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FULL PAPER

Coronary artery fistula in adults: evaluation with dual-source CT coronary angiography

¹K ZHOU, MD, ¹L KONG, MD, ¹Y WANG, MD, ¹S LI, MD, ¹L SONG, MD, ¹Z WANG, MD, ²W WU, MD, ³J CHEN, MD, PhD, ¹Y WANG and ¹Z JIN, MD

¹Department of Radiology, Peking Union Medical College Hospital, Beijing, China

²Department of Cardiology, Peking Union Medical College Hospital, Beijing, China

³Healthcare, CT Division, Siemens Ltd China, Beijing, China

Address correspondence to: Dr Yining Wang

E-mail: wangyining@pumch.cn

Objective: To evaluate the clinical value of dual-source CT (DSCT) coronary angiography in the diagnosis of coronary artery fistula (CAF) in adults.

Methods: A large cohort of 17,548 patients, who underwent DSCT coronary angiography in our hospital between January 2008 and October 2013, were retrospectively reviewed for CAF. The origin, course and drainage site of CAF and coexisting abnormalities were analysed. The conventional angiography results, treatments and follow-up DSCT images were also evaluated.

Results: A total of 33 CAFs from 17,548 patients were detected. The incidence of CAF was 0.19% by DSCT. CAF originating from the left coronary artery (LCA) was found in 14 (42.4%) patients, from right coronary artery (RCA) in 4 (12.1%) cases and from both LCA and RCA in 15 (45.5%) patients. The pulmonary artery was the most common site of drainage (28/33, 84.8%). 8 of the 33 (24.2%) cases were

associated with aneurysms. Six cases were associated with coronary artery atherosclerosis. Four patients underwent conventional angiography.

Conclusion: Coronary-pulmonary artery fistula in adults was found more often than in previous studies. CAF commonly originates from LCA or both LCA and RCA in adults. DSCT is a robust tool for investigating the origin, course and drainage site of CAF and coexistent abnormalities.

Advances in knowledge: A large adult patient cohort who underwent DSCT angiography was reviewed to assess CAFs. Coronary-pulmonary artery fistula in adults was found more often than in previous studies. CAF was observed to originate from the LCA or both coronary arteries in adults. DSCT could clearly depict the fistula origin, course, drainage site and coexisting abnormalities. Conventional angiography results, treatments and follow-up DSCT images were analysed.

Coronary artery fistulas (CAFs) are anomalous connections of the coronary arteries. The phenomenon was first described in 1865 by Krause.¹ CAF is considered as a major coronary anomaly by Ogden's classification.² Most CAFs are congenital. CAFs have an estimated prevalence of 0.002% in the general population; however, they are present in 0.05–0.25% of patients who undergo coronary angiography.^{3–5} The traditional diagnosis tool for CAFs is conventional angiography. With the advent of 64-slice multidetector CT in chest and cardiac imaging, the number of incidentally found CAFs has been increasing. The advanced electrocardiogram (ECG)-gated technique of dual-source CT (DSCT) could provide high diagnostic accuracy for the assessment of coronary artery disease.

According to prior studies, CAF arises from the right coronary artery (RCA) in approximately 50% of patients.^{6–8} In particular, 70% of the CAFs in children (mean age, 2.9 years)

originated from the RCA.⁹ In this study, we focused on adult patients. A large cohort of adult patients who underwent DSCT angiography was reviewed to assess CAFs. The CAFs and coexisting abnormalities were analysed.

METHODS AND MATERIALS

Patients

A total of 17,548 patients who were suspected of having coronary artery disease underwent contrast-enhanced CT angiography (CCTA) from January 2008 to October 2013 in Peking Union Medical College Hospital, Beijing, China. The CCTA reports and images were retrospectively reviewed. Four experienced radiologists reviewed the axial, maximum intensity projection and volume rendering technique images of all 17,548 patients through the picture archiving and communication system. All the images were reviewed only once because of the large amount of work.

