A patient with ischemic symptoms presents with left coronary artery-to-right ventricle fistula and agenesis of the right coronary artery

İskemik semptomları olan hastada sağ koroner arter agenezisinin eşlik ettiği sağ ventriküle fistül yapmış sol koroner arter

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Summary– Coronary artery fistulas are rare vascular anomalies characterized by abnormal communication, devoid of a capillary system between the coronary artery and the major vessels or cardiac chambers. In this report, we present a 14-year-old male patient with ischemic symptoms, a left coronary artery to right ventricle fistula and agenesis of the right coronary artery. The anatomy of the coronary arteriovenous fistula was determined in detail through a dual source CT coronary angiography. The patient underwent open cardiac surgery because of ischemic symptoms and a residual fistula was detected after the surgery.

Özet– Koroner arter fistülleri kalbin dört boşluğu veya büyük damarları ile koroner arterler arasında kapiller sistemi olmayan anormal bağlantı ile kendini gösteren nadir görülen damar anomalileridir. Bu yazıda, iskemik semptomları bulunan, sağ koroner arter agenezisinin eşlik ettiği sağ ventriküle fistül yapmış sol koroner arteri olan 14 yaşında bir hasta sunuldu. Koroner arteriyovenöz fistülün anatomisi çift kaynaklı bilgisayarlı tomografi ile ayrıntılı olarak gösterildi. Hastaya iskemik semptomları nedeniyle açık kalp cerrahisi uygulandı. Cerrahi sonrası fistülün tam kapanmadığı görüldü.

Coronary artery fistulas (CAFs) are rare congenital anomalies that indicate communication between the coronary arteries and the cardiac chamber or large vessels. CAFs have been detected in 0.3-0.8% of the patients that have undergone coronary angiography (CAG).[1,2] The fistulas most commonly originate from the right coronary artery (RCA) (~52% of cases), the left anterior descending (LAD) artery (~30% of cases), or the left circumflex (LCx) artery (~18% of cases).[3]

Coronary artery fistulas anomalies are difficult to detect clinically. They are usually asymptomatic during the first two decades, especially if hemodynamically small. However, medium or large fistulas can be associated with angina, myocardial infarction, endocarditis, or congestive heart failure.

Cardiac catheterization and angiography are the main diagnostic techniques used to identify CAFs. However, these techniques often do not reveal the origin of the course or the drainage site of CAFs.[4] More detailed structural information can be provided using multidetector computed tomography (MDCT). In particular, three-dimensional (3D) MDCT in conjunction with multiplanar reformation has been used to generate more a detailed structural analysis.

Abbreviations:

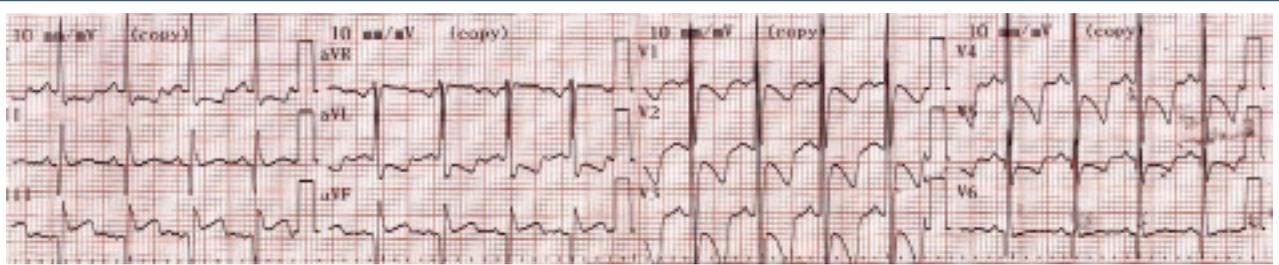|	
CAFs	Coronary artery fistulas
CAG	Coronary angiography
CTA	CT angiography
LAD	Left anterior descending artery
LCx	Left circumflex artery
LMCA	Left main coronary artery
MDCT	Multidetector computed tomography
RCA	Right coronary artery
TTE	Transthoracic echocardiography

Figure 1. Electrocardiogram shows inferolateral ST depression and T wave changes.

