


Heart Failure as the Initial Presentation of Anomalous Left Coronary Artery From the Pulmonary Artery

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Abstract

Coronary arteries arising from the pulmonary artery have an incidence of 0.002% in the general population. We present a 29-year-old woman who presented to our hospital with acute decompensated heart failure and atrial fibrillation with a rapid ventricular rate. She underwent a cardiac catheterization to rule out ischemic disease, which revealed retrograde contrast flow through the left coronary artery from the right coronary artery. A coronary computed tomography (CT) angiogram was pursued which showed the presence of an anomalous left coronary artery arising from the pulmonary artery (ALCAPA). For the management of her atrial fibrillation, she was electrically cardioverted. She was discharged on guideline-directed medical therapy for her heart failure, with a cardiac surgery referral for the surgical fixation of her ALCAPA.

Keywords

ALCAPA, anomalous coronary artery, acute decompensated heart failure, atrial fibrillation

Introduction

Coronary artery origin from the pulmonary artery has an incidence of 0.002% in the general population, and for this reason, the diagnosis may be overlooked.^{1,2} Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) usually leads to significant morbidity and mortality in infants if not discovered early, but a minority of patients develop collateral circulation from the right coronary artery (RCA) and may remain asymptomatic or minimally symptomatic into adulthood.

Case Presentation

A G2P2 29-year-old woman presented with a 1-week history of shortness of breath. She noted 2-pillow orthopnea, weight gain of 7 kg in 6 months, and worsening leg edema. Initial vital signs were a blood pressure of 135/81 mm Hg, heart rate of 101 beats per minute, respiratory rate of 18 breaths per minute, and oxygen saturation of 97% on room air. Physical examination revealed a young woman who was tachypneic with vesicular breath sounds, tachycardic with an irregularly irregular rhythm. She had 1+ pitting edema of the lower extremities, there was no jugular venous distension (JVD) nor hepatojugular reflux.

Past medical history was significant for paroxysmal atrial fibrillation, a stroke 6 months prior, asthma, major depressive disorder, generalized anxiety disorder, and binge-eating disorder. Six months preceding her presentation, she had come into the emergency department for palpitations. Her electrocardiogram (EKG) at the time revealed atrial fibrillation, and she was electrically cardioverted. Three days later, she was diagnosed with a left middle cerebral artery stroke. A transthoracic echocardiogram (TTE) was done at that time which showed an ejection fraction (EF) of 50% to 55% and a grade-3 diastolic dysfunction. She was discharged on a Holter monitor, which revealed rate-controlled atrial fibrillation.

Her home medications were apixaban 5 mg twice daily, albuterol as needed, montelukast 10 mg daily, atorvastatin

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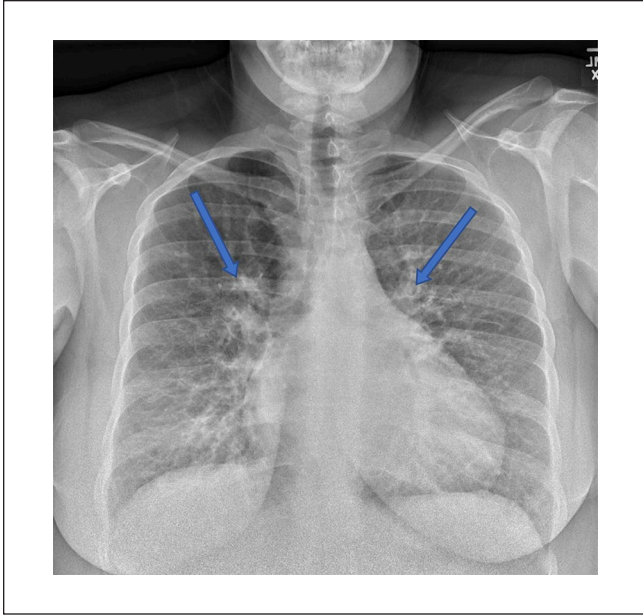


Image 1. Chest X-ray showed cardiomegaly with perihilar hilar vascular congestion (arrows).

80 mg daily, and newly added metoprolol tartrate 25 mg twice daily.

Two troponin levels came back less than 0.01 ng/mL, and proBNP was noted to be 2788 pg/mL.

Chest radiography (CXR) (Image 1) showed cardiomegaly with perihilar vascular congestion. Electrocardiogram (Image 2) showed atrial fibrillation with rapid ventricular rate at 116 beats per minute with prolonged QTc.

