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Anomalous Right Coronary Artery: A Case Report

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ABSTRACT

Background: Anomalous coronary arteries (ACAs) are rare but potentially life-threatening abnormalities of coronary circulation. Most variations are benign; however, some may lead to myocardial ischemia and/or sudden cardiac arrest.

Case Report: We present the case of a patient with a significant medical history of hypertension, hyperlipidemia, type 2 diabetes, obesity, and gastroesophageal reflux disease who presented to the emergency department with atypical chest pain. She underwent a cardiac catheterization that showed an anomalous right coronary artery originating near the anterior left coronary artery sinus and coursing between the pulmonary artery and aorta. The patient was deemed a poor surgical candidate, was discharged home on medical management with beta blocker therapy, and was instructed to restrict her physical activity.

Conclusion: Treatment of significant anomalies should be guided by the nature of the anomalous vessel. Symptomatic patients with ACAs have 3 treatment options: medical management, coronary angioplasty and stent deployment, or surgical correction. These treatment options remain controversial. Some clinicians advocate revascularization, but the long-term benefits of revascularization therapies have not yet been demonstrated.

INTRODUCTION

Anomalous coronary arteries (ACAs) are rare but potentially life-threatening abnormalities of coronary circulation. Most variations are benign; however, some may lead to myocardial ischemia and/or sudden cardiac arrest.[1] In previous studies, the reported incidence of coronary anomalies discovered in patients undergoing coronary angiography ranged from 0.64%-1.6%.[2] We present the case of a patient with anomalous right coronary artery (RCA) circulation who presented to the hospital with atypical angina, review the literature, and discuss the clinical significance of this anomaly.

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CASE REPORT

A 53-year-old Caucasian female with a significant medical history of hypertension, hyperlipidemia, type 2 diabetes, obesity, and gastroesophageal reflux disease and who had received an orthotopic kidney transplant for thrombotic thrombocytopenic purpura presented to the emergency department with atypical cardiac chest pain. She complained of intermittent chest discomfort that had persisted for 2 months. She described the pain as 5 of 10 in severity, substernal, lasting less than 1 minute, nonradiating, and ache-like, resolving spontaneously but becoming acutely worse overnight with minimal exertion. She had never taken sublingual nitroglycerin to relieve her pain, and her electrocardiogram (EKG) on presentation showed normal sinus rhythm with ST and T wave abnormalities potentially indicating anterior and inferior ischemia seen in III, aVF, and V1-V3 (Figure 1). Her 2D echocardiogram 2 months prior to admission had shown a normal ejection fraction (60%) with normal diastolic function. She was admitted to cardiology for unstable angina and underwent a cardiac catheterization that showed an anomalous RCA originating near the anterior left coronary artery sinus and coursing between the pulmonary artery and aorta (Figures 2 and 3). The patient was deemed a poor candidate for surgery, was discharged home on medical management with beta blocker therapy, and was instructed to restrict her physical activity.

DISCUSSION

Splanchnic mesoderm gives rise to all components of the heart, and the mesoderm differentiates into the cardiogenic area during the third week of embryogenesis.[3] The cardiogenic area then forms a pair of endocardial tubes that fuse to form the primitive heart tube.[3] Anomalies of coronary circu-

