

# Multi-vessel spontaneous coronary artery dissection in a patient with aortic dissection: a case report

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Received 28 December 2021; first decision 3 February 2022; accepted 22 June 2022; online publish-ahead-of-print 28 June 2022

## Background

Spontaneous coronary artery dissection (SCAD) is a rare cause of acute coronary syndrome that is often misdiagnosed.

## Case summary

We describe a case of multi-vessel SCAD in a 73-year-old patient with no evidence of fibromuscular dysplasia that is presented with Type A aortic dissection after undergoing an ascending aorta and aortic arch replacement with stent placement in the abdominal aorta. The use of percutaneous coronary intervention with cutting balloons and drug-eluting stent implantation helped wean the patient off extracorporeal membrane oxygenation successfully.

## Discussion

To our knowledge, this is the first reported case of multi-vessel SCAD presenting concomitantly with aortic dissection. More research is needed to help understand the pathophysiology of the two conditions as well as possible links between them.

## Keywords

Case report • Spontaneous coronary artery dissection • Aortic dissection • Connective tissue disease • Percutaneous coronary intervention • Cutting Balloon

**ESC Curriculum** 3.1 Coronary artery disease • 3.2 Acute coronary syndrome • 7.1 Haemodynamic instability

## Learning points

- Multi-vessel spontaneous coronary artery dissection (SCAD) can occur in extreme cases of physical stress like aortic dissection and could be related to connective tissue disorders.
- When patients with SCAD are hemodynamically unstable, percutaneous coronary intervention (PCI) is indicated.
- Often using a cutting balloon to relieve the intramural haematoma can facilitate PCI and prevent proximal/distal propagation.

## Introduction

Spontaneous coronary artery dissection (SCAD) is a rare cause of the acute coronary syndrome (ACS). It is defined as the separation of the layers of a coronary artery with or without intimal disruption

(intramural haematoma) in the absence of trauma, atherosclerosis, or iatrogenic cause.<sup>1</sup> Despite great advances in understanding its pathophysiology, it continues to be underdiagnosed and mismanaged. In this case report, we describe a rare occurrence of aortic dissection associated with multi-vessel SCAD.

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Handling Editor: Andrew Peter Venezis

Peer-reviewers: Amr Idris; Andreas Mitsis

Compliance Editor: Polyvios Demetriades

Supplementary Material Editor: Fabienne Vervaat

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## Timeline

Day 0 (12:35 p.m.)	Admitted to hospital—diagnosed with Type A aortic dissection
Day 0 (8 p.m.)	Emergency surgery
Day 0 (10 p.m.)	Unable to wean off cardiopulmonary bypass—VA-ECMO started
Day 0 (10 p.m.)	Echo—new apical regional wall abnormality
Day 0 (10:30 p.m.)	Urgent coronary angiogram—multi-vessel Type 2 SCAD
Day 0 (11 p.m.)	PCI to proximal-mid LAD with cutting balloon and stenting, other vessels managed conservatively
Day 3	Decannulated from VA-ECMO
Day 7	Transferred from ICU to ward
Day 30	Discharged home
Month 6	Clinic follow-up. Patient remains well. No cardiac symptoms. Follow-up imaging showed a stable appearance of the repaired aorta
Month 10	Follow-up angio showed patent LAD stent with mild in-stent restenosis and complete resolution of LCx and RCA SCAD. Echo showed normal ejection fraction (EF) and new severe central mitral regurgitation

