Mysterious window: A right coronary artery-left ventricular fistula diagnosed by multiple imaging methods

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Running title: YI YU *et al*. right-sided coronary artery fistula terminating into left ventricle: a case report

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Abstract

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Prevalence of primary coronary cameral fistula (CCF) is extremely rare, especially for CCF with the drainage site into the left ventricle. We describe a 45-year-old patient with chest tightness in whom a giant aneurysm associated with proximal right coronary artery (RCA), and the distal end of RCA terminated into the left ventricle through a fistula was discovered by echocardiography. Dual-source computer tomography revealed the CCF-related giant RCA aneurysm. The drainage site and coexistent abnormality could not be visualized well by coronary artery angiography because of the severely diluted contrast medium into the aneurysm. Finally, the patient received surgical treatment and is asymptomatic now.

Keywords: Coronary cameral fistula, Cardiac imaging, Case report

Introduction

Coronary cameral fistula (CCF) is an unusual vascular anomaly that communicates between one of the coronary arteries and a cardiac chamber [1]. The most common sites for a coronary fistulous connection are the right ventricle, right atrium, and left ventricle. A giant coronary artery aneurysm associated with a CCF is also seldom, found in only 0.02% of the cardiac surgical population [2]. The right coronary artery-left ventricular fistula is extremely rare [2]. Surgical repair is necessary to occlude fistulas in most cases, even when the patient is asymptomatic at the time of confirmed diagnosis. Thus, an appropriate imaging algorithm to assess CCF is essential to assist with the decision making of surgery.

Case presentation

A 45-year-old male was admitted on December 6, 2015 in our hospital due to chest tightness. Chest x-ray examination revealed moderate cardiomegaly. Electrocardiography showed high left ventricular voltage and T-wave abnormalities in precordial leads. Grade 3/6 systolic and diastolic murmurs were detected in the apical region during physical examination. A transthoracic echocardiography (TTE) revealed a giant aneurysm in the proximal segment of RCA (inner diameter: 80 mm) from the parasternal long-axis view (Fig. 1A), dilated opening of the right coronary artery (RCA, 25mm), right atrium and right ventricle were depressed by the giant aneurysm (Fig. 1B), and the distal end of RCA entered into the left ventricle through a 8 mm fistula. The fistula was located behind the posterior lobe of the mitral valve,

there was no mitral valve stenosis. Apical five-chamber and aortic root short axis view of the heart displayed the termination of the coronary artery fistula from the right coronary artery to the left ventricle. This fistulous communication terminated in the posterior surface of the ventricle after passing along the atrioventricular groove. Color Doppler imaging confirmed the RCA fistula draining into the left ventricle. Pulse wave Doppler showed predominantly diastolic blood flow at the fistula orifice (Fig. 1C). The winding and tortuous course of RCA could be detected from the subcostal views (Fig. 1D). A dual-source computed tomography（DSCT）scan was advised to further characterization of the coronary artery and assessment of fistula. It is a superior tool for determining the associated coronary artery lesions. The morphological characteristic of CCF was evaluated that a giant RCA aneurysm (Fig. 2A) associated with CCF and the distal end of which drained to the left ventricle (Fig. 2B). The patient underwent conventional CAG, and dilated RCA ostium and a giant right coronary artery aneurysm were viewed by CAG. However, the drainage site and coexistent abnormality could not be visualized with satisfaction (Fig. 3A) in comparison with the left coronary artery visualization (Fig. 3B). Subsequently, the patient received surgical excision of the giant aneurysm and repair of the fistula. During the operation, the fistula was found to originate from the RCA and drained into the left ventricle. The surgery removed the large coronary artery aneurysm and followed by angioplasty. The fistula was closed with a Teflon patch. After cardiac surgery, the symptoms of the patient disappeared and myocardial perfusion

scintigraphy did not show evidence of ischemia. Follow-up echocardiograghy after 3 months after operation showed small residual shunt of the fistula and the volume of the left ventricle was reduced from 298 milliliter to 201 milliliter.

Discussion and conclusion

The patient had a single fistula, to be more precise, it was a right-sided CCF. The RCA was involved, associated with a giant coronary artery aneurysm and a drainage entered in the left ventricle. This finding was finally confirmed by surgery. CCF refers anomalous connections of the coronary artery. Murmur is often one of the first clinical symptoms of fistulas. Our patient initially presented with chest tightness and heart murmur. According to prior studies, CCF arises from the RCA and the drainage site into the left ventricle is extremely rare [2]. Zhou K et al. [3] found that most aneurysms detected in their study (75%) were adjacent to the drainage site into the pulmonary artery. Canga et al. [4] reported a same observation 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. There has been increased use of noninvasive imaging techniques for detecting CCF [5-6]. Echocardiography is considered as the first line technique for imaging CCF, while CT, cardiac magnetic resonance （CMR） and CAG are more useful tools for detailed characterization and differential diagnosis [7]. Although TTE revealed the dilated RCA with a giant aneurysm in the proximal

