A case report of a coronary artery fistula to coronary sinus with giant aneurysm: risk does not end with repair

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Background

Isolated coronary arteriovenous fistulas are extremely rare, accounting for 0.08–0.4% of all congenital heart disease. Closure of the fistula is recommended in cases of large dimensions, relevant left–right shunt, or ischaemic events. Thrombosis of the coronary aneurysms may occur as a postoperative complication.

Case summary

We report a case of a coronary fistula between the circumflex artery and coronary sinus with giant aneurysm. After a failed percutaneous closure attempt, the patient was surgically treated without major postoperative complications. Despite therapeutic anticoagulation and antiplatelet therapy, she presented at clinical follow-up with thrombosis of the dilated coronary artery without signs or symptoms of ischaemia.

Discussion

Management of coronary artery fistula may be challenging in cases in which initial percutaneous closure is unsuccessful. This particular case also highlights the importance of close follow-up, despite optimal therapy, to detect potentially lethal complications related to the low flow in the dilated coronary aneurysm.

Keywords

Coronary fistula • Aneurism • Surgical treatment • Coronary thrombosis • Antiplatelet • Anticoagulation • Case report

Learning points

• Closure of a coronary fistula is recommended in cases of large dimensions/aneurism, relevant left–right shunt, or ischaemic events, but the management can be challenging.
• Intracoronary thrombosis and myocardial ischaemia are two potential lethal complications in the postoperative period; therefore, anticoagulant/antiplatelet therapies are recommended.
• Close follow-up is crucial to detect early complications that may develop despite optimal medical therapy.
Introduction

Isolated coronary arteriovenous fistulas are extremely rare, accounting for 0.08–0.4% of all congenital heart disease. There are different forms and are usually classified by the site of drainage [i.e. cardiac chamber, coronary sinus (CS) and its tributary veins, or great vessels]. Rarely they drain into the CS and they may be frequently associated with aneurysmal dilatation. In adulthood, presentation is generally beyond the second decade of life, with symptoms ranging from mild dyspnoea to ischaemic events or heart failure related to a left to right shunt. Closure of the fistula is recommended in case of large dimensions, relevant left to right shunt (Qp/Qs > 1.5), or ischaemic events. Thrombosis of the coronary aneurysm may occur as a postoperative complication; therefore, anticoagulation and antiplatelet therapies are recommended.

Timeline

| Day               | Events                                                                 |
|-------------------|------------------------------------------------------------------------|
| 2007–2016         | Some episodes of chest pain and fainting episodes                       |
| September 2016    | Heart computed tomography (CT) with evidence of coronary fistula from left coronary artery to coronary sinus (CS) |
| 22 September 2017 | Coronary angiography with confirmation of voluminous fistula between circumflex artery and CS with long and tortuous course. Qp/Qs: 2/1 |
| 25 September 2017 | Ergometric test positive with lateral ST depression                     |
| 29 September 2017 | Percutaneous closure attempt fails due to device instability and a significant residual shunt |
| 13 November 2017  | Surgical closure of the fistula—start anticoagulant therapy            |
| 15 November 2017  | Signs of myocardial ischaemia with depressed left ventricular (LV) function [ejection fraction (EF) 45%]: postoperative coronary angiography highlights the exclusion of the fistula without any relevant alteration of the coronary arteries—start antiplatelet therapy |
| December 2017     | Transthoracic echocardiogram shows the presence of a hyperechogenic formation within the dilated circumflex artery—start dual antiplatelet therapy |
| February 2018     | Angio CT confirmed the presence of a thrombotic formation, partially perfused, in the circumflex artery |
| November 2019     | Patient is in good health conditions without signs of ischaemia and the echocardiogram shows recovery of the LV function (EF 55%) |

