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Letter to the Editor · **Open Access** ·

A rare case of single right coronary artery arising from the right sinus of Valsalva with severe three-vessel disease

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A 63-year-old man was admitted to our observation unit with atypical angina found to be non-adherent with his anti-hypertensive medication regimen for one month. His medical history was notable for hypertension for ten years and current tobacco smoking. He was hypertensive with otherwise, stable vital signs. Cardiac auscultation revealed normal S1 and S2 without a murmur. The ECG revealed nonspecific ST changes and T wave inversion in lateral leads (V5–6, I and aVL). The cardiac enzymes [troponin T, creatine kinase (CK), and creatine kinase myocardial B fraction (CK-MB)] were within the normal limit. His blood pressure was initially 163/105 mmHg which later improved to 100–120/70–85 mmHg after starting on low dose carvedilol. Echocardiogram revealed normal left ventricular size, wall thickness, and function, aortic sclerosis, mild tricuspid regurgitation, and impaired relaxation. He underwent sestamibi lexiscan myocardial perfusion imaging study which revealed mild to moderate inducible myocardial ischemia in the inferolateral and lateral walls with no scar and hypokinesis of those regions.

Consequently, cardiac catheterization was performed revealing all three major coronary vessels arising from the right coronary cusp (Figure 1A&B). An AL catheter was used to engage the right coronary artery (RCA). A severe left anterior descending coronary artery (LAD) artery lesion and a severe ostial lesion in the first diagonal branch were noted. The left circumflex coronary artery (LCx) artery had three sequential, severe, mid to distal lesions. The RCA was completely occluded proximally. Cardiac tomography angiogram was done to further evaluate the specific course of the vessels and revealed single coronary artery (SCA) originating from the right coronary cusp (Figure 2). This coronary artery was a short, large caliber trunk that trifurcated early into the RCA, LAD, and the LCx arteries. The left anterior descending artery had a subpulmonic course with mid LAD segmental myocardial bridging and a lesion in the mid LAD. The LCx had a retroaortic course. Dense calcification of the mid coronary precluded accurate estimation of the disease severity. The RCA was dominant and there was proximal total occlusion visualized. The patient was managed with elective three vessel coronary bypass surgery. This included a left internal mammary artery to the distal LAD, saphenous vein graft to the first obtuse marginal artery, and a saphenous vein graft to the distal RCA. The patient had no complications during his hospital course and was discharged from the hospital in stable condition.

This case demonstrates a rare congenital coronary artery anomaly found incidentally with a presentation of atypical angina and three-vessel coronary artery disease (CAD). Our patient was found to have a single coronary artery (SCA) arising from the right cusp of Valsalva and then branching into three separate ostia representing RCA, LAD and LCx. The incidence of this variation is between 0.011% and 0.066%.[1–7]

Coronary anomalies are congenital malformations of coronary vessel anatomy, usually affecting the origin and course of the vessel. An anomaly is defined as an abnormal variant that occurs in less than 1% of the general population. About 20% of all coronary artery anomalies are life threatening.[8] These anomalies typically stay undiagnosed until postmortem review, usually asymptomatic until sudden cardiac death. Less than 30% of adult patients present with

