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Visual Diagnosis Pearl

Anomalous Origin of the Right Coronary Artery with an Inter-Arterial Course

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A 66-year-old female with a history of essential hypertension and hyperlipidemia was referred to cardiology clinic for intermittent episodes of substernal chest pain. Her electrocardiogram (ECG) revealed normal sinus rhythm with poor R-wave progression. Her last episode of chest pain was a week prior, rated 9/10 in severity, and resolved in 15 minutes. Prior cardiac workup, including a stress echocardiography, was unrevealing. Re-evaluation of her symptoms included a coronary computed tomography (CT) angiogram which showed a narrowed, medialized, malignant and high origin of the right coronary artery (RCA) and arising from the left coronary cusp, with an inter-arterial course of approximately 9 mm (Figure 1). The patient underwent unroofing of the right coronary artery with repair of the aortic valve with resuspension of the commissures, a modified-MAZE procedure, and ligation of the left atrial appendage. The procedure was well tolerated, and she has remained free of chest pain for the past four years.

DISCUSSION

The detection of anomalous aortic origin of coronary arteries (AAOCA) has increased with the advent of advanced imaging and has a detection incidence range of <1% to >5% on invasive and non-invasive studies. Noninvasive angiography has played a significant role in the increased diagnoses of this rare condition. ECG-gated cardiac CT angiography is now considered the “gold standard” for evaluation of coronary artery anomalies. The presence of an RCA that originates from the left sinus of Valsalva is a congenital abnormality with a prevalence of 0.2% to 0.5% in published autopsy series.[1,2] Anomalous origin of the RCA with an inter-arterial course (ARCA-IA), in which the artery traverses the space between the ascending aortic artery and pulmonic artery, is associated with an increased risk of sudden cardiac death. One series of 11 patients, aged 53 +/- 18 years, showed that episodic chest pain was the most prevalent symptom (64%) and was typically present for more than 6 months prior to diagnosis.[3] Myocardial ischemia in these patients can be due to either fixed effects, such as ostial or arterial abnormalities, or dynamic effects result-

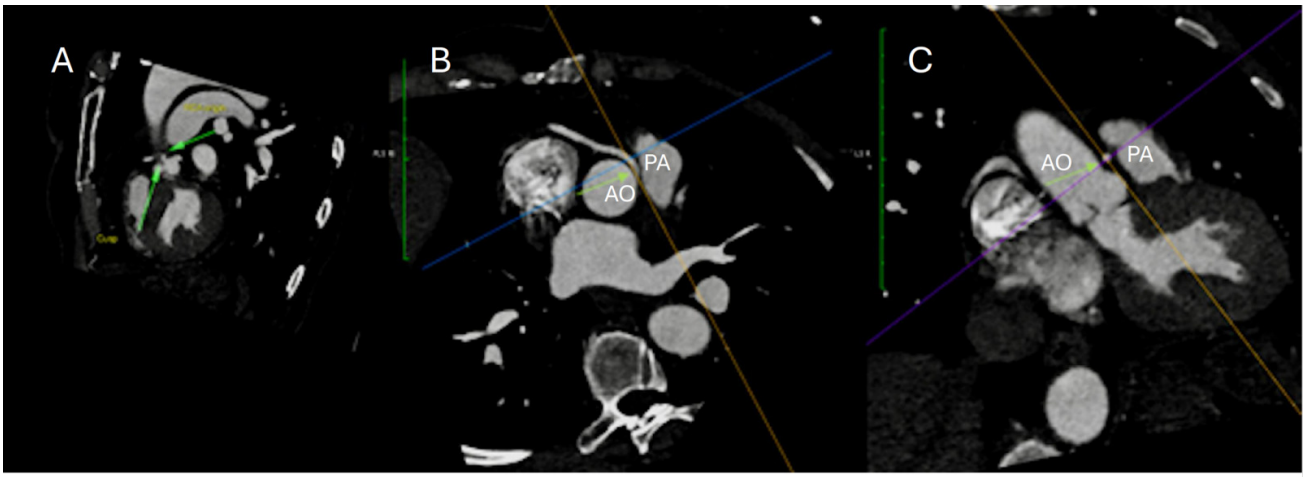

Figure 1 A: Axial contrast-enhanced CT image at the level of the great vessels demonstrating anomalous origin of the right coronary artery from the left coronary sinus (green arrows)
Figure 1 B: Coronal reformatted image showing the inter-arterial course of the anomalous right coronary artery (green arrow) between the ascending aorta (AO) and main pulmonary artery (PA)
Figure 1C: Short axis anomalous right coronary artery (green arrow) highlighting inter-arterial course of the right coronary artery between the aorta (AO) and pulmonary artery (PA)

ing from an RCA path that traverses the aortic wall or myocardium. The previous belief that ischemia results from the compression of the RCA between the pulmonic and aortic arteries has more recently been called into question. Treatment decisions are based on anatomical features such as the length of an intramural segment and slit-like ostia. Surgical unroofing is performed most often and offers durable responses with no related mortality. Ostioplasty, which creates an RCA ostium in the right Sinus of Valsalva (SOV), may be performed alone or in combination with unroofing, and may be necessary when there is an acute take-off angle of the RCA from the aorta. Other surgical options include coronary artery bypass grafting (CABG) with transfer of the RCA ostium. CABG may be indicated in cases of complex anatomy or co-existing clinically significant coronary atherosclerosis. Another technique includes a combination of unroofing, ostiotomy, and reimplantation of the RCA to the right SOV, which is technically complicated.[1,2]

The case of AAOCA described here follows a typical pattern, with involvement of the RCA, which is seen 5-10 times more commonly than a similar anomaly of the left coronary artery. The patient's predominant symptom was intermittent chest pain without dyspnea on exertion, which is seen in only a subset of patients. A period of diagnostic delay followed by surgical management with an unroofing procedure that provides excellent results are also commonly seen.[1]

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