Case presentation

A 52-year-old woman was referred to our Unit after a routine medical check-up that revealed a cardiac murmur. The patient reported having suffered from episodes of chest pain and fainting in previous years. There were no other relevant elements in her medical history. The physical examination revealed a continuous cardiac murmur (3/6) along the sternal border, mostly eared on the base, and no other pathological signs. The electrocardiogram (ECG) detected a counterclockwise rotation of the QRS axis on the longitudinal plane without any sign of ischaemia (Figure 1). Transthoracic echocardiography found a severe dilation of the left coronary artery ostium with an enlarged convoluted coronary artery on the posterior aspect of the left atrium. The coronary artery was draining into a dilated CS with a high flow left–right shunt suggesting the presence of a coronary fistula (Figure 2A and Supplementary material online, Video S1). The computed tomography (CT) coronary angiogram (Figure 2C and D and Supplementary material online, Video S3) confirmed the diagnosis and showed an enlarged left coronary artery with a convoluted segment and giant aneurysm of the circumflex artery (diameters up to 35 mm). The preoperative magnetic resonance revealed preserved contractile left ventricular (LV) function and no late gadolinium enhancement in the myo-pericardium, excluding the presence of tissue damage. Coronary catheterization excluded stenosis of the coronary arteries and the aneurismatic circumflex tract draining into the CS through a fistulous connection was measured at 9 mm in diameter (Figure 2B). There was a significant left to right shunt (Qp/Qs: 2/1), with a slightly increased mean pulmonary pressure (26 mmHg; normal range: 14–25 mmHg). After collegial discussion, the patient was scheduled for elective percutaneous closure. The interventional cardiologist attempted to close the fistula with a 10 mm Amplatzer device, but due to a significant residual shunt and device instability, the percutaneous procedure was abandoned in favour of a surgical closure (Supplementary material online, Video S2).

The surgical operation was conducted through a median sternotomy, using standard bicaval cannulation. The aorta was cross-clamped, and cold blood cardioplegia was used for myocardium protection. After the right atriotomy we were not able to achieve good visualization of the fistulous drainage site in the CS. Therefore, we proceeded with the incision of the posterior aspect of the circumflex artery, exposing the fistulous tract (Figure 3B and C) that was closed with a Prolene 5/0 continuous suture (Figure 3D). The intraoperative transoesophageal echocardiography showed a mild systolic dysfunction of both ventricles.

The postoperative course was uneventful until Day 2 when the patient developed signs of myocardial ischaemia with an elevation of the ST segment in infarolateral leads and cardiac troponin T (peak of 1151 ng/L; normal values < 14 ng/L). Echocardiography showed moderate LV dysfunction [ejection fraction (EF) 48%] with hypo-kinesis of the infarolateral wall and apical portion of the septum. We performed a postoperative coronary angiography that showed the successful closure of the fistula without any relevant alteration of the coronary arteries (Supplementary material online, Video S5). The patient was started on antiplatelet (aspirin) and oral anticoagulation therapies [International Normalized Ratio (INR) target range: 2.5–3.5]. The remaining postoperative course was uneventful, and the patient was discharged in good clinical condition. At discharge, ECG...
showed negative T waves in the inferolateral site (V3–V6) (Figure 4). At 1-month follow-up, a routine transthoracic echocardiogram demonstrated the presence of a hyperechogenic formation within the dilated circumflex artery, for which a dual antiplatelet therapy (DAPT) was introduced. The angio CT performed after 3 months confirmed the persistence of a thrombotic formation with the presence of a residual lumen inside that allows perfusion of the circumflex artery (Figure 5) without any consequence on the LV function or clinical/ECG signs of ischaemic origin. At last follow-up, 2 years after the operation, the patient was in good clinical conditions, without cardiovascular symptoms or signs of ischaemia. The LV function was completely recovered (EF 55%) (Supplementary material online, Video S4).

Discussion

We present a case of a coronary arteriovenous fistula between the circumflex coronary artery and the CS. It is an extremely uncommon pathology, and only a few treated cases have been reported in the literature.3 Despite the high risk of rupture and heart failure in their natural history, the therapeutic indications are still debated. The main indications for a fistula closure are the medium or large diameter, the presence of a significant left–right shunt, and symptoms.2–4 Surgical closure is characterized by minimal morbidity and good outcomes.3 However, in older patients with severe dilated coronary artery, the risk of thrombotic complications may be high, making the medical management the best option.4 In our case, the management was challenging because a large distal fistula, with a significant left–right shunt, was associated with severe aneurysmal upstream dilatation. The first attempt with percutaneous closure failed due to the instability of the device and the presence of a relevant residual shunt. Surgical closure of the fistulous tract was necessary, but it was complicated by an acute coronary syndrome 2 days after surgery occurred. The acute coronary syndrome in the setting of a negative coronary angiography was probably caused by the reduced blood flow following the fistula closure inside the dilated coronary aneurysm. In cases of aneurysmal dilatation with a z-score >10, like for other diseases such as Kawasaki syndrome, a long-term anticoagulant/antiplatelet therapy is recommended14,6,7 to avoid thrombosis. Our patient developed a partial coronary thrombosis 1 month after surgery, although antiplatelet and anticoagulant therapy with a therapeutic range of INR was started.
from the beginning. The treatment was then potentiated with a second antiplatelet medication (DAPT); nonetheless, she presented with the progression of thrombosis that almost occluded the dilated coronary tract upstream of the fistula leaving only a small perfused lumen inside the thrombus. Despite correct therapy, the progression of the thrombosis occurred without causing complete occlusion of the circumflex artery, which could have caused ischaemic consequences. With this case report, we would like to emphasize the importance of a collaborative environment to select the most appropriate strategy to treat such complex patients. Although we followed all guidelines recommendations, the changes in the blood characteristics (flow, velocity, turbulence) caused by the fistula closure caused an intracoronary thrombosis of the aneurismatic segment. Therefore, after coronary fistula closure with an aneurismatic segment, emphasis must be placed on the priority of an early-, mid-, and long-term follow-up due to the