Transthoracic echocardiogram (Image 3) showed an EF of 32%, left ventricular hypertrophy, moderate diffuse hypokinesis, grade-2 diastolic dysfunction, right ventricular enlargement, and severe left atrial dilation.

Cardiac catheterization demonstrated a very large caliber dominant RCA which fully collateralized the left coronary artery (LCA) system. No coronary artery disease was noted. Contrast from the RCA via collaterals was seen to flow retrograde through the LCA all the way to the left main coronary (LMC) and reflux into a vessel other than the aorta, suspicious for an LCA with an anomalous origin from the pulmonary artery. The catheterization revealed an elevated LVEDP (35 mm Hg), moderate pulmonary artery hypertension (mean pressure of 50 mm Hg), and a small left to right shunt (QP/QS) of 1.1.

A computerized tomography angiography (CTA) of the coronary arteries (Images 4-6) showed an anomalous LMC artery arising from the undersurface of the main pulmonary artery. Both the left ventricle and left atrium were noted to be dilated. The main pulmonary artery was mildly dilated at 3.3 cm.

The Pediatric Cardiology/Adult Congenital Heart Service was consulted, who confirmed that the patient had efficient

collateral circulation from the right to the left coronary system and that her current symptoms and signs of heart failure were likely related to her cardiomyopathy and atrial fibrillation. For this reason, surgical intervention was not warranted at this time but could be considered in the future. She underwent transesophageal echocardiogram (TEE) followed by electrical cardioversion. She remained in sinus rhythm and was discharged. She was discharged on guideline-directed medical therapy for nonischemic cardiomyopathy and paroxysmal atrial fibrillation.

Discussion

In the case of ALCAPA, *coronary blood flow* becomes limited, and ischemia will occur as pulmonary vascular resistance and pressure naturally decrease during the first few months of age. If the decrease is sudden in early infancy, then ischemia, left ventricular hypokinesis, and heart failure will develop, and the infant will present with a clinical picture of dilated cardiomyopathy.³

ALCAPA has multiple presentations. If the decrease in pulmonary vascular resistance is very gradual, then collateral perfusion from the RCA (arising from the aorta) to the LCA and its distribution will occur, and this may reduce or eliminate the development of ischemia, left ventricular hypokinesis and symptoms. When collateralization develops extensively, left ventricular function may be preserved and lead to asymptomatic survival to adulthood as in our patient. In between these 2 clinical presentations is the rarer patient with borderline collateralization who presents with heart failure or arrhythmias in later childhood. Papillary muscle ischemia and dysfunction leading to mitral incompetence may also occur and complicate the clinical picture at any age, and it is assumed that arrhythmias are generally more likely with this diagnosis.⁴

(Electrocardiogram) often demonstrates anterolateral ischemia in symptomatic patients and CTA or magnetic resonance imaging (MRI) can delineate the coronary artery anatomy.⁵⁻⁷ Still, the gold standard for diagnosis of an anomalous coronary artery is coronary angiography. Surgical treatment options depend on the clinical symptoms and degree of left ventricular ischemia or hypokinesis. Surgical procedures have included ligation of the proximal LCA with or without coronary artery bypass grafting, surgical reimplantation of the anomalous left coronary into the ascending aorta, or the Takeuchi procedure. This procedure connects the orifice of the anomalous LCA from the main pulmonary artery to the ascending aorta by means of an intrapulmonary tunnel that crosses the lumen of the main pulmonary artery.⁸

Our case highlights the importance of suspecting congenital anomalies in relatively younger (<30 years old) patients who present with heart failure without any significant risk factors. A delayed presentation at 29, implied adequate collateral circulation in our patient, which was confirmed on catheterization. Early diagnosis of anomalous coronary

