

A rare case of a congenital anomalous origin of the coronary arteries: All in one

Nadir bir vaka olarak koroner arterlerin konjenital çıkış anomalisi: Hepsi bir arada

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ABSTRACT

Congenital anomalous origin of the coronary arteries is a rare condition with an incidence of 0.6-1.3% in angiographic series. The majority of patients with an anomalous origin of the coronary artery is asymptomatic. They have higher risk of sudden cardiac death if there is external compression of the coronary arteries by the great arteries. There still is not consensus on the management in these patients. In this case report, we present a rare case of a coronary anomaly where both the right coronary artery and the left main coronary artery arise from the right coronary sinus and there is a single ostium.

Keyword: Coronary anomalous, Right coronary sinus, Single coronary ostium

ÖZET

Koroner arterlerin konjenital çıkış anomalisi nadir bir durum olup anjiyografi serilerinde insidansı %0,6-1,3 olarak gösterilmiştir. Koroner arter anomalisi olan hastaların çoğunda bulgu olmamasına rağmen, büyük arterlerin koroner arterlere dışarıdan bası oluşturması durumunda yüksek ani kardiyak ölüm riskine sahiptirler. Bu hastaların tedavisinde hala bir fikir birliği bulunmamaktadır. Bu olgu sunumunda, sağ koroner arterin ve sol ana koroner arterin sağ koroner sinüste, tek bir çıkış yolundan köken aldığı nadir bir koroner anomali vakasını sunduk.

Anahtar Kelimeler: Koroner anomali, Sağ koroner sinüs, Tek koroner ostium

Introduction

Congenital anomalous origin of the coronary arteries is a rare but well-described cause of myocardial ischemia and sudden death. Previous studies have shown that the incidence of congenital anomalous origin of the coronary arteries is 0.6-1.3% in angiographic series and 0.3% in autopsy series [1]. Coronary arteries with anomalous origins can result in episodic or obligatory myocardial ischemia and have been implicated in chest pain, syncope, myocardial ischemia, malignant ventricular arrhythmia, and sudden cardiac death. It is still controversial whether surgical treatment is necessary in asymptomatic patients.

Case Report

Here we present a 64-year old male patient who applied to our clinic with complaints of dyspnea, orthopnea, and pretibial edema for 2 months. His exercise capacity was the New York Heart Association (NYHA) class 3. He had a history of diabetes mellitus, atrial fibrillation and a 20 pack-year history of smoking. On physical examination, bilateral respiratory rales, 2+ bilateral pitting pretibial edema and irregular pulse were found. On electrocardiography, atrial fibrillation with a rate of 140 per minute was detected. Moderate depression of left ventricle systolic function and dilatation of the left ventricle were revealed on transthoracic echocardiography. Coronary angiography was performed to determine the etiology of the left ventricle systolic dysfunction. Left sided coronary arteries were not visualized at the left coronary sinus. During the visualization of the right coronary artery, both right coronary artery and left main coronary artery were found to arise from the right coronary sinus and from a single ostium. This was confirmed by aortography (Figure 1). Upon further investigation, multi-slice computer tomography (MSCT) demonstrated that both left and right coronary artery originated from the right coronary sinus and from a single ostium with no compression by the great arteries (Figure 2 A-D).

Discussion

A congenital anomalous origin of the coronary arteries is a rare condition. Single coronary artery anomalies (CAA) are unusual

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Submitted/Gönderilme: 04.01.2014 Accepted/Kabul: 24.03.2014

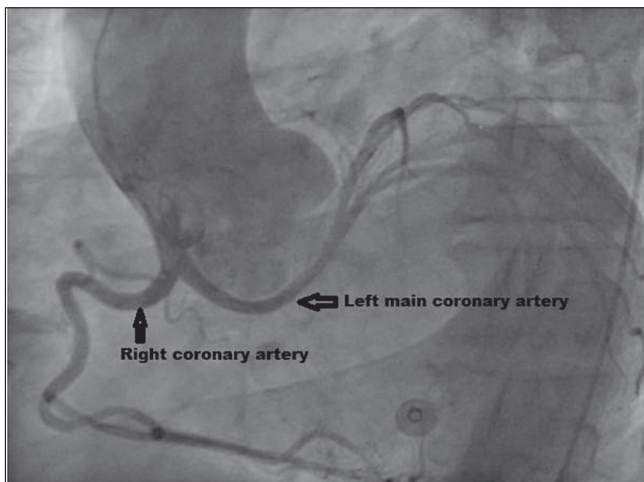


Figure 1. Aortography showed that right and left coronary artery originated from the right coronary sinus.

