Case Report

Huge Arteriovenous Fistula between a Giant Aneurismal Right Coronary Artery and Coronary Sinus

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Abstract

Coronary arteriovenous fistulas are rare and may be congenital, traumatic, or iatrogenic. We report one of these rare congenital anomalies with a giant aneurismal right coronary artery (RCA) in a middle-aged lady who was asymptomatic for a long time.

Keywords: Aneurysm, arteriovenous fistula, congenital heart defect

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Introduction

Coronary arteriovenous fistulas are rare and may be congenital, traumatic, or iatrogenic.[1,2] The reported incidence is 0.1%–0.2% among all cardiac catheterizations and of 0.002% in the general population.[3] The right coronary artery (RCA) is the most common site of origin, however, the coronary sinus (CS) is rarely involved as a draining site. We report one of these rare congenital anomalies of a giant aneurismal RCA in a middle-aged lady, who was asymptomatic for a long time.

Case Report

A 56-year-old lady presented with dyspnea on exertion for one month (NYHA class II-III). There was no family history of congenital heart disease or other inherited abnormalities. The vital signs were normal. Physical examination was unremarkable, except for a grade III continuous heart murmur at the left sternal border in the fourth intercostal border. Chest X-rays showed only mild cardiomegaly. Transthoracic echocardiography and a Doppler study confirmed a dilated CS filling via fistulous connection from the RCA (Figure 1). Coronary angiography revealed a dilated right coronary ostium and a huge RCA with a large arterio-venous fistula drained into the CS. There was no significant atherosclerothic lesion in the coronary arteries (Figure 2). No significant step up in the right side of the heart or pulmonary hypertension was seen in cardiac catheterization.

Surgical method

She underwent a cardiopulmonary bypass operative procedure under general anesthesia. Figure 3 shows the surgical view of the dilated RCA. Cardioplegic cardiac arrest was induced after aortic cross clamping via antegrade and retrograde routes. Right atriotomy was done during infusion of antegrade cardioplegic solution. The large fistulous connection was found near the ostium of the CS and transiently occluded by slight finger compression. The giant RCA was unroofed over both ends. Dilated RCA ostium was closed by an autologous untreated pericardial patch. To avoid distortion of the CS, the entry of the fistulous connection was occluded by the same technique. Because of the retrograde filling of RCA with its other branches and the risk of turbulent flow and *in-situ* thrombosis, the RCA was completely unroofed and major branches were sutured with the aid of 4-0 prolene sutures. A three graft coronary artery bypass was performed and three saphenous vein grafts were anastomosed to the right ventricle, posterolateral branches, and posterior descending artery (Figure 4). Transesophageal echocardiography showed no residual arteriovenous connection and preserved myocardial function. The postoperative period was uneventful and the patient left the hospital as functional class I.

Discussion

Coronary arteriovenous fistulas are rare and may be congenital, traumatic or iatrogenic.[1,2] The reported incidence is 0.1%–0.2% among all cardiac catheterizations and 0.002% in the general population.[3] The RCA or its branches are the most common site of the coronary fistula (55%) and both right and left coronary system are involved in 5% of cases.[4] Low pressure structures including the right-sided heart chambers, pulmonary artery, superior vena cava, and CS are the most common draining sites of coronary fistulas.[4] Most fistulas have single communication but multiple fistulas have also been reported.[4,5] Although a left-to-right shunt exists in more than 90% of patients, the shunt size most often is small and usually has no significant effect on myocardial flow.[4] The clinical presentation is dependent on the degree of the shunt, the size of the fistula, and potential complications, which include endocarditis, myocardial ischemia secondary to embolization or steal phenomenon, high output heart failure and rupture, and sudden cardiac death.[5–8] Since the first successful surgically treated coronary fistula by Bjork and Crafoord in 1947, several interventional techniques have been presented including off-pump proximal and distal ligation of the fistula, tangential arteriorrhaphy without cardiopulmonary bypass machine (CPB), and transcatheter embolization with various types

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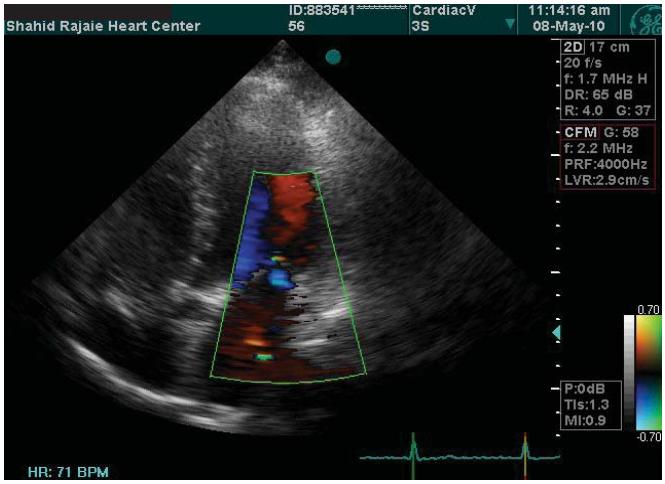

Figure 1. Trans-thoracic echocardographic view of the fistula.