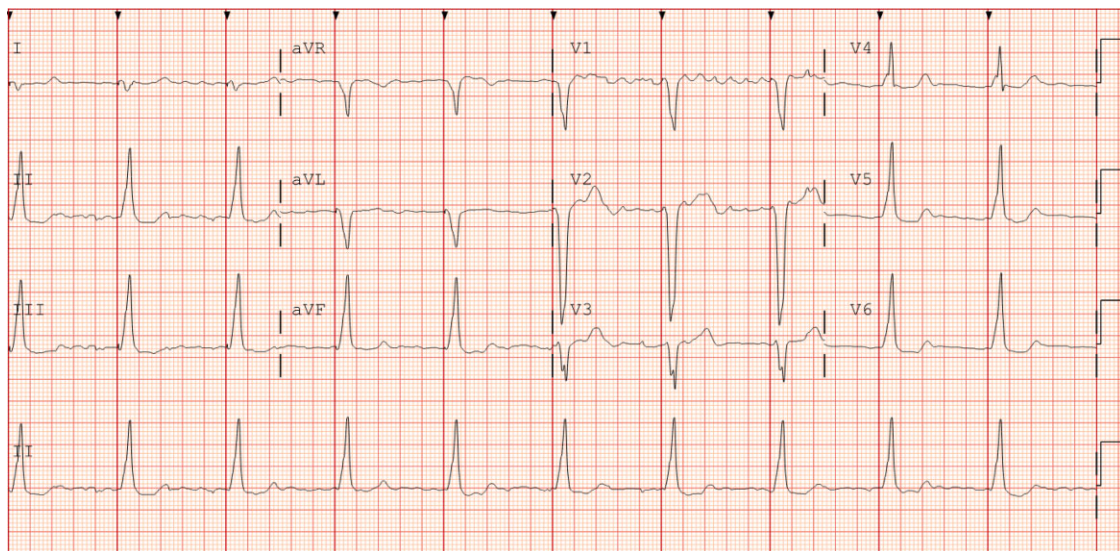
## Case report

A 73-year-old female presented to the hospital with sudden onset of severe stabbing chest pain radiating to the back. Her past medical history included hypertension, dyslipidaemia, transient ischaemic

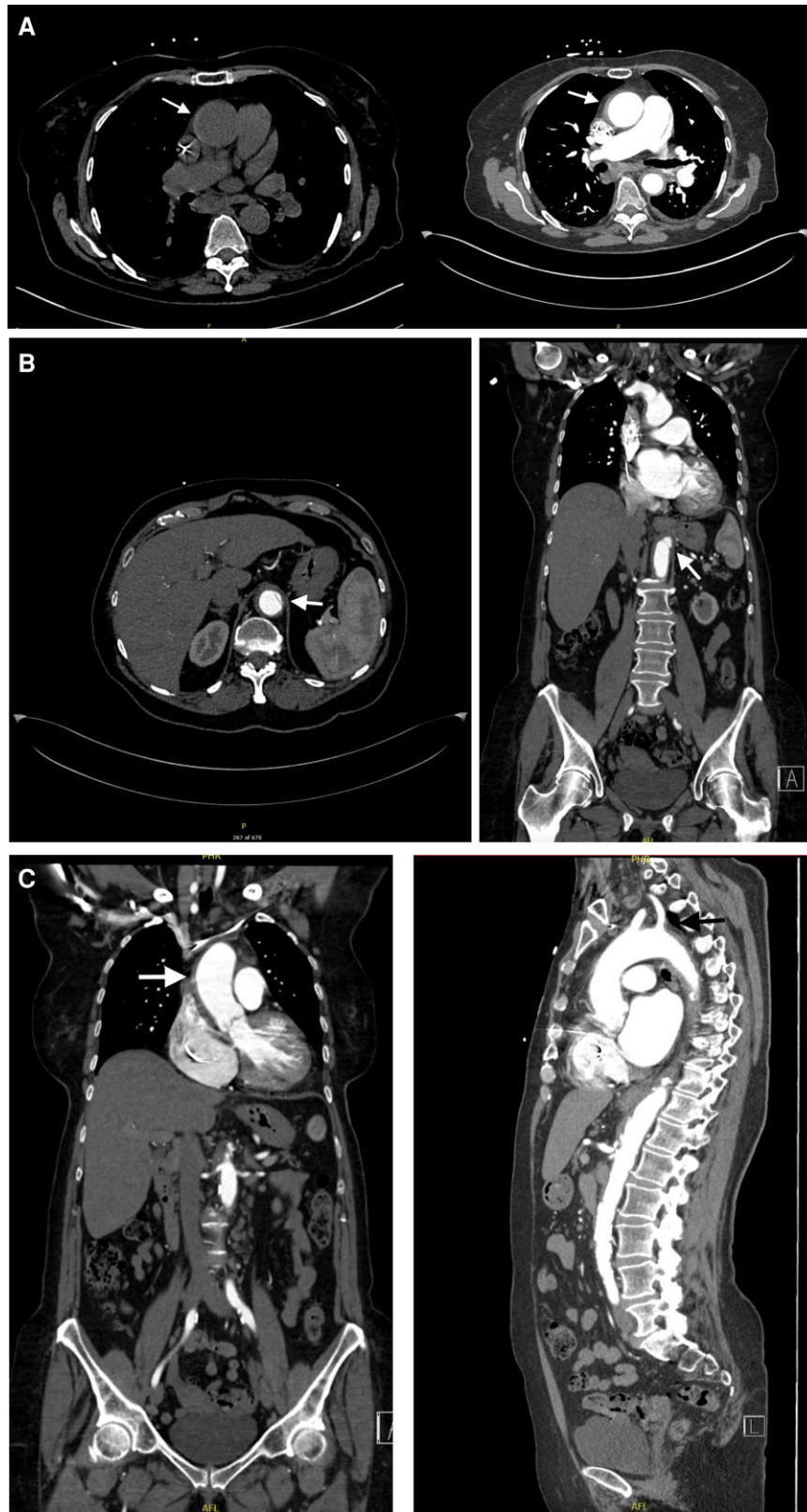
attacks with minor deficits (mild short memory deficits and gait instability), atrial fibrillation requiring AV nodal ablation and a VVI single chamber pacemaker, there was no family history of aortopathies or any other familial disorders. On arrival at the emergency department, she was found to be hemodynamically stable without a significant arm blood pressure difference or any new neurological findings. Her echocardiogram (ECG) showed a ventricular-paced rhythm (Figure 1). Because of the classic characteristics of her pain (sudden, tearing, and radiating to the back), an acute aortic syndrome was suspected and an emergent computed tomography (CT) angiogram of the chest, abdomen, and pelvis was performed which showed an extensive type A (DeBakey Type I) intramural haematoma in circumferential contiguous fashion from a level just beyond the aortic valve, extending distally to a level between the take-off of the renal artery and the inferior mesenteric artery with a focal non-flow-limiting dissection flap anteriorly just below the level of oesophageal hiatus (Figure 2), which could have been the entry site tear. There was no visible site of entry tear observed in the ascending aorta with all major branches being filled by the true lumen.

