First Yemeni Coronary Congenital Anomaly Visualized with Multidetector Computed Tomography: Case Report

Yemen'de Multidetektör Bilgisayarlı Tomografi ile Saptanan İlk Koroner Konjenital Anomali Olgusu

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ABSTRACT Congenital coronary artery anomalies are rare and usually lack clinical significance. There are, however, some malignant cases that may cause even myocardial infarction or sudden death. Till two decades ago, invasive coronary angiography was the gold standard procedure to study congenital coronary anomalies, although it still had important limitations. In the last years, several studies showed the usefulness of the non-invasive modalities for the detection of coronary vessel anomalies, such as magnetic resonance imaging, and especially multidetector computed tomography (MDCT). In this case report we present the first Yemeni coronary congenital anomaly visualized with MDCT. We also present a review of the literature and discuss the recent approved clinical practice guidelines.

Key Words: Multidetector computed tomography; coronary vessel anomalies; coronary artery disease

ÖZET Doğumsal koroner arter anomalileri çok nadir olup, genellikle klinik açıdan önemsizdirler. Ancak miyokard infarktüsüne ya da ani ölüme yol açabilecek bazı malign vakalar da bulunmaktadır. Yirmi yıl öncesinde invaziv koroner anjiyo, bu doğumsal koroner anomalilerinin saptanmasında, bazı yetersizliklerine rağmen altın standart işlem idi. Son yıllarda bu konuda manyetik rezonans görüntülemenin, özellikle de multidetektör bilgisayarlı tomografinin (MDCT) invaziv olmayan yöntemler olarak daha yararlı olduğunu gösteren çalışmalar yayınlanmıştır. Bu olgu raporunda, biz Yemen'de MDCT ile görülebilen ilk koroner konjenital anomali vakasını sunuyoruz. Olgu eşliğinde literatür bilgisinin de tartışmasını yapmış bulunmaktayız, böylece güncel onaylanmış klinik uygulama kılavuzlarından da bilgiler verilmiştir.

Anahtar Kelimeler: Multidetektör bilgisayarlı tomografi; koroner damar anomalileri; koroner arter hastalığı

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In humans, coronary arteries and their main branches have an epicardial course running over the cardiac musculature. A coronary artery can be classified as "normal" when it is observed in > 1% of an unselected population, as "normal variant" when, although it has a relatively unusual morphological feature, it is seen in > 1% of the same population and as an "anomalous" artery when it has a morphological feature seen in < 1% of that population.[1]

Congenital coronary artery anomalies are a heterogeneous group of diseases. In the majority of cases they lack clinical significance and are merely epiphenomena found accidentally during necropsies, while performing in-

vasive or non-invasive coronarography, or during surgical interventions. Subjects are usually asymptomatic, but may become symptomatic after some decades of life, especially if there are associated precipitating factors. However, in some cases there are "malignant" anomalies that may be responsible for chest discomfort, malignant arrhythmias, fatal or non fatal acute myocardial infarction, syncope and sudden death. Older patients may have both coronary artery disease (CAD) and coronary vessel (CV**s**) anomalies and in these cases it is difficult to clarify the exact mechanism of myocardial ischemia.

The procedure used to define the normality of CVs may be a bias. In fact, invasive coronary angiography (ICA) is performed in symptomatic patients, while necropsies are done for medico-legal purposes. This bias explains why myocardial bridges are rare in patients referred for cardiac surgery (0.2-0.3%) or ICA (0.4-4.9%), while they are very frequent during autopsy (5.4-85.7%).[2] Such disparate autopsy prevalence rates may result from the selection and preparation of the hearts, variation in definitions and probably also to ethnicity.[3] On the other side, coronary anomalies of origination and course are rare during autopsy (0.17% of the cases), while their incidence is higher in the population of patients referred for coronary angiography (0.6-1.3%).[1]

To date, ICA is the gold standard technique for the evaluation of CVs. However, ICA has some limitations as it provides a few 2D view images of the CVs and sometimes fails to clearly visualize the relationship between the CVs and the surrounding structures. With ICA it is not always easy to selectively engage the anomalous CV, which may lead to the erroneous assumption that the CV is occluded. In addition, with this traditional 2D technique it is more difficult to understand the course of the CV within the heart and discern the anterior versus the posterior direction of the anomalous vessels.

In the last few years, several studies showed the usefulness of non-invasive modalities for the detection of CV anomalies, such as magnetic resonance imaging (MRI) and especially multidetector computed tomography (MDCT). MDCT provides an unlimited number of 2D reformatted images as well as 3D images of the single vessel making it possible to have a 3D depiction of the whole heart.[4] In the literature, there are several papers where MDCT was successfully used to visualize anomalous CVs.[5,6]

In this paper, we describe the anomalous origin and course of a coronary artery in a young patient referred for chest pain and suspected CAD.