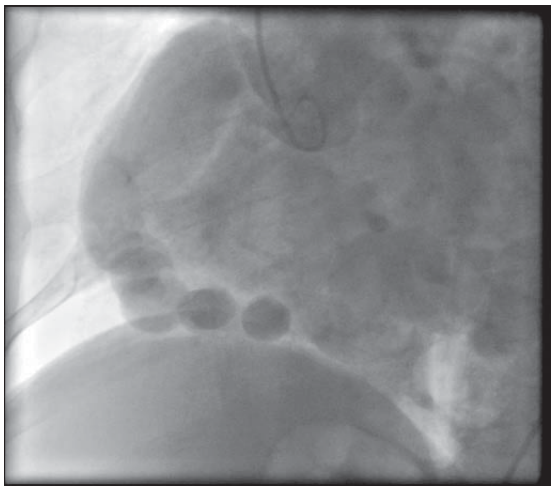

Figure 2. Selective RCA angiographic view.

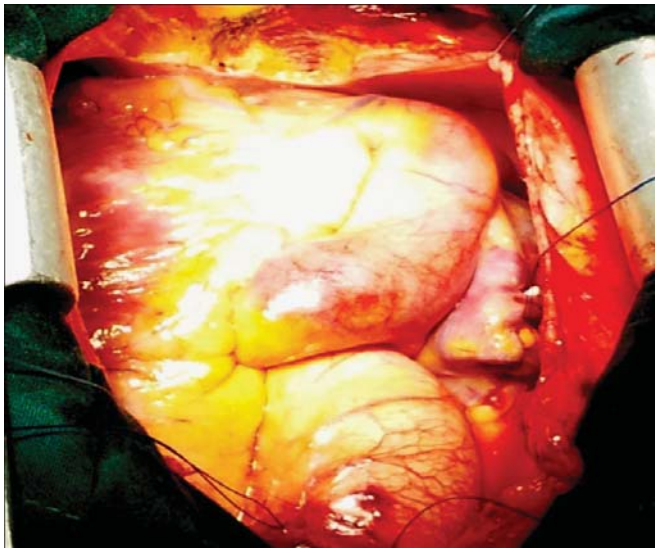

Figure 3. Surgical appearance of the huge RCA.

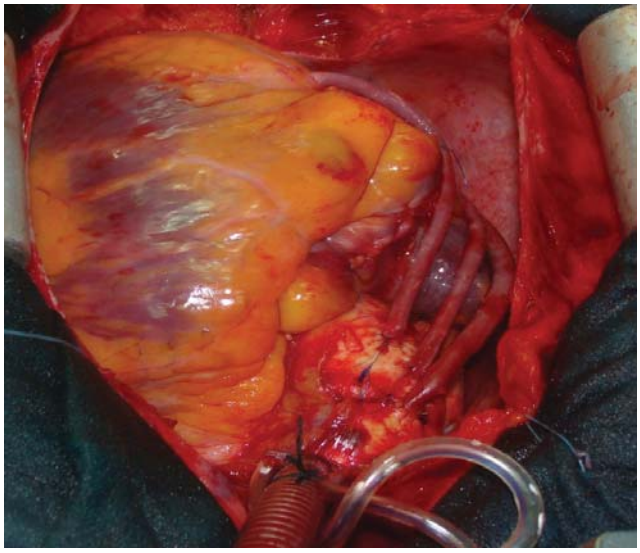

Figure 4. Unroofed aneurismal RCA and three vein graft.

of devices (such as stainless steel coils, detachable balloons, and covered stents).[3,5,7,9] Despite the increasing use of these modalities, the intracardiac closure of the fistula by CPB with or without coronary artery bypass graft remains the standard method.

We preferred to use the standard surgical technique because the patient had right dominant coronary anatomy with good distal branches. On the other hand, the risk of device embolization and residual defect were high, with percutaneous intervention using an occlusion system for such a large arteriovenous communication. Also, it might have compromised the RCA distal branches. Since the tortuous and aneurismal RCA had a potential risk of turbulent flow, clot formation and propagation of thrombosis may have lead to perioperative myocardial infarction, we occluded the origins of three major branches and performed 3 saphenous vein bypass grafts.

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