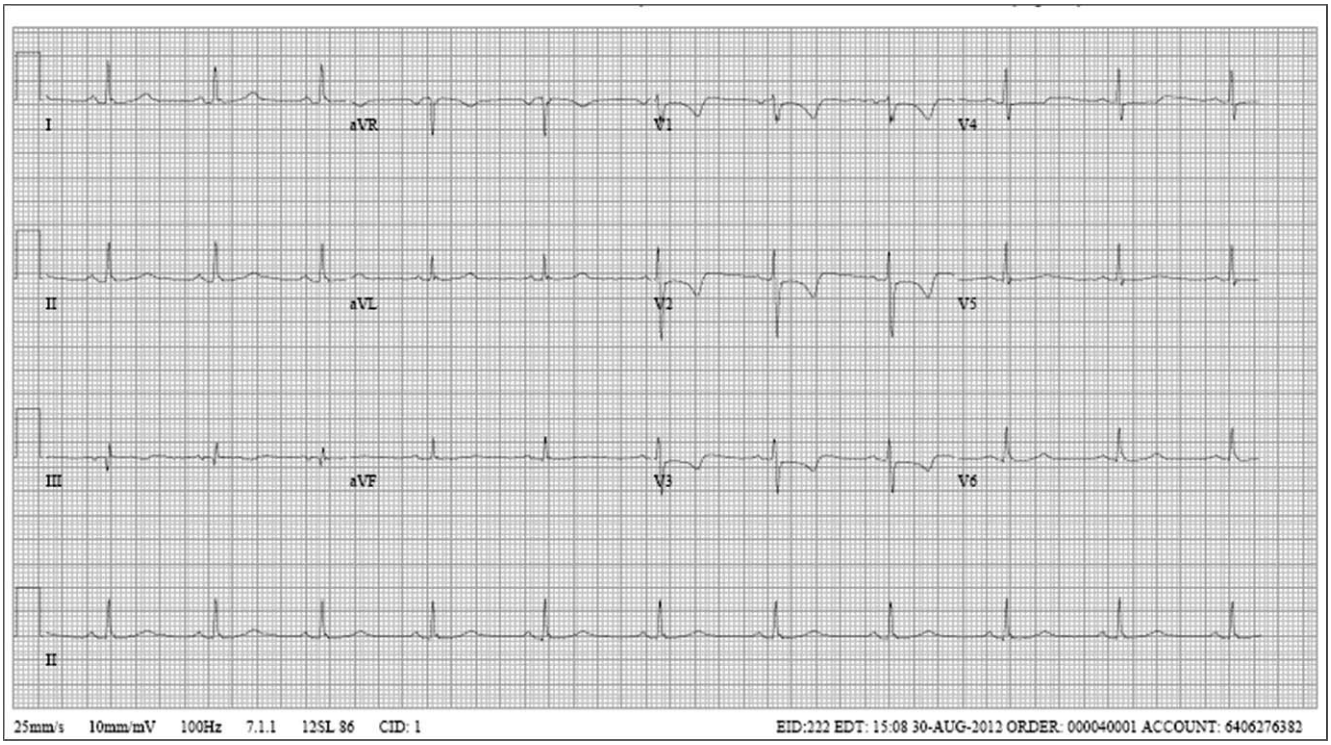

Figure 1. The patient's electrocardiogram upon presentation to the emergency department.

lation result from processes that disrupt the normal differentiation and specialization of the primitive heart tube.[4] Position of the endothelial buds or septation of the truncus arteriosus may give rise to anomalous origins of coronary arteries.[5] Few anomalies present with symptoms or serious clinical sequelae that require surgical correction; most are discovered incidentally during angiography.[2] White and Edwards first described the anomalous origin of the RCA as a rare congenital abnormality in 1948.[6]

Anomalous RCAs that originate from the left coronary sinus occur in 0.05%-0.1% of the general population.[7] Although ACAs occur with low frequency, a high risk of sudden cardiac death because of myocardial ischemia and resultant arrhythmia are associated with them, even in the absence of atherosclerosis.[8] Although compression of the RCA by the great vessels is suggested as the main explanation of symptoms, the pathophysiological basis for this aforementioned explanation remains unclear.[8] Others suggest that the angle at which the left coronary sinus and anomalous RCA meet creates a slit-like orifice that collapses with exertion or exercise.[8] Noninvasive techniques such as magnetic resonance imaging (MRI) and multislice computed tomography (CT) can clearly delineate the anatomy and have replaced angiography as definitive diagnostic tools.[9,10] Multislice CT has been recommended because it offers excellent spatial resolution and identifies most coronary anomalies, but it uses ionizing radiation and potentially nephrotoxic agents.[11] The use of cardiac MRI for studying congenital anomalies has generated great interest among cardiologists; however, current studies are insufficient to recommend MRI as the imaging method of choice for ACAs.

Treatment of significant anomalies should be guided by the nature of the anomalous vessel. Treatment options remains controversial, with some clinicians advocating revascularization. Symptomatic patients with ACAs have 3 treatment options: medical management, coronary angioplasty and stent deployment, or surgical correction. Stenting generally is not recommended. Several surgical options are available, including directly reimplanting the anomalous artery, surgically unroofing the intramural coronary segment from the ostium to the exit point at the aortic wall, or creating a new ostium at the end of the anomalous artery's segment, a procedure known as osteoplasty.[12,13] Revascularization using direct reimplantation of the anomalous RCA into the right coronary sinus is the preferred method of surgical treatment for this abnormality per the surgical literature.[14] Current literature does not demonstrate any long-term benefits of revascularization.

