Coronary Artery Fistulas in Adults: Evaluation with Coronary CT Angiography

Yetişkinlerde Koroner Arter Fistülleri: Koroner BT Anjiyografi ile Değerlendirme

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Öz

Bu çalışmada, kurumumuzda koroner BT Anjiyografi (BTA) uygulanan erişkin hastalarda koroner arter fistüllerinin (KAF) prevalansını ve tiplerini, özellikle köken, seyir, terminasyon ve ilişkili anomalilere odaklanarak değerlendirmeyi amaçladık. Ağustos 2006-Ağustos 2020 tarihleri arasında kurumumuzda EKG-gated koroner BTA yapılan 2462 hasta çalışmaya alındı. KAF tanısı alan 11 hastanın kayıtlarını bulduk. Değerlendirme, fistül orifis ve drenajını, koroner arter hastalığının varlığını, ilişkili konjenital veya edinilmiş anomalileri ve komşu yapılarla ilişkilerini içeriyordu. 2462 hastanın 11'inde (%0.44) (7 kadın, 4 erkek; yaş aralığı 35-73 yaş, ortalama= 55.2 yaş, SD±11.38) KAF'lara koroner BTA tanısı konuldu. 7 vaka tek fistül damarı ve dört vaka birden fazla fistül damarı vardı. 11 CAF arasında, 11 Sol Ana Koroner Arter kaynaklı ve altı Sağ Koroner Arter kaynaklı dahil olmak üzere 17 fistül vardı. 17 fistüle ait drenaj bölgelerine ilişkin dağılım sıklığı şu şekildeydi: dokuz pulmoner arter, olgu iki sol ventrikül, iki sağ ventrikül, iki koroner sinüs, bir sol bronşiyal arter, bir sol atriyum. 3D rekonstrüksiyonlu kardiyak BTA; bölge, kaynak sayısı ve drenaj bölgeleri ve ilişkili anomaliler dahil olmak üzere KAF'lerin karmaşık anatomisini doğru bir şekilde değerlendirmek için güvenli ve kolay şekilde kullanılabilir. Bu bilgiler tedavi planlaması ve hastaların takibi açısından önemlidir.

Anahtar Kelimeler: Koroner Anjiografi, Koroner Arter Fistülleri, Koroner BT Anjiografi

Abstract

In this study, we aimed to evaluate the prevalence and types of coronary artery fistulas (CAFs) with a special focus on origin, course, termination, and associated anomalies in adult patients who underwent coronary CT Angiography (CTA) at our institution. 2462 patients who underwent ECG gated coronary CTA at our institution between August 2006 and August 2020 were included. We found the records of 11 patients who had the diagnosis of CAF. The assessment included the vessels of origin, the drainage vessels, the presence of coronary artery disease, combined congenital or acquired anomalies, and the relations with the adjacent structures. The CAFs were diagnosed with coronary CTA in 11 of 2462 patients (0.44%) (7 women, 4 men; age range 35-73 years, mean=55.2 years, SD±11.38). There were 7 cases of single fistula vessel and four cases of multiple fistula vessels. Among 11 CAFs there were 17 fistula vessels involved, including 11 Left Main Coronary Artery originated vessels and six Right Coronary Artery originated vessels. There were 17 drainage sites and distribution frequency were as follows: nine cases with pulmonary artery two cases with left ventricle, two cases with right ventricle, two cases with coronary sinus, one case with the left bronchial artery, one case with left atrium. Cardiac CTA with 3D reconstruction can be used to accurately assess the complex anatomy of CAFs, including site, the number of origins and drainage sites, and associated anomalies. This information is important for planning the treatment and the follow-up of the patients.

Keywords: Coronary Angiography, Coronary Artery Fistulas, Coronary CT Angiography

