

Surgical management of a giant right coronary artery aneurysm with coronary arteriovenous fistula: case report

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Received 19 May 2020; first decision 2 June 2020; accepted 30 July 2020; online publish-ahead-of-print 9 September 2020

Background

Coronary artery aneurysms (CAAs) are uncommon, and giant aneurysms (>2 cm) are even more unusual. Coronary atherosclerosis and Kawasaki disease are the leading causes for this pathology. The treatment for this condition is controversial because the evidence is based on case report series.

Case summary

We describe the case of a 77-year-old female patient who presented with heart failure symptoms. She was diagnosed with a giant saccular aneurysm arising from the right coronary artery (RCA) ostium and a fistula between the RC and the left anterior descending artery (LAD) to the coronary sinus. And an atrial septal defect (ASD) and severe tricuspid regurgitation were also found. The patient underwent surgery through a medium sternotomy, the aneurysm was opened and resected under cardiopulmonary bypass. The RCA was ligated at the distal end of the aneurysm, and a saphenous vein graft bypass was performed. A coronary arteriovenous fistula from the distal portion of RC and LAD artery to a severely enlarged coronary sinus was found and corrected with an autologous pericardial patch. Closure of the ASD was performed with a pericardial patch and a tricuspid ring annuloplasty was done. Post-operative course was uneventful.

Discussion

There are few cases of giant coronary aneurysms associated with fistulas reported in the literature. Despite the endovascular percutaneous techniques available to treat these patients, we believe that surgical treatment was the best option for this particular case. We consider that surgical treatment is a very good option for giant CAAs associated with AV fistulas that are not susceptible for current endovascular available devices. The literature lacks evidence regarding the best approach for these cases, and we think that invasive treatment should be tailored according to the heart's anatomy and patient risk.

Keywords

Coronary artery aneurysm • Coronary artery fistula • Cardiopulmonary bypass • Case report

Learning points

- We consider surgical management is the best approach for giant coronary artery aneurysms in cases when complications such as rupture and death are very likely.
- Various surgical strategies have been adopted, such as reconstruction, resection, and isolation with concomitant coronary bypass. In cases with fistula, closure of the fistula is needed.

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Handling Editor: Julien Adjedj

Compliance Editor: Christian Fielder Camm

Peer-reviewers: Marcelo Haertel Miglioranza; Nikolaos Bonaros

Supplementary Material Editor: Peregrine Green

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Introduction

A coronary artery aneurysm (CAA) is defined as a segmental coronary dilatation that exceeds the diameter of the normal adjacent coronary artery or the diameter of the largest coronary artery 1.5 times.¹ It is a rare condition with an incidence of 1.5–5%² in the general population and it affects more frequently the right coronary artery (RCA).³ In adults, arteriosclerosis is responsible for 50% of the cases, but other causes include endovascular trauma, autoimmune disease, and other inflammatory disorders.⁴ These aneurysms tend to grow over time and are considered as 'giant' when they exceed 2 cm in diameter.⁵ The majority of the patients are asymptomatic but complications such as thrombosis, rupture, and fistula formation (26%) can occur.⁵

A coronary artery fistula (CAF) can be either a congenital or acquired pathology. Congenital fistulas have an incidence of 0.2%⁶ in the general population and can be associated with aneurysms in 19% of the cases.⁷ In this report, we present the very unusual case of an elderly patient with a giant, 9 cm CAA, associated with an uncommon condition, a fistula from both, the RCA and the LAD artery to the coronary sinus.

Timeline

2009	Diagnostic of a coronary arteriovenous fistula.
2017	Started to have heart failure symptoms.
2018	Started diagnostic test. Computed tomography, angiography, echocardiogram.
2019 Day 1	Admission for surgery
Day 2	Transfer from ICU to floor.
Day 5	Departure home
Post-operative 6, 12, and 18 months from surgery	Asymptomatic. New York Heart Association I.

Case presentation

We describe the case of a 77-year-old female, who for the last 12 months, had been presenting progressive dyspnoea and orthopnoea associated with asthenia. The patient denied the presence of angina-like chest pain or other symptoms. Her past medical history was relevant for hypertension and atrial fibrillation, she was treated with Rivaroxaban 20 mg once daily and with angiotensin converting enzyme (ACE) inhibitors (Enalapril 5 mg once daily). On physical examination, she presented a grade II systolic murmur, rales at the lung bases and grade II oedema of the lower limbs. The patient had been diagnosed in 2009 with a CAF and a CAA, but she never had surgical treatment until she was referred to our institution.

