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CASE REPORT
Coronary heart disease

Spontaneous coronary artery dissection of the proximal left circumflex artery: a case report

Aleksandra Pineda *, Josh Martin , Aniket Puri, and Bijan Jahangiri

Christchurch Hospital, 2 Riccarton Ave, 8011 Christchurch, New Zealand

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Background Spontaneous coronary artery dissection (SCAD) has gained attention as an important cause of acute coronary syndrome and sudden cardiac death (SCD) among women. Management strategies of SCAD differ from those of atherosclerotic disease. There is an elevated risk of complications and suboptimal outcomes in patients with SCAD undergoing percutaneous coronary interventions (PCIs).

Case summary A 48-year-old woman without any traditional cardiovascular risk factors was admitted with severe central chest pain with associated dyspnoea and diaphoresis. The patient had a strong family history of SCD, affecting three female members in their 40s and 50s. Cardiac troponins were elevated. Coronary angiogram showed moderate to severe stenosis of the proximal circumflex coronary artery. Optical coherence tomography confirmed SCAD with sub-intimal haematoma. Despite significant stenosis in the proximal segment of a relatively large artery, a decision was made not to proceed with PCI. The follow-up angiogram demonstrated normal coronaries. Magnetic resonance imaging of renal arteries showed features suggestive of fibromuscular dysplasia affecting the right renal artery. Subsequent genetic counselling and gene testing were unremarkable.

Discussion Conservative management of SCAD is recommended because the large majority of SCAD lesions heal naturally, whereas PCI is associated with increased risk of complications and adverse outcomes. Whether SCAD is associated with the sudden death events in our patient's family remains unclear. It certainly raises concerns as to an inheritable condition. In the absence of post-mortem findings in her family members, we can only speculate about this representing a possible inheritable form of SCAD.

Keywords Case report • Spontaneous coronary artery dissection • Sudden cardiac death • Optical coherence tomography

Learning points

- Distinction between spontaneous coronary artery dissection (SCAD) and atherosclerotic lesions is important, given suboptimal outcomes in patients with SCAD undergoing percutaneous interventions.
- In most cases, a conservative management strategy is recommended.

Introduction

Spontaneous coronary artery dissection (SCAD) accounts for 2–4% of all cases of acute coronary syndrome, 24–36% of myocardial infarctions (MIs) in women <50 years and is a common cause of MI associated with pregnancy.[1] Spontaneous coronary artery dissection is also gaining attention as an important cause of sudden cardiac death (SCD).[2] Management strategies of SCAD differ significantly from

* Corresponding author. Tel: +64 03 3640640, Email: Aleksandra.Pineda@cdhb.health.nz
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those of atherosclerotic disease, and it is important to distinguish these two conditions given the markedly elevated risks of complications and suboptimal outcomes in patients with SCAD undergoing percutaneous interventions.[1]

Timeline

Time	Events
Day 1	Patient presented with Canadian Cardiovascular Society (CCS) Class IV angina. Diagnosis of non-ST-segment elevation myocardial infarction.
Day 3	Coronary angiography showed lesion of the proximal circumflex coronary artery. Optical coherence tomography confirmed spontaneous coronary artery dissection. The patient was managed medically (received dual antiplatelet therapy, metoprolol, and cilazapril).
Day 6	Patient remained clinically stable and was discharged from hospital.
11 weeks later	Repeat coronary angiography showed normal coronaries.

Case presentation

A 48-year-old woman was admitted to hospital with severe central chest pain with associated dyspnoea and diaphoresis. She reported an argument with her brother the previous day. She did not have any traditional cardiovascular risk factors; however, there was a strong family history of SCD that affected three female members in their 40s and 50s. She also had a prior history of anxiety and panic attacks. Her current medications included an intrauterine device releasing Levonorgestrel.

Her physical examination was unremarkable and her electrocardiogram (ECG) normal. Cardiac troponins were elevated [high-sensitivity troponin I 780 ng/L (normal range <16 ng/L)]. Her echocardiogram showed good ejection fraction and no wall motion abnormalities. A diagnosis of non-ST-segment elevation myocardial infarction was made, and a coronary angiogram (CAG) was performed. The CAG showed a moderate to severe stenosis of the proximal circumflex coronary artery (LCx), with good blood flow into the distal vessel (*Figure 1*). The remaining coronary segments, as well as other epicardial arteries, appeared normal but very tortuous; the appearance was suggestive of Type 2A SCAD. Optical coherence tomography (OCT) confirmed an intramural haematoma (IMH) without communication between true and false lumen (*Figure 2*). No intimal tear was identified. Despite significant stenosis in the proximal segment of a relatively large artery, a decision was made not to proceed with percutaneous coronary intervention (PCI). The patient was commenced on dual antiplatelet therapy (aspirin and clopidogrel) for 3 months; this was followed by a long-term aspirin therapy, as well as metoprolol and cilazapril. She remained clinically stable and was subsequently discharged from hospital.