CASE REPORT

A 14-year-old male presented with a short history of angina and exertional dyspnea. On auscultation, a grade 3-4 continuous murmur could be heard over the right inferior sternal border. An electrocardiogram showed inferolateral ST depression and T wave changes (Figure 1), while a chest radiography showed mild cardiomegaly and slightly increased pulmonary vascularity.
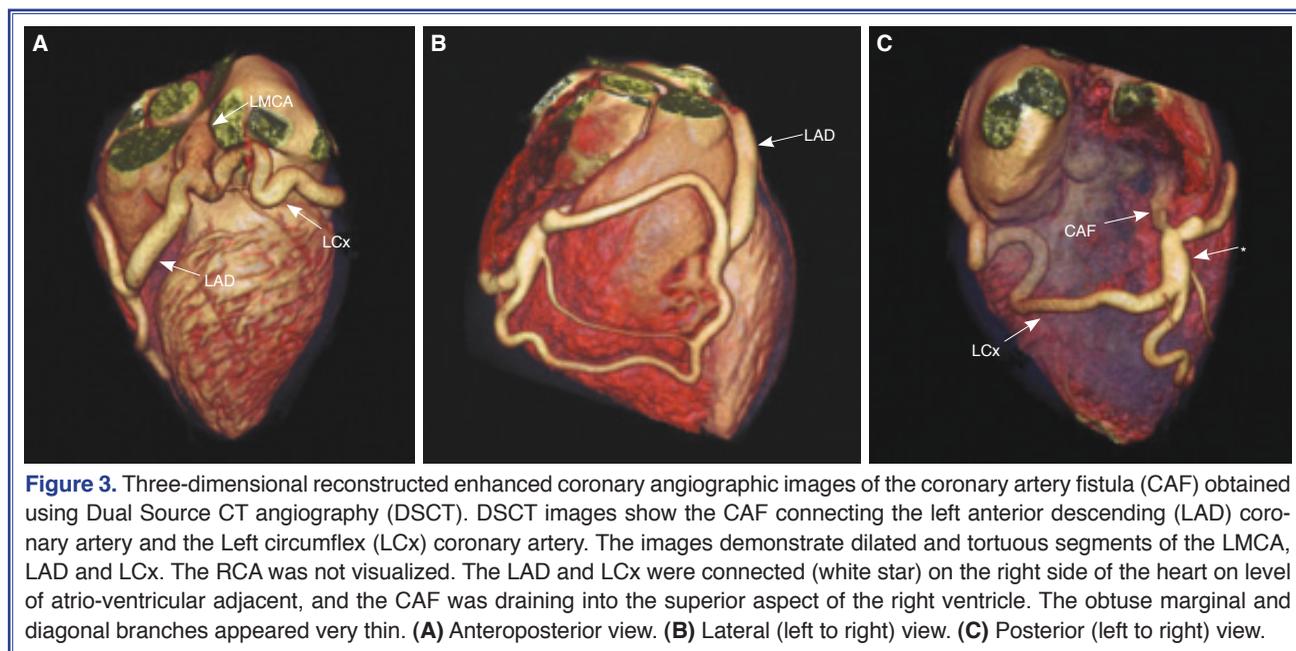
Transthoracic echocardiography (TTE) showed an abnormally large left main coronary artery (LMCA) with right ventricle continuous flow. The RCA agenesis also was detected by TTE. Given the patient's ischemic symptoms and our clinical suspicions of CAF, a selective CAG was performed. The conventional angiography showed a highly dilated and tortuous LMCA, LAD artery and LCx artery. The LAD and LCx were connected on the right side of the heart. However, the drainage site of the fistula was not clear, and the RCA was not visualized (Figure 2). We subsequently performed dual source CT angiography (CTA) to determine the detailed anatomy and the drainage site of the fistula. The patient was pretreated with atenolol (0.8 mg/kg) two hours before the procedure. Following administration of 50 ml of iodinated contrast medium (Ultravist 370, Schering AG, Berlin, Germany) to an antecubital vein at 5 ml/s, a diagnostic coronary CTA was performed on a dual-source CT system (DSCT; Somatom Definition; Siemens Medical Solutions, Erlangen, Germany) using retrospective ECG gating. The following imaging parameters were implemented: 128 rows x 0.6 collimation; rotation time, 330 ms; helical pitch, 0.28; tube voltage, 120 kV; and tube current, 80 mA. Scanning yielded an anteroposterior view of the chest which was used to position the imaging volume for coronary artery imaging. The imaging volume extended from the carina to below the diaphragm face of the heart.

