ESC European Society of Cardiology

European Heart Journal - Case Reports (2021) **5**(11), 1–5
doi:10.1093/ehjcr/ytab323

CASE REPORT
Cardiac Imaging

Case report of an acute coronary syndrome in a patient with artery coronary fistulae

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Received 2 January 2021; first decision 23 February 2021; accepted 26 July 2021; online publish-ahead-of-print 30 August 2021

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Background Coronary artery fistulae are rare vascular anomalies. Although they are usually asymptomatic, the presence of symptoms might present a challenge in the diagnostic and therapeutic management.

Case summary We present a patient with chest pain whose initial tests were normal, but coronary artery fistulae were found. Single-photon emission computed tomography test showed ischaemia due to coronary artery fistulae and cardiac computed tomography helped in the planning of the percutaneous closure.

Discussion CCT is emerging as an optimal non-invasive tool to characterise the morphology and course of coronary artery fistulae and may be essential for its accurate diagnosis and planning for percutaneous closure.

Keywords Case report • Coronary fistulae • Percutaneous closure • Coronary computed tomography • Steal phenomenon • SPECT

Learning points

- Coronary artery fistulae should be considered as differential diagnosis of chest pain when other common causes have been ruled out.
- Percutaneous closure of coronary artery fistulae is a safe and effective treatment in selected cases after an exhaustive study of the vessels' anatomy.
- Cardiac computed tomography is an optimal tool for identifying the morphology and course of the CAF and is useful while planning the percutaneous closure.

Introduction

Coronary artery fistulae are a rare cause of chest pain, they are usually asymptomatic and found incidentally, hence the presence of symptoms could complicate the diagnosis.[1] The most common cause of coronary atery fistulae is congenital, although iatrogenic, and other causes, have also been reported. Steal syndrome, thrombosis, heart failure, or arrhythmias have been described as frequent complications.[2] We report the case of a young male with chest pain due to a steal phenomenon by artery coronary fistula. After having demonstrated ischaemia and anomalous channels with multiple complex microvessels on cardiac computed tomography (CCT), percutaneous closure of the fistula was performed.

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Handling Editor: Edoardo Conte
Peer-reviewers: Pruthvi C. Revaiah; Antonios Karanasos
Compliance Editor: Rahul Mukherjee
Supplementary Material Editor: Mariame Chakir

Timeline

Time	Event
Day 1	The patient had chest pain. Admission in the emergency room
	Initial tests were normal, except a treadmill test that was positive
Day 2	Coronary angiography showed coronary artery fistulae
Day 3	The single-photon emission computed tomography was positive
Day 4	A cardiac computed tomography (CCT) was performed to plan interventional procedure
Day 5	Coronary artery fistula was closed
Day 6	Patient hospital discharge
2 months later	Control CCT showed the absence of flow in the fistula and the correct location of the microcoil device
	The patient remained asymptomatic

Case presentation

A 42-year-old male patient presented in the emergency room with an oppressive, prolonged, and non-radiating chest pain at rest of recent onset. Physical examination was normal. The patient was receiving long-term treatment with atorvastatin for dyslipidaemia with no other personal or family medical history. Initial tests, including electrocardiogram (EKG) and chest radiography (CXR) were normal. An echocardiogram showed normal biventricular function, without dilatation of the right chambers or other pathological findings. Laboratory results were also normal, including D-dimer testing and high-sensitive troponin serial measurements. The patient underwent a treadmill test, which was terminated at Stage 1 of the Bruce protocol due to significant early electrocardiographic changes (3 mm ST-segment depression in inferior leads and V5–V6), achieving a workload of 5.4 METs; nevertheless, symptoms did not occur during exercise. On account of these results, together with chest pain characteristics, coronary angiography was performed for suspicion of coronary artery disease. Unexpectedly, it revealed a tortuous fistula originating in the proximal segment of the left anterior descending (LAD) coronary artery, with an anterior trajectory and a narrow neck. Exercise stress imaging using Tc-^{99}m Sesta single-photon emission computed tomography (SPECT) was requested to evaluate a possible steal phenomenon of the fistula. This revealed mild–moderate ischaemia of the anterior, anteroseptal and inferior walls (*Figure 1*), and electrically positive ischaemia on EKG during stress SPECT. Coronary computed tomography (CCT) was then performed to explore the drainage of the fistula prior to eventual percutaneous closure. Cardiac computed tomography confirmed coronary angiogram findings, allowing the accurate visualization of the course and morphology of the fistula. *Figure 2* shows CCT images revealing the complexity of the fistula from proximal LAD to main pulmonary artery (PA), which also extended to the aortic arch and the left subclavian artery, but the origin was single and narrow.