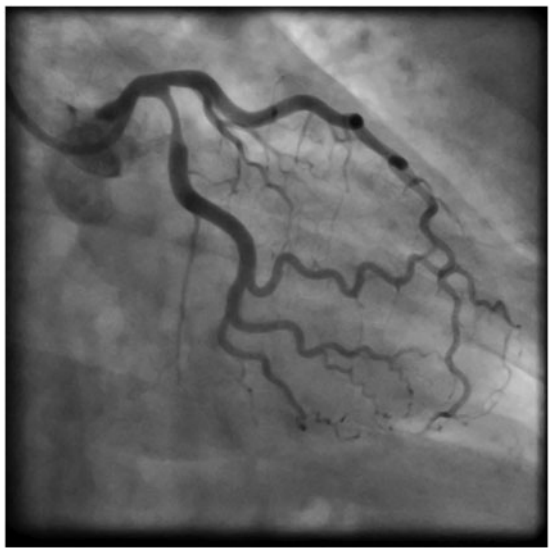

Figure 1 Coronary angiography demonstrating spontaneous coronary artery dissection of the proximal circumflex coronary artery.

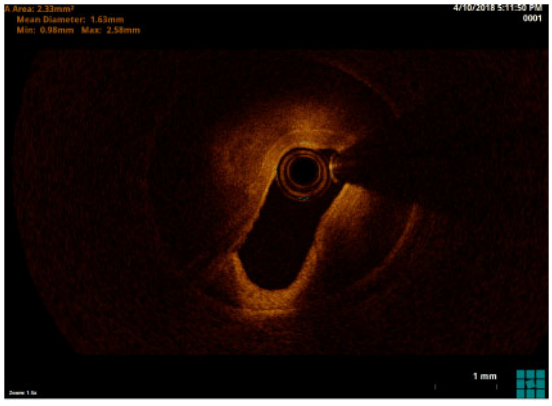

Figure 2 Optical coherence tomography showing a circular haematoma of the proximal circumflex coronary artery.

Eleven weeks following the index event a repeat CAG was performed, demonstrating completely normal coronaries (*Figure 3*). There was no evidence of remaining lumen reduction in proximal LCx.

Given the patient's family history of SCD, further testing was arranged. A 12 lead ECG was normal with no evidence of QT prolongation, ventricular pre-excitation, or arrhythmia. Cardiac magnetic resonance imaging (MRI) was performed and did not show any features of an inheritable cardiomyopathy or ischaemic scar. Genetic counselling and subsequent gene testing were performed for known

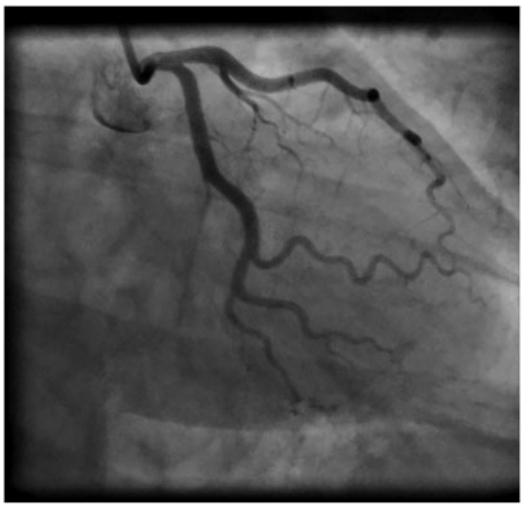

Figure 3 Coronary angiography demonstrating normal proximal segment of the circumflex coronary artery.

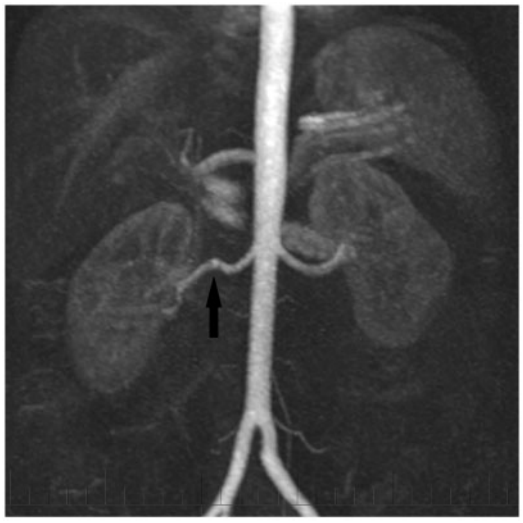

Figure 4 Magnetic resonance imaging demonstrating focal area of irregularity of the right renal artery (arrow) suggestive of fibromuscular dysplasia.

variants associated with aortopathy and SCD and were unremarkable. Magnetic resonance imaging of the renal arteries showed a focal area of irregularity of the right renal artery suggestive of fibromuscular dysplasia (FMD) affecting the right renal artery (*Figure 4*).

