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A Giant Right Coronary Artery of Diffuse Ectasia Induced by a Right Ventricular Fistula

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ABSTRACT

Coronary artery fistula (CAF) is a rare entity. Sometimes it may associate with mild diffuse or segmental coronary ectasia. CAF with giant coronary artery is exceptionally rare. We present a unique case of a 49-year-old female patient with a giant right coronary artery of diffuse ectasia coexisting with a fistula draining into the right ventricle. To our best knowledge, CAF with diffuse coronary ectasia of such giant size has never been reported. The patient was treated successfully by resection of the dilated right coronary artery, fistula closure, and coronary artery bypass grafting.

INTRODUCTION

Coronary artery fistula (CAF) was first described by Krause in 1865 [Dodge-Khatami 2000]. It is considered to be a major coronary malformation with an incidence of 0.002% in the general population and 0.4% in all cardiac malformations [Mangukia 2012]. It can be congenital or acquired from trauma, infectious diseases, and iatrogenic injury. CAF most commonly affects the right side of the heart and sometimes associates with mild diffuse or segmental coronary ectasia. Herein, we report a rare case of a giant right coronary artery (RCA) of diffuse ectasia with a right ventricular fistula, which was successfully treated by surgical closure and revascularization.

CASE REPORT

A 49-year-old Chinese female, without any medical and family history of trauma, infectious diseases, hypertension, or vascular malformations, presented with recurrent dyspnea on exertion for half a year. Physical examination revealed a continuous grade II-III murmur in the left fourth intercostal space. Laboratory tests were unremarkable.

Electrocardiography showed sinus rhythm with incomplete right bundle branch block as well as left ventricular hypertrophy. Chest roentgenogram showed mild pulmonary congestion and cardiac enlargement. A giant dilated RCA associated with a right ventricular fistula was detected by transthoracic echocardiography. The diameters of the RCA ostium and the fistula were 28.9 mm and 8.5 mm respectively. During further assessment, computed tomographic angiography (CTA) of the coronary artery confirmed the diagnosis. The RCA was uniformly ectatic. After a tortuous walk anterior to the right atrium, it smoothly tapered and finally drained into the right ventricle (RV) (Figure 1, B and E). In addition, the other coronary artery issues were excluded.

Surgical treatment was undertaken through median sternotomy with assistance of regular cardiopulmonary bypass. After pericardiotomy, a giant tortuous RCA injecting into the RV was noted (Figure 1, A). The RCA was uniformly ectatic and the diameter increased up to about 30 mm at the beginning. The aorta was clamped and antegrade cardioplegic cardiac arrest was provided. Following a longitudinal incision along the RCA, the fistula and the RCA ostium were exposed. Without any electrocardiographic changes, the fistula was closed by 5-0 sutures (Figure 1, C). Due to compromised distal perfusion, coronary artery bypass grafting (CABG) procedure was performed by anastomosis between the great saphenous vein and the distal RCA (Figure 1, C). The RCA ostium was closed by autologous pericardial patch and polyester felts. At last, the residual vascular cavity of the dilated RCA was closed by 4-0 sutures (Figure 1, D). The patient recovered without any complications and the postoperative CTA of the coronary artery showed patent vessel grafting and no residual dilated RCA and fistula (Figure 1, F). There was no evidence of recurrence or cardiac ischemic symptoms after 15-month follow-up.

DISCUSSION

The coronary artery fistula is a rare entity of abnormal connection between the coronary artery and other vessels or cardiac chambers. The incidence is about 0.002% in the general population and 0.4% in all cardiac malformations. It is mainly congenital while infectious diseases, trauma, and iatrogenic injury constitute a few cases. According to Chirantan's review, the RCA and pulmonary artery are the most common originating artery and draining site respectively [Mangukia 2012].

Sometimes mild, diffuse, or segmental coronary ectasia may be induced by CAF. In some rare reported cases, giant segmental coronary ectasia such as coronary aneurysm may occur [Jiang 2014]. However, such giant coronary artery of diffuse ectasia induced by CAF has never been reported before.

The clinical presentation of CAF depends on multiple factors including age, fistula size, draining site, and individual tolerance. CAF is usually asymptomatic in younger patients.

