Anomalous Origin of All Coronary Arteries from Right Sinus of Valsalva

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Anomalous coronary arteries are rare and often incidental findings. Most variants are benign. We present the case of a 75-year-old man with exertional dyspnea in whom the left anterior descending coronary artery arose from the right sinus of Valsalva, and the left circumflex coronary artery originated from the distal right coronary artery and supplied the obtuse marginal branch. No arteries originated from the left sinus of Valsalva. The patient was prescribed optimal medical therapy for atherosclerotic stenosis in his ramus intermedius. His symptoms were stable 3 years later. ***(Tex Heart Inst J 2020;47(2):170-2)***

Anomalous coronary arteries are rather rare but are well described in the medical literature. Variants are usually benign; however, some are associated with sudden cardiac death. Recognizing such anomalies is crucial in guiding management decisions. We report the case of a patient whose unusual coronary artery configuration has not been described.

Case Report

In January 2017, a 75-year-old man with a medical history including atrial fibrillation, hypertension, and previous tobacco use presented with progressive exertional dyspnea. Single-photon emission computed tomographic myocardial perfusion images during exercise showed a basal anterolateral perfusion defect and a normal left ventricular ejection fraction.

A coronary angiogram showed an ectopic vessel from the right sinus of Valsalva, giving rise to the left anterior descending coronary artery (LAD) and its branches, as well as the ramus intermedius artery (Fig. 1A). No artery originated from the left sinus of Valsalva. The distal posterior atrioventricular segment of the right coronary artery (RCA) gave rise to an ectopic vessel from which the obtuse marginal artery (OM1) and its branches originated (Fig. 1B). A stenotic lesion from acquired atherosclerotic disease in the ramus intermedius was determined to be the cause of the patient's ischemia.

A coronary computed tomographic angiogram (CTA) showed that the LAD shared an ostium with the RCA (Fig. 2), coursed anterior to the right ventricular outflow

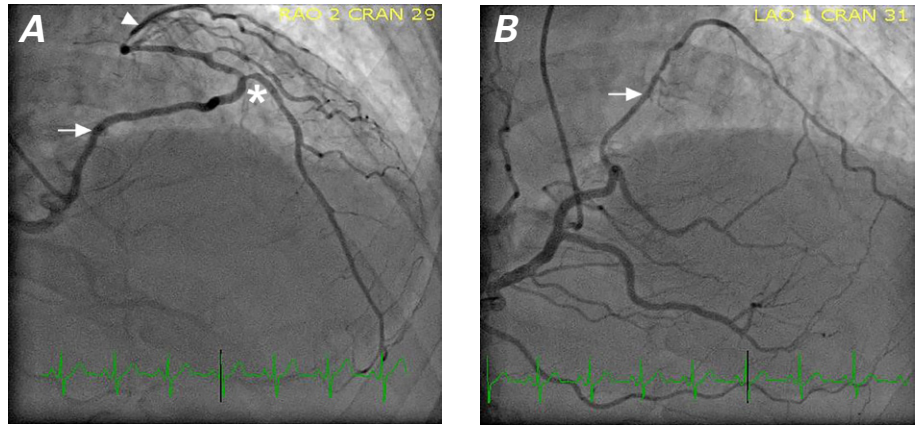

Fig. 1 *Coronary angiograms show* ***A****) an anomalous vessel originating from the right sinus of Valsalva (arrow), supplying the left anterior descending coronary artery (asterisk) and the ramus intermedius (arrowhead); and* ***B****) a vessel originating from the distal right coronary artery (arrow).*

Key words: *Abnormalities, multiple; coronary vessel anomalies/diagnosis/diagnostic imaging/physiopathology/therapy; myocardial ischemia/etiology; sinus of Valsalva/abnormalities; treatment outcome*

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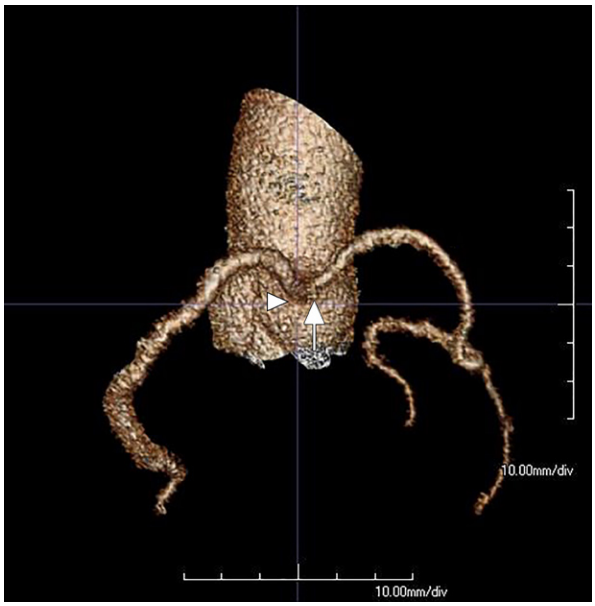