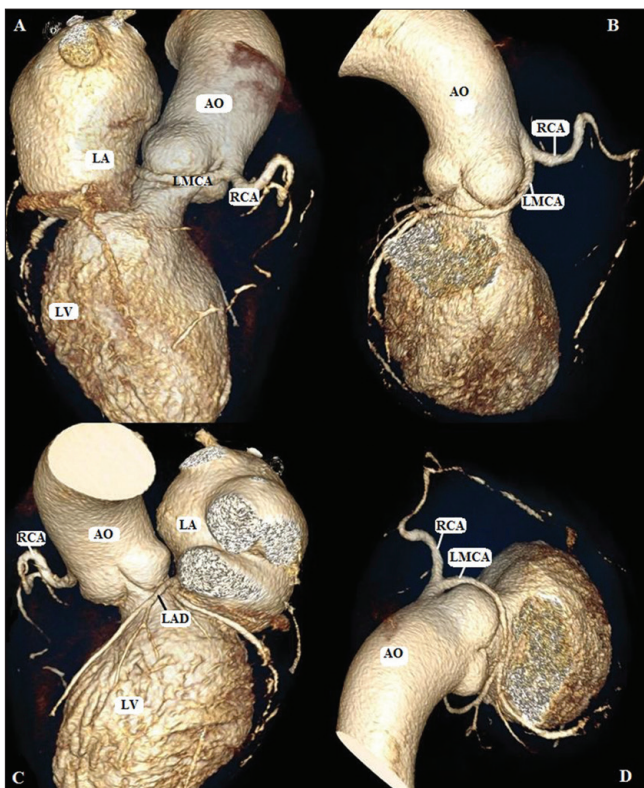


Figure 2. Multislice coronary computer tomography demonstrated that both left and right coronary arteries originated from the right coronary sinus and there was a single ostium with no compression by the great arteries. (LMCA: left main coronary artery, LAD: left anterior descending, RCA: right coronary artery, LV: left ventricle, LA: left atrium, AO: Aorta)

conditions defined as the coronary artery arising from a single coronary ostium [1]. Previous studies have demonstrated that single CAA incidence ranges from 0.24 to 0.66 % [1]. The anomalous origin of the left coronary artery from the right sinus with an interarterial course has a prevalence of 0.17% [2]. It is diagnosed by the presence of a single ostium in one coronary artery with a

lack of any other coronary artery origin from an ectopic area [3]. CAAs are classified according to origin, course, and termination of the coronary arteries [3-5]. Various congenital heart diseases such as patent foramen ovale, bicuspid aortic valve, tetralogy of Fallot, coronary arteriovenous fistula, truncus arteriosus, ventricular septal defect, transposition of the great vessels, and patent ductus arteriosus are associated with single CAAs [6].

Although the majority of patients with a single CAA are asymptomatic, serious complications such as angina and sudden cardiac death may result from this anomaly [7, 8]. These complications can occur due to interarterial course of the coronary artery and its compression by the great vessels [9, 10]. On the other hand, severe coronary stenosis, vasospasm, and the formation of a muscular bridge are also possible causes of these complications [11]. Electrocardiographic abnormalities, new onset of chest pain, dyspnea, and syncope may also present in these patients [12]. Previous studies have shown that CAA is the second most important cause of sudden cardiac death in young athletes [13]. Several hypotheses have been proposed to explain this phenomenon: expansion of the aortic and pulmonary roots during systole could cause compression of a common trunk with an anomalous course, or excessive angulation at the origin of an anomalous coronary could compromise the lumen when there is dilation of the aortic root.

The coronary artery anomaly is often diagnosed at autopsy due to asymptomatic presentation of the anomaly [14]. Patients with CAA are usually diagnosed incidentally during diagnostic coronary angiography. Although coronary angiography is the gold standard method for the evaluation of CAA, it is inadequate in detecting the relationship of anomalous vessels to great vessels and other structures of the heart. Therefore, MSCT and magnetic resonance imaging have recently been used for documentation of coronary anomalies and its relation to the great arteries [15-18]. These are also safe methods and give detailed information with three-dimensional reconstructions of the coronary arteries. There still is not a consensus on the management of anomalous coronary arteries with origin from the contralateral aortic sinus. Medical therapy, percutaneous coronary intervention (PCI) and surgery are therapeutic choices for patients with CAA. Surgery is indicated when the left coronary arteries arise from the right sinus and course between the aorta and pulmonary artery [16]. Surgical methods include direct repair of the anomalous origin in the aortic root or coronary artery bypass surgery. In symptomatic patients, surgery is often indicated, but in asymptomatic patients, surgery is more controversial. Although PCI is technically very difficult, there are some cases in the literature [11, 19]. In our patient, there was no external compression to the coronary arteries. Therefore, the patient was discharged with optimal heart failure therapy.

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