At her arrival, a coronary computed tomography (CT) angiogram was performed that showed the presence of an arteriovenous fistula from the distal RCA and LAD artery to a dilatated coronary sinus

(Figure 1). She also had a giant sacular aneurysm arising from the proximal coronary artery, right after the coronary ostium that was 9 cm in diameter. The remaining portion of the right artery was tortuous and dilatated with a diameter of 12 mm (Figure 2). A coronary angiography was performed and showed the same results as the CT scan and showed no evidence for coronary obstructive arterial disease (Figure 3). A transthoracic echocardiogram showed normal left ventricle ejection fraction, severe tricuspid regurgitation (TR), a patent foramen ovale and a 10 mm ASD.

Due to the fact that the patient had symptoms of heart failure because the fistula generated a volume overload of the right cavities and the aneurysm represented a high risk for rupture and/or thrombosis, we considered the patient should be treated. The case was discussed by the Heart Team, medical and endovascular options were considered, but according to the anatomy of the lesions we decided that the best option was to perform an open procedure. The patient was taken to the operating theatre, standard invasive monitoring with transoesophageal echocardiogram was used. We considered there was a high risk for rupture during opening, so the right axillary artery and right femoral vein were cannulated, and the chest was opened through a standard median sternotomy. The right superior vena cava was also cannulated, the patient went into cardiopulmonary bypass and the heart was arrested with Del Nido cardioplegia. A giant sacular aneurysm, 9 cm × 9 cm × 8 cm in diameter, was found emerging from the right coronary ostium and was located between the aortic root and right atrium. A coronary arteriovenous fistula located between the distal portion of the RCA and LAD artery to three large veins draining into a severely enlarged coronary sinus was also found. The aneurysm was open and resected, the RCA was ligated at the distal end of the aneurysm and a saphenous vein bypass graft was performed (Figure 4). The fistula and the atrial septal defect (11 cm × 13 cm) were corrected with an autologous pericardial patch (Figure 5) and a ring annuloplasty was performed to correct the severe TR. The procedure was finished, the patient was taken out of bypass and taken to the intensive care unit (ICU). She had an uneventful post-operative course and recovered completely. The patient has been monitored at 6, 12, and 18 months, at the moment she is asymptomatic and under medical management with enalapril, metoprolol, and acetylsalicylic acid (ASA).

Discussion

Coronary artery aneurysms are an uncommon condition, although there has been an increase in their diagnoses since the current cardiac diagnostic imaging has improved. Giant CAAs are rarely seen,⁸ and their main causes include atherosclerosis, Kawasaki disease, complications of percutaneous transluminal coronary angioplasty, and endocarditis.⁴

The possible complications of a CAA include the risk of thrombosis, embolization, rupture, and vasospasm. In cases where there is fistula formation, congestive heart failure and infectious endocarditis can occur. The natural history and prognosis of giant CAAs remains controversial. In this case, the giant aneurysm was compressing the right atrium, and there was a severe volume overload to the right cavities caused by the AV fistula and the large ASD that was found. These findings explained the symptoms that the patient presented.

