Anomalous Origin of the Left Main Coronary Artery from the Right Coronary Artery

with a Preaortic Course

Omer Yildiz, MD
Kanber Ocal Karabay, MD
Canan Akman, MD
Vedat Aytekin, MD

We report the case of a 51-year-old woman who presented with stable angina pectoris and Canadian Cardiovascular Society class II functional capacity. An electrocardiogram during a treadmill exercise test showed substantial ST-segment depression in the inferolateral leads. Coronary angiograms revealed an anomalous origin of the left main coronary artery from the opposite sinus of Valsalva and an interarterial course between the ascending aorta and pulmonary artery. Although this phenomenon is dangerous, the patient refused further examination. We discuss the diagnosis and treatment of patients who have an anomalous origin of a coronary artery from the opposite sinus of Valsalva. ***(Tex Heart Inst J 2015;42(3):243-5)***

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From: *Departments of Cardiology (Drs. Aytekin, Karabay, and Yildiz) and Radiology (Dr. Akman), Sisli Florence Nightingale Hospital, 34381 Istanbul, Turkey*

Dr. Yildiz is now at the Department of Cardiology, Koc University Hospital, 34365 Istanbul, Turkey.

Address for reprints: *Omer Yildiz, MD, Maltepe Mah. Davutpasa Cad. No:4, 34010 Topkapi, Istanbul, Turkey*

E-mail: *omer.yildiz@e-mail.com.tr*

Coronary artery (CA) anomalies are defined in accordance with their origin, course, and termination.[1] Anomalous origin of a CA from the opposite sinus of Valsalva (ACAOS) with an interarterial course of that artery is a rare congenital defect that might have great clinical impact, particularly myocardial ischemia and sudden cardiac death.[2,3] We discuss the case of a patient in whom this condition was discovered.

Case Report

In January 2014, a 51-year-old woman was admitted to our cardiology department with anginal chest pain. Her risk factors for atherosclerosis were a history of smoking and well-treated arterial hypertension. During a treadmill exercise test, an electrocardiogram (ECG) showed substantial ST-segment depression in the inferolateral leads. Coronary angiograms revealed that the right coronary artery (RCA) was in a normal position, but that the left main coronary artery (LMCA) originated from the RCA and branched into the left anterior descending and left circumflex coronary arteries (Fig. 1).

Because the origin and course of anomalous CAs must be defined, we performed cardiac 64-slice multidetector computed tomography (MDCT), which confirmed the above diagnosis. Moreover, the LMCA followed an intramural course and was severely compressed between the aorta and pulmonary artery. An image of the maximal cross-sectional diameter showed severe lateral compression of the lumen at the intramural segment (Fig. 2A) in comparison with the more proximal extramural LMCA (Fig. 2B).

We recommended intravascular ultrasonography (IVUS) and stress echocardiography, to identify the potential causes of myocardial ischemia and evaluate the ischemia. However, the patient refused further examination and was instead prescribed β-blockers.

Discussion

Typically, CA anomalies are asymptomatic and are diagnosed incidentally during coronary angiography or at autopsy. In certain cases, CA anomalies are associated with sudden cardiac death. In the absence of atherosclerosis, some subgroups of CA anomalies, called ACAOS, can lead to myocardial ischemia and sudden cardiac death