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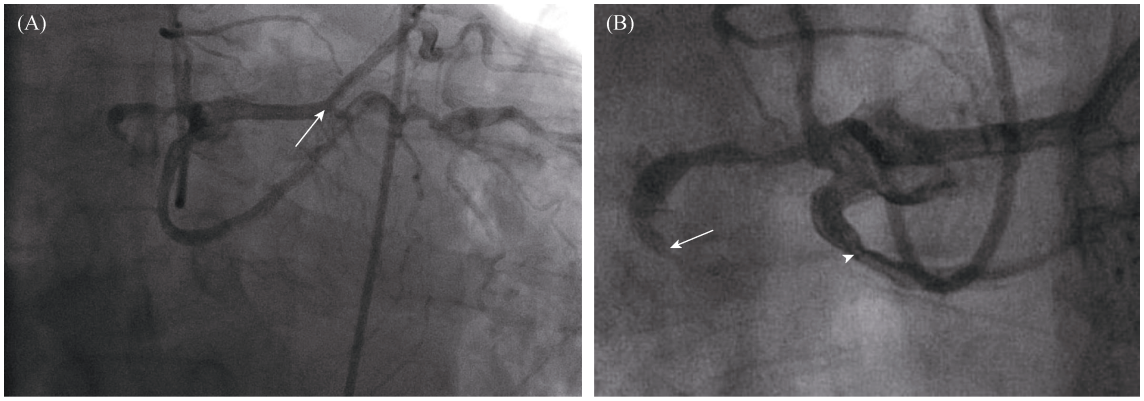

Figure 1. Cardiac catheterization images. (A): All three coronaries arising from the right coronary cusp. There is LAD artery lesion and a severe ostial lesion in the first diagonal branch (arrow); (B): the LCx artery had three sequential, severe, mid to distal lesions (starting at the arrow head), and RCA was completely occluded proximally (arrow). LAD: left anterior descending coronary artery; LCx: left circumflex coronary artery; RCA: right coronary artery.

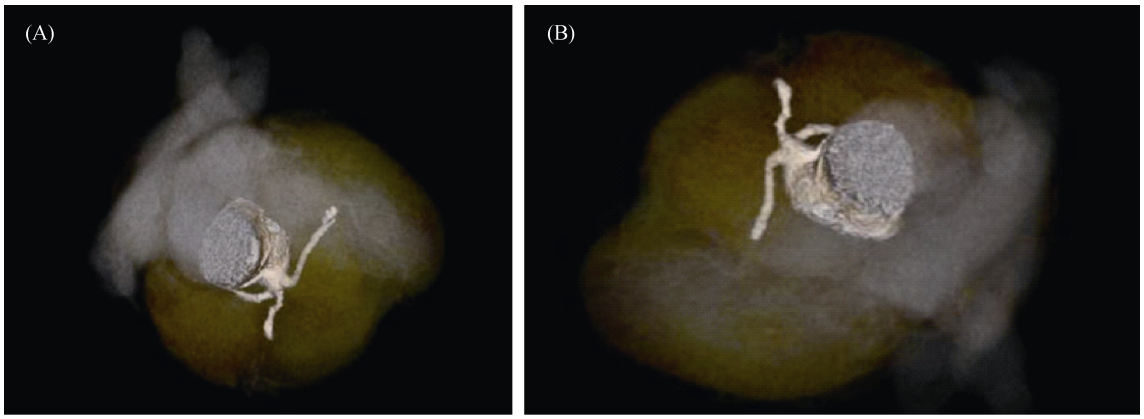

Figure 2. Cardiac tomography imaging results. (A): Cardiac tomography imaging showing SCA originating from the right coronary cusp; an artery that has short, large caliber trunk that trifurcates early into the RCA, LAD, and the LCx arteries; (B): three dimentional reconstruction of cardiac tomography showing the LAD has a subpulmonic course. The LCx had a retroaortic course. Dense calcification of the mid coronary precluded accurate estimation of the disease severity. The RCA is dominant with proximal total occlusion. LAD: left anterior descending coronary artery; LCx: left circumflex coronary artery; RCA: right coronary artery; SCA: single coronary artery.

symptoms of palpitations, exertional dyspnea, angina, syncope or fatigue. This is principally due to increased angulation at the origin of the coronary artery of the LAD or its branches as it courses between the aorta and pulmonary trunk causing external compression, occurring mainly during exercise.[9] This is termed a malignant course.

