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Fistula between the right coronary artery and coronary sinus: a case report and literature review

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Introduction

Coronary artery fistula is a rare organic heart disease. Its incidence rate is approximately 0.002%. Right coronary artery (RCA)-coronary sinus fistula is rare and most likely accounts for

7% of all coronary artery fistulas (1, 2). Most patients may present with chest pain, palpitations, syncope, and a continuous murmur at the precordium, accompanied by local thrill or a systolic and diastolic dual-phase murmur (3, 4). We report a recent, rare case involving a fistula between RCA and coronary sinus.

Case Report

A 14-year-old boy with a chief complaint of recurrent syncope on moderate exercise was referred to our department. Physical examination revealed a grade 4/6 continuous murmur in the second-to-fourth intercostal space at the left sternal border. Electrocardiogram showed sinus rhythm. Chest X-ray revealed heart shade enlargement. Transthoracic echocardiography (TTE) revealed left ventricular enlargement. RCA, which was markedly dilated and had an opening diameter of approximately 10 mm (Fig. 1a, Video 1), tortuously coursed to the atrioventricular sulcus of the heart bottom along the right ventricular anterior wall surface and entered the coronary sinus medially and posteriorly to the coronary vein (Fig. 1b, c; Video 2). The fistula diameter was approximately 6 mm. Color Doppler imaging detected a dual-phase shunt in the fistula (Vmax, 4.2 m/s; peak gradient, 69 mmHg) (Fig. 1d). Multidetector coronary angiography (MDCT) revealed that RCA, which was significantly dilated, was interrupted by the coronary sinus (Fig. 1e, f). Intraoperative findings revealed that RCA, which was dilated at the beginning portion, coursed to the right ventricular anterior wall surface, accompanied by significant broadening and a fistula into the coronary sinus (at the proximal