Fig. 2 *Computed tomographic angiogram (3-dimensional reconstruction) shows the anomalous origin of the left anterior descending (arrow) and the right (arrowhead) coronary arteries.*

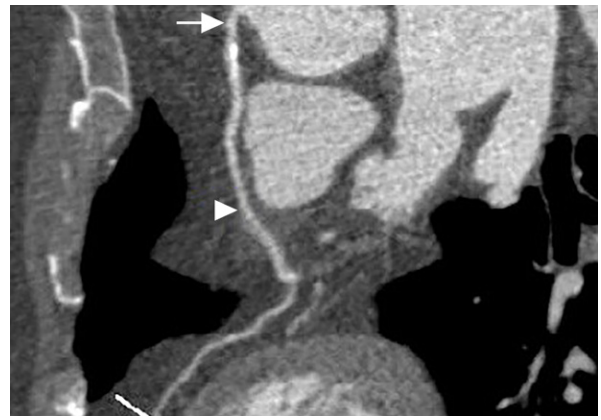

Fig. 3 *Coronary computed tomographic angiogram at the level of the aortic root shows the origin of the ectopic left anterior descending coronary artery (arrow) beginning at the right sinus of Valsalva, and coursing anterior to the right ventricular outflow tract and main pulmonary artery trunk (arrowhead).*

tract and main pulmonary artery trunk, and supplied the ramus intermedius (Fig. 3). The second ectopic vessel (from the RCA to the OM1), although distinct on coronary angiograms, was too small to identify on 3-dimensional reconstructed CTA images.

Because of the patient's complex coronary anatomy, we avoided intervention and prescribed guideline-directed medical therapy for stable atherosclerotic coronary artery disease (CAD), with outpatient monitoring. He was doing well 3 years later.

Discussion

The estimated incidence of anomalous origin of the coronary arteries is 1% to 2% in the general population.[1] Many or most anomalies are benign; however, some, such as anomalous origin of the left coronary artery from the pulmonary artery, have been associated with poor prognosis starting in childhood.[2,3] Anomalous origin of a coronary artery from the opposite sinus (ACAOS), single coronary artery, and large coronary fistulas can cause angina, exertional syncope, dyspnea, palpitations, and ventricular arrhythmias.[2,4] Of note, sudden cardiac death is frequently the initial manifestation of anomalous left coronary artery arising from the opposite sinus (L-ACAOS).[5]

A higher prevalence of anomalous coronary arteries is found in individuals with Turner syndrome—most often absent left main coronary artery. In the general population, the prevalence of this anomaly is less than 1%.[6,7] Anomalous origin of the LCx from the right sinus or RCA is typically a benign variant.[8]

Several mechanisms may underlie the increased risk of sudden death in patients with ACAOS: an intramural course, a slit-like orifice of the anomalous artery, compression between the pulmonary artery and aorta, or repeated trauma to the endothelium that causes ischemia during exercise and increases susceptibility to arrhythmias.[9] Coronary anomalies may also accelerate atherosclerosis in older patients by causing turbulent blood flow consequent to an acute takeoff angle of the vessel from the aorta.[10] Diagnosis typically occurs when patients undergo coronary angiography because of atypical chest pain or when coincidental CAD is diagnosed.[4] Coronary CTA is the gold standard for diagnosis.

Recognizing coronary anomalies is crucial, especially in individuals who have obstructive CAD and those who need valve repair.[11] Delineating coronary anatomy is also essential in managing acute coronary syndrome because of technical challenges in delivering stents.[12]

All branches of our patient's coronary arteries originated from the right sinus of Valsalva. The first ectopic vessel gave rise to the LAD and ramus intermedius; and the second, originating from the distal RCA, gave rise to the OM branch. To our knowledge, this configuration has not been reported before.

Because of technical challenges, we decided against performing percutaneous coronary intervention. The patient's symptoms resolved with guideline-directed medical therapy. If they worsen or if acute coronary syndrome occurs, we will consider coronary artery bypass grafting. Similar coronary anomalies have been treated successfully with use of medical therapy, coronary angioplasty, or surgery, depending on the location of the lesion.[4]

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