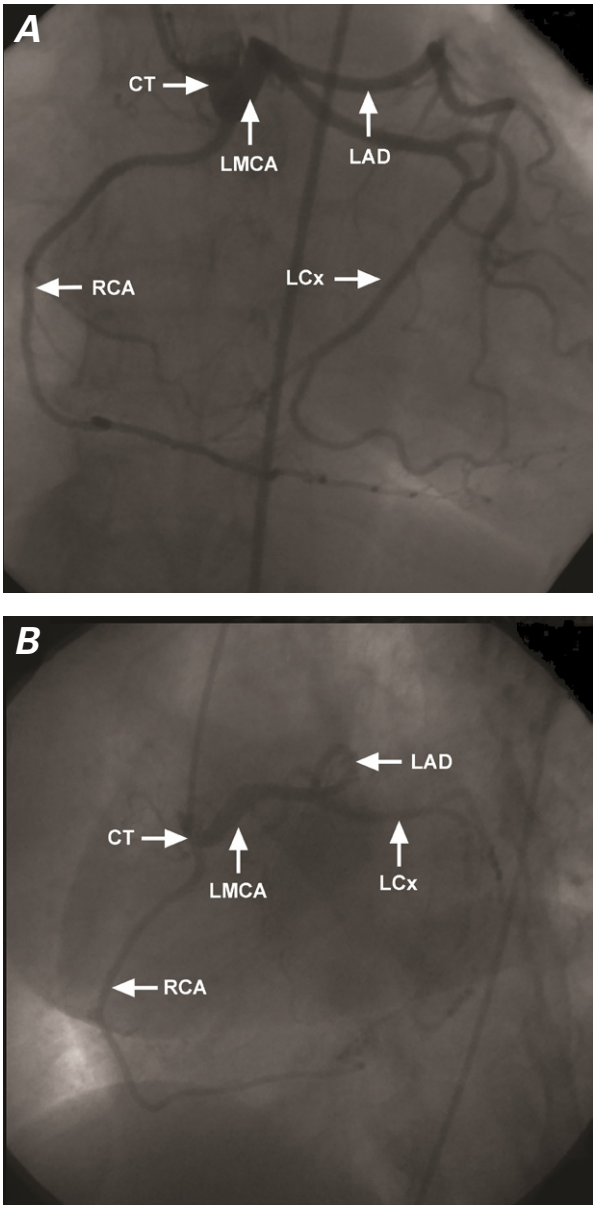

Fig. 1 *Coronary angiograms of a 51-year-old woman in **A**) anteroposterior and **B**) left anterior oblique views show that the entire coronary system arises from a single ostium in the right sinus of Valsalva. The right coronary artery (RCA) splits from a common trunk (CT) and continues in normal position. The left main coronary artery (LMCA) crosses to the left of the CT, and the preaortic course—"between the aorta and pulmonary artery"—leads to the left anterior descending coronary artery (LAD) and left circumflex coronary artery (LCx).*

