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Case Report

Atypical Presentation of an Anomalous Right Coronary Artery

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A B S T R A C T

Anomalous right coronary artery is a rare congenital disorder, which is usually asymptomatic and diagnosed incidentally. Symptoms usually manifest in the young adulthood and show a wide range of manifestations, including sudden cardiac death. The symptoms are mainly exertional because of the intramural course of anomalous coronary, making it vulnerable to compression during activity. We report a case of a patient with anomalous right coronary artery presenting atypically at an older age with angina occurring even at rest. This case was diagnosed on a coronary angiography and treated successfully with coronary unroofing.

Keywords: Cardiology; Internal medicine; Anomalous coronary arteries

Introduction

Anomalous origin of the right coronary artery arising from the left coronary sinus and taking an interarterial course between the great vessels is a rare diagnosis, with a reported incidence between 0.026% and 0.250%[1]. With most cases being asymptomatic, the anomalous right coronary artery is typically diagnosed incidentally on cardiac imaging[2]. However, cases can also present with sudden cardiac death, when they are found to have anomalous coronaries during autopsy. Most of the symptomatic cases have been reported to occur before the age of 35 with exertional angina, and some even presenting with sudden cardiac death[3]. We report a case with this rare diagnosis that presented atypically with angina, occurring even at rest, and had a late presentation at an older age.

Case Presentation

A 46-year-old female presented with a year-long history of intermittent episodes of retrosternal chest pain, radiating to the jaw, neck and the left arm. These episodes occurred both during the activity and at rest. They were severe enough to disrupt her sleep and worsened progressively over time, leading to multiple office and emergency department visits for the patient. Electrocardiogram showed normal sinus rhythm without any ST segment changes, and troponins were normal. Lexiscan stress test revealed normal myocardial perfusion with no obvious ischemia and no transient ischemic dilatation (TID). Echocardiogram revealed normal ejection fraction of 55-60% without any regional wall motion abnormalities. Left heart catheterization showed normal left main coronary artery, left anterior descending and left circumflex artery, however, the RCA could not be engaged. Eventually, coronary CT angiogram was done which showed an anomalous right coronary artery originating from the left coronary cusp with an intramural course, with reformatted images revealing compression of the proximal RCA between the proximal pulmonary artery and the aorta **(Figures 1 and 2)**. The patient had already failed medical management by the time this diagnosis was made, as she had been tried on different drugs including antianginals, non-steroidal anti-inflammatory drugs across her multiple office visits.

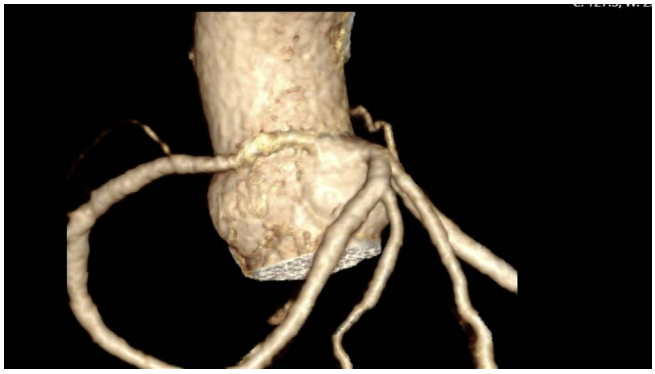

Figure 1: Coronary CT angiogram showing anomalous origin of the right coronary artery.

