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Congenital Giant Right Coronary Artery to Pulmonary Artery Fistula and Congenital Left Main Coronary Artery Atresia

A Case Report

Ya-hui Ding, MD and Hai Zou, MD

Abstract: Both of congenital giant RCA, RCA to PA fistula, and CLMCA-A are one of exceedingly rare vascular abnormalities. We present the case of a 34-year-old man with multiple congenital coronary anomalies above, and a successful surgical intervention was proceeded for our patient.

This is a case of a 34-year-old man presented with apical systolic murmur and exertional chest pain. Furthermore, a dilated left heart and abnormal pulmonary blood flow was observed in this patient. Coronary CTA and selective CAG showed a 15-mm diameter RCA with a fistula to pulmonary trunk and CLMCA-A. During cardiac surgery, the coronary abnormalities were confirmed and the outlet of the fistula was closed. After surgery, less chest pain, respiratory infection, and left heart failure were observed in this patient.

This case illustrates that the surgery of closing the fistula between the giant RCA and pulmonary trunk could improve the symptoms and signs for this patient. It could be predicted that simplified operation could be a promising therapy for patient with giant RCA to PA fistula and CLMCA-A. Moreover, CABG should be taken full account of when myocardial ischemia was found in the blood-supply area of LAD.

(*Medicine* 95(8):e2900)

Abbreviations: 3D-CTA = 3-dimensional reconstruction of computed tomographic angiography, AA = ascending aorta, CABG = coronary artery bypass grafting, CAF = coronary artery fistula, CAG = coronary angiography, CLMCA-A = congenital left main coronary atresia, CTA = computed tomographic angiography, LA = left atrium, LAD = left anterior descending coronary artery, LCX = left circumflex artery, LMCA = left main coronary artery,

Editor: Leonardo Gilardi.

Received: October 29, 2015; revised: January 23, 2016; accepted: January 31, 2016.

From the Department of Cardiology, Zhejiang Provincial People's Hospital, Hangzhou, Zhejiang Province, China.

Correspondence: Hai Zou, Department of Cardiology, Zhejiang Provincial People's Hospital, Hangzhou, Zhejiang Province, China (e-mail: haire1993@163.com).

Authors' contributions: Y-hD planned the article and contributed to data collection, discussing content, writing, and reviewing the article; HZ conceived the study and participated in its design, study supervision, and helping to writing the article.

There was no financial support or funding for this case report.

Informed patient consent was obtained for publication of this case report. The authors have no conflicts of interest to disclose.

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ISSN: 0025-7974

DOI: 10.1097/MD.0000000000002900

LV = left ventricular, PA = pulmonary artery, RCA = right coronary artery, SAQ = Seattle Angina Questionnaire.

INTRODUCTION

Both of congenital giant RCA, RCA to PA fistula, and CLMCA-A are one of exceedingly rare vascular abnormalities. We present the case of a 34-year-old man with multiple congenital coronary anomalies above, and a successful surgical intervention was proceeded for our patient.

CASE REPORT

A 34-year-old man was referred to the hospital for symptoms of chest pain, which was mild and usually occurred after exercise, and short breath for 3 years, and an apical sign of systolic murmur for 7 years. Furthermore, the chest pain was worse for a history of 3 months. There was no relevant cardiovascular history such as hypertension, valve disease, and no family history of heart disease.

Echocardiography showed a globular dilated left heart (left ventricular end-diastolic diameter 63 mm and left atrial diameter 42 mm), the dilated RCA (~15 mm), an abnormal flow in PA, and ejection fraction 70%. Coronary CTA revealed an obvious dilated and tortuous RCA, the mild dilated LAD, and a connection between RCA and PA (Figure 1). The origin of LMCA and LCX was not found (Figures 2 and 3). The diameter of the proximal RCA was ~14 mm, and there was no atheromatous plaque in coronary arteries.

He was admitted and performed selective CAG. His hemodynamics was stable (temperature 36.2°C, heart rate 96 beats per minute, supine left arm blood pressure 131/73 mm Hg). His weight was 81 kg, height was 175 cm, and body mass index was 26.45 kg/m². At his apex, a III/VI systolic murmur heard was been auscultated. A rest 12-lead electrocardiogram showed sinus rhythm at the rate of 88, no ST-segment depression, and no T-wave change. The laboratory test results were as follows: hemoglobin 150 g/L, total cholesterol 7.07 mmol/L, low-density lipoprotein cholesterol 4.62 mmol/L, triglyceride 2.39 mmol/L, brain natriuretic peptide 41.9 pg/mL, cardiac troponin-I 0.00 µg/L. Moreover, the score of SAQ was 41.76.

CAG showed an obvious dilated tortuous RCA, and coronary collaterals from distal segment of RCA to LAD (Figure 4). But the opening of LMCA was not found yet, even after directly injecting contrast medium into the left sinus of Valsalva. Selective fistula angiography showed a large fistula with some branch draining into PA (Figure 5). Right heart catheterization showed that the blood pressure of PA was 27/16 mm Hg. The opening of LMCA was not found by pulmonary angiography.