Table 1. Admission symptoms of 33 patients with coronary artery fistula

Symptoms	Number	Prevalence (%)
Pre-cordial discomfort	27	81.8
Mild chest tightness	20	60.6
Palpitation	13	39.4
Unstable angina	4	12.1
Severe chest tightness after exercise	1	3.0
Lower extremity oedema	1	3.0

Image acquisition

CT data were acquired with retrospective ECG-gated cardiac CT scans by DSCT (Somatom® Definition; Siemens Healthcare, Forchheim, Germany). The scan parameters were set as follows in 2008: tube voltage of 120 kV, tube current of 400 mAs, collimation of 32.0×0.6 mm, slice acquisition of 64.0×0.6 mm, by means of a z-flying focal spot; and the gantry rotation time was 330 ms. The ECG-pulsing window was set at about 25–80% of the R-R interval for all patients. A pitch of 0.20–0.28 was adopted for the lowest estimated heart rate during scanning. The scan direction was craniocaudal from the level above the coronary ostia to the level below all cardiac structures around the diaphragm. A two-phase contrast media injection protocol was adapted to the scan duration. The first phase consisted of 60 ml of contrast agent (iopamidol, 370 mgI ml^{-1}) at a rate of 5.0 ml s^{-1} , and the second phase consisted of 50 ml of saline at a rate of 5.0 ml s^{-1} . Injection was performed through an antecubital vein via an 18-gauge catheter, using a dual-head power injector (Empower, EZEM, Westbury, NY). Since 2010, the above protocol has been performed only for patients with arrhythmia. For patients without arrhythmia, an automatic electrocardiographic tube current modulation (ECG-pulsing; Siemens Healthcare) technique [pulsing from 60% to 75% R-R interval if $\text{HR} < 70$ beats per minute (bpm); 35–75% R-R interval if $70 \text{ bpm} < \text{HR} < 90 \text{ bpm}$; 35–50% R-R interval if $\text{HR} > 90 \text{ bpm}$; scan from 30% to 80% R-R interval] was performed, and the tube voltage was 100 kV.

Image reconstruction

33 patients were diagnosed with CAF. All images were post-processed using a dedicated Multi-Modality Work Place (Siemens Healthcare). The images were reconstructed with a slice thickness of 0.75 mm, an increment of 0.5 mm and using a medium soft-tissue convolution kernel (B26f). The CAFs were evaluated with the original transverse images, multiplanar reformations, curved multiplanar reformations and maximum intensity projections as well as volume-rendered images. The coronary CT angiography (CTA) images were evaluated by two experienced radiologists who were blinded to each other's results. The reconstructed images from the multiphases with the fewest artefacts were chosen. Both readers were unaware of either coronary CTA or any other examination results.

RESULTS

CAF was diagnosed in 33 of 17,548 patients. The incidence of CAF was 0.19% by DSCT. Among the 33 cases of CAF, 18 patients were male and 15 patients were female, and their

mean age was 56.9 ± 12.7 years. 28 patients had mild symptoms such as pre-cordial discomfort, mild chest tightness and palpitation (84.8%), whereas 4 patients had unstable angina (12.1%).

Figure 1. 67-year-old male with no symptoms. (a, b) Volume rendering and maximum intensity projection images show dilated and tortuous fistula between first diagonal artery (D1) and right ventricle. The drainage site of the coronary artery fistula is shown clearly (arrowhead). AO, aorta; LAD, left anterior descending artery.

Figure 2. 57-year-old female with left anterior chest pain. (a–d) Volume rendering, maximum intensity projections and multiplanar images show dilated and tortuous fistula originating from the conus branch and left anterior descending artery (LAD). The coronary pulmonary fistula passes lateral to the pulmonary artery and forms a network with aneurysmal dilatation (arrows) before it enters the main pulmonary artery (MPA). The high-density flow jet is shown clearly. An atherosclerotic plaque is shown at the proximal segment of the LAD, near the origin of the coronary artery fistula vessel. AO, aorta; RCA, right coronary artery.

One patient had serious symptoms of severe chest tightness after exercise and lower extremity oedema (3.0%), and was demonstrated to have a large CAF vessel between the left anterior descending artery (LAD) and the right ventricle (Table 1).

