Anomalous Origin of the Right Coronary Artery from the Pulmonary Artery in a Morbidly Obese Patient Presenting with Chest Pain

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Conflict of interest: None declared

Patient: Female, 58
Final Diagnosis: Anomalous origin of RCA from PA with reversible myocardial ischemia of the inferior wall
Symptoms: Chest pain • shortness of breath
Medication: —
Clinical Procedure: —
Specialty: Cardiology

Objective: Congenital defects/diseases
Background: Anomalous origin of the right coronary artery from the pulmonary artery (ARCAPA) is a rare congenital anomaly with an incidence of 0.002%.
Case Report: A 58-year-old African American female with a history of diabetes mellitus, hyperlipidemia, and hypertension was evaluated for shortness of breath and chest heaviness. On physical examination, she was found to be morbidly obese. Her blood pressure was 160/90 mmHg. There were no carotid bruits or jugular venous distension. Cardiac auscultation showed distant heart sounds with no audible murmurs. Lower extremity examination showed +1 edema with weak pedal pulses. ECG showed non-specific ST segment and T-wave changes. Echocardiogram demonstrated dysfunction grade 1 with preserved ejection fraction. An adenosine nuclear study showed an area of reversible ischemia of the inferior wall. Selective left coronary angiography showed the left coronary artery (LCA) originating from the left sinus of Valsalva. From the LCA, the left anterior descending and the left circumflex coronary arteries arose in a typical course. The right coronary artery (RCA) was visualized in a retrograde fashion via collaterals originating from the left coronary system and it drained into the pulmonary artery. On aortic root angiography, the origin of the RCA could not be determined. The patient’s surgical risk was deemed unacceptably high and she was not considered a surgical candidate. Her symptoms were controlled conservatively.

Conclusions: By reporting this case, we shed some light on a rare congenital anomaly involving the coronary arteries. Variable presentations have been described for ARCAPA, however, many patients remain asymptomatic. Diagnosis can be confirmed by coronary angiography. Surgical correction is the definitive treatment.

MeSH Keywords: Coronary Vessel Anomalies • Coronary Vessels • Pulmonary Artery

Full-text PDF: https://www.amjcaserep.com/abstract/index/idArt/910820
Background

Congenital abnormalities involving coronary arteries are relatively rare [1]. Among these abnormalities, anomalous origin of the right coronary artery from the pulmonary artery (ARCAPA) stands as an exceedingly rare anomaly with an incidence estimated to be 0.002% [2].

Case Report

A 58-year-old morbidly obese African American female with a history of diabetes mellitus, hyperlipidemia, hypertension, chronic kidney disease, and smoking was referred for evaluation of shortness of breath and chest heaviness. She described her pain as “it feels like acid reflux”. Her family history was of no relevance, with no record of premature coronary heart disease or sudden cardiac death.

On physical examination, the patient was determined to be morbidly obese with a body mass index (BMI) of 51.8 kg/m$^2$. Her blood pressure was 160/90 mmHg. There were no carotid bruits or jugular venous distension. Cardiac auscultation showed distant heart sounds with no audible murmurs or gallops. Lower extremity examination showed +1 edema with weak pedal pulses.

Her current medications included nisoldipine 20 mg daily, losartan/hydrochlorothiazide 100/25 mg per day, atenolol 25 mg per day, and lansoprazole 30 mg per day.

The patient’s ECG showed non-specific ST segment and T-wave changes. Initial echocardiogram demonstrated left ventricular hypertrophy, left atrial enlargement, and diastolic dysfunction grade I with preserved ejection fraction. An adenosine nuclear study showed an area of reversible ischemia of the inferior wall. Subsequently, she underwent right and left cardiac catheterization.