According to these results, percutaneous closure of the fistula was planned using two detachable coils (Target^{TM} Helical Ultra 1.5 mm × 3 cm and 2 mm × 4 cm) delivered through a Prowler$^{®}$ select LP microcatheter in the course between the proximal LAD and the main PA to close the fistula, leading to total occlusion of all fistulous channels with optimal final results (*Figure 3A and B*). The patient was discharged without additional medications. A control CCT performed 2 months later demonstrated the absence of flow in the fistula and the correct location of the microcoil device (*Figure 4*). The patient remained asymptomatic with no evidence of ischaemia.

Discussion

We present an uncommon case of symptomatic complex small coronary arteriovenous fistulae with a single origin, but multiple drains (pulmonary and aortic arteries) causing myocardial ischaemia confirmed by SPECT. This multiple drain condition is much rarer than CAFs with a single drain (10% vs. 90%) and can complicate the clinical management but, in this case, a coil embolization was successfully performed. Coronary artery fistulae have been defined as precapillary anastomoses between a coronary artery and a major or medium vessel such as the PA, vena cava, or a cardiac chamber, among others. According to recent publications, the prevalence of CAFs in patients undergoing CCT angiography is reported to be 0.9%,[2] which is higher than the previously reported prevalence of 0.3% on invasive angiography.[3] The most common origins involve the right coronary artery draining into the right ventricle followed by the LAD draining into the PA and the most frequent cause is congenital, but some can also be secondary to cardiac trauma or complications of procedures such as cardiac surgery or endomyocardial biopsy.[4]

Coronary artery fistulae are usually asymptomatic, and thus, the diagnosis is often incidental, especially in young patients. However, our case shows a young male who debuted with ischaemic symptoms, which are mainly caused by low coronary flow as blood takes the path of lowest resistance, in this case to the PA. This concept is known as a 'steal phenomenon'. Up to 63% of patients at older ages may present symptoms such as angina, arrhythmias, heart failure in the presence of large left-to-right shunt and, more rarely, infective endocarditis.[5] Clinically, the site of the fistula termination is more important than the origin. For example, if it drains into the right cardiac chambers or systemic veins it may cause a left-to-right shunt and, therefore, right-sided volume overload, whereas drainage into left chambers may cause a left-sided volume overload. Our patient had normal cardiac chambers, as shown on the echocardiogram.

Regarding diagnosis, coronary artery fistulae can be suspected on physical examination by a continuous murmur in mid-chest, but this finding is unusual. Electrocardiogram and CXR are usually unhelpful, although EKG can sometimes show left ventricular overload, or ischaemic changes, and CXR may present right-sided chamber dilatation in cases of large shunts. Echocardiography might help to detect coronary artery fistulae by showing abnormally coloured signals in cardiac chambers, or can provide evidence of large volume fistulae.[6]

