ESC European Society of Cardiology

European Heart Journal - Case Reports (2019) **3**, 1–5
doi:10.1093/ehjcr/ytz157

**CASE REPORT**
*Coronary heart disease*

# Left anterior descending coronary artery compressed by a giant coronary fistula aneurysm: a case report

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*Received 24 January 2019; first decision 16 April 2019; accepted 31 August 2019; online publish-ahead-of-print 24 September 2019*

**Background** Ischaemic chest pain can be originated by different causes. Among all, coronary fistulas are rarely the reason. Such entities are usually asymptomatic and can be diagnosed by echocardiography or coronary angiography. In an even rarer scenario, coronary fistulas might dilate and form an aneurysm.

**Case summary** We report the case of a 62-year-old patient who was initially referred to the emergency department for stable angina. Coronary angiography and computed tomography scan showed a giant aneurysm relating to a coronary fistula with a course from the circumflex coronary artery to the superior vena cava. The aneurysm was critically compressing the left anterior descending coronary artery. It was confirmed and resolved by surgery.

**Discussion** Giant aneurysms of a coronary fistula are very uncommon entities. We describe a rare case of angina caused by extrinsic compression of the left anterior descending artery from a giant aneurysm of a coronary fistula.

**Keywords** Case report • Giant aneurysm • Coronary fistula • Coronary angiography

## Learning points

- This may be the first reported giant aneurysm case of a coronary fistula, in which left anterior descending coronary artery compression was the main mechanism of ischaemia.
- Surgical management is a safe option in cases of a giant aneurysm of a coronary fistula.
- A coronary artery extrinsic compression by fistula aneurysm should be considered as a possible rare cause in the differential diagnosis of angina.

## Introduction

A coronary artery fistula (CAF) results in abnormal communication between a coronary artery and other cardiovascular structures. The incidence of CAF in patients undergoing coronary angiography (CA) is 0.1–0.2%, representing 0.4% of all cardiac malformations, with the majority of cases having a congenital origin.[1,2]

A CAF is manifested by a continuous murmur at the lower or midsternal border. CAF are usually small, but if they become enlarged they can be detected by two-dimensional or transoesophageal echocardiography.[3] The most common CAF are those originating in the

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Handling Editor: Vijay Kunadian
Peer-reviewers: Mohamed Hassan, Rafael Vidal-Perez, and Dejan Milasinovic
Compliance Editor: Amir Aziz
Supplementary Material Editor: Peysh A. Patel

right or left coronary artery and draining into the right ventricle or right atrium. In very rare cases, they drain into the superior vena cava (SVC).[1] Coronary artery fistula is usually asymptomatic, but when symptoms are manifested, angina and heart failure are the most common.[2] These symptoms appear when ischaemia is present, which is itself produced by the coronary steal phenomenon and/or fistulous tract degeneration. It is recommended that only haemodynamically significant CAF be closed.

We report a rare case of a CAF with a giant aneurysm that compressed the left anterior descending coronary artery (LAD), thus causing ischaemic chest pain.

## Timeline

| Time | Events |
|---|---|
| December 2017 | The patient reported weekly episodes of exertional angina with moderate intensity physical activity in the previous 6 months. |
| 19 January 2018 | The patient came back to our hospital due to worsening symptoms and was diagnosed with unstable angina. Admission in cardiology department. |
| 22 January 2018 | Coronary angiography identified a coronary fistula with a giant aneurysm. |
| 23 January 2018 | Computed tomography angiography confirmed the diagnosis. |
| 26 January 2018 | The patient was transferred to the corresponding surgery centre for the intervention. |
| 31 January | Surgical treatment. |
| 8 February | Computed tomography angiography showed aneurysm exclusion. |
| 15 February | Patient left hospital free of symptoms. |
| 6 months after surgical intervention | She remains asymptomatic. |

