Isolated Anomalous Right Coronary Artery From the Pulmonary Artery in Adulthood: Anatomical Features and Ischemic Burden

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A 51-year-old woman was admitted via the acute medical take after a prolonged episode of sharp central chest pain radiating to her neck and shoulder. The ECG was normal (Figure 1), as were subsequent consecutive troponin I measurements. Additional risk stratification with an exercise tolerance test was performed in consideration of her heavy smoking habit. At 85% of maximal predicted heart rate she developed chest pain and breathlessness with associated 2-mm inferolateral ST-segment depression (Figure 2).

Her coronary angiogram demonstrated unobstructed vessels but an unexpected anomalous origin of the right coronary artery (RCA) from the pulmonary artery (ARCAPA) with retrograde filling from the left system (Figure 3; online-only Data Supplement Movie I).

A cardiovascular magnetic resonance (CMR) was requested to further delineate the anatomy and the ischemic burden. Turbo spin echoes T1-weighted images confirmed the origin of the RCA from the main pulmonary artery and its course between the aorta and the pulmonary trunk (Figure 4). Adenosine stress perfusion images showed an inducible perfusion defect in the RCA territory (Figure 5; online-only Data Supplement Movie II). Left ventricular function was within normal limits, and there was no late gadolinium enhancement (ie, no myocardial scarring). After the CMR, the patient was referred to a cardiac surgeon for vessel reimplantation.

Anomalous origin of the RCA from the pulmonary artery is a rare congenital coronary artery malformation. There are <100

![Figure 1. Presentation ECG.](image-url)
cases reported in the literature.\textsuperscript{1–4} Age of presentation can be from infancy to late adulthood. Presentation can be with angina, heart failure, myocardial infarction, or sudden death and found at autopsy. A large proportion of cases are asymptomatic. Even in asymptomatic individuals, surgery is often the therapy of choice given the risks of an acute event; in 1 literature review, 15\% of patients with isolated ARCAPA presented with a cardiac arrest or acute myocardial infarction.\textsuperscript{1}

Anomalous origin of the RCA from the pulmonary artery is known to be associated with other congenital heart diseases; there have been case reports of ARCAPA associated with a ventricular septal defect, aortopulmonary window, bicuspid aortic valve, tetralogy of Fallot, coarctation of the aorta, and patent ductus arteriosus.\textsuperscript{1–3} Patients with associated cardiac anomalies are diagnosed early in life compared to patients with isolated ARCAPA.

Diagnosis has most commonly been via angiography and echocardiography or post mortem.\textsuperscript{1–3} Cardiac magnetic resonance has been shown to be a useful noninvasive tool in determining the anatomy of anomalous coronary arteries and their course; anomalous coronary arteries can also be followed up with CMR after surgical reimplantation.\textsuperscript{1} The mechanism for adenosine stress perfusion defects can be explained by the coronary steel phenomenon. This case is the first to report and illustrate how CMR with stress perfusion can provide both structural and functional information to guide the management of ARCAPA.

Figure 2. Exercise ECG with inferolateral ST-segment depression.

Figure 3. Coronary angiography showing retrograde filling of RCA from left system injection and subsequent reflux into main pulmonary artery.

Disclosures

None.

References

Figure 4. Turbo spin echoes T1-weighted images confirming the origin (A) of the RCA from the main pulmonary artery and the proximal (B) and mid (C) course between the aorta (*) and the pulmonary trunk (**).

Figure 5. Adenosine stress perfusion images showed an inducible perfusion defect in the RCA territory (arrow).
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