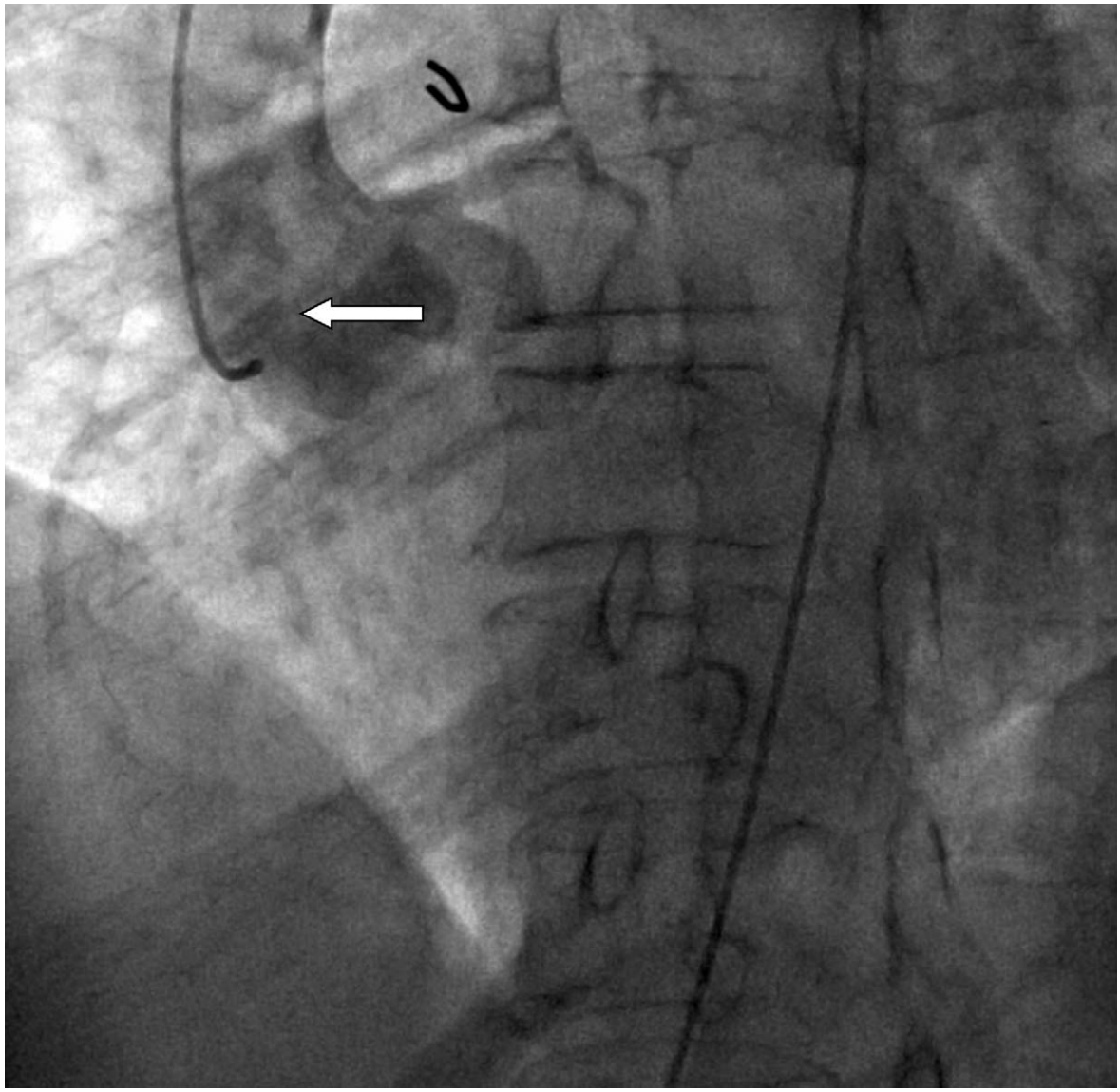

Figure 2. Nonselective cardiac catheterization of anomalous right coronary artery with origin near the left main in the anterior left coronary artery sinus.

The Japanese approach to this condition is far more conservative, as demonstrated by a study of 56 patients who had anomalous arteries and were treated medically with beta blockers. Side effects of the conservative approach included hypotension and arrhythmias on exertion (9%).[15] No deaths had occurred at a 5-year analysis of the study.[15] The Congenital Heart Surgeons' Society established a North American registry in 2009 to study large cross-institutional cohorts of patients with anomalous arteries, hoping to generate evidence-based guidelines for patient management. This research is still ongoing.[16]

During her hospitalization, our patient had another EKG. It was unchanged from her prior EKG, and echocardiogram was normal with a preserved ejection fraction of 65%. Other noninvasive testing was not pursued by the primary team. Medical management was effective in resolving her chest discomfort. Her EKG remained unchanged at subsequent outpatient follow-up. She remains free of chest pain with her medical management.