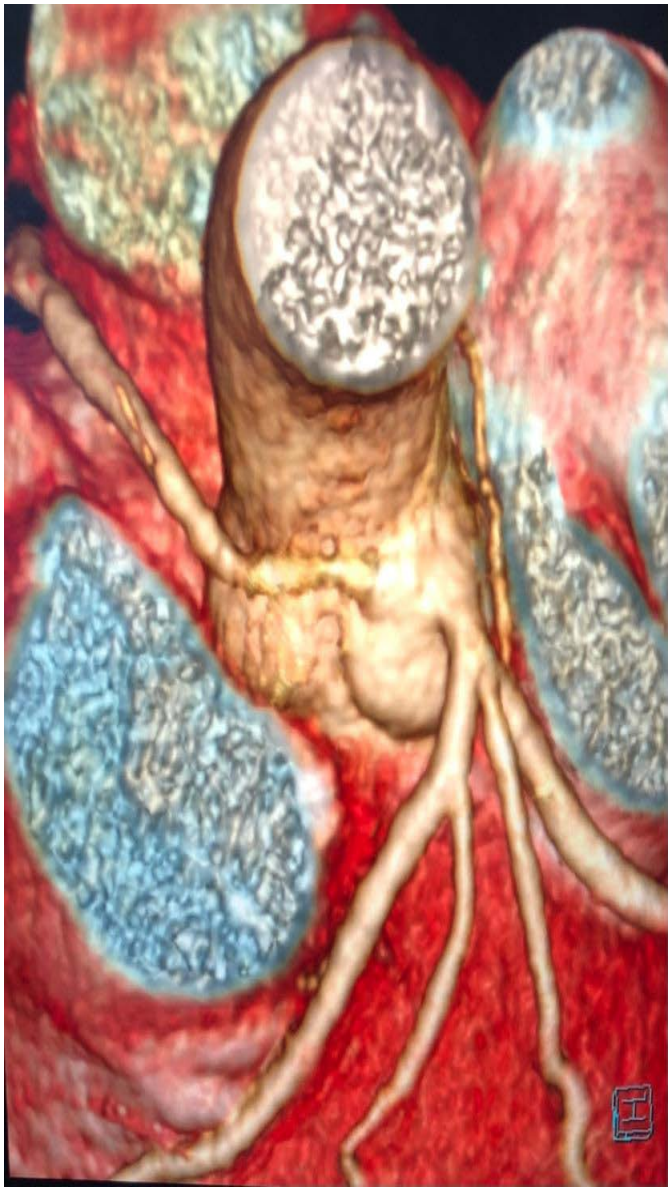

Figure 2: Coronary CT angiogram showing anomalous origin of the right coronary artery (3D image).

Patient was then referred to cardiothoracic surgery. She got coronary unroofing of the abnormal origin of the anomalous origin of the right coronary artery. Her intraoperative course was complicated by severe biventricular dysfunction on the first attempt off cardiopulmonary bypass (CPB) which resolved on the second attempt. Post operative course was unremarkable and she was discharged on post operative day four. At one and three month follow up visits, patient reported marked improvement and resolution of her symptoms.

Discussion

Anomalous origin of a coronary artery (AAOCA) can be defined as the origin of coronary artery occurring at or above the incorrect sinus of Valsalva **(Figure 3)**. These are further classified based on the course they take as inter-arterial, subpulmonic, pre-pulmonic, retroaortic, or retrocardiac. Although both them are very rare in incidence, anomalous inter-arterial RCA has higher reported incidence (six times more) than the anomalous inter-arterial LCA[4]. Inter-arterial coronaries, between the aorta and pulmonary artery, typically take an early intramural course between the intimal and adventitial layers of the aortic root[5]. Our case had anomalous origin of right coronary artery originating from the left coronary cusp, taking an interaterial course with an intramural segment.