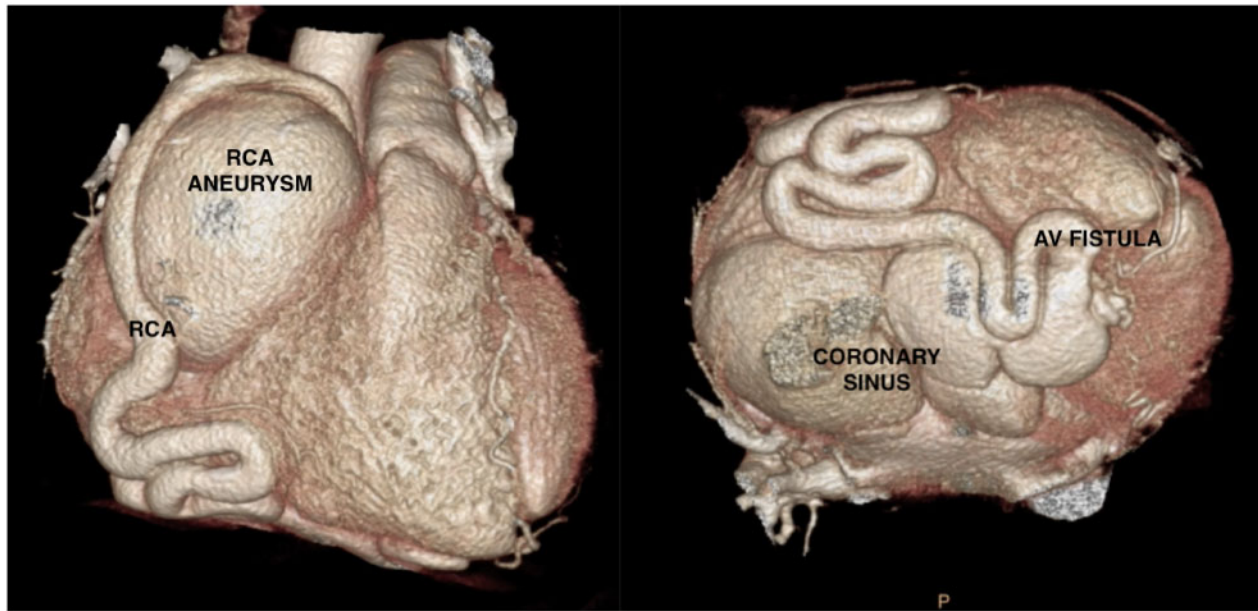


Figure 1 Computed tomography angiogram. Arteriovenous fistula from the right coronary artery to coronary sinus.

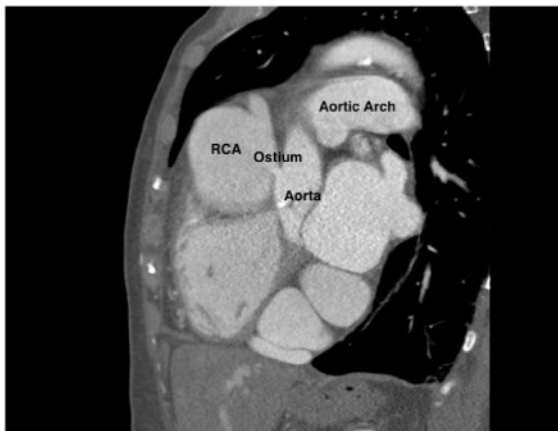


Figure 2 Computed tomography angiogram. Giant sacular aneurysm arising from the proximal right coronary artery just after the coronary ostium.

Because of the rareness of giant CAAs, treatment cannot often be standardized. Simple observation may be appropriate for small aneurysms that produce no symptoms. Conservative treatment tries to prevent thrombo-embolic complications with the use of anti-coagulant and antiplatelet drugs. However, surgical procedures need to be considered in large aneurysm cases that produce significant symptoms,⁹ as was the case for this report. The strategy to treat these patients includes, medical intervention, endovascular procedures, and open surgical techniques. Covered stents with polytetrafluoroethylene (PTFE), stents covered with saphenous vein graft

material, coil deployment, and atrial septal defect occluders (Amplatzer) have been used to treat giant CAAs, depending on their size and location.¹⁰

Polytetrafluoroethylene-covered stents were introduced in the late 1990s and have been used for treatment of CAAs with some success.¹¹ In one case, a PTFE-covered stent was used in a patient with a 16-mm × 22-mm giant CAA in the LAD just distal to a focal stenosis. The stent excluded the aneurysm and eliminated the corresponding stenosis. A follow-up angiogram continued to show exclusion of the aneurysm. Bhindi *et al.*¹² presented a case of progressive dilation of an LAD aneurysm that was successfully treated with a single covered stent. Szalat *et al.*¹³ reported successful treatment of a giant right CAA with a PTFE-coated stent and presented a review of patients who had surgical treatment of their CAAs, compared with those treated with covered stents. In this comparison, stent patients who had aneurysms larger than 10 mm had a higher restenosis rate than did those with a CAA smaller than 10 mm. Therefore, the less invasive percutaneous approach to CAA exclusion seems reasonable for aneurysms greater than 5 mm but less than 10 mm.

Surgical treatment is most widely used to avoid complications, such as extension, thrombosis, rupture, and coronary embolization. Various surgical strategies have been adopted, such as reconstruction, resection, and isolation with concomitant coronary bypass when blood flow is compromised. When a fistula is also present, it should be closed when possible, direct or patch suture should be performed.¹⁴

In the symptomatic patient not suitable for stent insertion, surgical excision or ligation of the aneurysm with bypass graft of the affected coronary arteries may be the procedure of choice. Coronary artery aneurysm that present with life-threatening complications such as heart chamber compression or fistula formation with severe shunts