CONCLUSION

This case illustrates an example of an anomalous RCA originating from the left coronary sinus in a middle-aged patient presenting with chest pain. The preferred treatment for these patients is conservative medical therapy.

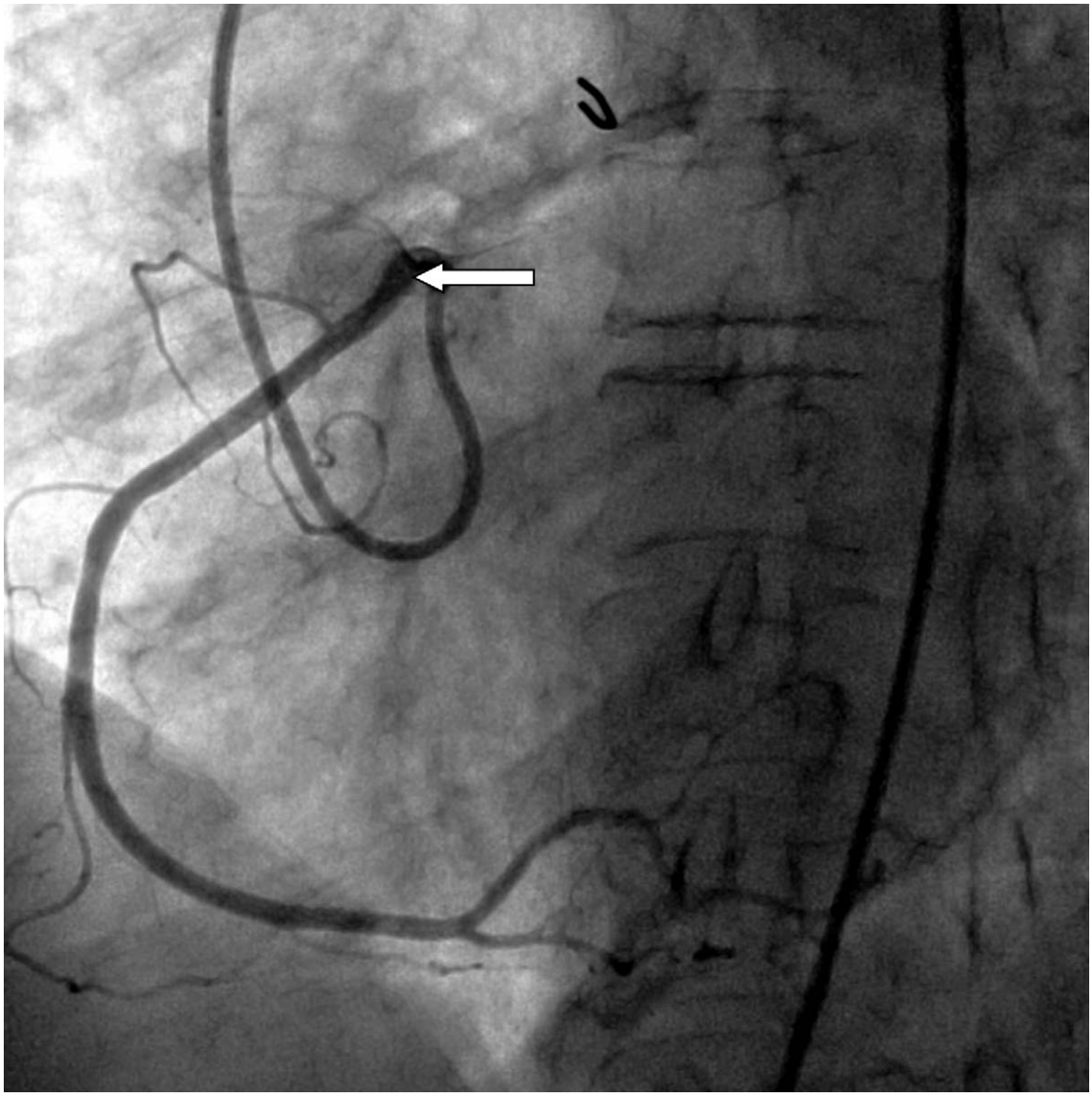

Figure 3. Selective engagement of anomalous right coronary artery.

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