segment, and the distal end of this artery entered into the left ventricle through a fistula. Echocardiography is noninvasive and convenient, the efficacy in assessing CCF can also be challenging in adult patients owing to potential poor acoustic window and awareness related knowledge on CCF by the operator. Specifically, small origins of CCF are generally difficult to detect by TTE, because of low spatial resolution and poor penetration through the chest wall. This is most notable when assessing CCF with tortuous feeding coronary arteries, distal portals, or multiple fistulas. In addition, echocardiography cannot accurately provide information regarding associated coronary artery lesions. DSCT is also an useful method which generally provides more diagnostic information regarding CCF than CMR. It enables accurate assessment of morphological features, quantitative features, and associated coronary artery lesions in patients with CCF [8]. It could provide motion-free high-resolution images by using an electro-cardiographically 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 [8], and the volume-rendered images acquired from three- dimensional CT data sets can provide an overview of the heart as well as its vascular anatomy and help surgeons to understand anatomical complexity before surgery [9]. With contour and heterogeneous contrast enhancement, the origin, course and drainage site of CCF coexistent abnormalities were clearly observed in this case. But it is difficult to supply following information for the surgery including the precise diameters of the feeding coronary artery and drainage site. Shi K et al. [2]

enrolled 34 patients with coronary artery fistulas analyzed by DSCT morphologically and quantitatively and compared the analyses with surgical results. They concluded that DSCT served as an alternative noninvasive imaging method that enables accurate assessment of morphological features, quantitative features, and associated coronary artery lesions in patients with coronary artery fistulas. DSCT is capable of accurately evaluating associated lesions, including arteriosclerotic plaques, coronary artery aneurysms, and myocardial bridges. Thus, DSCT is helpful for determining surgical strategies. At present, CMR suffers limitations like the long acquisition time, low spatial resolution, and many other contradictions, which we think far outweigh the lack of radiation exposure when evaluating CCF and associated coronary artery lesions.

As we all known, CAG is the golden standard for the diagnosis of coronary artery disease, but it is not the optimal imaging technique for the evaluation of CCF due to following reasons. First, the exact three-dimensional course of the artery is difficult to obtain by conventional angiography. Second, the drainage site and aneurysm could not be visualized satisfactorily with CAG because of the severely diluted contrast medium in this case [10] and the limited angle of projection, particularly in complex cases or when there are multiple fistulas. Third, there is a great rupture risk of aneurysmal-CCF if a mechanical contrast media injector was used, moreover, filling the large coronary aneurysm with the contrast media using manual injection was impossible.

In conclusion, CCF associated with a giant right coronary artery aneurysm and a drainage entering in the left ventricle in this patient is an extremely rare finding. The successful diagnosis of our case relied on the comprehensive utilization of multiple imaging techniques.

Authors'contributions

(I) Conception and design: Y Yu; (II) Administrative support: YG Li ; (III) Provision of study materials or patients: QS Wang, W Li ; (IV) Collection and assembly of data: J Sun, Q Wang; (V) Data analysis and interpretation: C Li; (VI) Manuscript writing: All authors; (VII) Final approval of manuscript: All authors.

Ethics approval and consent to participate

The study was approved by the Ethics Committee of Xinhua Hospital.

Consent for publication

Written informed consent was obtained from the patient.

Competing interests

The authors declare that they have no competing interests.

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Figure Legends:

Fig. 1 Echocardiography revealed the right coronary artery-left ventricular fistula. (A) Transthoracic echocardiogram (TTE) revealed a giant aneurysm in the proximal segment of right coronary artery from the aortic root short axis view. (B) Color Doppler echocardiography detected the opening of the right coronary artery (RCA) and a giant aneurysm in the proximal segment of RCA, pressing the right atrium and right ventricle. (C) TTE revealed a winding and tortuous course of RCA from the subcostal views. (D) The fistula was detected behind the posterior lobe of the mitral valve by color doppler echocardiography. Pulse wave doppler showed the diastolic blood flow at the fistula orifice.

TTE: transthoracic echocardiogram; RCA: right coronary artery;

Fig. 2 DSCT assessed the morphological characteristic of fistula. (A) Maximum intensity projection demonstrates the dilated RCA with scattered calcified plaques. A representative right-sided CCF terminating into an LV. (B) The volume rendering shows dilated and tortuous fistula originating from the right coronary artery. The RCA with aneurysmal dilatation (arrows) is shown at the proximal segment by DSCT.

DSCT: dual-source computed tomography; RCA: right coronary artery; CCF: coronary cameral fistula; LV: left ventricular.

Fig. 3 Coronary artery angiography was performed for the evaluation of CCF before surgical planning.

(A) The drainage site and aneurysm were not well visualized with CAG, the severely diluted contrast medium could be seen in the giant aneurysm. (B) The left coronary artery was clearly showed.

CAG : coronary artery angiography；CCF: coronary cameral fistula