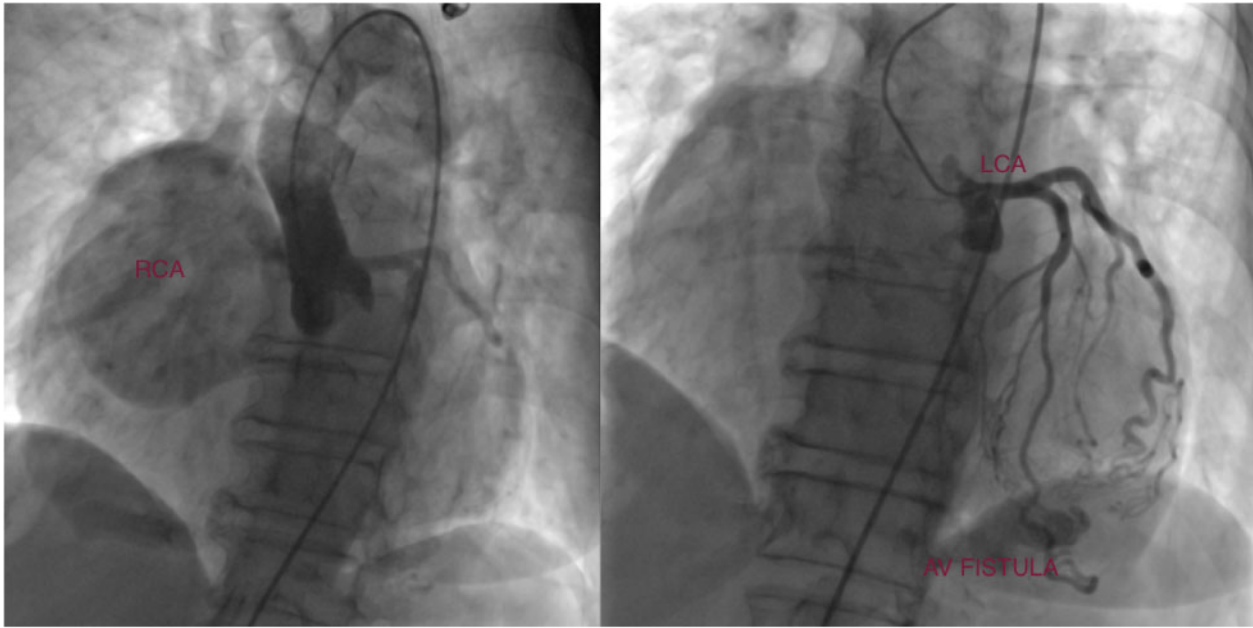


Figure 3 Coronary angiography.

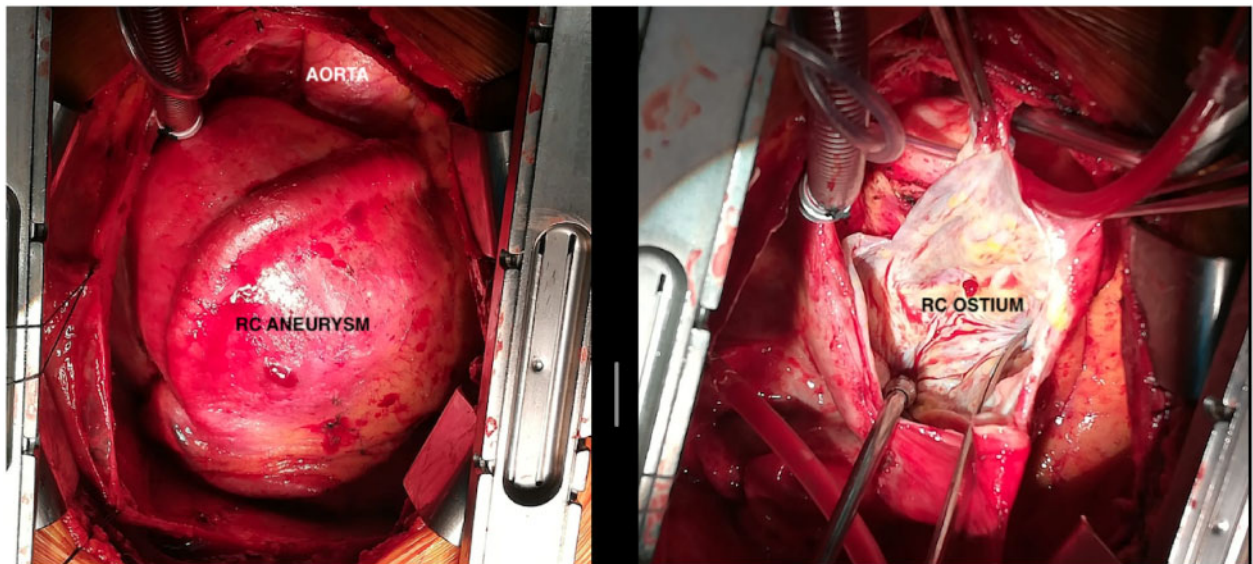


Figure 4 Right coronary aneurysm close and open with the right coronary ostium. Saphenous vein graft bypass.

require prompt surgical intervention. Some authors argue that CAA of at least three to four times the size of the original vessel diameter is an absolute indication for surgical intervention because of the propensity for complications such as compression, rupture, or thrombosis.⁸ In our case the patient was symptomatic, complicated with CAF, and in danger of rupturing if left untreated. When a giant CAA is very large, axillar, or femoral artery cannulation for cardiopulmonary bypass may be necessary to relieve the compression of ventricle

inflow tract for the resection of giant aneurysm,¹⁵ like it was in In our case.

