

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

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Received 23 April 2020; first decision 11 May 2020; accepted 4 August 2020; online publish-ahead-of-print 27 October 2020

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, anti-coagulant/antiplatelet therapies are recommended.
- Close follow-up is crucial to detect early complications that may develop despite optimal medical therapy.

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Handling Editor: Monika Arzanauskaite

Peer-reviewers: Christoph Sinning and Christoph Jensen

Compliance Editor: Max Sayers

Supplementary Material Editor: Vassilios Parisi Memtsas

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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].¹ Rarely they drain into the CS^{1,2} 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 the left to right shunt. Closure of the fistula is recommended in case of large dimensions, relevant left to right shunt ($Q_p/Q_s > 1.5$), or ischaemic events.^{2–4} 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. Q_p/Q_s : 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 counter-clockwise 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 (Q_p/Q_s : 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 inferolateral 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-kinesia of the inferolateral 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.⁵ 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 recommended^{4,6,7} 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

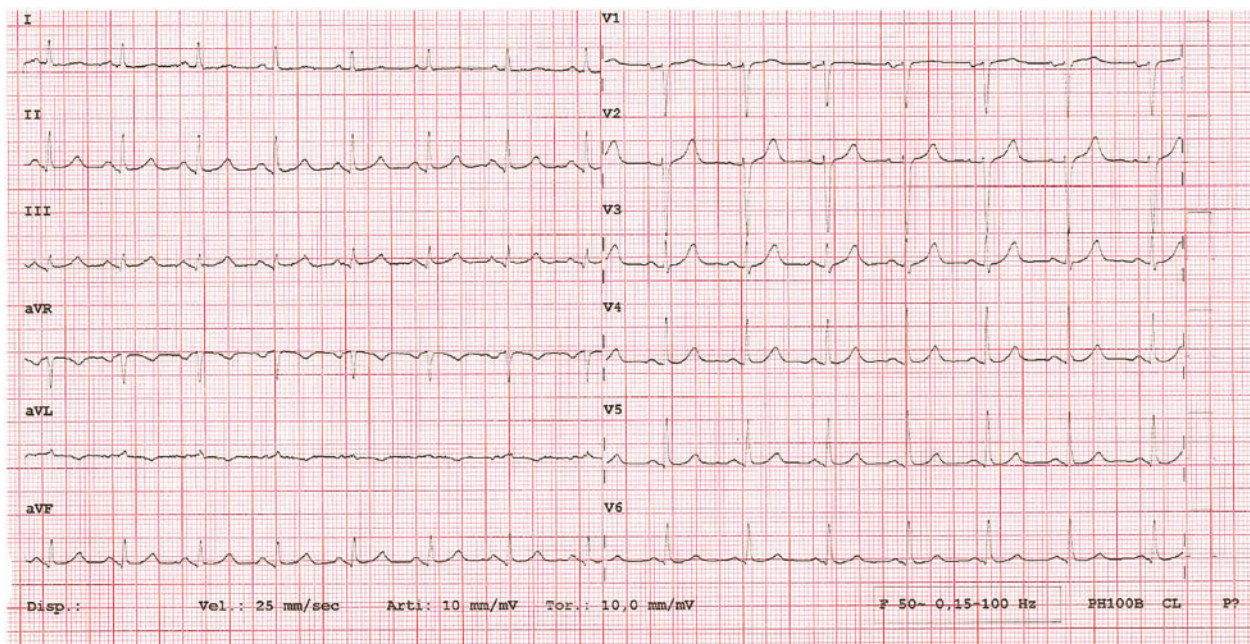


Figure 1 Preoperative electrocardiogram.

