The Heart Surgery Forum 2022-5077
25 (6), 2022 [Epub December 2022]
doi: 10.1532/hsf.5077

Online address: http://journal.hsforum.com

Right Coronary-Left Ventricular Fistula and Its Course: A Case Report

Sihan Miao,[1,2] Haige Zhao,[1] Shengjun Wu,[1] Liang Ma[1]

[1]Department of Cardiovascular Surgery, The First Affiliated Hospital, College of Medicine, Zhejiang University, Hangzhou, China; [2]College of Medicine, Zhejiang University

ABSTRACT

Background: Coronary fistula is a relatively rare cardiac disease. The incidence of coronary fistula flowing into the right heart structure is much higher than in inflow into the left heart structures. Opportunities for surgical intervention and treatment strategies for these patients still need to build more consensus.

Case presentation: Here, we report a case of right coronary artery-left ventricular fistula combined with a giant coronary artery aneurysm during the treatment course.

Conclusion: Because of its rarity as well as its specificity of coronary artery fistula, each case requires an individualized evaluation. More consideration also can be given to the choice of surgical approach.

INTRODUCTION

Coronary fistulas (CFs) are rare diseases. Compared with other coronary arteries, fistula occurring in the right coronary artery accounts for a larger proportion [Fragakis 2015]. During the progression of the disease, most patients progress from asymptomatic to symptomatic with the increase of age [Agarwal 2017]. To date, the treatment modalities of coronary fistula includes percutaneous catheter closure, surgical procedures, and drug control [Chia 1981]. Surgical intervention is a definitive and effective treatment modality for patients presenting with clinical symptoms.

CASE PRESENTATION

A 34-year-old female with right coronary artery-left ventricular fistula was admitted to the hospital, due to tachycardia and cardiac murmur. On physical examination, the patient was found to have regular beats on the right side and below the larynx, with the same rhythm as the heart. A continuous murmur of grade III could be heard in the third intercostal space at the left sternal border, and electrocardiogram suggested sinus tachycardia. Echocardiography suggested a right coronary artery-left ventricular fistula. (Figure 1)

For clarity, coronary computed tomographic angiography as well as coronary angiography was refined. It was found that the right coronary artery was opened at the right coronary sinus, extended outward, and bypassed to the left atrioventricular groove into the left ventricular lateral wall intramuscular layer, which communicated with the left ventricle. The lumen significantly was dilated, about 3.0 cm at the widest. (Figure 2)

A huge right coronary was found on coronary angiography, and the contrast medium could not sufficiently show the whole lumen, which was suspected ventricular fistula. (Figure 3)

A median incision of the anterior chest was made, the sternum and pericardium were opened, and the extracorporeal intubation was routinely completed. Intraoperatively, the right coronary artery could be seen to be significantly thickened. The right coronary artery was opened on the surface of the left ventricle, exploration revealed a left ventricular fistula of approximately 1.5 cm, and autologous pericardial suture was given to close the incision. (Figure 4)

After extending the incision and exploring the remaining right coronary artery to confirm the absence of remaining fistula ostia, the right coronary artery incision was continuously sutured. Postoperative anticoagulation was performed with

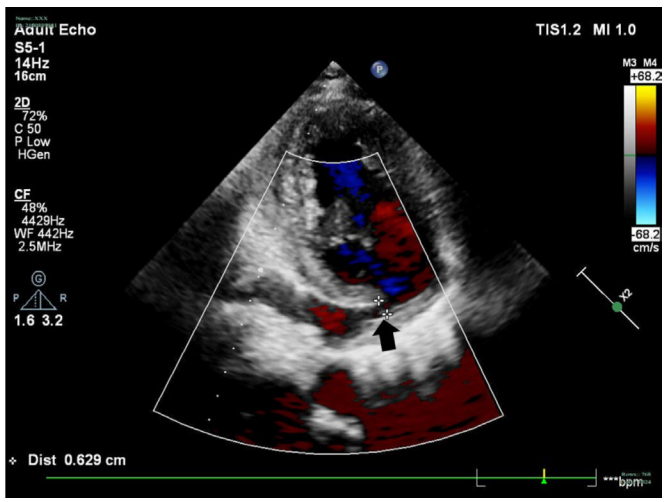

Figure 1. Black arrow indicates the location of the fistula from the right coronary artery to the left ventricle, which measured approximately 0.6 cm.

