Single coronary artery from the right sinus of Valsalva
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Abstract

We describe a case of a single coronary artery originating from the right coronary sinus (LCA) and right coronary artery (RCA) in a 74-year old woman, with a non-ST elevation acute myocardial infarction (NSTEMI). Diagnosis was made by coronary angiography which ruled out stenosis, and showed normal LCA and RCA branching. The connection path of LCA, with the opposite cusp, was defined retroaortic by multislice computed tomography (CT). The variants of this coronary anomaly, together with their clinical implications and pathophysiology of acute myocardial infarction (AMI) are discussed. Multislice CT is fundamental for clinical decision making.

Introduction

Coronary artery anomalies (CAA) are found in 0.64-5.6% of patients undergoing coronary angiography. They are usually associated with other cardiac anomalies and are classified as anomalies of origination and course, anomalies of intrinsic coronary arterial anatomy, anomalies of coronary termination and anomalous anastomotic vessels. Of particular interest are the anomalies arising from coronary artery from the wrong sinus (incidence 1.07%). The origin of the left coronary artery (LCA) from the right sinus is even rarer (incidence 0.15%), but its diagnosis is very important due to the connection with sudden death in young athletes. Basso et al. reported a series of 27 athletes in whom 85% of sudden deaths were due to the origin of the LCA from the right sinus. Extreme exercise can play a role in triggering such deaths. We describe a 74-year old woman with a single coronary artery arising from right sinus who suffered an acute coronary syndrome.

Case Report

A 74-year old woman was admitted to our department with chest pain that had lasted one hour and dyspnea following emotional distress. On examination, blood pressure was 125/80 mmHg, pulse rate was 88/min and regular, and a systolic murmur 2/6 Levine was audible. The echocardiogram (ECG) showed sinus rhythm and prominent ST elevation in lead V2. During the echocardiogram, akinesia of medium septum, septal apex (Wall Motion Score Index: 1.36) and mild mitral regurgitation were detected. Chest pain, ECG and echocardiogram changes subsided spontaneously 30 min after admission. The peak CK-MB was 7.2 ng/mL (<5 ng/mL), Troponin I was 0.62 ng/mL (<0.04 ng/mL) and, therefore, a diagnosis of a non-ST segment myocardial infarction (NSTEMI) was made. Coronary angiography (CA) showed the entire coronary circulation arising from the right Valsalva sinus, from a single ostium (Figure 1A). After a common main stem (CMS), the artery divided into a dominant RCA, and into a left main stem (LMS) that finally bifurcated into the left anterior descending (LAD) and left circumflex artery (LCX). The coronary arteries were normal without any significant stenosis and without any extrinsic compression. Contrast enhanced CT confirmed the diagnosis (Figure 1B). The large RCA (5mm) and LAD showed no signs of stenosis, coursing behind the aortic bulb and towards the interventricular sulcus (Figure 2). Dipyridamole-echo stress test at maximal heart rate was negative for inducing ischemia. The patient was discharged with the following medications: simvastatin, ramipril, diltiazem and clopidogrel. At follow up at 36-months the patient was in good health without any recurrence of acute coronary syndrome.

Discussion

Ectopic left coronary artery has 5 possible paths arising from the right coronary sinus: anterior to the pulmonary artery, intra-septal, between the aorta and pulmonary artery, posterior to the aorta, and retrocardiac. The anomaly of the course between aorta and pulmonary artery is associated with sudden death and a more severe prognosis. The pathophysiological mechanism is not the compression, scissor-like mechanism of the coronary by aortic and pulmonary roots. The vessel has an aberrant course within the aortic wall and is often hypoplastic and exposed to a lateral compression over the entire proximal intraparietal tract. Tachycardia and increased pulsatility and stroke volume during exercise may induce ischemia, and this could explain the association between this anomaly and sudden death during strenuous physical activity. The clinical manifestations are variable and include: dyspnea, palpitations, angina pectoris, diziness and syncope depending on anatomical relationships, mainly with the aorta and pulmonary artery. Many patients are asymptomatic and the diagnosis is incidental. When the course is between the aorta and pulmonary artery, the coronary anomaly usually occurs early with syncope, chest pain or sudden death. In the other paths, it is often diagnosed by the presence of angina pectoris, induced by a co-existing atherosclerotic plaque that can be effectively treated with coronary angioplasty.

Our patient presented two particularities: this is the oldest age at which such a diagnosis has been made, and the combination of a...
benign course of ectopic coronary artery, with an acute myocardial infarction, in the absence of coexisting coronary atherosclerotic lesions. The ventricular wall motion abnormalities present on admission indicated that the LAD location was the site of ischemia. Emotional stress, which precedes the onset of angina, is the perfect scenario for Prinzmetal's angina. Indeed, the catecholaminergic state can induce a coronary spasm. Although the initial clinical presentation (transient ST-segment elevation in V2) was compatible with such a diagnostic hypothesis, the spasm was not inducible during CA. Moreover, the myocardial necrosis (not usually associated with variant angina), the negative echo-stress test and the absence of any recurrence at follow up, suggest the mechanism of coronary spasm as possible, but not likely. Stress also causes tachycardia, increased pulsatility and increased stroke volume precipitating a condition of ischemia in the tract of coronary artery closest to the aortic root (LMS at the bifurcation in LAD and LCX), by lateral compression of aortic expansion on the ectopic coronary. This compression may be possible even if the retroaortic coronary artery does not lie within the aortic wall, and the presence of such an anomaly has only been described in one case. Finally, the CA excluded a myocardial bridge, coronary hypoplasia, ostium abnormalities (atresia, stenosis) or an atherosclerotic significant stenosis as a cause of myocardial necrosis. However, a transient thrombosis, due to disruption of a plaque, cannot be excluded as a cause of the NSTEMI. A lower degree of initial stenosis may not be visible by CA.

Selective angiography remains the gold standard for diagnosis, but frequently fails to identify the proximal course of coronary arteries while multi-slice CT gives us more detailed information not only on the relationship with the surrounding anatomical structures, but also on the proximal tract of the coronary artery anomaly. This information is crucial for clinical decision making. In our case the patient was considered at low risk of arrhythmia and sudden death and was, therefore, treated conservatively. A careful analysis of clinical presentation together with angiography combined with multislice coronary CT is useful to optimize therapy and minimize the risk to the patient.

Figures 1 and 2

Figure 1. (A) Coronary angiography, left anterior oblique view. CMS, common main stem; LMS, left main stem; LAD, left anterior descending artery; LCX, left circumflex; RCA, right coronary artery. (B) Same projection to the multi-slice CT scan (LightSpeed VCT 64-slice Scanner, GE Healthcare) showing a single coronary artery.

Figure 2. (A) Lateral view. Shows the retroaortic course of anomalous coronary artery. IVP, posterior interventricular coronary artery. (B) Caudal left anterior oblique view.

References