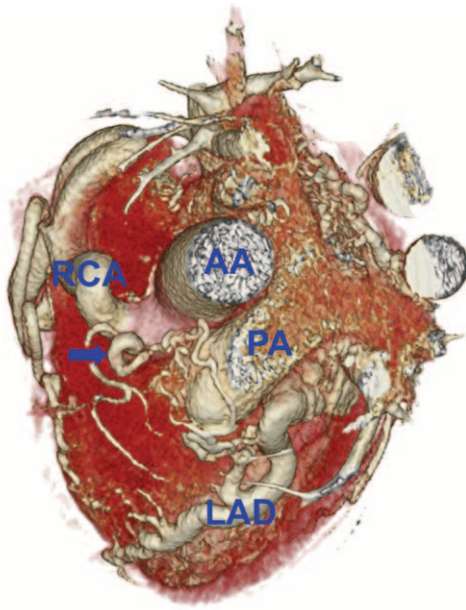


FIGURE 1. 3D-CTA demonstrated the obvious dilated and tortuous RCA, the mild dilated LAD, and a connection (indicated by the blue arrow) between RCA and PA. 3D-CTA = 3-dimensional reconstruction of computed tomographic angiography, LAD=left anterior descending coronary artery, PA=pulmonary artery, RCA=right coronary artery.

Closure of CAF was performed through a median sternotomy. During cardiac surgery, dilated left atrium and ventricular, a giant tortuous RCA with 15 mm in diameter, and a CAF, which divided into 3 to 4 branches and get together again, were found (Figure 6). After heparinization with 3.0 mg of heparin kg/body weight, the vena cava and ascending aorta were cannulated. After institution of cardiopulmonary bypass, a longitudinal incision was made in pulmonary trunk, and a fistula's opening with 10 mm in diameter was found above the pulmonary valve. Suture closure with 5–0 Prolene suture of

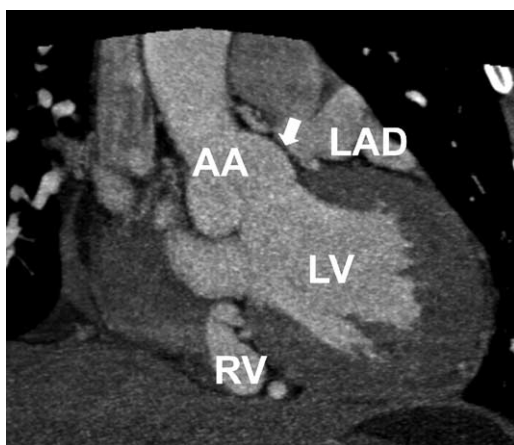


FIGURE 2. The coronal section of CTA showed that the origin of LMCA is atresic (indicated by the white arrow). CTA = computed tomographic angiography, LMCA=left main coronary artery.

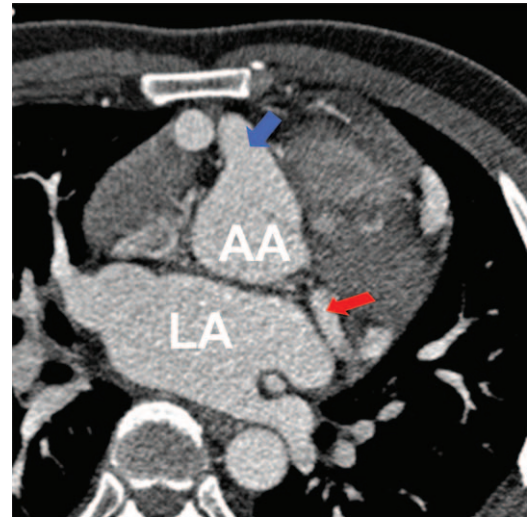


FIGURE 3. The cross-section of CTA showed that the dilated opening of RCA (indicated by the blue arrow). The origin of LCX (indicated by the red arrow) is not clear. CTA=computed tomographic angiography, LCX=left circumflex artery, RCA=right coronary artery.

the distal opening of CAF was performed. There was no bleeding in pulmonary artery after restoring coronary perfusion. We also explored the distribution of LAD, and there was no obvious LMCA on aorta or pulmonary trunk.

After surgery, he was treated with metoprolol succinate tablet 47.5 mg per day for heart failure. No statin or other lipid-lowering drugs were used for treating dyslipidemia.

During follow-up, a lower incidence of chest pain episodes and respiratory infections was reported. Echocardiography, which was repeated at 5 weeks after the operation, showed left ventricular end-diastolic diameter 50 mm, left atrial diameter 36 mm, and ejection fraction 62%. There was no shunt found. Meanwhile, the repeated SAQ score of the evaluation of chest pain was higher than before (Table 1).