Received August 16, 2022; accepted September 26, 2022.

Correspondence: Liang Ma, #79 Qingchun Road, Hangzhou, Zhejiang Province, China, 31003, Telephone +86 0571 87236841 (e-mail: ml1402@zju.edu.cn).

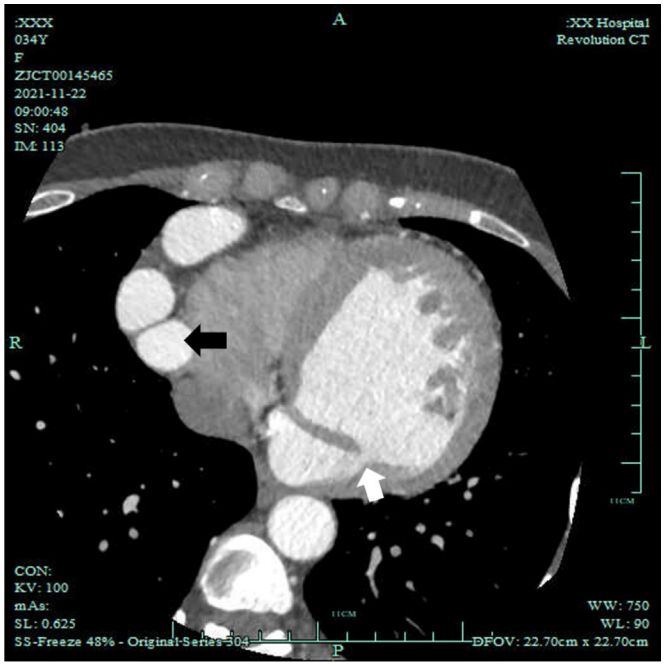

Figure 2. Black arrow shows markedly widened right coronary artery. White arrows indicate the site of right coronary left ventricular fistula.

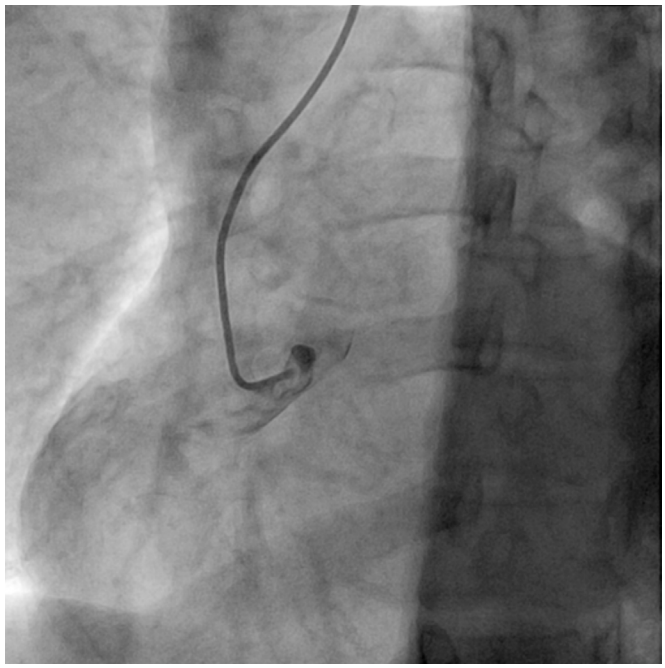

Figure 3. A single frame coronary angiogram can reveal a markedly enlarged right coronary artery.

Figure 4. The black arrow is the right coronary left ventricular fistula ostium, which is approximately 1.5 cm in size, and the white arrow area is the significantly dilated right coronary artery.

