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CASE REPORT

BEGINNER

CLINICAL CASE

Anomalous Origin of a Right Coronary Artery from the Pulmonary Artery in a Middle-Aged Woman

Yogamaya Mantha, MD,[a] Mark Feldman, MD,[a] Andrew Assaf, MD,[a] Rakushumimarika Harada, MD,[a] David Musselman, MD,[c] Michinari Hieda, MD, MS, PhD,[a,b] Darryl Kawalsky, MD[c]

ABSTRACT

A 53-year-old female patient presented for evaluation of a murmur. The examination revealed a 2/6 systolic ejection murmur in the left upper sternal border. Transthoracic echocardiography with color Doppler showed increased blood flow around the apex of the right ventricle. Further imaging revealed the right coronary artery emerging from the pulmonary artery. (**Level of Difficulty: Beginner.**) (J Am Coll Cardiol Case Rep 2020;2:464–7) © 2020 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/).

PRESENTATION

A 53-year-old female patient presented for evaluation of a murmur. The examination revealed a blood pressure of 145/82 mm Hg, and a grade of 2/6 systolic ejection murmur, heard best at the left upper sternal border. Heart sounds S_1 and S_2 were normal without cardiac gallops or diastolic murmurs.

LEARNING OBJECTIVES

- Investigate the cause of unexplained murmurs using multimodality imaging.
- Consider the possibility of adult congenital heart disease.
- Understand the pathophysiology of myocardial ischemia secondary to abnormalities in coronary circulation.
- Apply the American Heart Association/American College of Cardiology guidelines and treatment recommendations for appropriately anomalous right coronary artery from the pulmonary artery.

MEDICAL HISTORY. The patient had hypertension, which was treated with daily lisinopril/hydrochlorothiazide (10/12.5 mg).

DIFFERENTIAL DIAGNOSIS. Differential diagnoses of a systolic ejection murmur are broad and include ventricular septal defect, aortic valve stenosis, aortic bicuspid valve, pulmonary stenosis, or anomalous right coronary artery from pulmonary artery (ARCAPA).

INVESTIGATIONS. Electrocardiography showed moderate T-wave inversions in the inferior and anterior lateral leads (**Figure 1**). Cardiac nuclear stress testing using an A2A adenosine receptor agonist

From the [a]Internal Medicine Department, Texas Health Presbyterian Hospital of Dallas, Dallas, Texas; [b]Division of Cardiology, Institute for Exercise and Environmental Medicine, University of Texas Southwestern Medical Center, Dallas, Texas; and the [c]Cardiovascular Department, North Texas Heart Center, Dallas, Texas. Dr. Hieda is supported in part by American Heart Association Strategically Focused Research Network grant 14SFRN20600009-03, by American Heart Association postdoctoral fellowship grant 18POST33960092, and by the Harry S. Moss Heart Trust. All other authors have reported that they have no relationships relevant to the contents of this paper to disclose.

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(regadenoson injection) revealed no evidence of myocardial ischemia. Transthoracic echocardiography (TTE) revealed a normal left ventricular (LV) ejection fraction with intact aortic and pulmonary valves and high Doppler flow around the apex of the right ventricle suggesting the possibility of a ventricular septal defect (**Figure 2**). However, transesophageal echocardiography results excluded a ventricular septal defect. In order to elucidate the cause of the occult flow seen in the Doppler TTE, a multislice computed tomography angiogram (MSCT) was performed which revealed that the origin of the RCA was from the main pulmonary artery (**Figure 3**). Subsequent left and right heart catheterizations revealed normal LV systolic function, a very high LV end-diastolic pressure of 30 mm Hg with otherwise unremarkable left heart hemodynamics, mild pulmonary hypertension, and a moderately large left-to-right shunt (Qp/Qs of 1.45) in addition to massively dilated coronary arteries with flow from the left coronary system to the right coronary system. Given this finding, the patient underwent ligation of the RCA at its origin from the pulmonary artery with a saphenous vein bypass graft from the aorta to the RCA in its midportion (**Figure 4**).

MANAGEMENT. According to the latest American Heart Association/American College of Cardiology guidelines, ligation of the RCA is Class I recommendation for symptomatic patients with ARCAPA and a Class IIa recommendation in asymptomatic patients with ventricular dysfunction or myocardial ischemia attributed to ARCAPA (1). Given this finding, the patient underwent ligation of the RCA at its origin from the pulmonary artery with a saphenous vein bypass graft from the aorta to the RCA (**Figure 4**).