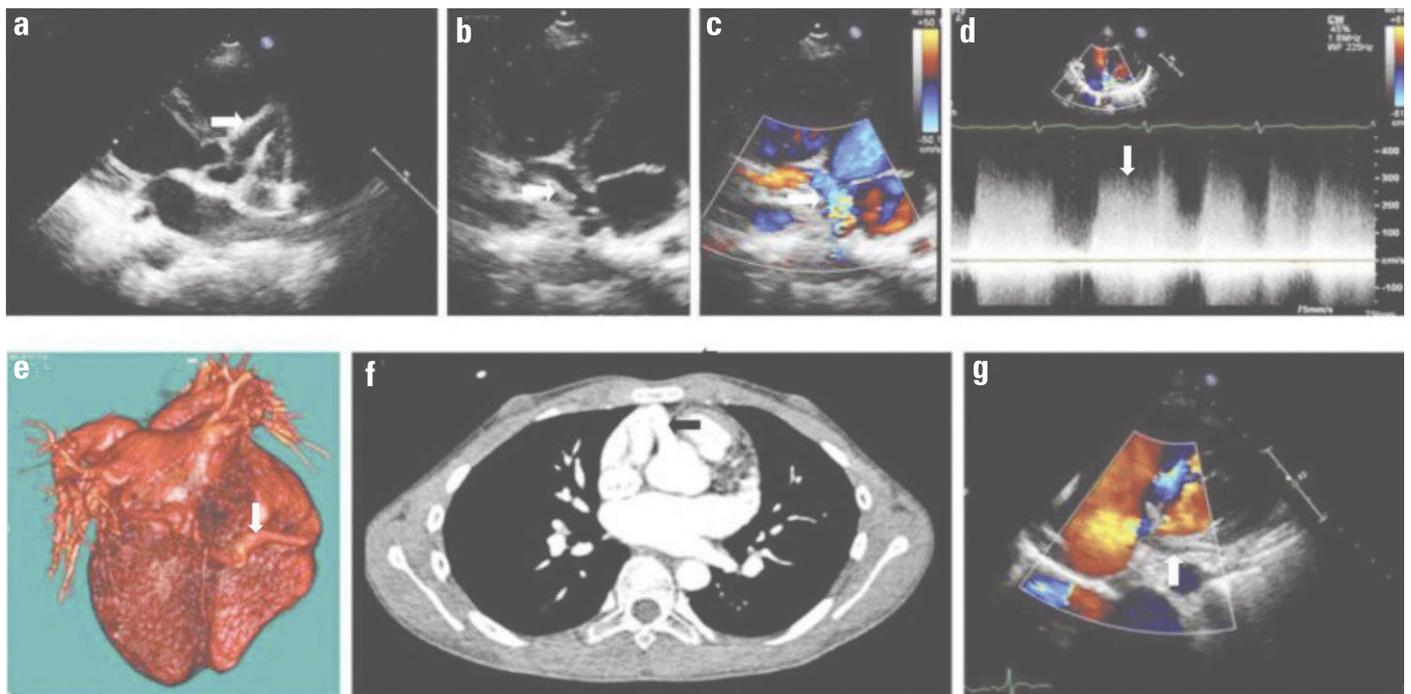


Figure 1. (a) TTE, left ventricular long-axis view showing the dilated RCA (arrow); (b, c) TTE, no standard of the aorta short-axis view showing different segments of the fistula (arrows); (d) TTE (color Doppler imaging), dual-phase shunt in the coronary artery fistula (arrow); (e) MDCT volume-rendering reconstruction of the significantly dilated RCA (arrow); (f) MDCT multiplanar reconstruction demonstrating a connection between RCA and coronary sinus (arrow); (g) TTE, four-chamber view displaying no shunt at the primary coronary artery fistula postoperatively

coronary sinus orifice). The fistula diameter was approximately 5–6 mm. The fistula between RCA and coronary sinus was closed using intermittent small incisions into RCA and continuous sutures reducing the volume of RCA. Postoperative TTE revealed that no shunt was detected in the original coronary artery fistula (Fig. 1g) and RCA patency. The patient recovered well postoperatively.

Discussion

Coronary artery fistula refers to the left and right coronary artery or its branches being directly connected to the chambers of the heart, pulmonary blood vessels, and coronary vein sinus (5). The disease makes the coronary artery blood flow directly enter the heart without flowing through the myocardial capillaries, which decreases the distal coronary blood flow and causes the coronary artery “steal” phenomenon and myocardial ischemia. Common complications include heart failure, infective endocarditis, myocardial infarction, and aneurysm rupture (6, 7). The diagnosis of the disease relies primarily on clinical manifestations and echocardiography. Two-dimensional ultrasound clearly shows the origin and course of the coronary artery and abnormal angioectasias. Color Doppler blood flow imaging shows that a ring-like, anechoic area in the fistula is accompanied by a diastolic or dual phase of colorful mosaic blood flow into the corresponding chambers of the heart. Doppler blood flow imaging not only shows the origin and the location of the bloodstream injection of the coronary artery fistula but also has a positive value in the differential diagnosis of coronary artery fistula.

The present case was a typical case of a fistula between RCA and coronary sinus. The results of echocardiography were in accordance with MDCT and intraoperative findings. Although echocardiography may diagnose most coronary artery fistulas because of the influence of the acoustic window or fistula size, it is occasionally not accurate for detecting certain coronary artery fistulas. However, coronary artery angiography can directly reveal the blood vessel distribution and the fistula drainage situation, which may help physicians definitely diagnose and effectively treat the coronary artery fistula (8). Therefore, we recommend that patients suspected of having a coronary artery fistula who are not clearly diagnosed by echocardiography should undergo coronary angiography. The possibility of natural closure of a congenital coronary artery fistula is very small. For patients with other congenital heart diseases, surgical treatment should be particularly performed as early as possible to avoid some related complications (9, 10).

Conclusion

Fistula between RCA and coronary sinus is a very rare organic heart disease. TTE can be used to diagnose most coronary

artery fistulas and is consistent with surgical findings. Once the disease is clearly diagnosed, surgical treatment should be performed as early as possible to close the fistula and reduce damage to the coronary circulation.

Video 1. RCA was markedly dilated and had an opening diameter of approximately 10 mm.

Video 2. RCA tortuously coursed to the atrioventricular sulcus of the heart bottom along the right ventricular anterior wall surface and entered the coronary sinus medially and posteriorly to the coronary vein.

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