Angina and sudden cardiac death caused by anomalous right coronary artery origin from above the left coronary cusp

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Anomalous right coronary artery (RCA) origin, an extremely rare but well described condition and cause of myocardial ischemia, arrhythmia, and syncope, has recently become recognized as a cause of sudden cardiac death in the young.¹ In contrast to the previously reported cases of RCA anomalies, we report this unique case of angina and sudden cardiac death caused by anomalous RCA origin from above the left coronary cusp. It is unique in that it occurred in a female, had a late onset of presentation, and was associated with no other cardiac abnormalities. The patient underwent successful revascularization.

Case

The patient was a 67-year-old female who presented with a three-month history of angina pectoris and one episode of sudden cardiac death while shopping at a nearby supermarket that was successfully resuscitated. She had no other risk factors for coronary artery disease. Acute myocardial infarction was ruled out. Physical examination was unremarkable with normal blood pressure and heart rate. EKG, CXR, and echocardiogram as well as CBC, renal, lipid and hepatic profile all were normal. A stress MIBI SPECT scan was positive for large inferior reversible defect (Figure 1). Cardiac catheterization and coronary angiogram revealed an anomalous RCA origin from above the left coronary cusp (Figure 2), with its proximal segment between the aorta and pulmonary trunk and a normal course thereafter. This artery was dominant and free of any significant lesions. The left main coronary artery was short, wide, and free of any significant lesions, and in addition to the left circumflex

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Figure 1. Thallium SPECT scan long axis view showing stress-induced (upper panel) large inferior and infero-apical defect with reversibility during rest (lower panel).

(LCX) and left anterior descending (LAD) arteries, it gave off another vessel, possibly a conus branch, which ended abruptly. The LAD and LCX were free of any significant lesions. No other abnormalities or congenital lesions were detected. The left ventricle was normal in size and contractility with a calculated ejection fraction of 70%. The cardiac output, right and left sided heart pressures were normal. No shunt or oxygen saturation step up was present. A decision was made to relieve her symptoms with surgical correction (options were discussed either to anastomose a bypass graft to proximal-mid RCA or reimplant the RCA into the right sinus of Valsalva). She underwent successful single vessel coronary artery bypass surgery with right internal mammary artery (RIMA) to mid RCA. The patient remained asymptomatic after her surgery. A follow-up Stress MIBI SPECT scan (Figure 3) was normal and currently, she is leading a normal life.

Discussion

Anomalous coronary arteries have been the subject of more than 500 articles and studies over the past 35 years.² Anomalous origin of the coronary arteries, recently recognized as a potentially serious form of congenital heart disease,³ is extremely uncommon. The prevalence of this anomaly was recently estimated to range from 0.3% to 2.85%⁴⁻⁵ among different age groups with a male to female ratio of 4:1. Numerous types of anomalies have been described, with the most frequent anomaly found to be of the origin of the LCX (50%), followed by an abnormal origin of the RCA (31%), and left coronary tree (19%). RCA originating from the left sinus of Valsalva was recognized in 0.02% of coronary angiograms performed by Donaldson and Rafael, and 0.17% by Chaitman et al.⁶ The relationship of the anomalous coronary artery to the great vessels was the following: a retroaortic course (69%), interarterial (12%), to the anterior free wall (12%) and septal in (6%). The anomalous origin of RCA from the left sinus of Valsalva was initially thought to be a benign condition and has only recently become recognized as a cause of angina, myocardial infarction, arrhythmias and sudden cardiac death. The mechanism of cardiac death may be decreased myocardial perfusion by the anomalous vessel due to a combination of tangential takeoff of the vessel with a resultant slit-like ostium, along with initial investment in the aortic adventitia with diastolic entrapment and compression as it courses between the aorta and pulmonary trunk. Both factors are both thought to be

Figure 2. Left anterior oblique view using left Judkins catheter showing anomalous RCA from left coronary cusp and coursing between the pulmonary artery anterior and aorta posterior.

Figure 3. Post-operative thallium SPECT scan long axis view showing normal myocardial perfusion during both stress (upper panel) and rest (lower panel).

Figure 4. Right coronary artery originating from above the left coronary cusp. The proximal portion curves between the aorta (located posteriorly) and the pulmonary trunk (located anteriorly).

at play (Figure 4). Among the previously published cases of anomalous RCA arising from above the left coronary sinus, there were other associated congenital, structural, coronary or valvular abnormalities, manifested at an early age and more commonly in males. We present a case of angina-sudden cardiac death caused by anomalous RCA as a unique case of a late onset of presentation, in a female with normal aortic valve cusps. The diagnosis can be suspected by transesophageal echocardiogram,⁷ magnetic resonance angiography (MRA)⁸ and definitive diagnosis established by coronary angiography. The preferred treatment is surgical correction by either anastomosing a bypass graft to RCA, or re-implanting the RCA to the aorta.⁹

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