The patient was noted to have a normal Rubidium positron emission tomography perfusion 18 months prior to her presentation which was done to investigate exertional chest pain. She was rushed to the operating room and underwent a valve-sparing replacement of the ascending aorta and the aortic hemiarch with the placement of a distal aortic stent (frozen elephant trunk) with resuspension of the aortic valve. Cardiopulmonary bypass weaning was unsuccessful, and she was found to have a new apical regional wall motion abnormality (echo images available in [Supplementary material online](#)). Central veno-arterial extracorporeal membrane oxygenation was started, and the patient was transferred to the intensive care unit.

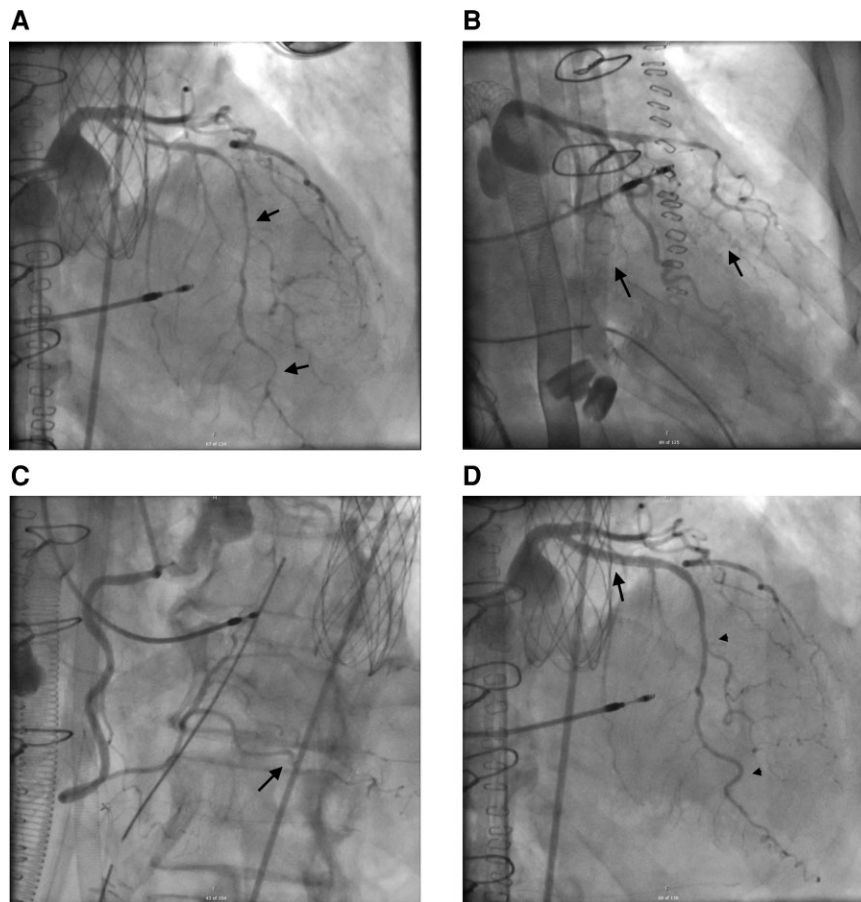
She was then taken to the cardiac catheterization lab for an urgent coronary angiogram. This was done through a right femoral approach and showed the angiographic appearance of type II