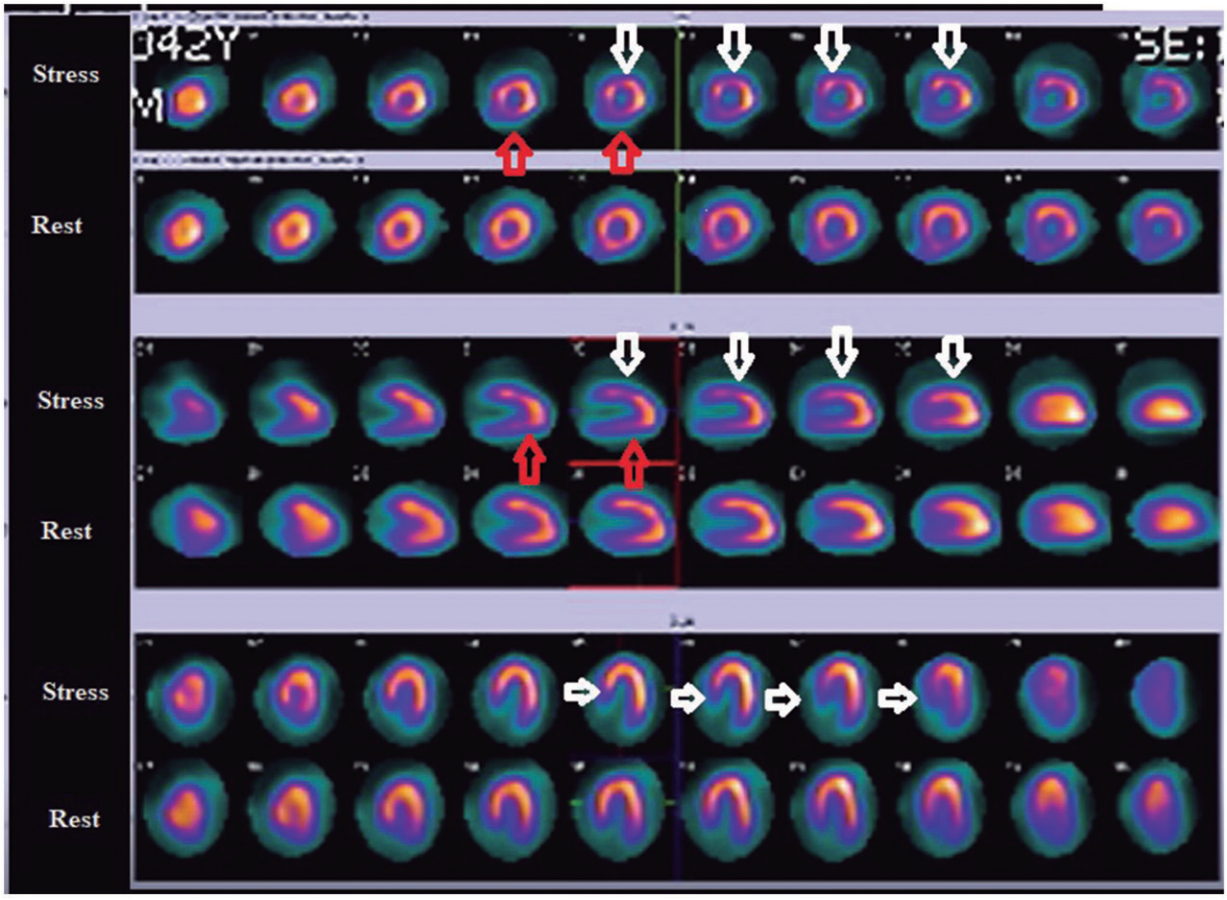

Figure 1 Stress myocardial perfusion imaging. Stress myocardial perfusion imaging examination showing a mild–moderate reversible cardiac perfusion defect in the territory of the left anterior descending artery (the anterior and anteroseptal walls, white arrows) and a moderate reversible cardiac perfusion defect in the inferior wall (red arrows) and on rest single-photon emission computed tomography images. Stress single-photon emission computed tomography (upper panels) also shows a mild fixed inferior wall perfusion abnormality resulting from diaphragmatic attenuation.