Discussion

Spontaneous coronary artery dissection has gained attention as an important cause of acute coronary syndrome and SCD among women.[2] It has unique risk factors, such as female sex, pregnancy, and postpartum period. An association with exogenous female sex hormones has also been reported. Commonly described precipitants include extreme physical and emotional stress. Underlying arteriopathies (particularly FMD) and systemic inflammatory diseases also contribute to a risk of SCAD.[1] Three angiographic types of SCAD have been proposed: Type 1, with an apparent dissection flap and linear filling defect; Type 2, with a presence of a smooth stenosis that might be bordered by normal segments (Type 2A), or extending to the tip of the coronary artery (Type 2B); and type 3, which mimics atherosclerotic lesion. Coronary angiogram also identifies patients with increased vessel tortuosity, which is a potential SCAD risk factor.[3] The main limitation of CAG is that it does not visualize the vessel wall. Therefore, an IMH may not be apparent on angiography.[2] Additional use of intracoronary imaging techniques facilitates the diagnosis of SCAD in uncertain cases. Both intravascular ultrasound (IVUS) and OCT allow for the identification of IMH and intimal disruption. In particular, OCT provides high-resolution images leading to diagnostic certainty and is particularly useful for such an indication.[2,4] However, OCT requires a firm contrast injection, which poses a risk of hydraulic worsening of dissection; comparatively, IVUS does not require contrast and is widely available.[2]

Questions remain regarding optimal interventional and medical management of SCAD. Currently, a conservative management strategy is recommended as the great majority of SCAD lesions (70–97%) heal naturally through haematoma resorption, whereas PCI is associated with increased risk of complications and adverse outcomes. Additionally, SCAD occurs most often in distal vessels, which poses a technical limitation to PCI.[5]

Implantation of long coronary stents, to prevent the propagation of IMH when compressed by the stent, has been described as a successful approach.[1] However, this approach might increase the risk of restenosis or stent thrombosis caused by stent strut malapposition after resorption of haematoma. Alternative interventional strategies with successful outcomes are balloon angioplasty alone, to restore coronary flow, or cutting balloon fenestration of the IMH to decompress the false lumen. Using a multi-stent approach to seal the distal and proximal end of SCAD, and the use of bioresorbable stents have also been proposed.

Current recommendations favour PCI or revascularization by means of coronary artery bypass surgery in case of haemodynamic instability, ongoing ischaemia or left main stem SCAD, emphasizing the necessity of an individual approach to each patient.[1]

Spontaneous coronary artery dissection identified in our patient posed a therapeutic dilemma. The severity of the stenosis in the proximal segment of a relatively large coronary artery, causing MI and good availability of the lesion, were factors that prompted us to treat the lesion with PCI. However, the awareness of the increased risk of complications, particularly extension of the dissection proximally to the left main stem and distally, resulted us in adopting a 'hands off' approach. Our patient was haemodynamically stable with no signs of ongoing ischaemia, which motivated our decision of conservative management. If the coronary flow had been threatened, we would have also considered using glycoprotein IIb/IIIa inhibitors. However, the role of these inhibitors in SCAD management is controversial

due to increased bleeding risk and therefore the potential risk of worsening the IMH.

The repeat CAG 11 weeks after the index event demonstrated complete angiographic 'healing' of the dissection, this supports the decision for conservative management.

Whether SCAD is associated with the sudden death events in our patient's family remains unclear. The history of sudden death affecting relatively young women in this family certainly raises concerns as to an inheritable condition and testing in our patient has not identified an alternate explanation. Our patient does have MRI features suggesting FMD involving the right renal artery; this condition has been associated with SCAD, but the genetics of both conditions are not well understood. A recent series identified approximately 8% of SCAD patients with a molecularly identifiable disorder associated with vascular disease (mostly Ehlers–Danlos syndrome).[6] Unfortunately, further information including post-mortem findings among the patient's family members was not available, and in the absence of such information alongside genetic markers we can only speculate about this representing a possible inheritable form of SCAD.

Lead author biography

Aleksandra Pineda graduated from the Medical University of Lodz (Poland) and subsequently underwent her specialty training in Internal Medicine and Cardiology in Germany. Currently, she is doing a fellowship in Interventional Cardiology at Christchurch Hospital (New Zealand).

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.

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