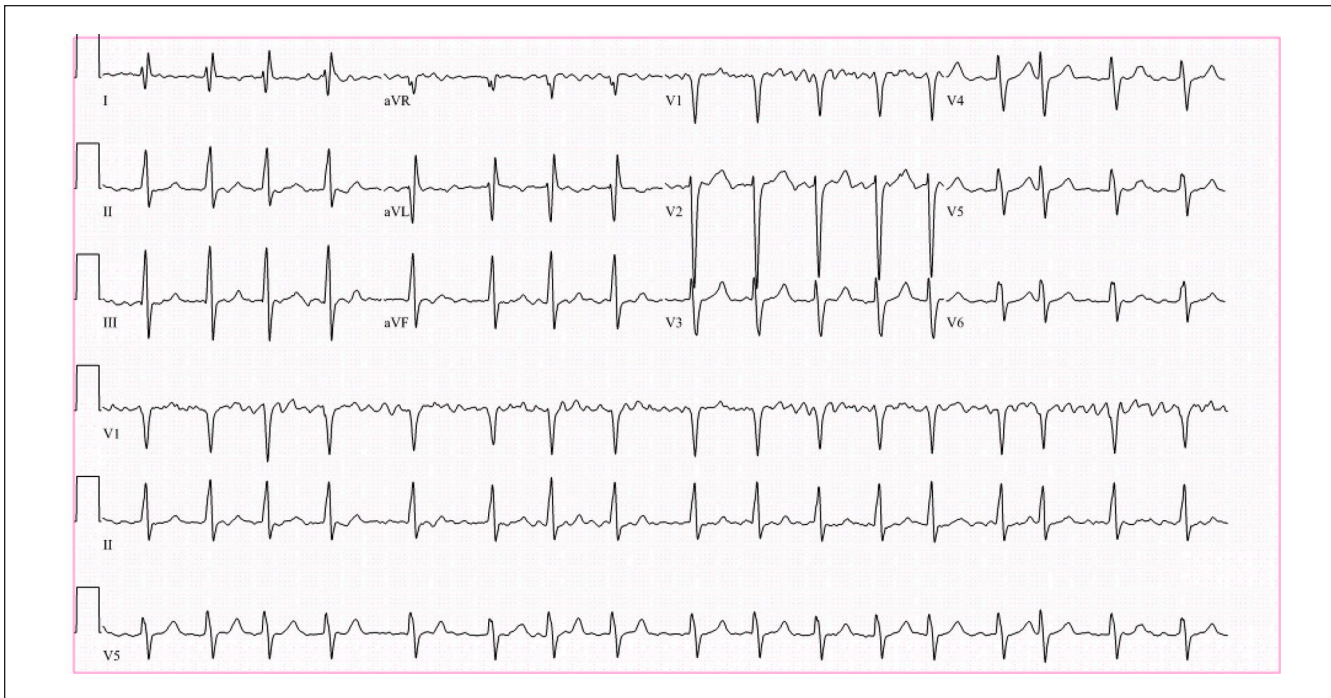


Image 2. EKG showed atrial fibrillation with rapid ventricular fibrillation.
Abbreviation: EKG, electrocardiogram.

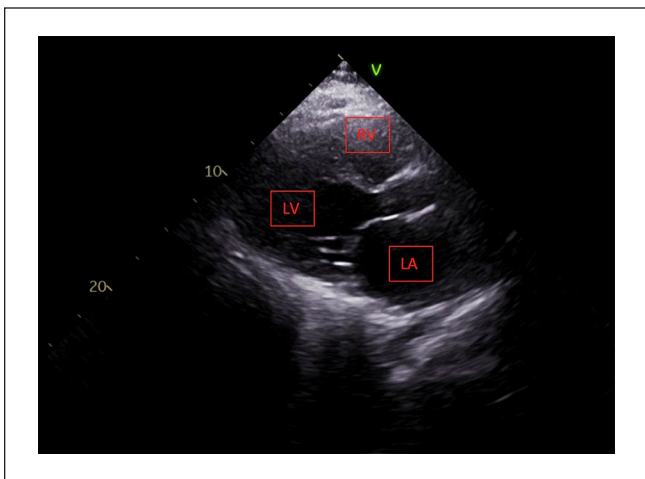


Image 3. TTE showing left ventricular hypertrophy, right ventricular enlargement, and severe left atrial dilation.
Abbreviation: LA, left atrium; LV, left ventricle; RV, right ventricle; TTE, transthoracic echocardiogram.

arteries is necessary to ensure that complications, such as congestive heart failure and atrial fibrillation (driven by either the heart failure or inadequate blood supply to the left atrium) do not occur, such as in our patient.