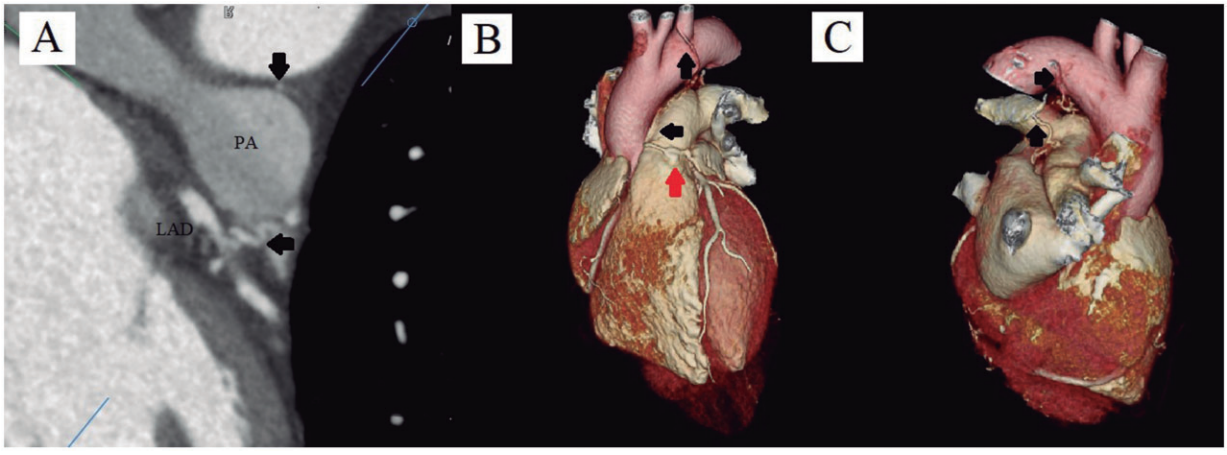

Figure 2 Coronary fistula trajectory. Coronary computed tomography showing the coronary artery fistula. (*A*) Multiplanar reconstruction demonstrating the fistula origin in the proximal left anterior descending artery (black arrow). (*B*) Volume rendering reconstructions showing an aneurism located anterior to the pulmonary artery (red arrow) and the fistula course up to the left subclavian artery (black arrow). (*C*) Different fistulous courses by the right pulmonary artery and the aortic arch (black arrows). LAD, left anterior descending; PA, pulmonary artery.

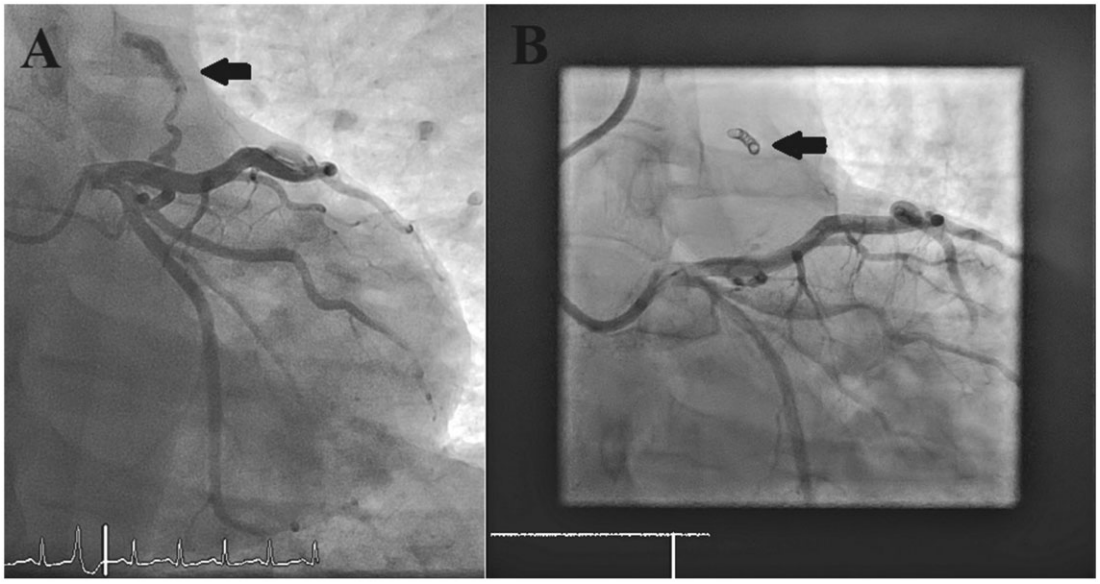

Figure 3 Coronary angiography. (*A*) Coronary angiography showing the fistula (black arrow) between the proximal left anterior descending artery and the main pulmonary artery. (*B*) Absence of flow in the fistula course after microcoil implantation (black arrow).

