Reversible myocardial ischaemia caused by ectopic left circumflex coronary artery: a case report

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Background
Coronary artery anomalies (CAAs) are congenital disorders associated with variable manifestations and pathophysiological mechanisms. Anomalies can be asymptomatic or cause chest pain, myocardial infarction, or even sudden cardiac death.

Case summary
We describe a 34-year-old man with a history of a single episode of chest pain. An ectopic origin on the part of the left circumflex (LCX) coronary artery from the proximal right coronary artery (RCA) was evident upon coronary computed tomography angiography. A positron emission tomography perfusion study revealed a stress-induced perfusion defect in the anomalous LCX territory (infero-posterior wall). The patient experienced dyspnoea and ST-segment depression in electrocardiography, suggestive of myocardial ischaemia during the maximal bicycle ergometer stress test. No mechanical compression or stenosis was seen upon invasive coronary angiography. The left ventricular perfusion normalized after the initiation of beta-blocker medication.

Discussion
Patients with CAAs especially benefit from a multimodality assessment of the vascular territories. In our case, the myocardial perfusion of the infero-posterior wall normalized after treatment with beta-blockers. This may be due to increased coronary vasodilation capacity and myocardial flow reserve, as well as reduced oxygen consumption. Beta-blockers may represent a viable option in low-symptomatic CAA patients with perfusion defect and no ostial stenosis or compression.

Keywords
Case report • Left circumflex coronary artery • Coronary anomaly • Myocardial ischaemia • PET perfusion

ESC Curriculum
2.1 Imaging modalities • 2.4 Cardiac computed tomography • 2.5 Nuclear techniques

Learning points
- Ectopic left circumflex coronary artery origin with the retro-aortic course can cause perfusion defects. In this case, the left ventricular perfusion normalized after treatment with beta-blockers.
- Multimodality imaging was needed to exclude ischaemic coronary artery disease or mechanical compression of the coronary artery.
- Especially patients with coronary artery anomalies might benefit from the assessment of vascular territories with hybrid imaging.

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Introduction

The normal anatomy of the coronary artery has two main coronary branches: the left main coronary artery (LCA) and the right coronary artery (RCA). The left circumflex (LCX) artery and left anterior descending (LAD) artery originate from a common trunk of the LCA. However, the coronary anatomy can vary. The prevalence of an anomalous origin on the part of the LCX from the RCA has been reported to be ≈0.1%.1 Some of these coronary artery anomalies (CAAs) can lead to symptoms, such as chest pain, myocardial infarction, and even sudden cardiac death. Although LCX anomalies are generally considered to present as benign and asymptomatic, in some cases, they have been reported to induce myocardial ischaemia.2 Invasive approaches for CAAs are considered if the ostium is compressed or stenotic.3–5

Timeline

| Week(s) Before | Event |
|---------------|-------|
| 9 weeks       | The patient experienced right-sided chest pain while driving a car. Symptoms lasted about an hour and relieved spontaneously before any medical examination. The patient contacted occupational healthcare and was referred to a cardiologist for further studies. |
| 5 weeks       | Transthoracic echocardiography was normal (left ventricular ejection fraction was 60%). An ectopic left circumflex artery (LCX) origin was detected in the diagnostic computed tomography coronary angiography. |
| 3 weeks       | Significant myocardial ischaemia was detected in the ectopic LCX territory in the 15O-water positron emission tomography (PET) perfusion stress study. |
| 4 weeks       | The patient underwent a maximum capacity bicycle ergometer test and had demonstrable myocardial ischaemia on his electrocardiogram. |
| Day 1         | Long-acting beta-blocker (metoprolol 23.75 mg/day) and isosorbide nitrate (20 mg/day) were started. |
| Day 2         | The patient discontinued the use of isosorbide nitrate after the first dose due to overall weakness. |
| Day 21        | The left ventricular perfusion was normalized in the control 15O-PET perfusion stress study. |
| Day 28        | Invasive coronary angiography was performed with no signs of vessel compression. |
| Day 31        | The patient is asymptomatic and back to work. |