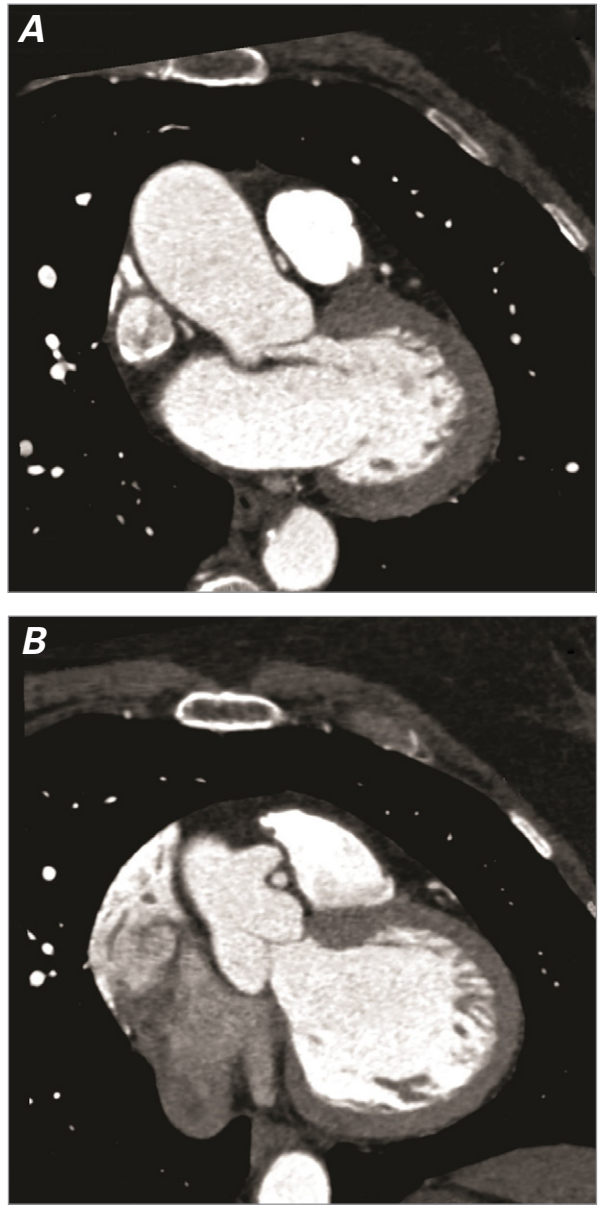

Fig. 2 *Cardiac 64-slice multidetector computed tomogram (cross-sectional view) shows **A**) severe lateral compression of the lumen of the intramural segment of the left main coronary artery in comparison with **B**) the more proximal extramural section.*

on the basis of anatomic malformation, especially in young persons.[2,3]

In a continuous series of 6.3 million 18-year-old recruits who underwent intense military training for 8 weeks, researchers at the Armed Forces Institute of Pathology recorded 21 cardiac deaths that were related solely to ACAOS of the left coronary artery (left ACAOS), and not that of the RCA (right ACAOS).[2,4,5]

Potential mechanisms of stenosis that evoke the aforementioned symptoms include the acute angle of takeoff and kinking of the anomalous CA, intramural proximal intussusception of the ectopic artery at the aortic root wall, mechanical compression of the anomalous CA when it courses within the aortic wall, flap-like closure of the anomalous coronary orifice, and spasm of the anomalous CA as a result of endothelial injury.[2,3]

After IVUS evaluation, an intramural aortic course without an interarterial course has been reported to lead to stenosis in certain types of ACAOS.[2] Therefore, precise images of the anomalous anatomy and an evalu-

ation of mechanisms that underlie the ischemia are essential to the appropriate management of the condition.

Intravascular ultrasonography is the preferred method of determining the mechanisms of ischemia in anomalous origins of a CA.[2] Although coronary angiography is typically used for evaluating CA disease, MDCT is the method of choice for detecting and classifying the complex anatomy of CAs in the absence of atherosclerosis.[3,6]

To treat patients with ACAOS properly, the myocardial ischemia must be documented. Exercise stress echocardiography has high sensitivity in diagnosing myocardial ischemia in ACAOS.[7] In addition, ECG-gated single-photon-emission computed tomography is useful in evaluating myocardial ischemia that is caused by coronary anomalies.[2]

Intramural proximal intussusception of the ectopic artery at the aortic root wall,[2] which is always present in patients with left ACAOS and a specific abnormal course (typically called an interarterial course or "between the aorta and the pulmonary artery"), is associated with a poor prognosis.[2-4,8] In such cases, the potential mechanisms of ischemia should be evaluated with use of IVUS, and the origin and course of the anomalous CA investigated with use of MDCT.

Subsequent examination should include nuclear stress testing to examine effort-induced ischemia and scars, and to establish a baseline for follow-up evaluation in the case of eventual intervention.[2] Despite our limited knowledge of such anomalies, the need for intervention might be justified solely on the basis of IVUS results, as discussed above.[2]

We diagnosed our patient's left ACAOS with use of coronary angiography. Furthermore, we used MDCT to identify the interarterial course of the LMCA between the ascending aorta and the pulmonary artery. We intended to perform IVUS and stress echocardiography. When our patient refused further testing and examination, we recommended that she avoid vigorous exercise and sports. To optimize her outcome, we prescribed β-blockers.

If there is evidence of a mechanism of myocardial ischemia and of myocardial ischemia with a possibly fatal outcome in patients with left ACAOS, surgical repair should be considered for effective treatment. In patients with right ACAOS, coronary angioplasty with stent deployment is an option, but it can be justified only by IVUS results. Otherwise, observation and medical therapy, especially with β-blockers, is preferred in this group of patients.

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