

A Rare Case of a Coronary Artery Anomaly Detected on Multidetector Computed Tomography

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Abstract

An anomalous aortic origin of the coronary artery arising from the opposite sinus with an interarterial course is a rare condition that is associated with a high risk of sudden cardiac death during or after strenuous exertion. We herein report the case of a 47-year-old woman presenting with chest pain, syncope and palpitations who presented with a rare coronary artery anomaly on multidetector computed tomography coronary artery (MDCT-CA) with prospective electrocardiogram (ECG) gating.

Key words: coronary anomalies, coronary computed tomography, sudden death

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Introduction

According to the literature (1-3), coronary anomalies are found in approximately 1% of patients among the general population undergoing coronary angiography and approximately 0.3% of autopsy cases. Some coronary anomalies cause cardiovascular morbidity and mortality. These include coronary arteries originating from the contralateral coronary sinus, single coronary sinus or the pulmonary artery. Van Camp and coworkers (4) reported that coronary anomalies cause 11.9% of deaths in US high school and college athletes. According to the Sudden Death Committee of the American Heart Association (5), coronary anomalies cause 19% of deaths in athletes. Moreover, Burke and colleagues (6) reported that among 14- to 40-year-old individuals, coronary anomalies are involved in 12% of sports-related sudden cardiac deaths versus 1.2% of non-sports-related deaths. Coronary artery anomalies may present with symptoms such as angina, shortness of breath and syncope. More severe presentations include the clinical syndromes of either myocardial infarction, congestive heart failure or sudden cardiac death. Overall, symptoms are present in 1/3 of patients. The frequency of symptoms is somewhat related to

the extent of the myocardium at risk (7). Anomalous coronary arteries are believed to cause a reduced regional myocardial blood flow, with a mechanism depending on the particular anomaly.

Although some anomalies may manifest only under exceptional conditions, such as extreme exertion, no means of testing this hypothesis are available and standard clinical submaximal stress-test protocols are frustratingly inadequate for identifying the presence and prognosis of most anomalies (5, 8, 9). Indeed, long-term Holter monitoring (for arrhythmias and ST-segment changes) maybe more informative (5, 10).

Multislice computed tomography (11) has also been recommended; it offers excellent spatial resolution and identifies most anomalies of the coronary course. However, it uses ionizing radiation and potentially nephrotoxic or allergenic contrast agents.

Case Report

A 47-year-old Caucasian woman with a history of diabetes mellitus type 2, hypercholesterolemia, hypertriglyceridemia and active smoking was referred to our hospital for chest pain, syncope and palpitations. On admission to the

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Figure 1. Exercise electrocardiography test. **A:** ECG obtained before exercise (control), **B:** severe ST-segment depression developing during exercise, **C:** ECG obtained during recovery.

emergency department, her chest pain was relieved.

The findings of a physical examination, electrocardiogram (ECG) and echocardiogram were normal.

A treadmill exercise test was interrupted due to chest pain, and an electrocardiogram showed exercise-induced ST-segment depressions of up to 3 mm (Fig. 1). Subsequently, low pressure occurred during recovery.

After the stress test, the patient underwent coronary angiography, which revealed the absence of stenosis or thrombi, although it aroused suspicion of a congenital coronary origin anomaly (Fig. 2A).

Multidetector computed tomography coronary artery (MDCT-CA) with prospective ECG gating (64×2, Definition AS, Siemens Healthcare, Forchheim, Germany) was performed. The presence of obstructive coronary stenosis was excluded (Fig. 2B-D). Volume-rendering reconstruction showed the right coronary artery (RCA), left anterior descending artery (LAD) and retroaortic coronary artery (RAA) arising separately from the right Valsalva sinus (Fig. 2E, F). Multiplanar reconstruction revealed that the LAD had an acute take-off angle with a proximal epicardial segment and an intramyocardial course through the interventricular septum beneath the right ventricular infundibulum,

which then coursed in the distal anterior interventricular sulcus (Fig. 2G).

The retroaortic coronary artery arising with an acute take-off angle gave off two branches. One branch coursed in the proximal anterior interventricular sulcus, while the other coursed-in the atrioventricular sulcus as a normal circumflex coronary artery (Fig. 2H).

Discussion

The condition of an anomalous origin of the left main coronary artery in the right aortic sinus is further classified based on the course of the anomalous artery: between the aorta and the pulmonary trunk; anterior to the pulmonary trunk; posterior to the aorta; and posterior to the right ventricular outflow tract within the interventricular septum (12-15).

In patients with an origin of the left coronary artery in the contralateral coronary sinus, all instances of sudden death involve the presence of either an acute take-off angle, an intramural coronary segment or coursing of the anomalous coronary artery between the pulmonary trunk and the aorta (16).

An acute take-off angle may cause the anomalous artery to collapse. Compression of the coronary artery occurs at the onset of diastole with aortic root distension and may explain the increased frequency of sudden death observed when the left coronary artery courses between the aorta and the pulmonary trunk. It has been postulated that exercise-induced expansion of the pulmonary artery and aortic root may further compress the coronary artery lumen, leading to exercise-related death with a high probability.

In clinical practice, due to cardiovascular comorbidities, it is difficult to establish a causal relationship between symptoms and the type of anomalous origin.