DISCUSSION

Congenital coronary artery anomalies are rare in adults, with an incidence of 0.3% to 0.9% (2). Coronary artery anomalies arise in the embryonic stage of the fetus as a result of malrotation of the spiral septum or malposition of the coronary buds (3). The 4 types of anomalies of the coronary arteries originating from the pulmonary artery are: 1) the anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA); 2) the anomalous origin of the ARCAPA; 3) the origin of both coronary arteries from the pulmonary artery; and 4) an accessory coronary artery from the pulmonary artery (3). ALCAPA is the most common of these anomalies, with an incidence of 0.008% (1 in 12,500 persons), and ALCAPA presents mostly as heart failure in infancy (4). The incidence of ALCAPA is believed to be higher than that of ARCAPA due to the proximity of the left coronary bud to the pulmonary artery sinus (5).

The incidence of ARCAPA is 0.002% (1 in 50,000 persons) (6). The high pulmonary vascular resistance in neonates can permit forward perfusion of an ectopic RCA flow from the pulmonary artery. A decline in the pulmonary vascular resistance after birth will produce a steal phenomenon leading to heart remodeling and a subsequent left-to-right shunt of blood from the RCA to the PA. Patients may remain asymptomatic until adulthood. Presentations may include exertional chest pain, syncope, sudden cardiac death, heart failure, and myocardial infarction (7). Surgical correction is recommended even in asymptomatic patients, such as in the present case.

The pathophysiology of ARCAPA is dependent on the direction of blood flow in the coronary arteries and the rate of oxygen delivery to the myocardium (8). The anomalous artery has retrograde blood flow due to the pressure differences between systemic and pulmonary circulation, which leads to an intercoronary steal syndrome (left coronary artery [LCA]-to-RCA). The severity of myocardial ischemia is determined by the size of the left-to-right shunt, the degree of collateral circulation, and the myocardial oxygen demand (8). Any increase in myocardial oxygen demand results in exhaustion of the physiological reserve and may lead to ischemia, infarction, and

ABBREVIATIONS AND ACRONYMS

ALCAPA = anomalous left coronary artery from pulmonary artery

ARCAPA = anomalous right coronary artery from pulmonary artery

CMR = cardiac magnetic resonance

LCA = left coronary artery

LV = left ventricular

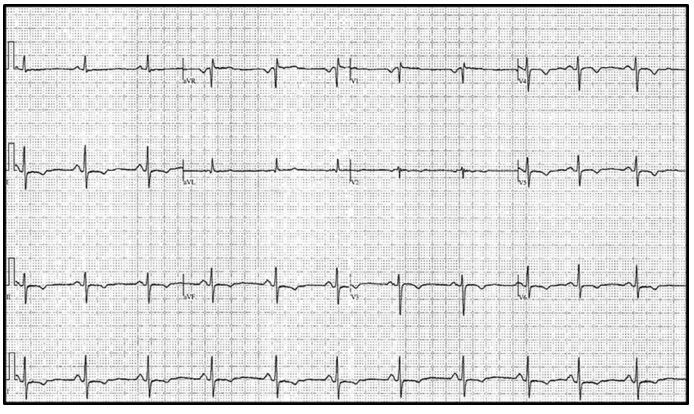
MSCT = multi-slice computed tomography

PA = pulmonary artery

RCA = right coronary artery

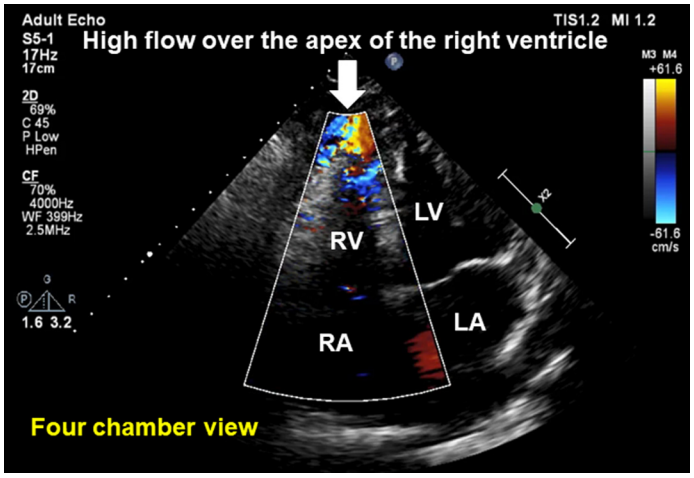
TTE = transthoracic echocardiogram

FIGURE 1 Electrocardiography Revealed T-Wave Inversions



Electrocardiography revealed T-wave inversions in the inferior and anterolateral leads.