## Case presentation

A 62-year-old woman without cardiovascular risk factors was referred to the emergency department (ED) with chest pain. The patient reported weekly episodes of exertional angina during the previous 6 months, which had caused her to moderate her physical activity. On admission, the patient was afebrile with a heart rate of 62 b.p.m. and blood pressure of 130/65 mmHg. Physical examination of the heart, lung, and abdomen revealed no abnormalities. Biochemical examinations (red blood cell count, liver function tests, plasma markers of coagulation, and blood cell count) were within the normal range, as were her C-reactive protein levels. An electrocardiogram (ECG) and chest radiograph were performed in the ED, and myocardial necrosis biomarkers were measured, with no abnormality being observed. The patient was eventually discharged with a diagnosis of stable angina. Acetylsalicylic acid 100 mg and beta-blocker 2.5 mg were added to her chronic treatment and a treadmill exercise test was requested. Days later, the patient returned to the ED with minimal exertional angina. Once more, physical examination, ECG, chest radiograph, biomarkers, and two-dimensional echocardiography failed to reveal any abnormal features. An exercise treadmill test (ETT), in which the Bruce protocol was employed, revealed an abnormal clinical and electrical response at the first stage; namely a 2-mm downsloping ST-segment depression in the precordial and inferior leads and a 1-mm ST-segment elevation in the lead aVR. At this point, the patient was admitted with a diagnosis of high-risk unstable angina and underwent a CA via left radial access within the following 48 h. Coronary angiography revealed a cavity (an aneurysm) compressing the proximal segment of the LAD and which seemed to connect with the SVC (*Figure 1*, Supplementary material online-video Slide 9). The contrast medium entered and exited the cavity rapidly. The proximal segment of the circumflex artery (Cx) was dilated but the right coronary artery showed no abnormalities. A final diagnosis was made possible by computed tomography angiography (CTA), which revealed a large arteriovenous fistula connecting the Cx with the SVC. The proximal orifice of the CAF was located three centimetres from the origin of the Cx. The proximal portion of the CAF was severely dilated, forming a giant aneurysm of 70 × 60 mm that caused a significant mass effect, displacing and compressing the LAD (*Figure 2*).

After discussing the case with the local heart team, a decision was made to seal the fistula by closing the entry and exit points in order to prevent a possible rupture. There are two possible mechanisms of ischaemia (and, by association, angina) in this case: LAD compression (the most likely in this case) and diversion of the blood flow away from the Cx (coronary steal phenomenon). The patient was transferred to the corresponding surgical facilities for the intervention.

Five days later, the patient underwent surgery. The exit orifice of the CAF was found to be located on the right posterolateral side of the SVC. The CAF was ligated in the distal middle portion, in the anatomical space between the aortic root and the SVC. The saccular aneurysm located in the proximal portion of the fistulous tract was sectioned and drained. Both the entrance hole, which connected directly with the Cx and the exit hole, which communicated with the distal fistulous tract and drained into the SVC, were located and sutured (*Figure 3*). Finally, aortocoronary bypasses to both the LAD and Cx were performed.

The post-operative period was uneventful. The control CTA showed a total exclusion of the fistula from the blood flow and a significant reduction of the aneurysm's volume. The proximal segment of Cx was seen to be thrombosed (*Figure 4*). The patient's symptoms improved significantly, to the point of her being asymptomatic at the time of discharge. Since then she has remained clinically stable.

## Discussion

Very few cases of an arteriovenous fistula with a giant aneurysm have been reported to date.[4] However, the presence of a giant aneurysm in a coronary fistula compressing a coronary artery is an extremely rare circumstance that, to our knowledge, has not been reported before.

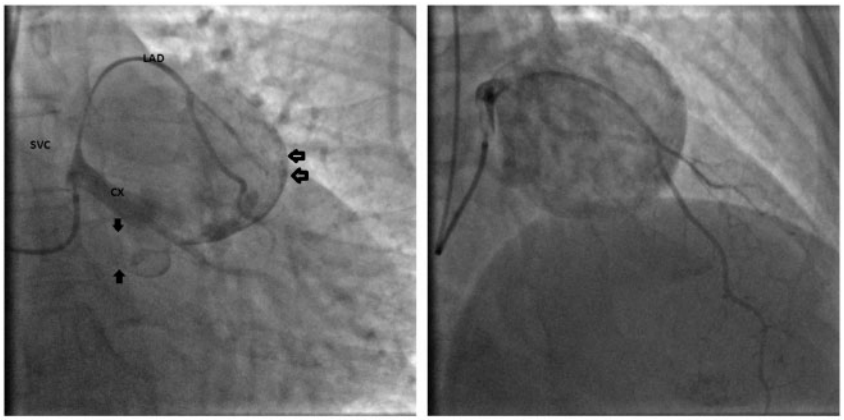


**Figure 1** Coronary angiography showed a cavity filled with contrast medium and compressing the left anterior descending coronary artery. Left: caudal view. Right: cranial view. Black arrows indicate edge of the aneurysm and filled-in black arrows indicate coronary fistula.