Introduction

The coronary artery fistulas (CAFs) are abnormal connections between coronary arteries and any of the four chambers of the heart or any of the great vessels (superior vena cava, pulmonary artery, pulmonary veins, or coronary sinus (1,2). Several studies report the presence of CAFs in 0.3% of the patients presenting with congenital heart disease, in 0.06% of children undergoing echocardiography, and in 0.13-0.22% of adults undergoing conventional coronary angiography (CAG) (3). Ethnic groups and gender do not affect the incidence of CAFs. They are usually congenital but acquired forms have been reported in the literature. Congenital forms are due to the persistence of intramyocardial trabecular connections formed by endothelial cells and blood lacunae that are formed initially within the cardiac venous plexus and subsequently with the epicardial coronary arteries. Acquired CAFs are due to infective endocarditis, aortic dissection, previous surgery, endomyocardial biopsy, coronary angioplasty, and bypass surgery, valve replacement, cardiac transplantation, trauma, permanent pacemaker placement, closed-chest ablation of accessory pathways, neoplasms, and iatrogenic management of Kawasaki disease (4-6). Previous studies showed that CAFs arise from the right coronary artery (RCA) in approximately 50% of the patients. The most common drainage sites detected with CAG, in decreasing frequency, are the right ventricle, right atrium, pulmonary artery, coronary sinus, left atrium, left ventricle, and superior vena cava (7,8). Possible complications of CAFs are dilatation of coronary artery, mural thrombosis, rupture, atherosclerotic deposition, aneurysm formation, calcifications, intimal ulceration, intimal rupture, medial degeneration, and side-branch obstruction (9,10).

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The traditional diagnosis tool for CAFs is CAG. In addition to the problems associated with invasiveness, the complex configuration of the anomalous vessels and their anatomic relations with the adjacent structures may be obscured on 2D fluoroscopic images. The technical difficulty that accompanies cannulation of all of the vessels associated with the CAF may limit the precise evaluation of the presence of CAFs. With the advent of 64-slice multidetector CT in the chest and cardiac imaging, the number of incidentally found CAFs has been increasing (11-13). Given that in the coronary CT angiography (CTA), 3D images are constructed that enable easier and more precise access to the vasculature, the prevalence of CAF determined using coronary CTA may be closer to the true incidence of CAF (14).

In this study, we aimed to evaluate the prevalence and types of CAFs with a special focus on origin, course, termination, and associated anomalies in adult patients who underwent coronary CTA at our institution.

Material and Method

This study was performed as a retrospective chart review. Ethics committee approval: İstanbul Yeni Yüzyıl University Ethics Committee permission was obtained with the letter dated 2020/07-482. In this study, 2462 patients who underwent ECG gated coronary CTA at our institution between August 2006 and August 2020 were included. We retrospectively reviewed coronary CTA reports using an electronic database and found the records of 11 patients who had the diagnosis of CAF.

Using a 64-detector row CT scanner (Somotom Sensation 64, Siemens Medical Solutions; Forchheim; Germany), coronary CTA with the synchronous electrocardiographic tracing of the patients was performed. Approximately 80 to 100ml of non-ionic contrast medium was injected at a rate of 4-6 ml/s, followed by 40-50 ml of saline solution chaser through a dual-head injector. Applied CTA parameters were as follows: collimation width 64x0.6 mm, tube potential 120 kV, tube effective current 650 to 850 mA, tube rotation time 330 ms, table feed 3.8 mm/rotation temporal resolution 83 to 165 ms, scanning time 8 to 13 seconds in inspiratory breath-hold and scan field from the tracheal carina to the diaphragm. The axial, coronal, sagittal, and oblique multiplanar reconstruction; thin-slab maximum intensity projection; and volume-rendered images were reformatted on an on-line Workstation (Wizard; Siemens Medical Solutions, Erlangen; Germany). Multiplanar reconstructed views were reformatted using a thickness of 0.6 mm with 0.4 mm intervals and the image thickness in maximum intensity projection reconstructions was either 5- or 6-mm. Different retrospective ECG gated reconstruction temporal window settings, usually between 40% to 80% of consecutive R waves (R-R interval) in gated ECG were applied, and most reconstructions were best achieved in mid-to end-diastole (from 60% to 75% of R-R intervals).

Interpretation of CTA images of the patients was made independently by two radiologists who had 14 years and two years of experience in coronary CTA. In case of disagreement on interpretation, the radiologists would discuss the case with a third radiologist to reach a consensus.

We evaluated the characteristics of the CAFs on axial, reformatted, and volume-rendered images. The characteristics assessed included the vessels of origin, the drainage vessels, the presence of coronary artery disease, combined congenital or acquired anomalies, and the relations with the adjacent structures.