FIGURE 2 Transthoracic Echocardiography Revealed High Flow Over the Apex of the Right Ventricle



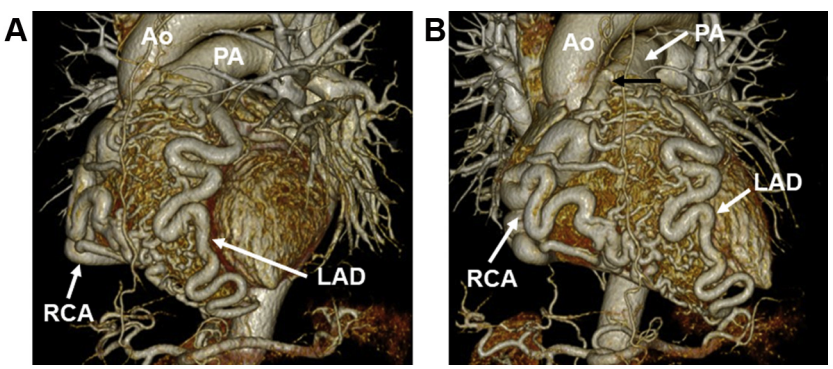
LA = left atrium; LV = left ventricle; RA = right atrium; RV = right ventricle.

sudden cardiac death (4). Due to the reduced ventricular workload and oxygen demands of the right ventricle compared to that of the left ventricle, ventricular ischemia is less prominent in ARCAPA than in ALCAPA. However, ARCAPA patients with a right dominant coronary circulation have increased adverse outcomes than patients with a left dominant circulation (8,9). Imaging studies in the present patient also showed dilated RCA and LCA, which can be explained by either LCA-to-RCA shunting or fistula formation due to hypoxic conditions with the production of hypoxia-inducible factor (HIF)-1 and vascular endothelial growth factors (VEGF). It is possible that the patient developed a fistula between the RCA and LCA in the setting of oxidative stress which led to the cascade of arterial remodeling and signal production (10). HIF-1 and/or VEGF is the key factor that regulates this cascade (10).

Although ARCAPA is usually diagnosed using echocardiography with color Doppler, coronary angiography is necessary for a definitive diagnosis. Electrocardiography findings in ARCAPA can show LV hypertrophy or deep Q waves in the inferior leads, which reflect prior myocardial infarction (11). Both cardiac magnetic resonance (CMR) imaging and MSCT are imaging modalities for detecting coronary artery abnormalities. MSCT and CMR imaging are recommended because they allow more definite spatial resolution which can be helpful for surgery. Different surgical procedures have been postulated for the management of ARCAPA. Two of the procedures described are the ligation of the right coronary at its origin from the pulmonary trunk with or without saphenous vein grafting from the ascending aorta to the RCA, or reimplantation of the anomalous RCA from the pulmonary artery into the ascending aorta to recreate a dual coronary system (12,13).

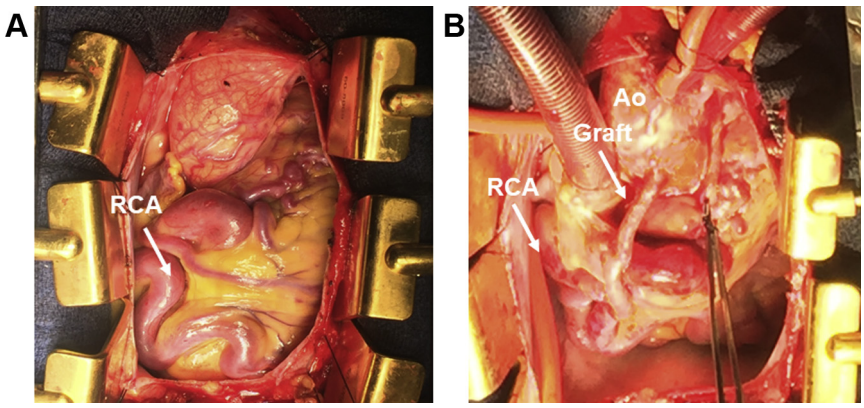
FOLLOW-UP. The patient tolerated the procedure well and was discharged 5 days after cardiac surgery. The patient has done well after the surgery for the past 2 years.

FIGURE 3 Computerized Tomography with Contrast



(A, B) Computed tomography with contrast revealed right coronary originating from the pulmonary trunk **(black arrows)** and left and right dilated coronary arteries **(white arrows).** Ao = ascending aorta; LCA = left coronary artery; PA = pulmonary artery; RCA = right coronary artery.

FIGURE 4 Surgeons View



(A) Dilated right coronary artery at its origin from the pulmonary artery. **(B)** Saphenous vein bypass from the aorta to the mid-right coronary artery. Ao = ascending aorta; LCA = left coronary artery; PA = pulmonary artery; RCA = right coronary artery.

CONCLUSIONS

This case highlights the rare condition of ARCAPA, which may lead to heart failure or sudden cardiac death. Because these complications are associated with ARCAPA, our patient underwent a corrective operation. MSCT and CMR imaging are useful tools for diagnosing adult congenital diseases.

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ADDRESS FOR CORRESPONDENCE: Dr. Darryl Kawalsky, North Texas Heart Center, 8440 Walnut Hill Lane, Suite 700, Dallas, Texas 75231. E-mail: dkawalsky@gmail.com.

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