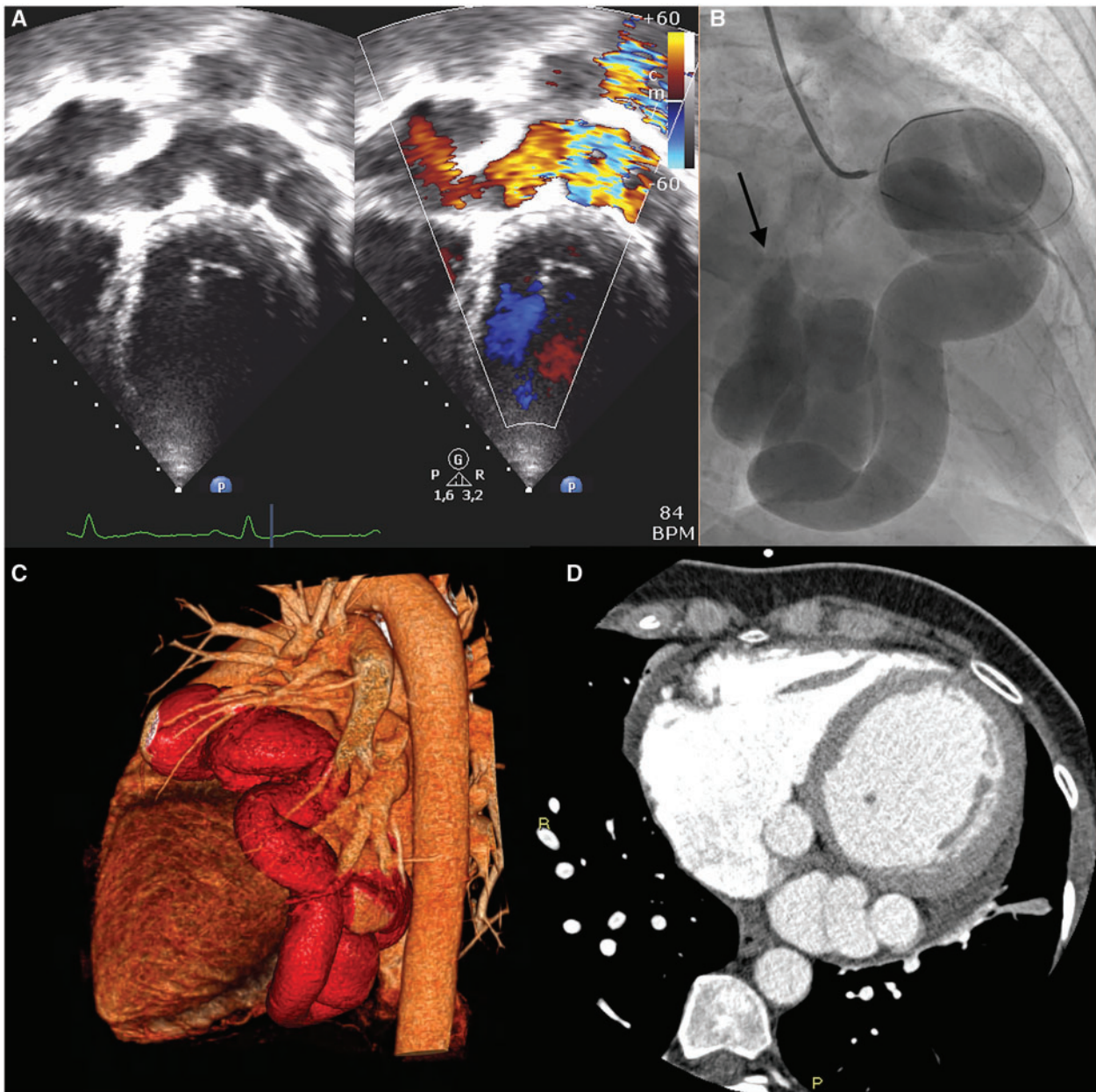


Figure 2 Preoperative study. (A) Transthoracic echocardiography visualization of giant circumflex artery with fistula communication to the coronary sinus. (B) Coronary angiography showing the aneurismatic coronary artery draining through the fistula in the coronary sinus (arrow). (C) Three-dimensional reconstruction from computed tomography scan showing the dilated circumflex artery (red). (D) Computed tomography scan transverse plane view of the convoluted giant circumflex artery.