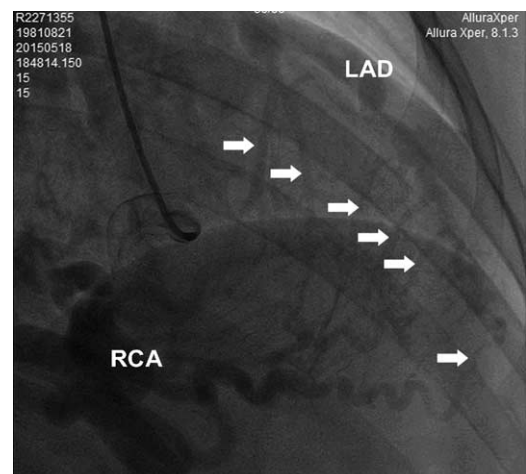


FIGURE 4. The selective right coronary angiography illustrated the obvious dilated and tortuous RCA with the coronary artery collaterals (indicated by white arrows) to LAD. LAD = left anterior descending coronary artery, RCA=right coronary artery.



FIGURE 5. The selective CAF angiography showed the fistula drain from proximal RCA to PA (indicated by the white arrow). The fistula divided into some branches and meet again. CAF=coronary artery fistula, PA=pulmonary artery, RCA=right coronary artery.

DISCUSSION

CAF, a rare coronary artery abnormality, usually occurred in isolation.¹ With the popularization of CAG and coronary CTA, more CAFs were discovered. The fistulas, which were usually dilated and tortuous, originated from RCA in ~52% cases, LAD in ~30% cases, and LCX in ~18% cases. Over 90% of fistulas drained into the right heart or PA that created a left-to-right shunt and increased the volume load of the right heart and PA. The size of the shunt depended on the size of fistulas, the pressure gradient between the coronary artery and the chamber into which fistulas drained. Due to the “steal” phenomenon of coronary arteries, myocardial ischemic could be found in CAF. CAF was usually asymptomatic in the first 2 decades; however, the symptoms, such as angina, thrombosis

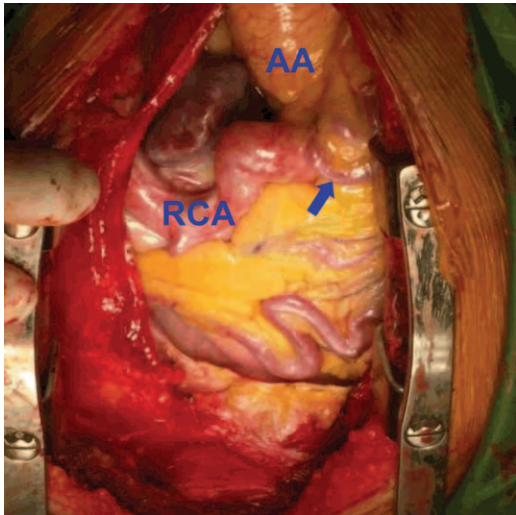


FIGURE 6. The obvious dilated and tortuous RCA was founded during the cardiac surgery. The blue arrow indicated the fistula draining from proximal RCA. RCA=right coronary artery.

TABLE 1. The SAQ Variables

	Before Surgery	At 5 Weeks After Surgery
Physical limitation	40	57.78
Anginal stability	25	100
Anginal frequency	60	80
Treatment satisfaction	58.82	82.35
Quality of life	25	58.35
Mean score	41.76	75.70

SAQ = Seattle Angina Questionnaire.

and embolism, cardiac failure, atrial fibrillation, rupture, endocarditis/endarteritis, and arrhythmia,² and complications of CAF increase after the first 2 decades.

The treatment of fistulas was recommend when these symptoms or signs appeared, including a large or increasing left to right shunt, myocardial ischemic, left ventricular dysfunction, left ventricular volume overload, congestive cardiac failure and for prevention of endocarditis/endarteritis. Catheter closure and cardiac surgery were common techniques to treat CAF.^{1–3} The treatment was based on clinical presentation, age of diagnosis, and the hemodynamic significance. Catheter closure was considered as an effective and safe alternative surgery. The termination point of fistula should be closed before occluding the artery fistula to avoid affecting the branches to normal myocardium.

CLMCA-A was an extremely rare coronary artery abnormality. The blood supply to LAD and LCX relied the coronary collaterals between RCA and LAD. And it could cause myocardial ischemia, left ventricular dysfunction, mitral incompetence, and heart failure.^{4,5} Our patient had a congenital giant RCA and a large fistula. A significant left-to-right shunt reduced the RCA blood flow and also affected the blood supply of LAD. As some branches in the fistula, the treatment of catheterization closure was not fit for him. However, to close the opening of CAF in PA by surgery was an appropriate treatment for him. Less chest pain and respiratory infection occurred after the operation. The blood supply of left coronary artery might only be draining from the coronary collaterals. The coronary CTA, selective CAG, pulmonary artery angiography, and cardiac surgery all proved that LMCA was not open onto the ascending aorta or PA.

CONCLUSIONS

In summary, this case reports a successful selective CAG and surgical intervention on a patient presented with multiple congenital coronary anomalies. However, long-time follow-up should be considered to look for possible myocardial ischemic due to coronary collaterals insufficiency which will be treated by CABG. Although the surgery in this case could improve symptoms and signs of this patient, it is necessary to extend further research to help find more effectively treatment for multiple congenital coronary anomalies.

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