Results

The CAFs were diagnosed with coronary CTA in 11 of 2462 patients (0.44%) (7 women, 4 men; age range 35-73 years, mean=55.2 years, SD±11.38). There were 7 cases of single fistula vessel and four cases of multiple fistula vessels. Among 11 CAFs there were 17 fistula vessels involved, including 11 Left Main Coronary Artery (LMCA) originated vessels and six Right Coronary Artery (RCA) originated vessels. The specific origin distribution frequency of fistula vessels was as follows: nine Left Anterior Descending (LAD) branches, six RCA branches, two left circumflex (LCX) branches. There were 17 drainage sites and distribution frequency were as follows: nine cases with pulmonary artery, two cases with left ventricle, two cases with right ventricle, two cases with coronary sinus, one case with the left bronchial artery, one case with left atrium.

Three of 11 patients with CAFs had lung parenchymal abnormalities such as bronchiectasis. Two of 11 patients with CAFs had coronary artery aneurysm before the fistula and one of the 11 patients had apically localized hypertrophic cardiomyopathy.

Seven of 11 patients underwent CAG, five of them at our institution, two of them at other institutions. Conventional angiography could reliably demonstrate the proximal part of the CAFs; however, the drainage sites and coexistent abnormalities could not be visualized well. For three patients that had CAFs, CAG did not show the drainage site.

Three of them received percutaneous coronary intervention and stent implantation because of severe coronary artery atherosclerosis.

One of the patients demonstrated large CAFs between RCA and pulmonary artery and LAD and RV received surgical treatment. The patient was followed up using coronary CTA after the operation (Table 1 and 2).

Table 1. Data of 11 patient with CAFs

	n
CAFs prevalence	11
Gender	
Woman	7
Man	4
Amount of fistulas	
Single	7
Multiple	4
Associated abnormalities	
Lung parenchyma	3
Coronary artery aneurysms	2
Hypertrophic cardiomyopathy	1
CAG	7/11
Proximal site	7/7
Distal site	3/7
Surgery	1

Table 2. Amount, origin, drainage of 11 patient with CAFs

	n
Amount of CAFs	17
Origin	
LMCA	
LAD	9
LCX	2
RCA	6
Drainage	
Pulmonary artery	9
Left ventricle	2
Right ventricle	2
Coronary Sinus	2
Left atrium	1
Left Bronchial artery	1

Discussion

In our study, the prevalence of CAFs determined with coronary CTA is 0.44% and the most common type of fistula is coronary artery to pulmonary artery. Zhou et al. showed that the incidence of CAFs found by coronary CTA was 0.19%, Lim et al. showed that the prevalence of CAFs determined with coronary CTA was 0.9% (8,11). Baltaxe et al. reviewed images from 1000 selective CAG examinations of patients older than 18 years and reported two (0.2%) cases of CAFs (15). Von den Brand et al. reviewed the records of 126.595 patients who underwent CAG, including the pediatric population, and reported 62 (0.05%) cases of CAFs (16). The prevalence of CAFs may be underestimated with CAG because cannulating all of the arteries with fistulous origins is technically difficult complex configurations of the anomalous vessels and their anatomic relations with adjacent structures can be obscured on 2D fluoroscopic images (8).

Origin and drainage sites of coronary artery fistula: Previous studies reported that CAFs arises from the RCA in 50% of patients and from both RCA and LMCA, the most common drainage site was the pulmonary artery. In contrast to previous findings, Zhou et al. observed that most CAFs originated from either LMCA or both LMCA and RCA and the most common drainage site was the pulmonary artery (8). Our study showed that the fistulas were mostly LMCA-originated, and the most common drainage site was the pulmonary artery. Seven of 11 patients underwent CAG, which failed to demonstrate drainage sites of three cases. The drainage sites may not be well visualized with CAG because of the severely diluted contrast medium. Lim et al. and Zhou et al. found that CAFs drained most commonly into the pulmonary arteries as in our study. According to CAG studies, the most common drainage site of CAFs was the right ventricle, which could be due to the technical limitation of CAG. The complex anatomy of CAFs would have been identified accurately only with CTA (8,11).

Coexistent abnormalities: In our study, one patient had a coronary to pulmonary fistula and three patients had lung parenchymal abnormalities. It was evident, however, that abnormalities in the lung parenchyma or the pulmonary vasculature were more frequent in cases of coronary to pulmonary and coronary to bronchial artery fistulas and coronary to SVC fistulas compared with other CAFs. This finding may support the hypothesis that abnormalities in the lung parenchyma or pulmonary vasculature may induce several types of acquired forms of CAF (8,17).