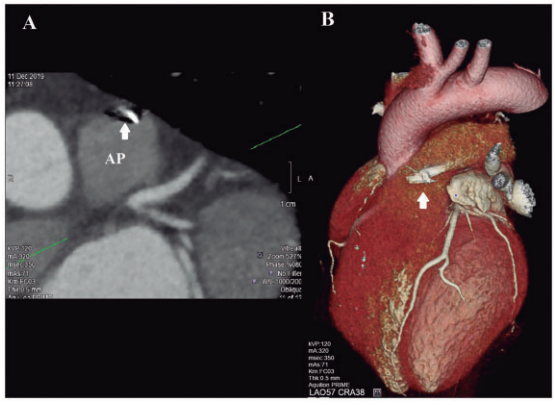

Figure 4 Follow-up cardiac computed tomography. (*A*) Multiplanar reconstruction showing proximal left anterior descending artery without evidence of fistula origin and the microcoil devices located anterior to main pulmonary artery (black arrow). (*B*) Volume rendering reconstruction showing the coils (black arrow) and the absence of fistula course. AP, pulmonary artery.

Although the gold-standard diagnostic technique is invasive coronary angiography, CCT is emerging as a novel, safer, non-invasive diagnostic technique, which also provides three-dimensional anatomical information. Cardiac computed tomography could be considered as a first-line test in low-intermediate risk patients as it can rule out coronary artery disease and can detect other coronary anomalies.[7] Furthermore, this technique can also diagnose postprocedural complications such as recanalization, thrombus, or device migration.[2] Based on our case, the use of CCT improved the assessment of fistulae, given its high spatial resolution to define anomalous channels with multiple complex microvessels and several draining sites, which were not well defined in the invasive angiography. Myocardial stress perfusion imaging by SPECT or cardiac magnetic resonance could also complement the diagnosis and unmask the presence of a steal phenomenon.[8] This is important, since treatment is only indicated in symptomatic, large, or haemodynamically significant fistulae. Antiplatelet or antibiotic prophylaxis may be given in asymptomatic cases.[2] In our case, we decided to close the coronary fistulae as ischaemia was demonstrated by stress SPECT in the LAD territory, despite the absence of coronary disease. This contrasts with the recent publication by Sherif *et al.*[9] who presented a patient with a coronary artery fistula where myocardial ischaemia was not demonstrated, so invasive treatment was not performed. Closure can be achieved either using a percutaneous approach, in cases of less tortuous or shorter courses and single fistulae,[10] or cardiac surgery, demonstrating a similar good prognosis. Percutaneous closure of coronary artery fistulae is an emerging alternative in patients with suitable anatomy and has shown to be safe and feasible in previous reported series.[4,11] Multiple devices have been used including coils, covered stents, vascular plugs, and atrial septal defect devices, depending on the size and shape of the anomalous vessel. In suitable cases, percutaneous closure with detachable coils is desirable to avoid a covered stent implantation that may be associated with future in-stent restenosis. In our case, a percutaneous transcatheter approach was chosen, because a single origin was identified of easy access.

Conclusions

We report the case of a young male with chest pain from an unusual cause: a steal phenomenon caused by a complex coronary

arteriovenous fistula. After having demonstrated anterior ischaemia and anomalous channels with multiple complex microvessels by CCT, percutaneous closure of the fistula was performed using micro-coils, with optimal early and long-term results and relief of symptoms.

Lead author biography

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Supplementary material

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

Acknowledgements

We thank Dr Beatriz López-Melgar for her contribution in the CCT imaging acquisition.

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

Consent: The authors confirm that written consent for submission and publication of this case report including images and associated text has been obtained from the patient in line with COPE guidance.

Conflict of interest: none declared.

Funding: L.F.-F. has received fundings from Comunidad de Madrid Government (AORTASANA-CM; B2017/BMD-3676), Fondo Social Europeo (FSE) and from Instituto de Salud Carlos III, Spain (PI15/02019).

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