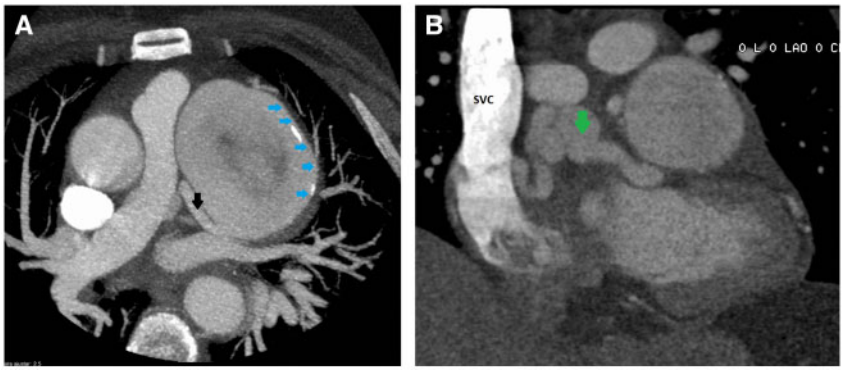


**Figure 2** Pre-operative computed tomography angiography. (*A*) Transverse axis. Blue arrows: edge of the aneurysm and black arrow: circumflex artery. (*B*) Frontal axis. Green arrow: coronary fistula.

Most CAF is asymptomatic, but some cases with large shunt can present symptoms such as heart failure or angina due to coronary steal.[5] The steal phenomenon is a known mechanism of myocardial ischaemia described in the medical literature. In this mechanism, the blood flow is diverted towards the vascular bed that offers less resistance—in this case, the walls of the fistula—thus resulting in the 'stealing' of nutrients and oxygen. However, the steal phenomenon associated with CAFs is one of the most confusing and inadequately discussed issues in the medical literature. Compensatory mechanisms take place when cardiac preload increases, and so it is difficult to ascertain to what extent this phenomenon is present in a given patient. In addition, other mechanisms may be involved, such as clotting, ectasia, or vascular wall deterioration. In the case in hand, several reasons led us to believe that the steal phenomenon was not the main cause of the patient's myocardial ischaemia. First, a physical examination did not reveal murmurs and the echocardiogram showed no dilation of right cavities or pulmonary hypertension. In our opinion, there was sufficient evidence to believe that the shunt was not haemodynamically significant. In addition, the ETT showed signs of widespread ischaemia, pointing to compression of the LAD by the aneurysm as the main cause. Moreover, the literature suggests that steal in itself is an improbable cause of critical myocardial ischaemia, as in the large majority of cases it is a consequence of coronary occlusive disease or fistulous tract degeneration.[5] However, although all these reasons supported extrinsic compression as a mechanism of our patient's ischaemia, the steal phenomenon could not be ruled out. In our opinion, our patient's fistula originated in a deviation during embryological development. Due to the characteristics of the fistula's venous wall, which make it unsuitable for a blood pressure circuit, it became dilated over the years and eventually formed a giant aneurysm that progressively compressed the LAD. At that point, when the extrinsic stenosis had become haemodynamically significant, ischaemia developed.

The patient's diagnosis was confirmed following two evaluations in the ED and, subsequently, a CA, which may be further evidence to favour including CTA in the diagnostic algorithm for patients with clinical suspicion of myocardial ischaemia.


**Figure 3** Three-dimensional reconstruction. (*A*) Anterior view. Pink: giant aneurysm, green: coronary fistula, blue: superior vena cava, and red: left anterior descending coronary artery. (*B*) Posterior view. Pink: giant aneurysm, green: coronary fistula, blue: superior vena cava, and red: circumflex artery. EH, plicated entrance hole; Eh, plicated exit hole; EO, exit orifice.