Case presentation

We present a case of a 34-year-old man who experienced a single episode of chest pain while driving his car. The patient had no past medical history or previous cardiac symptoms. The chest pain was predominantly right-sided and lasted about an hour. When the ambulance arrived, the patient was asymptomatic, and his electrocardiogram (ECG) was normal. Because there was no need for acute care, the patient was instructed to make an elective appointment with a physician. After a few days, the patient contacted his occupational physician, who referred him to a cardiologist. There were no abnormalities in the basic laboratory tests (haemoglobin 155 g/L, low-density lipoprotein 3.0 mmol/L, and creatinine 81 µmol/L) and no elevated levels of cardiac biomarkers (cardiac troponin T or creatine kinase isoenzyme MB mass). The patient’s heart rate was 73 beats/min, and his blood pressure level was 135/85 mmHg. The echocardiogram was normal, with a left ventricular ejection fraction of 60%. The regional wall motion was normal, and no valvular regurgitation or stenosis was found. The patient had no heart failure findings or symptoms at any stage.

Due to his work in the railways, he was granted sick leave and referred for computed tomography coronary angiography (CTCA) to rule out coronary artery disease (CAD). This CTCA was performed with a Siemens Somatom Flash (Siemens Healthineers, USA). There were no signs of stenosis or calcification in the coronaries. However, there was an ectopic LCX origin with a retro-aortic course from a small RCA (Figure 1). The influence of coronary ectopy on myocardial perfusion was further studied with 15O-water positron emission tomography (PET) during adenosine stress (0.14 mg/kg/min adenosine infusion for 6 min 35 s). A 15O-water bolus (875 MBq) was given at 1 min 35 s from the onset of the adenosine infusion. A dynamic 5 min PET scan was performed with a Siemens Biograph Vision (128 × 128 matrix with Gaussian filtering). During the adenosine infusion, the heart rate increased from 89 to 109 beats/min. The blood pressure did not change significantly from the rest value (130/89 mmHg) to that measured during the stress (128/60 mmHg). No ECG abnormalities were observed during the adenosine stress. The global perfusion was normal 2.8 mL/g/min (normal lower limit is 2.3 mL/g/min).6 However, a significant perfusion defect was seen in four segments of the infero-posterior wall (Figure 2A). In the PET/CTCA fusion image, the perfusion defect was localized to the ectopic LCX territory (Figure 3). A stress-only protocol was adopted to reduce the radiation dose.

The patient was referred to undertake a bicycle ergometer test with a ramp protocol (20 W/min) to detect potential ischaemia-induced electrophysiological and/or haemodynamic changes. The performance tolerance was normal (4-last-min mean load was 189 W). During the test, the patient experienced dyspnoea and concurrent down-sloping/ horizontal ST-depression in the ECG (Leads II, III, aVF, and V6, Figure 4). Symptoms and ECG changes resolved within 30 s after the stress. Because the CTCA obtained during diastole did not provide information regarding lumen diameter in systole, an invasive coronary angiography was performed to rule out mechanical compression of the anomalous coronary artery. No mechanical compression or stenosis was seen in LCX (Figure 5).