Figure 2 Preoperative study. (A) Transthoracic echocardiography visualization of giant circumflex artery with fistula communication to the coronary sinus. (B) Coronary angiography showing the aneurismatic coronary artery draining through the fistula in the coronary sinus (arrow). (C) Three-dimensional reconstruction from computed tomography scan showing the dilated circumflex artery (red). (D) Computed tomography scan transverse plane view of the convoluted giant circumflex artery.
Figure 3 Intraoperative view. (A) Giant aneurysmatic circumflex coronary artery. (B and C) Exposition of the fistulous drainage site into the coronary sinus through a posterior incision of the circumflex artery. (D) Continuous suture (Prolene 5/0) of the fistulous drainage site.

Figure 4 Discharge electrocardiogram with negative T waves in the inferior lateral site (V3–V6).
likelihood of unexpected postoperative findings and complications, such as coronary thrombosis and myocardial ischaemia.

**Lead author biography**

Giulia Poretti, MD, is currently a fourth year resident in the School of Cardiac Surgery at University of Verona, Italy. She was born in Varese, Italy, in 1990. She received an MD degree from Milan University Faculty of Medicine in 2015. For her interest in congenital heart disease, she is currently doing a 1-year internship at the Pediatric Cardiac Surgery Unit of the IRCCS Policlinico San Donato (Milan).

**Supplementary material**

Supplementary material is available at European Heart Journal - Case Reports online.

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**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 guidance.

**Conflict of interest:** none declared.

**References**

1. Congenital heart disease in the adult. In: Kirklin-Barratt-Boyes Cardiac Surgery, Vol. 1, 4th ed. Philadelphia: Elsevier/Saunders. p1123–1126.
2. Said SAM, Werf TVD. Dutch survey of coronary artery fistulas in adults: congenital solitary fistulas. Int J Cardiol 2006;106:323–332.
3. Hajj-Chahine J, Haddad F, El-Rassi I, Jebara V. Surgical management of a circumflex aneurysm with fistula to the coronary sinus. Eur J Cardiothorac Surg 2009;35:1086–1088.
4. Gowda ST, Latson LA, Kutty S, Prieto LR. Intermediate to long-term outcome following congenital coronary artery fistulae closure with focus on thrombus formation. Am J Cardiol 2011;107:302–308.
5. Cheung DLC, Au WK, Cheung HH-C, Chiu CSW, Lee WT. Coronary artery fistulas: long-term results of surgical correction. Ann Thorac Surg 2001;71:190–195.
6. Valente AM, Lock JE, Gauvreau K, Rodriguez-Huertas E, Joyce C, Armsby L et al. Predictors of long-term adverse outcomes in patients with congenital coronary artery fistulae. Circ Cardiovasc Interv 2010;3:134–139.
7. McCrindle BW, Rowley AH, Newburger JW, Burns JC, Bolger AF, Gewitz M et al; On behalf of the American Heart Association Rheumatic Fever, Endocarditis, and Kawasaki Disease Committee of the Council on Cardiovascular Disease in the Young; Council on Cardiovascular and Stroke Nursing; Council on Cardiovascular Surgery and Anesthesia; and Council on Epidemiology and Prevention. Diagnosis, treatment, and long-term management of Kawasaki disease: a scientific statement for health professionals from the American Heart Association. Circulation 2017;135:e927–e999.

**Figure 5** Computed tomography: thrombotic formation in the aneurysmatic circumflex artery.