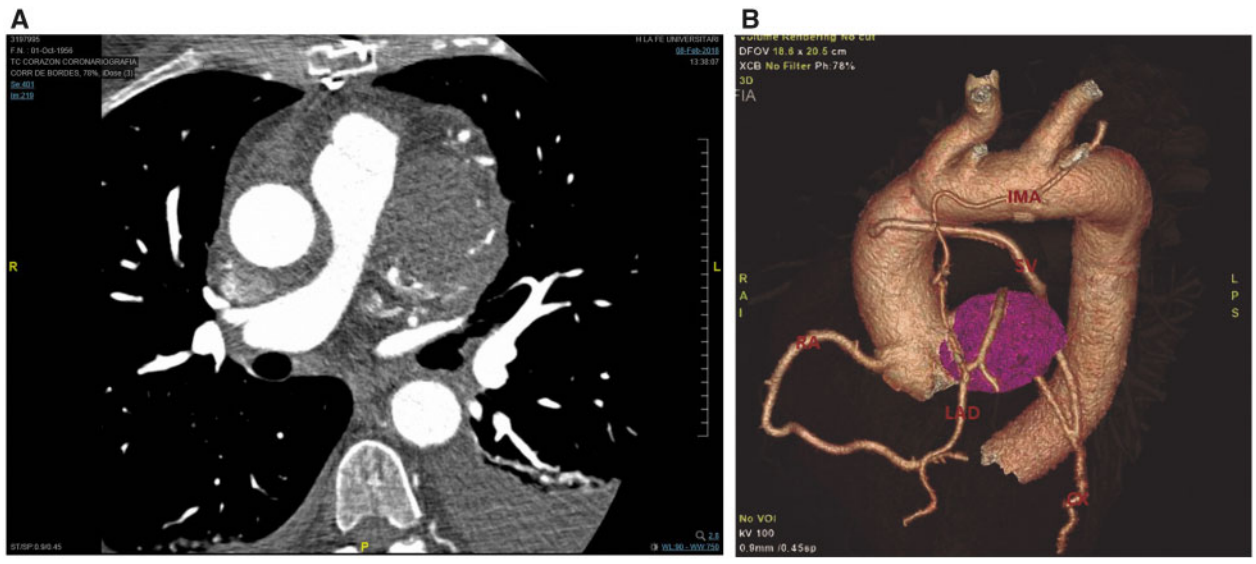


**Figure 4** (*A*) Post-operative computed tomography angiography. After the aneurysm had been resolved, computed tomography angiography confirmed total fistula exclusion and thrombosis of the proximal circumflex artery. (*B*) Post-operative three-dimensional reconstructions in volume rendering. Purple: residual aneurysm. IMA, internal mammary artery graft; RA, right coronary artery; SV, saphenous vein graft.

Therapeutic strategies in CAF are not clearly established, especially in cases with associated aneurysms. Generally, haemodynamically significant or symptomatic CAF should be closed, and early elective closure is recommended in symptomatic patients. Surgery is associated with low mortality and morbidity, and long-term outcome is excellent, with most patients remaining asymptomatic. Nevertheless, closure via cardiac catheterization is currently the method of choice, as it is a less invasive approach.[6–9]

In the case of our patient, the heart team opted for surgical treatment for two reasons: first, due to the size of the aneurysm, percutaneous intervention represented a high risk of rupture,[10] and we preferred to avoid having to perform urgent and risky surgery in the event of this occurring; and second, closing the fistula percutaneously would not have resolved the extrinsic compression by itself, as it would have been necessary to insert a long stent in the proximal LAD, which was displaced and compressed by the aneurysm, thus increasing the complexity of the procedure and the likelihood of a poor outcome.

Furthermore, the 2018 ESC/EACTS Guidelines for myocardial revascularization recommend equally (Class I) both coronary artery

bypass grafting and percutaneous coronary intervention in cases of one- or two-vessel disease with proximal LAD involvement.[11] However, in relation to our patient, whose case was considerably atypical, there was an important gap in the evidence, and we felt these recommendations should not be followed.

During the intraoperative phase, after the aneurysm had been drained and the fistulous tract sealed, the surgeon observed serious residual damage to the LAD and Cx walls due to the chronic compression, and so he decided to perform two aortocoronary bypasses. In fact, post-operative myocardial infarction due to reduced flow to the proximal coronary artery occurs in 11% of closed fistulas, which was another reason to perform the bypasses.[2]

In conclusion, this may be the first reported case of a CAF with a giant aneurysm in which LAD compression was the underlying mechanism of ischaemia. In light of this case, albeit a rare one, coronary artery compression by a fistula aneurysm should be considered as a possible cause of ischaemic chest pain.

## Lead author biography

Javier Bertolín Boronat was born in Valencia in 1987. His deep passion for science and long medical tradition in his family led him to study medicine, graduating from the University of Valencia in 2013. Currently, he practices as a Cardiology Resident doctor at the Doctor Peset Hospital in Valencia, Spain.

## Supplementary material

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

## Acknowledgements

The author would like to express his appreciation to all those who provided him the possibility to complete this report.

**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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