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

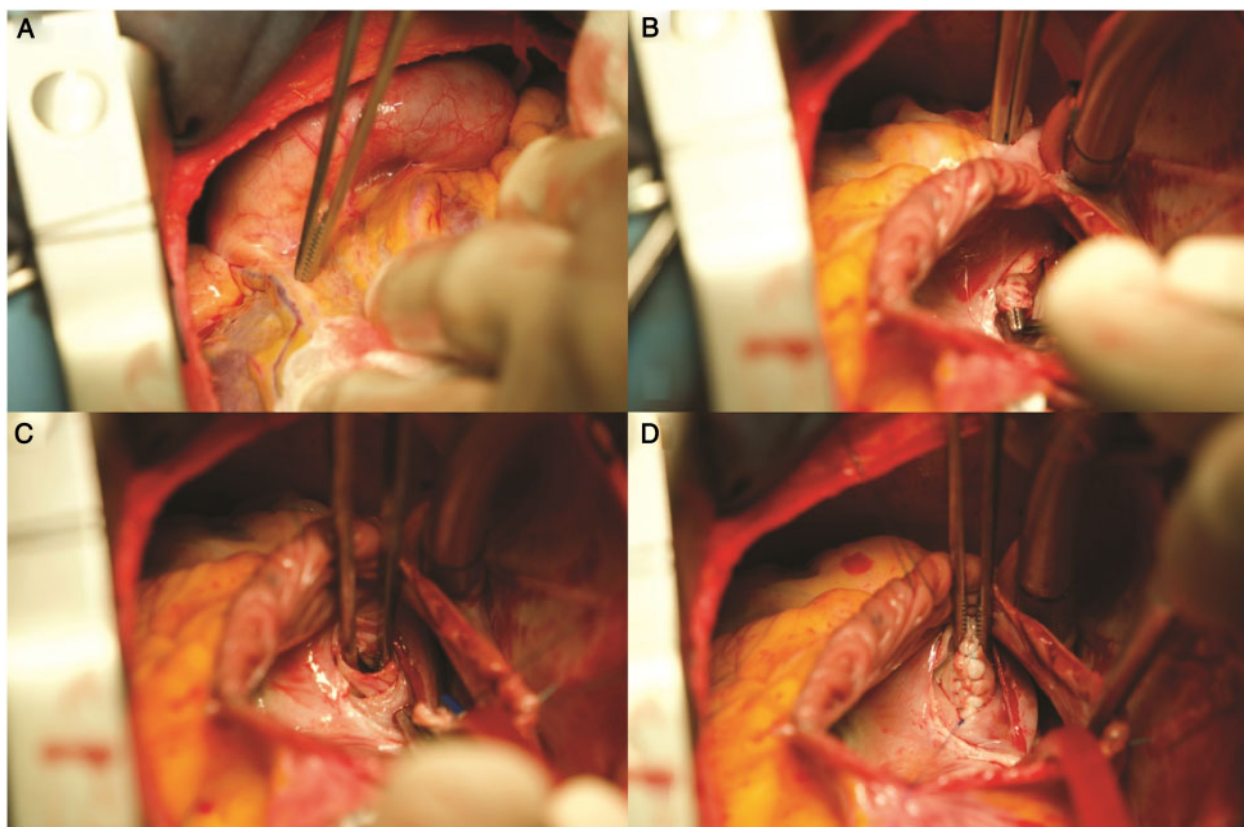


Figure 3 Intraoperative view. (A) Giant aneurysmatic circumflex coronary artery. (B and C) Exposition of the fistulous drainage site into the coronary sinus through a posterior incision of the circumflex artery. (D) Continuous suture (Prolene 5/0) of the fistulous drainage site.