Origin of coronary artery fistula

The origin of CAF from the left coronary artery (LCA) was found in 14 (42.4%) cases, from the RCA in 4 (12.1%) cases and from both LCA and RCA in 15 (45.5%) cases. Among the 14 cases that originated from the LCA, 12 cases originated from the LAD; one originated from the circumflex artery; and one originated from the left coronary sinus. Among the four cases that originated from the RCA, one originated from the proximal RCA; one from the conus branch; one from the right coronary sinus; and one from the posterior left ventricular branch. Among 15 cases originating from both LCA and RCA, 10 were from the LAD and proximal RCA, whereas 5 were from the LAD and conus branch.

Amount of coronary artery fistula vessels

In 16 (48.5%) patients, the fistula was shown as 1 vessel that could be traced (Figure 1); in 6 (18.2%) patients, the fistula was

shown as 2 vessels; and in 11 (33.3%) patients, the fistula was shown as innumerable multiple vessel networks (Figures 2 and 3).

Drainage site of coronary artery fistula

The pulmonary artery was the most common site of drainage (28/33, 84.8%) (Figures 2 and 3), followed by the right ventricle (2/33, 6.1%) (Figures 1 and 4), the right atrium (1/33, 3.0%), the aorta (1/33, 3.0%) and the left ventricle (1/33, 3.0%). The drainage site was most often the left lateral side of the pulmonary trunk ($n = 24$; 72.7%).

Coexistent abnormalities

8 of the 33 (24.2%) cases were associated with aneurysms. Most of the aneurysms were adjacent to the drainage site of the pulmonary artery (6/8, 75%) (Figure 2). The mean diameter of the aneurysms was 3.2 ± 1.3 mm. Six cases were associated with coronary artery atherosclerosis (Figures 2 and 3). Five of them showed stenosis of the LAD along the course where the fistula originated (5/6, 83.3%). Other coexistent abnormalities are shown in Table 2.

Conventional angiography, treatment and follow-up

Four patients underwent conventional angiography, which could reliably demonstrate the proximal part of the CAF and allow

Figure 3. 64-year-old male after PCI. (a-d) Volume rendering and maximum intensity projections images show a left anterior descending artery (LAD)-CB-main pulmonary artery fistula with multiple vessel networks (arrows). Through a curved planar reformation image with a very sharp kernel (B46f), the situation inside these two stents could be demonstrated clearly (stars). (e, f) Conventional angiography images before PCI show fistula vessels originating from the LAD and conus branch. A serious stenosis can be observed near the origin site of the CAF (stars). CB, conus branch; CX, circumflex artery; PCI, percutaneous coronary intervention; RCA, right coronary artery.

evaluation of the size or number of fistulas. However, the drainage sites and coexistent abnormalities could not be visualized well (Figures 3 and 4). Three of them received percutaneous coronary intervention and stent implantation because of serious coronary artery atherosclerosis (Figure 3). The fourth was demonstrated to have a large CAF vessel between the LAD and the right ventricle and received surgical treatment (Figure 4). All four patients were followed up by DSCT. No patients received endovascular treatment owing to CAFs.

DISCUSSION

Our study has shown that the incidence of CAF found by coronary CTA is 0.19% (33/17,548) in the large consecutive patient population, which is similar to the incidence found by conventional angiography.^{3,4}

According to previous studies, CAF arises from the RCA in 50% of patients, from the LCA in 42% of patients and from both RCA and LCA in 5% of patients; the most common drainage site is the right ventricle.⁶⁻⁸ In contrast to previous findings, we observed that most CAFs originated from either LCA or both LCA and RCA, and that the most common drainage site was the main pulmonary artery. Most patients in our study had mild symptoms, and their mean age was 56.9 ± 12.7 years. The fistulas were found to originate from either the LCA or both coronary arteries, and they drained into the main pulmonary artery. These properties may contribute to the characteristic of the CAFs in adult patients with

mild symptoms. Our findings are consistent with previous studies conducted by Sherwood *et al.*,¹⁰ Lim *et al.*¹¹ and Kim *et al.*¹² Sherwood *et al.*¹⁰ found that the majority of symptomatic CAFs originated from the RCA, whilst asymptomatic CAFs had a greater prevalence of origin from the LCA; moreover, Lim *et al.*¹¹ found that coronary to pulmonary artery fistula (CPAF) accounts for 76.8% of cases, and Kim *et al.*¹² found CPAF to account for 89.5% of CAFs. By contrast, Hu *et al.*⁹ found that 70% of CAFs in children (mean age, 2.9 years) originated from the RCA.