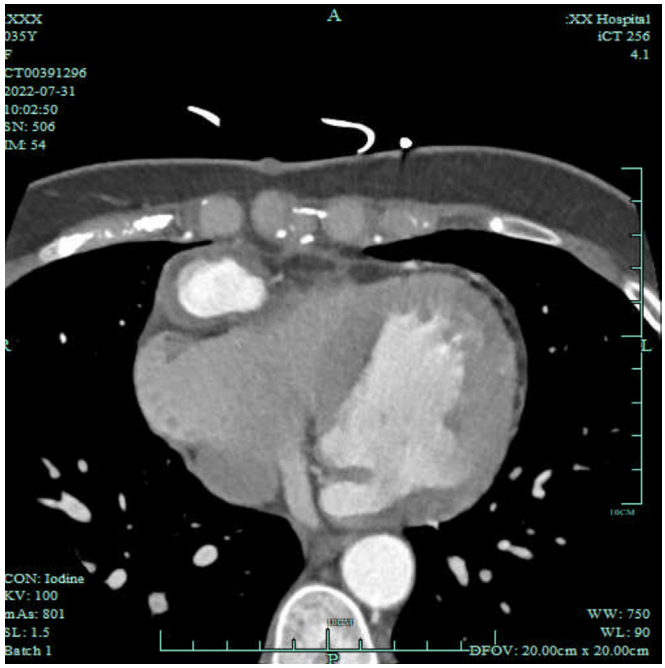

Figure 5. Images at a similar level to the preoperative examination demonstrated right coronary artery thrombosis with no significant blood flow at the surgical incision.

warfarin. Post-procedural coronary computed tomographic angiography suggested no significant transseptal shunt. Six months after operation, a right intracoronary thrombosis was formed. Nevertheless, the patient had no obvious discomfort symptoms. (Figure 5)

DISCUSSION AND CONCLUSION

CFs are rare cardiac disorder, most of which are congenital [Said 2013]. In related cases, the ostium of fistula entering the right heart structure is much more than the left heart structure [Fragakis 2015]. Among adults, a heart murmur was observed in about 37% of patients, and more patients came to visit a doctor because of remaining discomfort symptoms, such as chest pain and chest tightness [Agarwal 2017].

In our case, the coronary fistula originated from the right coronary artery flowing into the left ventricle. The patient did not have significant complaints of discomfort, but considering the possibility of an enlarged fistula with age, we surgically treated it.

Right to left shunting, caused by a larger fistula area, can lead to distal myocardial ischemia at the fistula site [Sommer 2008; Mohanty 2005], which explains why chest pain and dyspnea are the most common symptoms in patients with symptomatic CFs.

Because of the rarity of CFS and diversity of their fistula orifices, there is not a standard consensus on the modality of their treatment. Surgical treatment, percutaneous catheterization, or conservative management all have been reported, but surgery remains a relatively suitable treatment modality for symptomatic or larger coronary fistulas [Hong 2004; Hirose 1998]. Surgical treatment included fistula ligation, transcoronary closure, intracardiac repair, and coronary artery bypass grafting. Even though fistula ligation has been shown to be a safe and effective procedure [Mavroudis 1997], in this patient we did not choose this treatment option. Because this patient had a fistula opening into the left ventricle, after performing fistula ligation, there was a risk of aneurysm formation due to excessive left ventricular pressure. Therefore, we chose more stable intracardiac repair, opening the coronary arteries, and repair of the fistula from the coronary incision.

CFS often are accompanied by corresponding coronary artery dilation and even aneurysmal changes. A dilated coronary artery carries a higher risk of thrombosis when the fistula is manipulated with closure through a surgical or percutaneous catheter, thus there is a certain necessity for postoperative anticoagulation therapy in such patients. However, there is no standard protocol for anticoagulation in these patients, and anticoagulation with conventional warfarin or dual agent use of aspirin or clopidogrel has been reported [Nunez-Gil 2018; Nunez-Gil 2019].

It is mentioned in the available studies that the dilated coronary arteries had some degree of recovery to a normal diameter at long follow up after closure of the fistula ostia [Kulchetscki 2019]. Unfortunately, the number of complete patients with long follow up is not large. After more patients returned to normal life, only routine postoperative reexamination was performed, and no long-term follow-up coronary related examinations were performed. Meanwhile, the selection of the corresponding anticoagulation regimen also has not yet been standard.

The surgical procedure had a higher incidence of myocardial infarction and tricuspid regurgitation in the perioperative period, and the tricuspid regurgitation resolved during long-term follow up [Said 2013]. And, incomplete fistula repair significantly increased the incidence of infective endocarditis [Agarwal 2017]. Whether ischemic symptoms occur is an important factor of complications after surgical intervention in CFS. The monitoring of intraoperative electrocardiography as well as transesophageal echocardiography can, to some extent, monitor the occurrence or absence of ischemia [Upadhyay 2020].

As we discussed above, the choice of treatment modality for patients with CFS needs to be more individualized. Even surgically treated, there are options for different modalities.

ACKNOWLEDGEMENT

Funding: This report is supported by Youth Exploration Program, Zhejiang Province (LQ20H160026); General Program, National Natural Science Foundation of China (82070516).

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