Source images were reconstructed in both systolic and diastolic phases with a 0.75 mm slice thickness. Multiphase reconstruction data images were also utilized to assess the coronary artery segments. Images demonstrated dilated and tortuous segments of the LMCA, LAD and LCx (diameter of LMCA: 11.5 mm, LCx: 10 mm, LAD: 6.7 mm). The RCA was not visualized. The LAD and LCx were connected on the right side of the heart on level of atrio-ventricular adjacent, and the fistula coursed towards to the right ventricle (Figure 3 and 4). The diameter of the fistula tract was measured as 14x4 mm. The anatomy of the coronary arteriovenous fistula was then determined in detail by

Figure 2. Aortagram at age 14 showed that the LMCA, LAD and LCx were dilated and tortuous. The fistula (black arrows) displayed its origin quite well, but the course and drainage site were less clearly visualized. The RCA was not visualized.
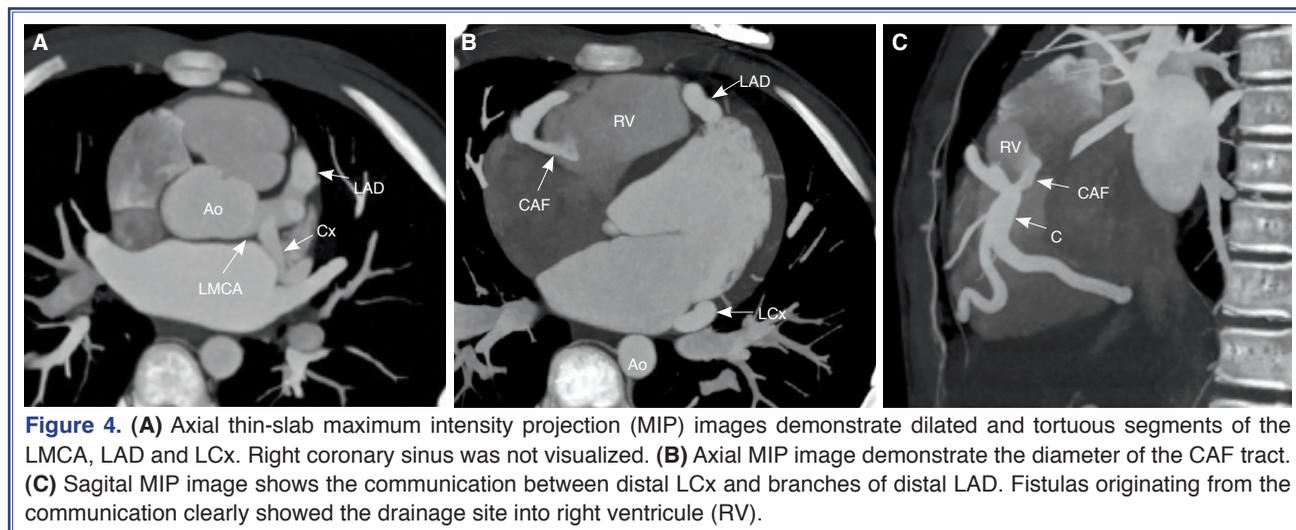
Figure 3. Three-dimensional reconstructed enhanced coronary angiographic images of the coronary artery fistula (CAF) obtained using Dual Source CT angiography (DSCT). DSCT images show the CAF connecting the left anterior descending (LAD) coronary artery and the Left circumflex (LCx) coronary artery. The images demonstrate dilated and tortuous segments of the LMCA, LAD and LCx. The RCA was not visualized. The LAD and LCx were connected (white star) on the right side of the heart on level of atrio-ventricular adjacent, and the CAF was draining into the superior aspect of the right ventricle. The obtuse marginal and diagonal branches appeared very thin. **(A)** Anteroposterior view. **(B)** Lateral (left to right) view. **(C)** Posterior (left to right) view.