Treatment: The treatment of patients with CAFs depends on the size and anatomic features of the fistula, the presence of symptoms, the patient's age, and the presence of other cardiovascular diseases. In our study, only one patient underwent a ligation operation. The patient had chest pain, aneurysmatic LAD, and RCA -the pulmonary artery and LAD-right ventricle fistulas. The coronary CTA helped the surgeons with preoperative planning, as well as follow up studies.

Amount of coronary artery fistulas: Previous studies reported that a single fistula is much more common in up to 90% of all CAFs (10). In our study four in 11 patients had multiple fistulas; these four patients had conventional angiography and three patients had conventional angiography that missed at least one of the fistulas that had been seen on coronary CTA. The missed fistulas were small in diameter. The coronary CTA, with its capability of showing the complex geometry of coronary artery anatomy in 2D and 3D images, is emerging as a valuable tool in the non-invasive diagnosis of CAF. Multiplanar reformations can accurately show the site of origin and the termination of abnormal blood vessels. Furthermore, volume-rendered images give an excellent overview of cardiac and vascular anatomy. These advantages of coronary CTA may explain the discrepancy between the results of our study and those of previous studies in which CAG has been used and may explain the increase in the number of incidentally detected CAFs as coronary CTA has become more popular (21).

Conventional coronary angiography has been a reference standard for the diagnosis of coronary

artery disease, but it is not optimal for the evaluation of CAFs. The exact complex anatomy of the fistulous tracts is difficult to obtain using CAG, and drainage sites may not be visualized with CAG. The coronary CTA may provide motion free high-resolution images using an electrocardiographically gated technique despite the limited hemodynamic information. The multiplanar reformation can demonstrate the sites of origin and termination of abnormal blood vessels, and volume-rendered images acquired from 3D CTA data sets can provide an overview of the heart, as well as its vascular anatomy, and help surgeons to understand the anatomical complexity before surgery. The coronary CTA can provide more information than CAG for the co-existing abnormalities (21-23).

Our investigation has several limitations that should be considered. First, this is a single-center retrospective study. Second, the study population was too small for the estimation of the actual prevalence of CAFs in the general population. The number of cases in our study is not sufficient for statistical evaluation. Third, only seven of our patients had undergone conventional angiography, so no comparative study with the gold standard could be performed. Only one of the 11 patients received surgical treatment, and no patients received endovascular treatment owing to CAFs. Therefore, we lacked the data to analyze the indications of surgical or interventional treatment of CAFs, and more cases are needed for further studies (Figures 1-6).

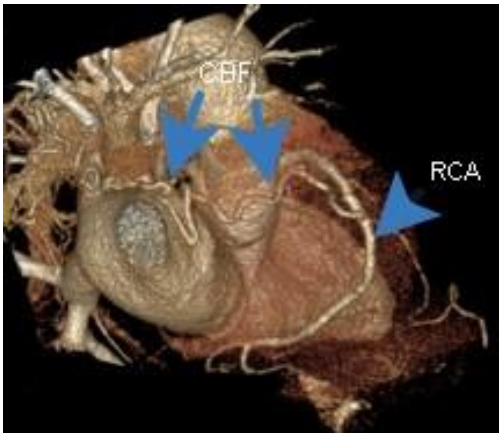

Figure 1. Three-dimensional volume rendered image from coronary CTA, showing that the coronary-to-bronchial artery fistula (CBF) originating from the right coronary artery (RCA) coursed through the right wall of the right atrium and connected to the left bronchial artery behind the right atrium.

In conclusion, the coronary CTA is a useful non-invasive imaging modality for the detection of CAFs. With the advent of multidetector CT in cardiac imaging, the number of incidentally found CAFs has been increasing. Given that in the CTA, 3D images are constructed that enable easier and more precise access to the vasculature, the prevalence of CAF determined using coronary CTA may be closer to the true incidence of CAF. Cardiac CTA with 3D reconstruction can be used to accurately assess the complex anatomy of CAFs, including site, the number of origins and drainage sites, and associated anomalies. This information is important for planning the treatment and the follow-up of the patients.