In the acute setting, we felt that our patient’s presenting symptoms and signs were secondary to tachycardia-induced cardiomyopathy, compounded by some degree of long-term ischemic cardiomyopathy secondary to the presence of an ALCAPA. As a result, controlling her atrial fibrillation and

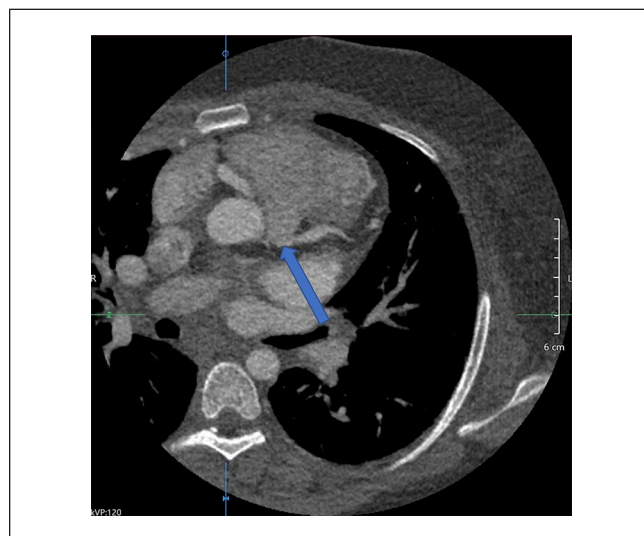


Image 4. A computerized tomography angiography of the coronary arteries showing the left main coronary artery arising from the undersurface of the main pulmonary artery (arrow).

management of her nonischemic cardiomyopathy became the focus of her current inpatient admission.

Our patient was seen outpatient 1 week following discharge. She did not have any acute complaints and remained compensated in sinus rhythm. Transthoracic echocardiogram performed 2 months after presentation revealed an improved EF of 50%, while maintaining sinus rhythm.

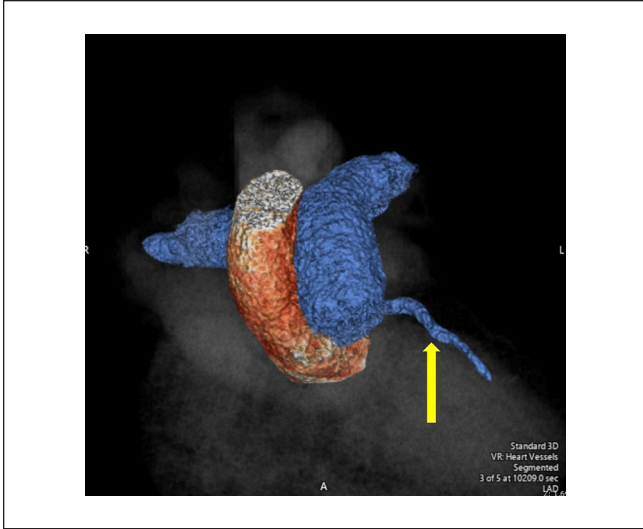


Image 5. A 3D reconstruction of the coronary arteries using CTA, with the arrow indicating LCA. Abbreviations: CTA, computerized tomography angiography; LCA, left coronary artery.

Conclusion

Anomalous coronary arteries that arise from the pulmonary artery are rare phenomena. Our case portrays the importance of including this disease in the differential diagnosis of younger individuals who present with worsening heart failure, without apparent instigating factors. Appropriate diagnosis with cardiac catheterization and management of anomalous coronary arteries is necessary to prevent complications, such as decompensated heart failure, cardiogenic shock, or myocardial infarction.

Declaration of Conflicting Interests

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Ethical Approval

Our institution does not require ethical approval for reporting individual cases or case series.

Informed Consent

Verbal informed consent was obtained from the patient for their anonymized information to be published in this article.

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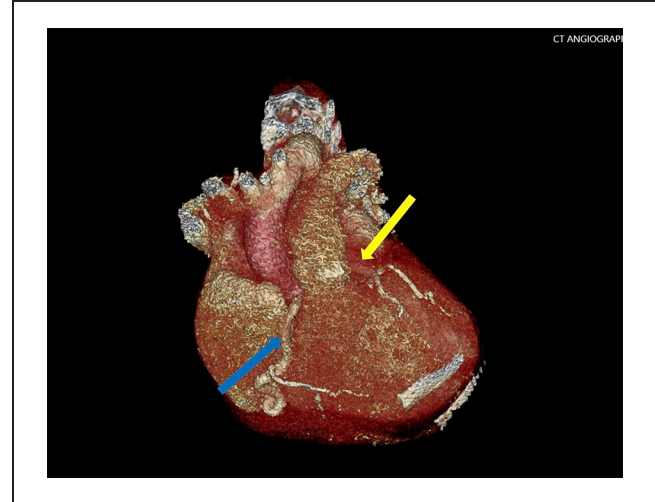


Image 6. 3D reconstruction of the heart with the left coronary originating from the pulmonary artery (yellow arrow) and the RCA originating from the aorta (blue arrow). Abbreviation: RCA, right coronary artery.

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