**Figure 1** 12 Lead ECG showing atrial fibrillation with ventricular-paced rhythm.



**Figure 2** (A) Unenhanced (left) and enhanced (right) axial CT view showing radio-opaque appearance of intramural haematoma within the wall of the ascending aorta (arrows). (B) Axial (left) and coronal view (right) showing entry site (dissection flap) in the abdominal aorta (arrows). (C) Coronal (left) view showing once again radio-opaque appearance of intramural haematoma in the ascending aorta without an entry site. Sagittal (right) view showing head and neck vessels arising from the true lumen (arrow)



**Figure 3** (A) Posteroanterior (PA) cranial view of the left coronary artery showing type II SCAD of the LAD (arrows). (B) Right anterior oblique (RAO) caudal view showing SCAD of the distal circumflex and ramus branch (arrows) (C) PA Cranial view of the right coronary artery showing type II SCAD of the distal RCA (arrow). (D) PA Cranial view of the left coronary artery (LCA) following LAD intervention with DES (arrow) showing excellent angiographic results with restoration of the normal calibre of the LAD (arrowhead).

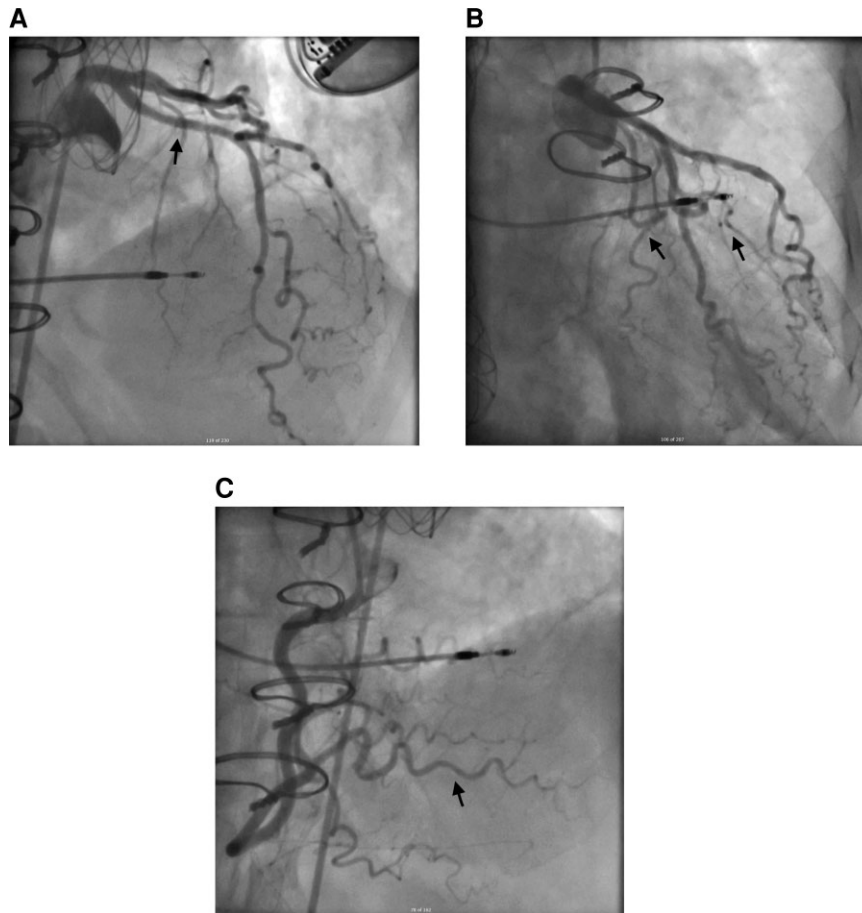
spontaneous coronary artery dissection (SCAD) of the proximal to distal left anterior descending artery (LAD), distal left circumflex artery (LCx), branch of ramus intermedius artery and distal right coronary artery (RCA) (Figure 3; cine clips available in [Supplementary material online](#)). Of note, the patient was not on any vasoactive agents at the time of the angiogram, which can cause intense diffuse vasospasm mimicking this appearance. A 2.50 × 10 mm FLEXTOME MONORAIL cutting balloon was inflated in the proximal and mid LAD to relieve the intramural haematoma followed by implantation of a 3.00 × 20 mm PROMUS ELITE drug-eluting stent, which yielded an excellent angiographic result (Figure 3). The rest of the vessels were treated conservatively. Intravascular imaging was not used given the typical angiographic appearance and the critical patient condition. Three days later, she was taken back to the OR and decannulated from ECMO. Fibromuscular dysplasia screening with CT angiogram of the head and neck and CT angiogram of the abdomen and pelvis were negative. She was discharged to inpatient rehab a week later where she underwent physiotherapy and occupational therapy before going home a month later; 10 months after discharge, she was complaining of shortness of breath on exertion and underwent a