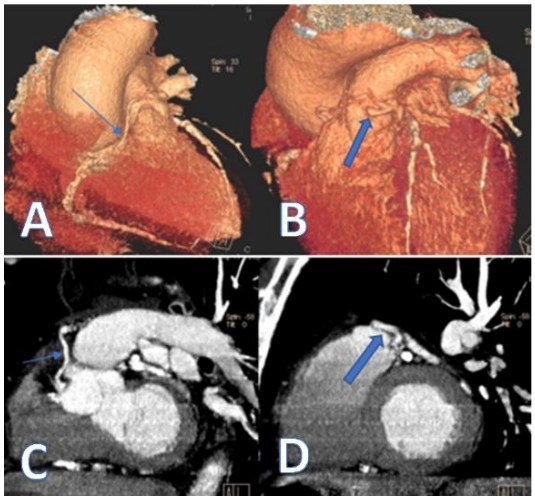

Figure 2. 59-year old man with anginal symptoms who was found to have two fistulas originating from the left coronary system and right coronary sinus and draining into pulmonary artery. **A,** Three-dimensional volume-rendered CTA shows right coronary sinus to main pulmonary artery fistula. **B,** Three-dimensional volume-rendered CTA shows the left coronary system to main pulmonary artery fistula. **C-D,** Maximum-intensity-protection CTA image reveals fistulas originating from right to left coronary system and draining into the main pulmonary artery.

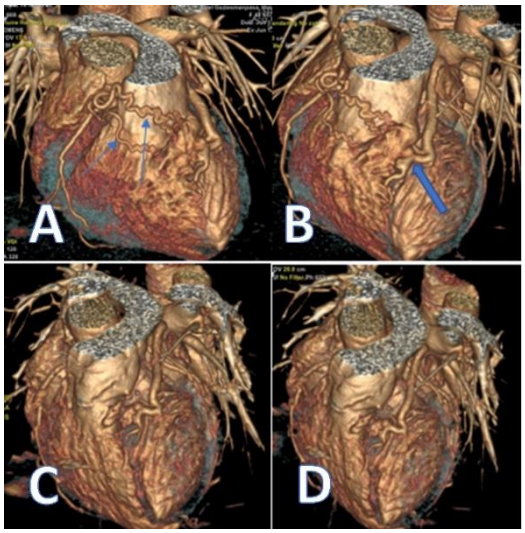

Figure 3. 45-year old women with angına symptoms. **A-B,** Volume rendering images show dilated and tortuous fistula originating from LAD and enters the right ventricle and multiple fistulas originating from RCA and enters the pulmonary artery (pro –op). **C-D,** After the operation CTA is used for the follow-up and fistulous completely resolved.

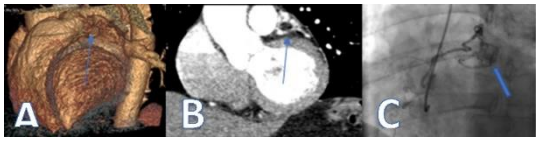

Figure 4. Volume- rendering and maximum intensity projections show tortuous fistula originating from LAD which enters pulmonary artery. **C,** Conventional angiography image shows the fistula.

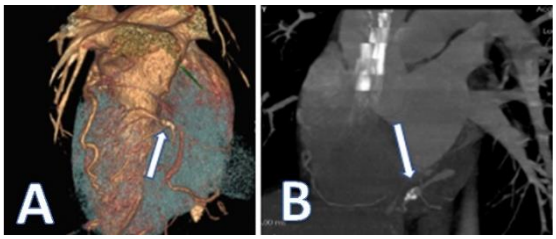

Figure 5. 49-year old man with no symptoms. **A-B,** Volume rendering and maximum intensity projection images show fistula between left circumcumflex artery and left ventricle.

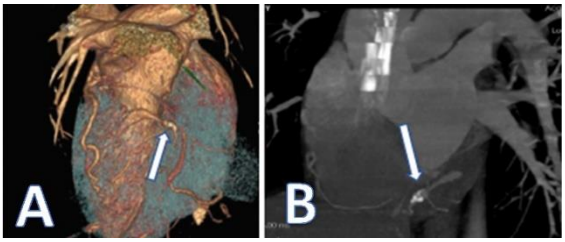

Figure 6. 35-year old man with no symptoms. **A-B,** Volume rendering and axial images show a totuous fistulous connection between LAD and pulmonary artery.

Ethics Committee Approval: İstanbul Yeni Yüzyıl University Ethics Committee permission was obtained with the letter dated 2020/07-482.

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