Selective left coronary angiography demonstrated short left main coronary artery (LCA) that originated appropriately from the left sinus of Valsalva. From the LCA, left anterior descending (LAD) and left circumflex (LCX) coronary arteries arose in a typical course (Figures 1A, 2A). Both LAD and LCX were dilated and tortuous without significant stenosis. The right coronary artery (RCA) was visualized in a retrograde fashion almost instantaneously via collaterals originating from the left coronary system (Figure 1B, Video 1). The RCA was markedly tortuous and dilated with no significant stenosis. It drained into the main pulmonary artery (Figure 2B, Video 2). On aortic root angiography, the origin of the RCA could not be determined (Figure 3, Video 3).

To better assess the significance of this anomaly, O$_2$ saturation run was obtained from her right cardiac chambers. All measurements are listed in Table 1. Using the Fick principle, systemic cardiac output was calculated to be 5.98 L/min, and the pulmonary flow was 5.98 L/min. The pulmonary to systemic flow ratio was of 1:1. No significant step-up in blood oxygen saturation was detected in the pulmonary artery. The patient was not considered a surgical candidate and her symptoms were controlled conservatively.
Discussion

Anomalous coronary vessels are relatively rare congenital defects. The incidence of such anomalies has been reported in the medical literature to range from 0.3% to 0.9% [1]. Among these abnormalities, anomalous origin of the RCA from the pulmonary artery (PA) (or ARCAPA) stands as an exceedingly rare anomaly with an incidence estimated to be 0.002%. In 1885, Brooks identified the first 2 cases of ARCAPA postmortem [3]. Since then, just over 100 cases have been reported [4]. In 1942, Soloff et al. described 4 anatomical variations for anomalous origin of the coronary arteries from the PA. Namely, anomalous origin of the left coronary artery (LCA) from the PA (ALCAPA), ARCAPA, both the LCA and the RCA originating from the PA, and an accessory coronary vessel originating from PA [5].

Other congenital heart defects have been reported concurrently in 22% of cases of ARCAPA, such as aorto-pulmonary window (30% of associated defects) and tetralogy of Fallot (19%) [5]. Investigations of the associated defects might lead to incidental identification of the anomalous vessels early in life. When isolated, however, many patients with ARCAPA remain asymptomatic, which suggests an under estimation of the true prevalence of this anomaly in the general population [6].

Figure 2. Selective left coronary angiography in LAO-caudal view shows: (A) LCA and (B) RCA filling retrogradely from LCA and terminating in the main pulmonary artery. LCx – left circumflex artery, OM – obtuse marginal branch, PA – pulmonary artery, RCA – right coronary artery.

Video 1. Selective left coronary angiography in RAO-30° view.

Video 2. Selective left coronary angiography in LAO-caudal view.
The symptomatology of ARCAPA largely correlates with the underlying pathophysiology. Potential factors that can determine the age of onset and the nature of symptoms include the direction of filling in the anomalous vessel, the formation of collateral vessels with the left coronary system, and the significance of the resulting left to right shunt [2]. In our case, retrograde filling of the RCA was demonstrated almost instantaneously on selective left coronary angiography. This was thought to be due to massive collateralization from the normally arising LCA. Additionally, the absence of notable step-up in blood oxygen saturation may explain the mild nature of our patient's symptoms.

**Table 1.** Hemodynamic parameters and quantitative coronary angiography measurements in a patient with anomalous origin of the right coronary artery from the pulmonary trunk.

| Chamber                        | Pressure | O₂ saturation |
|--------------------------------|----------|---------------|
| Right atrium (mean pressure)   | 16       | 65%           |
| Right ventricle                | 41/17    |               |
| Pulmonary artery               | 38/24    | 67%           |
| Pulmonary wedge mean pressure  | 20       |               |
| Left ventricle                 | 131/18   |               |
| Ascending aorta                | 126/72   | 94%           |

**Flows**

| Parameter                        | Value         |
|----------------------------------|---------------|
| Systemic cardiac output          | 5.98 L/min    |
| Cardiac index                    | 2.72 L/min/m² |
| Pulmonary blood flow             | 5.98 L/min    |
| Pulmonary/systemic flow ratio    | 1:1           |
| Ejection fraction                | 75%           |

**Quantitative coronary angiography measurements (mm)**

| Artery               | Diameter (mm) |
|----------------------|---------------|
| LAD (mean luminal diameter) | 7 mm         |
| RCA (mean luminal diameter)    | 8 mm         |

LAD – left anterior ascending artery; RCA – right coronary artery.
saturation in samples drawn from the pulmonary trunk questions the significance of left to right shunt. This could explain the relatively late presentation in our patient.