dual source CT CAG (see Video*).

The patient underwent open cardiac surgery because of ischemic symptoms at another hospital. The heart was not stopped during the operation. In addition, a cross clamp was not inserted and cardioplegia solution was not administered. The patient's coronary arteries were palpated and the drainage site of the fistula was detected manually. A right ventriculotomy was performed. More than one orifice of the fistula was seen in the right ventricle free wall and was closed completely by the surgical technique. Follow-up echocardiography revealed minimal residual fistula flow.

DISCUSSION

CAF is a rare congenital anomaly and composes the 0.2-0.4% of all cardiac anomalies.[5] Generally, patients with CAF are asymptomatic, however some may present with chest pain, heart failure or infective endocarditis.[4,6]

In spite of its invasive nature, CAG is a standard diagnostic method for CAF. However, the complex structure of abnormal veins, anatomic structures neighboring the fistula, course of the fistula, and the site of drainage may not be accurately detected due

Figure 4. (A) Axial thin-slab maximum intensity projection (MIP) images demonstrate dilated and tortuous segments of the LMCA, LAD and LCx. Right coronary sinus was not visualized. **(B)** Axial MIP image demonstrate the diameter of the CAF tract. **(C)** Sagital MIP image shows the communication between distal LCx and branches of distal LAD. Fistulas originating from the communication clearly showed the drainage site into right ventricule (RV).

to two-dimensional monitoring. MDCT is a more accurate non-invasive imaging method that can successfully show the complex structure of CAF in two or three dimensions.[7]

CAFs can be diagnosed as non-invasive by TTE, but the reliability of TTE in CAF diagnosis is limited and cannot show the fistula as a whole in many cases. TTE was able to show the abnormal dilated coronary arteries and the drainage site of the fistula in our patient. In addition, the RCA agenesis was also detected by TTE. CAG showed the origin of the fistula, but the course and drainage site could not be detected accurately. The use of the MDCT accurately showed the course and drainage site of the fistula.

MDCT has been used for coronary artery imaging since 2000. MDCT has been shown to provide clear anatomic images of high spatial resolution by minimizing cardiac motion artifacts using retrospective electrocardiogram-gated reconstruction methods.[8] Consequently, MDCT is an alternative method to CAG that shows the course, the drainage site, the origin of the fistula and the relationships between anatomic structures. Coronary CTA is a noninvasive diagnostic method that is singly sufficient for diagnosis in cases where coronary anomaly and coronary CAF are suspected.

There has been general agreement that all symptomatic patients with coronary fistulas should undergo closure of the fistulas as soon as possible after diagnosis, because of the increased morbidity and mortality associated with delay.[9]

Surgical closure of the fistula is a long term effective and safe method. However, closure of the fistula by catheterization is preferred to surgical method. This method could not be applied in our patient due to dilated and tortuous coronary arteries, so surgery had to be performed. Follow-up echocardiography revealed minimal residual fistula flow.

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