follow-up angiogram, which showed a patent LAD stent with mild in-stent restenosis and complete resolution of the LCx and RCA SCAD (Figure 4; cine images in the [Supplementary material online](#)). An ECG showed a normal ejection fraction (60%) with normal LV dimensions and thickened mitral valve leaflets with new severe central mitral regurgitation and a follow-up CT chest showed a stable appearance of the repaired aorta ([Supplementary material online](#)).

## Discussion

The arterial wall is formed of three layers; the single-cell layer lining endothelium called intima, the muscular media, and the outer connective tissue layer called the adventitia. It receives its own blood supply through small capillaries that run within these layers called 'Vasa Vasorum'. It is believed that the rupture of vasa vasorum is the main pathophysiology behind SCAD, particularly in cases of intramural haematoma.<sup>2</sup> SCAD has four different angiographic classifications: Type 1 (pathognomonic) where there is a clear double-lumen seen on angiography, Type II (most common) has no dissection flap





**Figure 4** (A) PA cranial view of the LCA showing mild in-stent restenosis of the LAD stent (arrow). (B) RAO Caudal view of the LCA showing resolution of LCx and Ramus branch SCAD. (C) PA Cranial view of the RCA showing resolution of the distal RCA SCAD.

and is characterized by a long segment of diffuse narrowing which may (Type IIB) or may not (Type IIA) reach the terminal end of the vessel, Type III mimics atherosclerosis, and Type IV with complete occlusion of the vessel.

SCAD is predominantly a disease of young females with no risk factors for atherosclerotic coronary disease;<sup>3</sup> however, it can infrequently affect older patients like in our case.

The association of fibromuscular dysplasia with SCAD is well described in the literature,<sup>4,5</sup> other conditions that have been linked to it include multiparity, tortuous coronaries, and emotional and physical stress.<sup>6,7</sup> SCAD in patients with connective tissue disorders like Marfan, Loeys–Dietz, and Ehlers–Danlos syndromes—which commonly predispose to aortic syndromes—is not common and usually presents at an early age.<sup>8,9</sup> The reported frequency of the combination of SCAD and inherited connective tissue disorders is less than 5%, but there are sporadic case reports in the literature.<sup>10–13</sup> Verstraeten *et al.* found that around 1 in 10 patients with SCAD will have a rare variant in genes known to be associated with aortic aneurysms and dissection; particularly, genes associated with Loeys–Dietz syndrome were more prevalent.<sup>14</sup>