Of note, this pattern of semi-instantaneous retrograde filling into the RCA from the LCA, which was demonstrated in our case, has been reported in a few cases of single coronary artery (SCA) [6]. However, in cases of SCA, direct communication between the RCA and the PA is extremely rare [7]. In one case reported in 1963, Murray described a patient with SCA where a connection between the RCA and the PA did occur [8]. Unlike our case, however, the communication was of a fistulous pattern and not a direct termination. In his review on SCA, Smith concluded that a coronary artery arising from the PA would make the diagnosis of SCA highly improbable [7]. Consequently, the diagnosis of ARCAPA in our patient was considered much more likely compared to SCA.

While patients with ALCAPA present in early childhood with congestive heart failure and myocardial ischemia, those with ARCAPA tend to follow a benign course and present later in life [9]. Versatile clinical presentations have been described in the literature. In their review, Modi et al. reported angina as the most common symptom of ARCAPA, for which 18% of patients come to initial medical attention [5]. Other manifestations included exertional dyspnea, congestive heart failure, and sudden cardiac death.

Often, ARCAPA is diagnosed incidentally during angiography performed to evaluate other cardiac conditions [10]. While conventional angiography is commonly performed to diagnose coronary anomalies, noninvasive imaging modalities including cardiac computed tomography angiography and/or cardiac magnetic resonance imaging can also be utilized to delineate the coronary anatomy and provide comprehensive anatomic information on the anomalous vessel [11,12]. In our patient’s case, this anomaly was diagnosed on selective left coronary angiography in the course of investigating atypical chest pain with abnormal nuclear stress test.

Once ARCAPA is identified, surgical correction is advocated as the definitive treatment to reduce the risk of sudden cardiac death associated with having a single coronary system [13]. The anomalous RCA can be either re-implanted to the aorta or ligated [14]. However, long-term outcomes of such procedures were not well studied in the majority of cases, with a few patients reported to develop thrombosis in the implanted artery after surgical repair [2,14,15]. Conservative medical management has also been described for this anomaly, especially when risk of surgery was deemed to outweigh benefits [16]. Our patient was a morbidly obese woman with multiple comorbidities that significantly increase the risk of corrective surgery. In patients undergoing cardiac surgery, morbid obesity is associated with a 60% increase in mortality compared with normal weight patients [17]. In addition, morbidly obese patients are at greater risk of postoperative complications, including renal failure, deep sternal wound infection, prolonged ventilatory support, and pneumonia [17,18]. These complications were of particular concern in our patient given her medical history of chronic kidney disease, diabetes mellitus, and smoking. Therefore, she was not considered a surgical candidate and aggressive modification of risk factors including weight loss, smoking cessation, and adequate control of her diabetes mellitus was recommended. Once the surgical risk is deemed acceptable, corrective surgery should be considered for this patient to establish dual coronary system and reduce the risk of sudden cardiac death. In the meantime, the patient’s symptoms are being satisfactorily controlled on conservative medical treatment.

Conclusions

By reporting this case of ARCAPA, we shed some light on a rare congenital anomaly involving the coronary arteries. While variable presentations have been described for this anomaly, many patients remain asymptomatic. Diagnosis can be confirmed by conventional coronary angiography. Surgical correction by either re-implantation or ligation of the RCA is the definitive treatment. However, conservative management might be considered in patients with very high surgical risk.

Conflict of interest.

None.

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