An anomalous main left coronary artery arising from the right Valsalva sinus with a septal course is traditionally considered to be benign (17, 18), while that with an interarterial course is considered to be malignant (19). However, there have been several reports in which a septal course, diagnosed on conventional coronary angiography (CCA), with symptoms (20) and a fatal outcome (21), was subsequently characterized as a mixed course based on the use of MDCT-CA, suggesting that the traditional dichotomous classification of either interarterial or septal encompasses a heterogeneous group of patients with different prognoses, thereby introducing the concept of a continuum (22).

In our case report, the evidence of a positive treadmill exercise test and cardiac-gated MDCT images allowed for a demonstration of the course of the coronary arteries, suggesting a mixed course. The mixed variant can lead to sudden death; therefore, it is important to characterize the patient's clinical profile and identify clinical markers that would enable the detection of this particular anomaly prior to a fatal event.

Several pathophysiologic mechanisms, in addition to the

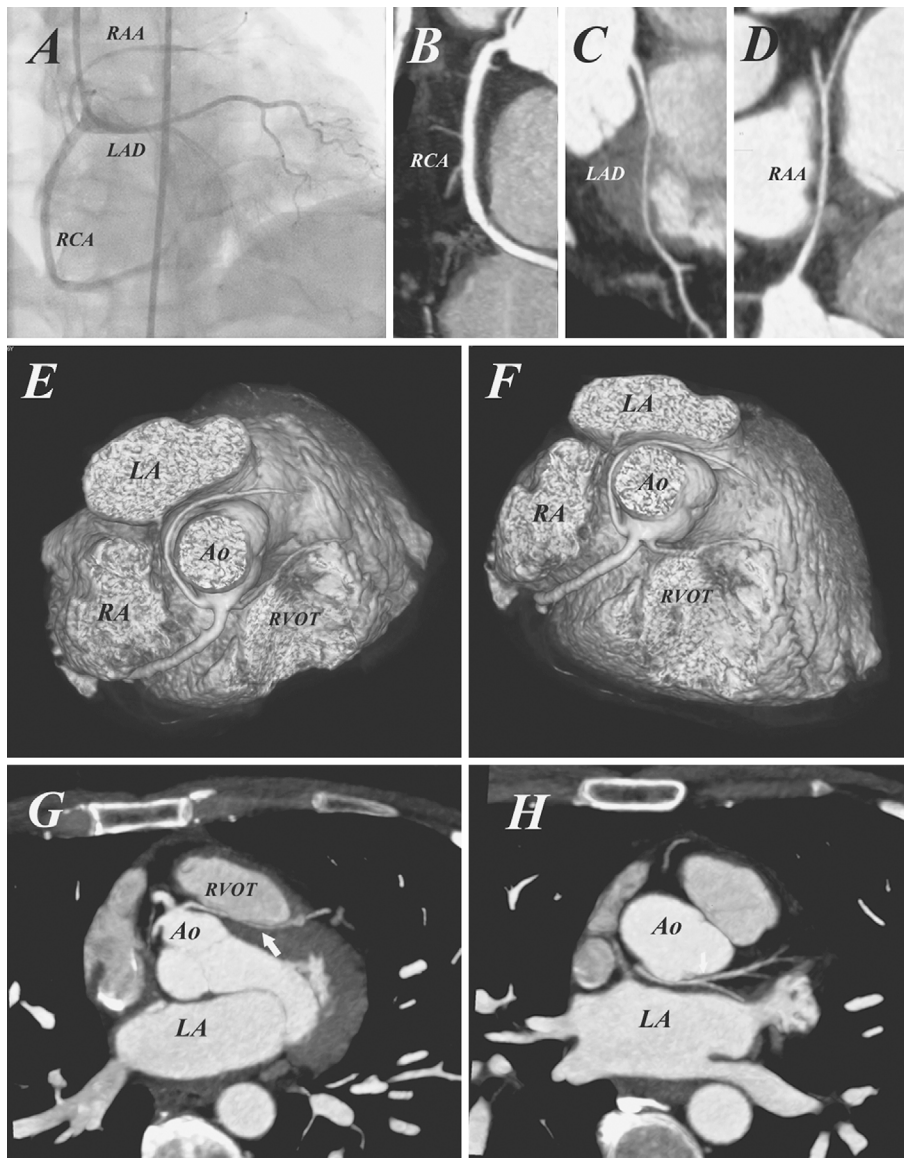


Figure 2. Coronary artery angiography (A) and multidetector computed tomography. MIP curve reconstruction of the RCA (B), LAD (C) and RAA (D). Volume rendering reconstructions (E, F); multiplanar reconstructions of the LAD (G) and RAA (H). RAA: retroaortic artery, LAD: left anterior descending, RCA: right coronary artery, LA: left atrium, RA: right atrium, RVOT: right ventricular outflow tract, Ao: aorta

course of the vessel, have been proposed to explain the increased risk of death associated with some subtypes of anomalous coronary arteries, including a slit-like orifice of the anomalous artery (23), the presence of a valve-like mechanism due to an ostial bridge (24) and (25) acute angulation of the arterial take-off angle with associated coronary kinking during exercise (24), a scissor-like effect on the proximal course of the vessel from the aorta and the pulmonary artery, particularly during exertion (26), and increased aortic wall distensibility with additional compression of an already narrowed ostium during exercise (27).

Obtaining a precise anatomic description of the coronary anomaly is key to making the diagnosis. Unless an accurate distinction is made, clinicians may never know the relationship between these variants and adverse cardiac events.

The authors state that they have no Conflict of Interest (COI).

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