CASE REPORT

The patient of this case report is a 35 years old active male who was referred to us after having performed ICA for suspected CAD. He did not have a family history of relevant disease, no past history of allergy, surgical intervention, diabetes, hypertension, and he did not take any kind of medication. He was a habitual khat chewer, but not smoker. Some months ago he began to complain of left sided chest pain aggravated by exercise and anxiety. Basal physical examination, blood pressure (120/80 mmHg) and blood tests were within normal limits. No abnormalities were found in the electrocardiogram; his heart rate was 72 beats per minute (BPM). Chest X-ray, trans-thoracic echocardiogram and maximal treadmill stress test were also normal. However, since chest pain became recurrent and of sudden onset, he was sent to perform ICA.

ICA showed that right coronary artery (RCA) was the dominant vessel. The left coronary artery (LCA) was anomalous as it jointly originated with RCA from the right sinus of Valsalva. Although several projections were performed, there were some doubts regarding the course of the LCA. No plaques were detected in the LCA and RCA.

To better evaluate the patient's coronary tree MDCT was performed (SOMATOM® Definition AS+ CT from Siemens Healthcare) by administering 80 ml of iodinated contrast medium (370 mg I/ml, Ultravist 370; Bayer Schering Pharma, Berlin, Germany) followed by 50 ml of saline solution at a rate of 5 ml/s through a 18 gauge cannula placed in the

antecubital vein. To reduce patient's heart rate, one hour before the test he was given 40 + 40 mg of Propranolol per os. Images were acquired during a 10 to 15 second breath-hold with retrospective electrocardiographic gating. Scanning was triggered to commence automatically when a predetermined level of contrast enhancement was detected in the ascending aorta. The tube voltage was 120kV, and the effective tube current was of 92 mA. Table feed was 5 mm per gantry rotation, with a pitch of 0.2. The radiation dose was of 403 mGycm.

After data acquisition, images were reconstructed especially at the 75% of the RR interval, with a thickness of 0.6 mm and increment of 0.3 mm. Original axial images, oblique and curved multiplanar reconstructions along the axis of the vessel of interest, and the cross-sectional images perpendicular to the vessel's centerline were used to evaluate coronary lumen.

All coronary segments were visualized and images were judged of good quality (Figure 1 and Figure 2). RCA was the dominant vessel and had no lesions. Also, the posterior descending coronary artery, three right ventricular branches and a posterolateral coronary artery were free from lesions. The LCA jointly originated with the RCA and then bifurcated in left anterior descending coronary artery (LAD) and left circumflex artery (LCX). Both LAD and LCX, as well as a diagonal branch and the obtuse marginal branch, were free from significant calcified or non-calcified plaques.

The results of MDCT clearly showed that there was not a CAD, but an anomalous origin of the LCA which passed posterior to the root of the aorta till the left side of the aortic arch where it divided into the LDA and LCX.

The patient was informed about his coronary artery anomaly and reassured that there wasn't any myocardial ischemia. No cardioactive treatment was administered and the patient was addressed to perform regular follow-up.

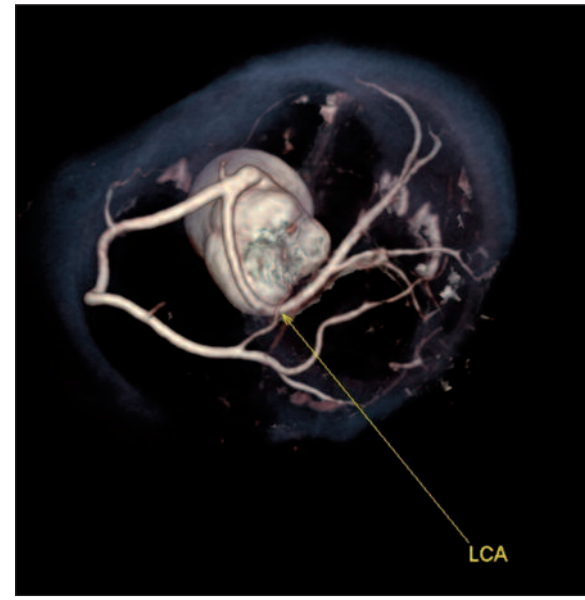

FIGURE 1: Volume rendering image of the heart with the origin of a single coronary artery from the right sinus of Valsalva.
(See color figure at
http://www.turkiyeklinikleri.com/journal/cardiovascular-sciences/1306-7656/)

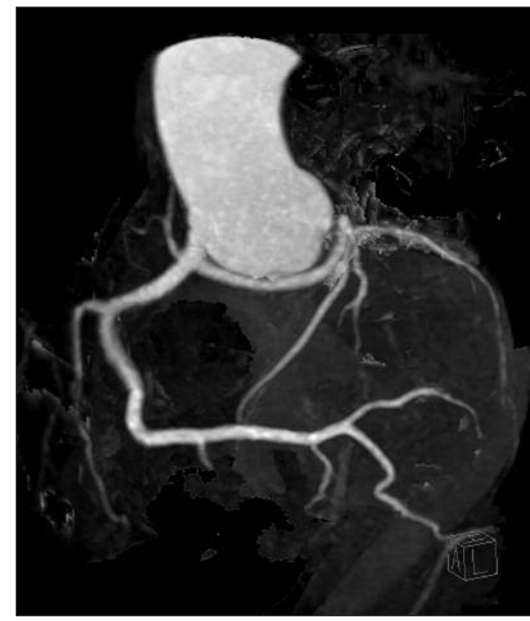

FIGURE 2: 3 D image of the coronary tree which originates from the right sinus of Valsava before bifurcating in the right and left coronary artery.