There is no consensus as to the optimal treatment of CAAs in terms of medical or surgical therapy. It has been suggested that conservative medical management with antiplatelet or anticoagulation drugs might be an option. However, given that three out of five patients with conservative management died in Keyser's meta-analysis¹⁶ and given the perioperative mortality of 0% in Beckmann's

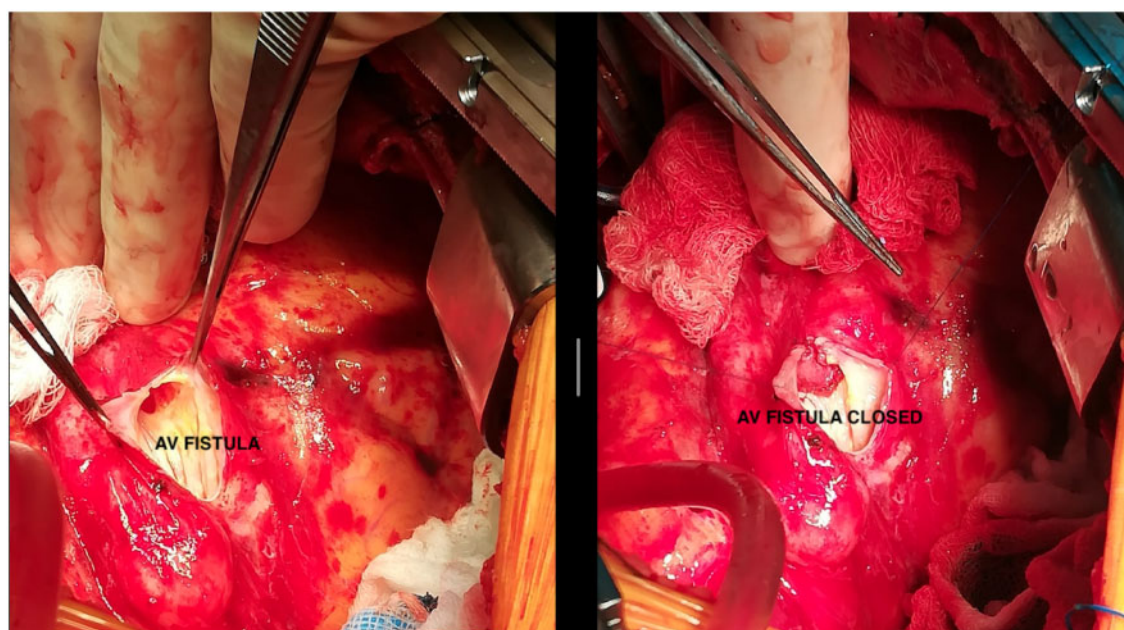


Figure 5 Coronary arteriovenous fistula open and corrected with an autologous pericardial patch.

study,¹⁷ we believe that surgical intervention is more appropriate to manage patients with CAA, especially if the patients are symptomatic and the CAAs are larger and/or associated with coronary artery disease.

Conclusions

We consider that surgical treatment is a very good option for giant CAAs associated with arteriovenous (AV) fistulas that are not susceptible for current endovascular available devices. The literature lacks evidence regarding the best approach for these cases, and we think that invasive treatment should be tailored according to the heart's anatomy and patient risk.

The patient was asked if she accepted the publication of her case, including preoperative and post-operative diagnostic and surgical images. The patient accepted and signed the informed consent.

Lead author biography



Albert Franz Guerrero graduated from Universidad Autonoma de Bucaramanga school of medicine in 2006. He started General surgery residency at Universidad industrial de Santander from 2008 to 2012. He then moved on to Cardiosurgery Fellowship at 'Colegio Mayor nuestra señora del Rosario' from 2012 to 2015. He then moved 1 year on to Congenital Heart surgery at

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Supplementary material

[Supplementary material](#) is available at *European Heart Journal - Case Reports* online.

Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as [Supplementary data](#).

Consent: The author/s confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient in line with COPE guidelines.

Conflict of interest: none declared.

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