It has been hypothesized that coronary artery anomalies can predispose to CAD due to the increase in sheering force at the ostium and proximal aspects of the vessel secondary to the angulation and the retro-aortic coarse.[10] Ischemia may or may not occur with coronary anomalies. The majority of coronary anomalies are not ischemic and have no clinical significance. When ischemia is found, it is likely either episodic ischemia or fixed ischemia. Coronary anomalies that cause episodic ischemia (secondary ischemia) usually present with intermittent, exertional, clinical symptoms and are typically not diagnosed until adulthood. Episodic ischemia occurs commonly in anomalous origin of a coronary artery from the opposite sinus; coronary artery fistulas; or myocardial bridging. Fixed ischemia (primary ischemia) usually presents with resting ischemic symptoms and may result in acute myocardial infarction, heart failure, ventricular arrhythmias, or sudden cardiac death. Fixed ischemia is caused by an anomalous left coronary artery from the pulmonary artery; coronary ostial atresia; or severe coronary ostial stenosis.[11,12]

Current literature on SCA is limited and the evaluation of CAD in this population is even more scarce. SCA has been reported in patients with obstructive CAD, non-obstructive CAD, and those without CAD.[7] One case series, retrospec-

tively evaluated 15 patients with SCA from 2010 to 2013. This study evaluated the current diagnostic and therapeutic management and clinical implications of CAD in patients with congenital SCA and found that SCA may be associated with transient transmural myocardial ischemia, non-sustained ventricular tachycardia, and aborted sudden cardiac death.[6] There have only been two cases of three vessel obstructive CAD in a SCA. In both cases the patients had an RCA branching from the LAD.[13] This is the first case to evaluate an RCA which branches into the LAD, LCx, and RCA with three vessel disease.

Although our patient's anomaly had a benign course the presence of CAD in the SCA supplying the entire myocardium precipitated angina symptoms. There is a predilection for the development of atherosclerosis in the posteriorly coursing anomalous vessel. In a review of 83 patients with coronary anomalies performed by Wilkins, *et al*.,[14] two patients with SCA arising from the right coronary sinus had significant atherosclerosis of their posteriorly coursing LCx arteries. This suggests that a posteriorly directed anomalous coronary artery arising from the right sinus of Valsalva is subject to unusual stress, predisposing it to development of atherosclerosis. More specifically, a greater degree of atherosclerosis of the proximal segment of the anomalous LCx with a retroaortic course has been documented, thus warranting further investigation with left heart catheterization and cardiac CT.[14] Identification of anomalous coronary arteries is frequently difficult with conventional coronary angiography because of the absence of three dimensional imaging but is crucial in initial identification and evaluation of anomalous coronaries. Multidetector CT (MDCT) is thus preferred because it can detect the course of the coronary arteries in relation to the great vessels and should be used for accurate delineation of the anomalous course of the coronaries and extent of CAD prior to surgical intervention.[15,16]

Serious tachyarrhythmia and myocardial ischemia are the indications for surgical intervention especially in anomalies that involve a large territory. These include coronary stenting, coronary artery bypass grafting or marsupialization of the coronary artery to prevent compression as seen in a malignant course,[9] or direct re-implantation of the ectopic artery at the aortic root.[4]

Coronary anomalies are rare, most often benign, and usually found incidentally. The clinical significance of coronary anomalies increases in patients with SCA. In patients with cardiac symptoms found to have a benign coursing SCA and CAD, consideration should be made to evaluate whether symptoms are due to SCA or obstructive CAD via stress testing, coronary angiogram, and MDCT. There are no specific guidelines for management and no randomized studies comparing the prognosis, perioperative mortality of patients with three vessel disease with normal coronary anatomy and patients with three vessel disease with SCA's of different Lipton Classes. The current consensus is that if there are significant three vessels CAD is found, the management is similar to patients with normal coronary anatomy. This article should prompt further investigation into the clinical significance of single coronary artery and three vessel diseases. A multicenter/international registry of patients with coronary artery anomalies and CAD may be helpful.

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The authors of this article have no conflicts of interest to declare

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