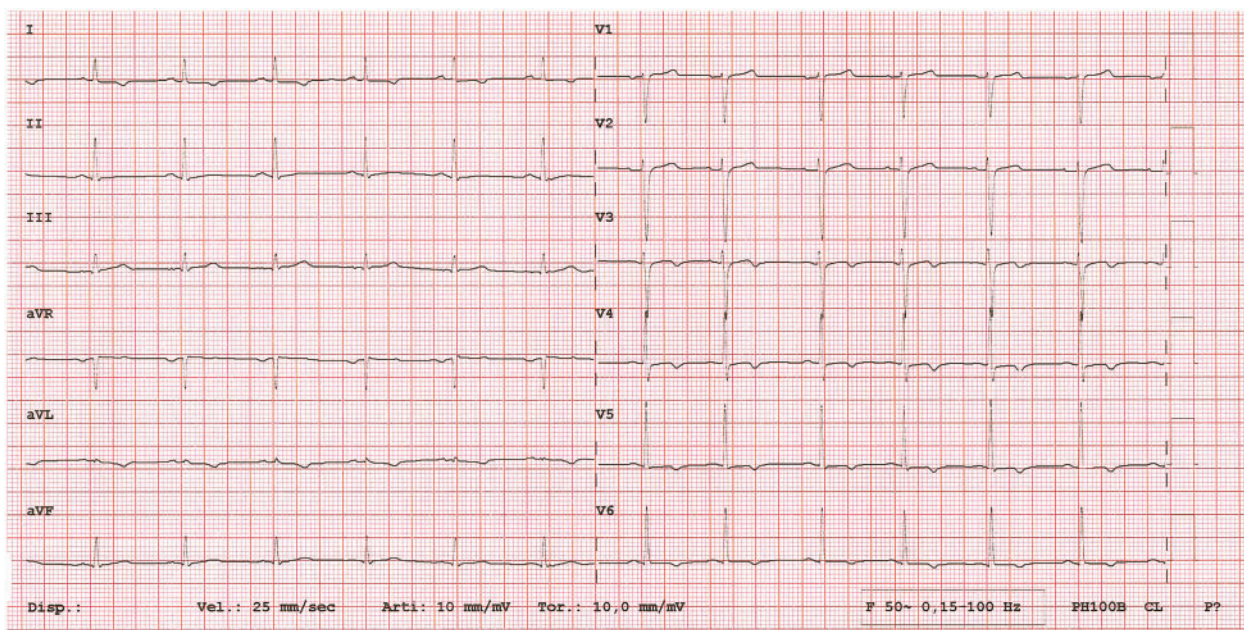


Figure 4 Discharge electrocardiogram with negative T waves in the inferior lateral site (V3–V6).

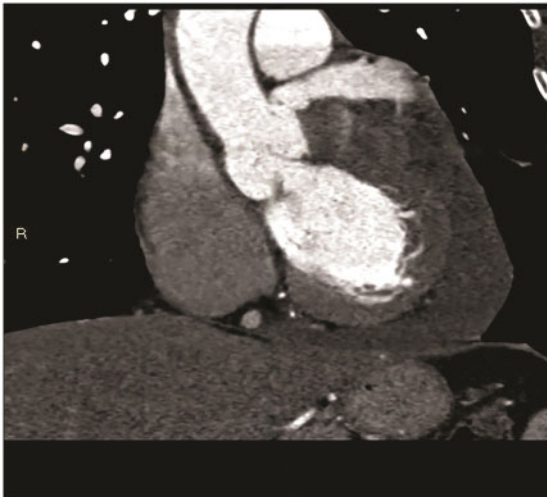


Figure 5 Computed tomography: thrombotic formation in the aneurysmatic circumflex artery.

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.

Acknowledgements

This study was supported by the IRCCS Policlinico San Donato, a Clinical Research Hospital recognized and partially supported by the Italian Ministry of Health.

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.

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