DISCUSSION

In Yemen there isn't a national registry of congenital cardiac anomalies. However, in the major hospitals of the country there are invasive and non invasive laboratories where patients with cardiac and in particular coronary artery anomalies are evaluated and treated. To our best knowledge this is the first patient with a coronary artery anomaly to be studied with MDCT.

In this case report, MDCT clearly visualized the coronary anatomy of a young patient with suspected CAD, detected the presence of the anomalous origin of the LCA, and excluded the presence of CAD.

Till now the "gold standard" technique to evaluate both normal and abnormal coronary arteries is ICA, and in more than a half century of experience it proved to be extremely reliable in the diagnostical processes of CAD patients. However, ICA is unable to give a 3D depiction of the vessel's course.[4]

Although MRI is a promising tool to visualize coronary arteries and, most importantly for young patients is radiation-free, for the moment it does not allow a comprehensive visualization of coronary arteries' course.

The introduction in the cardiac arena of MDCT has made imaging of the heart and, in particular, of epicardial coronary arteries feasible. In the last two decades MDCT has been used to study different groups of subjects, becoming in some cases the new "gold standard" technique instead of ICA, because of its ability to correctly visualize coronary arteries and, most interestingly, to obtain this information non-invasively.[4-8] The information provided by MDCT is very useful to invasive cardiologists and cardiac surgeons as it helps them to plan their interventions by directly seeing the exact course of the vessel and its relationship within the heart and with the other intra-thoracic organs and chest wall.[5]

In the recent American Appropriate Use Criteria Task Force for MDCT, the use of MDCT in the "assessment of anomalies of coronary arterial and other thoracic arteriovenous vessels" was pointed to be most appropriate (i.e. the test is acceptable and considered a reasonable approach to study the disease, and its expected incremental information, combined with clinical judgment, exceeds the expected negative consequences by a sufficiently wide margin) with a score of 9 out of 9.[8]

Furthermore, in the ongoing debate concerning the evaluation of asymptomatic young athletes who have a low probability to have an atherosclerotic coronary artery disease, the use of MDCT is under discussion. Some of these athletes, during or just after physical exertion, or in circumstances non-associated with sports (i.e., during routine daily activities or while sedentary or even asleep) may have an unexpected death. In an autopsy-based registry comprising 1866 young athletes (19 ± 6 years) the cause of sudden death in 56% of the cases was due to a cardiovascular disease. Sudden death was attributable to hypertrophic cardiomyopathy in 36% of the cases and to coronary artery anomalies in 19% of the cases (119 cases of coronary artery anomalies of wrong sinus origin and 24 cases of myocardial bridges).[9]

MDCT has however some important limitations that must be considered. ICA is still superior over MDCT because it has a higher spatial (<0.16 mm vs. 0.4 mm) as well as temporal resolutions (33 msec. vs. 140 to 200 msec. of the recent cardiac computed systems, or 83 msec. of the dual source systems). Another limitation of MDCT still present with the currently available 64 channel systems is related to patient's heart rate, which must be rhythmic and around or less than 60-64 BPM. Patients with atrial fibrillation or with a heart rate that cannot be reduced to 60-64 BPM, for the moment, are not eligible. The introduction of new tools like the "ECG-tube current modulation" and the "step and shoot" procedures and the 128, 256, 320 and 640 channel or dual source scanners, offers the possibility to study also patients with higher heart rates and with atrial fibrillation, making it possible to image the entire heart, not only, as it is now, in a single breath hold, but in a single heartbeat.[5] Moreover, almost 5% of patients have a un-evaluable MDCT scan either due to motion artifacts and involuntary motion of the diaphragm, or because of patients who are overweight, have respiratory problems, or they are not compliant with breathing commands.

Particular attention must be also given to the radiation dose delivered to patients. In the commonly used MDCT systems the amount of radiation, expressed as units of millisieverts (equivalent to millijoules per kilogram of tissue), absorbed by

patients during the test is 2-4 folds that of ICA. However, recent improvements in MDCT technology, like the introduction of the ''step and shoot'' protocols, reduced the radiation dosage significantly to almost equal to that of ICA.[10] Finally, it is worth noting that both ICA and MDCT use non-ionic contrast medium to visualize coronary artery lumen. For this reason, particular attention must be given in allergic patients and in patients with a pre-existing renal impairment.

In this paper, we describe the usefulness of MDCT in evaluating a young patient suspected to have CAD. In particular, MDCT was able to detect the anomalous origin of the LCA, which arose from the right sinus of Valsalva, and to exclude the presence of coronary plaques.

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