SCAD usually affects one coronary artery, and most commonly affects the LAD. Lempereur reported the first case of multi-vessel

SCAD in an FMD patient with ACS which was thought to have been triggered by severe mental stress.<sup>15</sup> To our knowledge, this is the first case of extensive multi-vessel coronary dissection associated with aortic dissection. The pathophysiology of both conditions has not been directly linked. It is possible that the immense physiologic stress of the aortic dissection and the operation caused the predisposed fragile coronaries of our patient to have extensive multi-vessel SCAD. It is less likely but possible that an underlying connective tissue disorder has gone undiagnosed in this elderly patient.

A CT angiogram of the whole aorta is recommended when suspecting an acute aortic syndrome.<sup>16,17</sup> This usually allows visualization of the proximal coronary arteries, and if gated properly, can further assess the rest of the coronary arteries; however, the image quality would not allow distinguishing SCAD from other forms of ACS. It is recommended that cases that present with ECG changes suggestive of ACS be treated as such and management should not be delayed to obtain a CT unless the patient is known to have a genetic syndrome that strongly predispose to aortic dissection like Loeys–Dietz syndrome.<sup>16</sup> Guidelines also recommend that patients with Loeys–Dietz syndrome or other genetic mutations predisposing to dissection get complete aortic imaging at the initial diagnosis and six months after to ensure there is no rapid growth of aorta

prompting intervention. There is no particular relevance to SCAD in the current workup recommendations for aortopathy workup.

Due to the relatively low success rate and the higher complication risk of PCI, most SCAD cases are managed conservatively with medical therapy.<sup>3</sup> However, when SCAD is thought to be the cause of hemodynamic instability or ongoing ischaemia, revascularization is recommended.<sup>1,18</sup> The use of cutting balloons with or without stenting has been reported to be beneficial in such cases.<sup>19</sup> We planned to implant a DES in the proximal LAD after using a cutting balloon in three different locations in the mid to proximal LAD. The other vessels were too distal and/or small for intervention. The rationale for using a cutting balloon is to relieve the luminal compression by the false lumen and to prevent the proximal propagation of the haematoma to the left main coronary artery. The excellent angiographic result and the rapid turnaround in the patient's clinical condition support the experts' opinion recommendation of intervention in such cases.

Diagnostic uncertainty is an important limitation of our case report. Although intracoronary imaging would have been helpful to confirm the diagnosis, we adopted an approach of treatment first and an image if needed given the patient's critical condition. The drastic improvement of flow following cutting balloon treatment supports the diagnosis of SCAD.

Our case illustrates a rare presentation of multi-vessel SCAD in a patient with extensive aortic dissection. No evidence for FMD was found in this patient, and it is unlikely but possible that she has an underlying connective tissue disorder. PCI with cutting balloon and stenting of the proximal LAD successfully helped wean the patient off ECMO. Follow-up angiography several months later revealed complete resolution of the LCx and RCA SCAD. Follow-up ECG showed a normal ejection fraction with new severe central mitral regurgitation.

## Conclusions

Multi-vessel SCAD can occur in extreme cases of physical stress like in our case of aortic dissection. More studies are needed to explore possible links between aortic dissection and SCAD and the utility of revascularization in high-risk cases.

## Lead author biography



Motasem Alyamani is an upcoming fellow of the Mass General Brigham (MGB) Complex Coronary Intervention programme starting July 2022. In 2012, he received his medical degree from Taibah University, Madinah, Saudi Arabia. He completed his Internal Medicine training at the University of Toronto and his cardiology and interventional cardiology training at the University of Alberta.

He is interested in conducting meta-analyses and investigating ways to reduce reperfusion injury in ST-Elevation Myocardial Infarction

(STEMI). He enjoys spending time with his family, reading novels, and watching movies in his free time.

## Supplementary material

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

## Acknowledgements

The authors would like to recognize the physicians, nurses, and other allied healthcare workers who were responsible for the care of this patient.

**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 of the case report including images and associated text has been obtained from the patient in line with COPE guidance.

**Conflicts of interest:** None declared.

**Funding:** None declared.

**Ethics approval:** Our institution does not require ethical approval for reporting individual cases or case series.

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