A multidisciplinary heart-team meeting was arranged to determine the optimal form of treatment. Due to the mild symptoms and the lack of coronary compression, surgical intervention was not considered appropriate. A long-acting beta-blocker (metoprolol 23.75 mg/day) and nitrate (isosorbide dinitrate 20 mg/day) were prescribed. No other medication was prescribed. Due to the side effects (overall weakness), the patient discontinued nitrate after its first administration.
Due to the patient’s occupation, the PET study was repeated 3 weeks after the onset of the medication. The radiation dosage of a single $^{15}$O-PET scan is low (effective dosage 0.8 mSv), and this is considered a justified re-evaluation method. The stress protocol and scanning devices were the same as during the first scan. The dosage of $^{15}$O was 917 MBq. The heart rate became raised from 62 to 102 beats/min during the adenosine stress. The blood pressure did not change significantly during the stress (126/63 mmHg) from the rest values (128/49 mmHg), and no significant ECG changes were detected. Global perfusion was 3.3 mL/kg/min, and the infero-posterior wall perfusion was normal (Figure 2B). The patient was asymptomatic and could return to work.
Figure 3  Hybrid-image (combined $^{15}$O-water perfusion map and computed tomography angiography) before the initiation of beta-blocker therapy. On the left, a posterior projection with decreased perfusion in the ectopic left circumflex territory (yellow–green). On the right, an anterior projection with normal perfusion in the left anterior descending territory. LCX, left circumflex; LAD, left anterior descending; RCA, right coronary artery.

Figure 4  The patient was referred to undergo a bicycle ergometer test with a ramp protocol (20 W/min). The electrocardiography sample is presented from all 12 leads. The first and fifth columns represent electrocardiography samples at baseline (in the sitting position). Maximum ST-depressions developed in Leads II, III, aVF, and V6 at peak exercise (Columns 2–3 and 6–7). Recovery electrocardiography samples are presented in Columns 4 and 8 at 6 min 50 s after stress, when the electrocardiography had normalized.
**Discussion**

Typically, perfusion defects associated with coronary arterial territories are based on general assumptions regarding the coronary anatomy and its typical vascular distribution pattern. In some cases, these assumptions may be inaccurate. For example, in our patient, if we had only assessed the perfusion map, without the CTCA information, this would have led to a misdiagnosis. For this reason, patients with CAAs especially benefit from a multimodal assessment of the vascular territories.

In our patient, invasive coronary angiography was performed after the imaging revealed signs of abnormal perfusion. However, no stenosis or mechanical compression of the vessel was detected. The perfusion defect diminished after 3 weeks of beta-blocker treatment. It has been shown, both in patients with CAD and healthy subjects, that beta-blockers increase the capacity for coronary vasodilation and myocardial flow reserve. The clinical effects of beta-blockers may be explained by their ability to reduce oxygen consumption but also by their vasodilation properties. It is unclear whether beta-blockers can be viewed as an alternative therapy to interventional procedures. However, in cases in which there is only slight ischaemia and mild symptoms, the initiation of beta-blocker medication may represent a viable option. Furthermore, it has been shown that middle-aged and older people with CAAs can often be treated conservatively.

Because the patient had only a single pain episode at rest, the pain may have been extracardiac in nature, and the abnormal origin of the LCX may have been simply a coincidence. Nonetheless, because of the person’s profession, cardiovascular disease had to be ruled out.

Based on this case and a previous case report of CAAs, it is important to note that, even though there is no evidence of compression of a coronary artery, there may well be reversible myocardial ischaemia. For this reason, functional perfusion imaging during stress should be added to standard anatomical imaging.

**Conclusion**

Patients with CAAs especially benefit from a multimodal assessment of the vascular territories. Our patient had ectopic LCX origin with an infero-posterior perfusion defect that proved reversible after the onset of beta-blocker medication.

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Saara Sillanmäki, MD, PhD, is working in the Department of Clinical Physiology and Nuclear Medicine, Kuopio University Hospital, Kuopio, Finland. Her research is focused mainly on cardiovascular physiology and imaging. In addition, she is working also in the Sleep Technology and Analytics Research (STAR) Group of the University of Eastern Finland. She is the present chair of the National Medical Association subdivision of Nuclear Medicine and Imaging.
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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 these cases and suitable for local presentation is available online as Supplementary data.

**Consent:** Written informed consent was obtained from the patient for anonymized information to be published in this article.

**Conflict of interest:** S.S. and M.H. have research collaboration with AstraZeneca and Precordior Oy. M.H. is an external lecturer and consultant of Siemens Healthcare. These collaborations have not influenced on writing the manuscript. Other authors have nothing to declare.

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