There are two interesting findings in our study. First, atherosclerosis of the coronary arteries occurred mostly near the origin site of CAFs (5/6, 83.3%). Second, most aneurysms detected in our study were adjacent to the drainage site into the pulmonary artery (6/8, 75%). Canga *et al.*¹³ made the same observations and further found that large fistulas originating from the proximal segments of coronary arteries might increase the likelihood of atherosclerosis and myocardial infarction. This phenomenon might be caused by both haemodynamic disorder and the pressure difference between the origin and drainage site.

Although conventional coronary angiography has been the reference standard for the diagnosis of coronary artery disease, it is not optimal for the evaluation of CAFs. First, the exact three-dimensional course of the artery is difficult to obtain by conventional angiography. Second, the drainage sites and aneurysms may not be well visualized with conventional angiography

Figure 4. 23-year-old male with chest tightness after exercise and lower extremity oedema. (a, b) Volume rendering and maximum intensity projection images show a left coronary artery (LCA)-right ventricle (RV) fistula with a very large vessel (arrows). Coexistent abnormalities were diagnosed as both ventricular enlargements and pericardial effusion (stars). (c) Conventional angiography shows a very large fistula vessel (arrows) and enlargement of the heart. (d) After surgery, the coronary artery fistula vessel has been closed, and a coronary artery bypass (arrow) between the left internal mammary artery and left anterior descending artery (LAD) can be observed clearly. There was a stenosis near the distal anastomosis.

because of the severely diluted contrast medium^{12,14,15} (Figures 2 and 4). Third, the invasive nature of conventional angiography has a mortality of 0.1%.¹⁶

In our study, coronary CTA helped surgeons with pre-operative planning as well as follow-up studies (Figure 4). DSCT could

provide motion-free high-resolution images by using an electrocardiographically gated technique, especially for coronary CTA with complex vascular anatomy, despite the limited haemodynamic information. The multiplanar reformation can clearly demonstrate the sites of origin and termination of abnormal blood vessels,^{16,17} and the volume-rendered images acquired

Table 2. Incidence of coexistent abnormalities found in 33 coronary artery fistula patients diagnosed by dual-source CT coronary angiography

Coexistent abnormalities	Number of abnormalities	Prevalence (%)
Aneurysms	8	24.2
Coronary artery atherosclerosis	6	18.2
Pericardial effusion	3	9.1
Left ventricular enlargement	2	6.1
Both ventricular enlargements	1	3.0
Pleural effusion	1	3.0
Pericardial cyst	1	3.0

from three-dimensional CT 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.^{8,14,18} Therefore, DSCT could provide more information than conventional coronary angiography for the diagnosis of co-existent abnormalities.

LIMITATIONS

Our investigation has a number of limitations that must be considered. First, this work is a single-centre retrospective study. Second, images of the 17,548 patients were reviewed only once because of the large amount of work, which may lead to biases. Third, the scan protocol was for normal coronary CTA, and CAFs communicating with the large vessels might have been missed owing to limitation of the scan range. Fourth, in our clinical practice, patients with arrhythmia were required to be examined by retrospective ECG-gated scan protocols, and 25–80% R-R intervals were used for acquisition windows. Thus, the radiation dose is relatively higher. Lastly, only four patients had undergone conventional angiography, so no comparative study with the gold standard could be performed. Only one patient received surgical treatment, and no patients received

endovascular treatment owing to CAFs. Therefore, we lacked data to analyse the indications of surgical or interventional treatment of CAFs, and more cases are needed for future studies.

CONCLUSION

Coronary–pulmonary artery fistulas in adults were found more often than in previous studies. CAF was observed to originate predominantly from the LCA or both coronary arteries in adults. In the meanwhile, DSCT could effectively depict its origin, course, drainage site and coexistent abnormalities.

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