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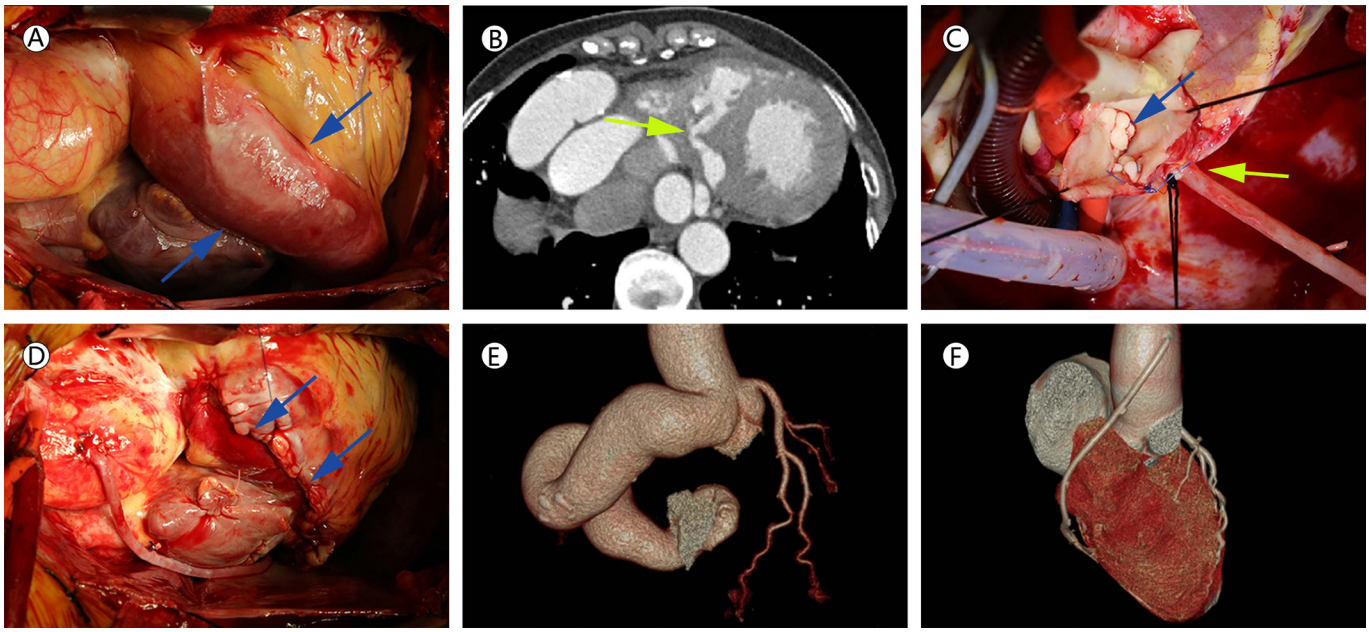

A, After pericardiotomy, the giant RCA of diffuse ectasia (blue arrows) and its tortuous journey was noted; B, The cross section of CTA showed the fistula (yellow arrow) between the RCA and RV; C, CABG procedure was performed by anastomosis between the great saphenous vein and distal RCA (yellow arrow) and the fistula was closed by 5-0 sutures (blue arrow); D, The dilated RCA was closed (blue arrow) and the CABG procedure was done; E, The three-dimensional reconstruction of the coronary artery showed the whole tortuous journey of the dilated RCA; F, Postoperative CTA of the coronary artery showed the patent vessel graft and no residual dilated RCA or fistula.

Symptoms tend to occur with increased age and the most common symptom is decreased exercise tolerance like exertional dyspnea and fatigue.

In the present case, there may be two possible reasons causing the symptom in this patient. The first reason is chronic myocardial ischemia due to coronary steal. Blood follows the lower-resistance pathway through the fistula rather than smaller arterioles and distal RCA. The dilation of RCA is a compensatory mechanism responding to the compromised myocardial perfusion. The second reason is the chronic pulmonary hypertension resulting from the left-to-right shunt, which can explain the pulmonary congestion on chest roentgenogram. Moreover, the continuity of murmur was caused by the continuous shunt throughout the cardiac cycle.

Natural history of CAF is relatively predictable because of the tendency to grow with age. If untreated, long-term complications such as cardiac enlargement, coronary steal, myocardial ischemia, endocarditis, valvular regurgitation resulting from papillary muscle dysfunction, and acute myocardial infarction due to spontaneous thrombosis [Shirai 1994] may emerge.

There is consensus about surgical treatment in symptomatic patients with CAF and various surgical approaches such as direct closure, pledgeted suture, and transcatheter embolization by coils can be applied. In addition, CABG is suggested if myocardial perfusion is compromised and the distal branch is of reasonable size.

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