary artery anomalies: an entity in search of an identity. *Circulation*. 2007;115:1296–1305.

4. 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;141:829–834.

5. Verhagen JMA, Kempers M, Cozijnsen L, et al. Expert consensus recommendations on the cardiogenetic care for patients with thoracic aortic disease and their first-degree relatives. *Int J Cardiol*. 2018;258:243–248.

6. Brothers JA, Frommelt MA, Jaquiss RDB, et al. Expert consensus guidelines: anomalous aortic origin of a coronary artery. *J Thorac Cardiovasc Surg*. 2017;153(6):1440–1457.

7. Khan HR, Hashim H, Bhandari M. Anomalous origin of the epicardial left coronary artery from the right coronary sinus revealed in a patient presenting with acute myocardial infarction. *J Ayub Med Coll Abbottabad*. 2016;28:420–422.

8. Tuo G, Marasini M, Brunelli C, et al. Incidence and clinical relevance of primary congenital anomalies of the coronary arteries in children and adults. *Cardiol Young*. 2013;23:381–386.

9. Rosier L, Zouaghi A, Barré V, et al. High risk of sustained ventricular arrhythmia recurrence after acute myocarditis. *J Clin Med*. 2020;9:848.

10. Nees SN, Flyer JN, Chelliah A, et al. Patients with anomalous aortic origin of the coronary artery remain at risk after surgical repair. *J Thorac Cardiovasc Surg*. 2018;155:2554–2564.

KEY WORDS anomalous coronary arteries, chest pain, perimyocarditis, ventricular tachycardia

APPENDIX For a supplemental figure and videos, please see the online version of this article.