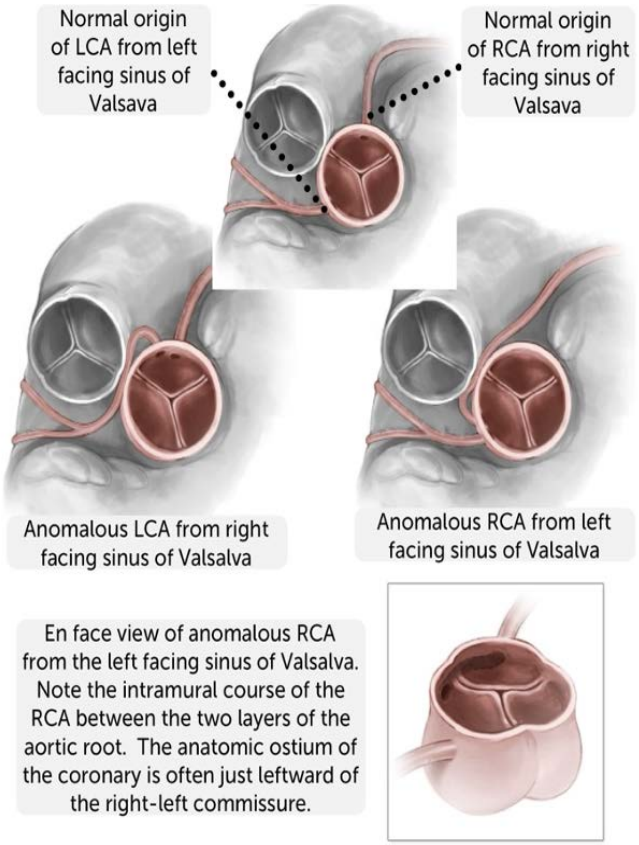

Figure 3: Description of normal and anomalous coronary artery origin.

Most of these cases are asymptomatic and only diagnosed incidentally. When symptomatic, anomalous RCA typically manifests before the age of 35 as ischemic symptoms during exertion[3]. Two mechanisms have been proposed. One, the elongated and narrow anomalous RCA is unable to provide enough perfusion due to the increased myocardial oxygen demand during exercise, creating a demand supply mismatch. Second, aorta dilates during exertion, which causes compression of the anomalous RCA against the pulmonary artery. The sharp angulation and slit like ostium of the anomalous RCA further makes it vulnerable to compression during aortic dilation.

Our case had an atypical presentation as the patient presented late for this congenital disorder with symptoms starting in late fourth decade. Furthermore, patient had angina occurring even at rest, rather than just during exertion. This case points that congenitally anomalous RCA should remain as a differential for the elderly or mid age presentation for angina. Angina at rest may be an indicator of a precarious coronary anatomy in our

case. Given the paucity of evidence and difficult measurement techniques, there are no clear guidelines stratifying the risk for anomalous RCA based upon the anatomy like length of intramural segment, degree of angulation etc. Further studies are needed to determine this risk. Coronary imaging through CT angiogram or MRA is not only the best test to diagnose this condition, but can also help in determining the key anatomical features of the anomalous coronary artery. These imaging features, along with the clinical presentation and shared decision making can play a role in determining the appropriate cases for definitive surgical intervention.

Different surgical techniques have been used for repair of anomalous coronaries, with coronary unroofing being the most commonly used technique[2] **(Figure 4)**. Other surgeries include coronary reimplantation, CABG, patch augmentation or combination of these. Coronary unroofing used in our case, involves opening up the intramural course of the anomalous coronary, to effectively create a new orifice in the correct sinus of Valsalva[6] **(Figure 4)**. in their larges study of 148 patients undergoing coronary unroofing, showed excellent early outcomes with no intraoperative and post operative mortality and 94% late survival rates[7].

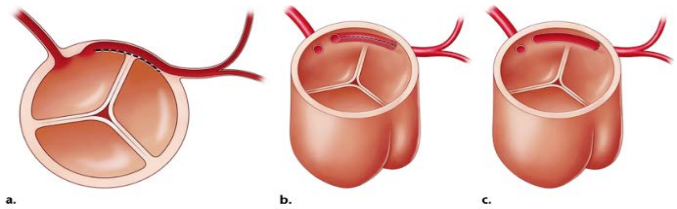

Figure 4: Diagram showing technique of coronary unroofing for an interaterial coronary with an intramural segment. The intramural course is opened up over its entire length (along the dashed line in a & b), thereby creating a wide neo-ostium without angulation or a slit like opening (c).

Conclusion

Anomalous RCA may present at an elderly age, atypically with angina occurring even at rest. Concrete understanding of the wide symptomatology of this rare disorder will help make physicians make early diagnosis and intervention to prevent malignant arrythmias and sudden cardiac death among these patients. Coronary unroofing is the definitive treatment, which has shown excellent intra and post operative outcomes.

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