Spontaneous coronary artery dissection, fibromuscular dysplasia, and biventricular stress cardiomyopathy: a case report

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Received 12 August 2021; first decision 30 November 2021; accepted 18 March 2022; online publish-ahead-of-print 25 March 2022

Background
Spontaneous coronary artery dissection (SCAD) is an important cause of acute coronary syndrome and is associated with fibromuscular dysplasia (FMD). The diagnosis of stress cardiomyopathy in patients with SCAD and FMD is uncommon, though an important consideration given the shared risk profile. Complications of severe left ventricular (LV) dysfunction associated with stress cardiomyopathy, such as LV thrombus, complicate the management of SCAD where anticoagulation is controversial in the context of SCAD-associated intramural haematoma.

Case summary
A 65-year-old female presented with non-ST elevation myocardial infarction with a recent diagnosis of hypertension but no other traditional cardiovascular risk factors. There was, however, a family history of early cardiac death from myocardial infarction affecting her mother. Echocardiography demonstrated severe biventricular dysfunction with circumferential akinesis of the mid to apical segments. Coronary angiography demonstrated type 2A SCAD involving the first diagonal artery. Cardiac magnetic resonance imaging (MRI) confirmed a diagnosis of stress cardiomyopathy with biventricular involvement, complicated by LV apical thrombus and a focal region of myocardial infarction. Vascular imaging confirmed the presence of FMD. Guideline-directed heart failure therapy in addition to clopidogrel and rivaroxaban was prescribed. Follow-up contrast echocardiography at six-weeks confirmed resolution of LV dysfunction and resolution of the LV thrombus with no adverse events.

Discussion
The dual diagnosis of SCAD and stress cardiomyopathy is uncommon. Cardiac MRI was useful for confirming the diagnosis of stress cardiomyopathy and the presence of LV thrombus, where anticoagulation may complicate the management of intramural haematoma in patients with concomitant SCAD and FMD.

Keywords
Spontaneous coronary artery dissection • Fibromuscular dysplasia • Takotsubo stress cardiomyopathy • Cardiac magnetic resonance imaging • Echocardiography • Case report

ESC Curriculum
2.2 Echocardiography • 2.3 Cardiac magnetic resonance • 3.2 Acute coronary syndrome • 6.5 Cardiomyopathy
Learning points

- Whilst the association between spontaneous coronary artery dissection (SCAD) and fibromuscular dysplasia (FMD) is well documented, stress cardiomyopathy may also occur concomitantly and shares a similar risk factor profile.
- Cardiac magnetic resonance imaging is useful for confirming the diagnosis of stress cardiomyopathy and may identify high-risk features (i.e. left ventricular thrombus, right ventricular involvement or outflow-tract obstruction).
- Extended vascular investigation in patients with SCAD with contrast enhanced magnetic resonance angiography or computed tomography angiography may identify other vascular abnormalities such as FMD or cerebral aneurysms.
- Use of anticoagulation (i.e. for left ventricular thrombus) remains controversial in cases of SCAD due to theoretical concerns for extension of intramural haematoma.

Introduction

Non-atherosclerotic presentations of myocardial infarction, such as spontaneous coronary artery dissection (SCAD) are increasingly recognized, particularly in women and in those with few traditional cardiovascular risk factors. The association between SCAD and arteriopathies such as fibromuscular dysplasia (FMD) is well described, and extended vascular screening is recommended in patients who present with SCAD.1 The concomitant presentation of SCAD and stress cardiomyopathy (i.e. Takotsubo) has also been described, and the two entities are thought to share a number of similar features, including patient demographics, predisposing factors, and clinical presentation.2,3 We present a case of concomitant SCAD, FMD, and stress cardiomyopathy, complicated by left ventricular (LV) thrombus.

Timeline

| Day 1: | Patient presents with non-ST elevation myocardial infarction. Managed as acute coronary syndrome. Severe left ventricular dysfunction identified on transthoracic echocardiography (ejection fraction 28%), appearances suggestive of biventricular stress cardiomyopathy. |
| Day 2: | Coronary angiography demonstrated severe stenosis of the first diagonal, with appearances consistent with intramural haematoma and type 2 spontaneous coronary artery dissection. Left ventriculogram demonstrated apical akinesis, inconsistent with isolated diagonal coronary artery territory. |
| Day 4: | Computed tomography (CT) renal angiogram confirming fibromuscular dysplasia involving external iliac arteries and left renal artery. |
| Day 5: | Cardiac magnetic resonance imaging confirmed diagnosis of stress induced cardiomyopathy with myocardial oedema involving the mid to apical segments. Severe left ventricular dysfunction with akinesis of the right ventricular apex consistent with biventricular involvement. Focal infarct identified in the diagonal coronary artery territory. Left ventricular apical thrombus detected (8 × 8 × 6 mm). |
| Day 6: | Discharged from hospital on guideline-directed heart failure therapy, clopidogrel and rivaroxaban. |

Case presentation

A 65-year-old female presented to the emergency department having been woken from sleep with severe chest pain. She reported that she had recently been experiencing frontal headaches and had subsequently been diagnosed with hypertension confirmed by ambulatory blood pressure monitoring. First-line antihypertensive therapy with candesartan had been commenced. She had no other traditional cardiovascular risk factors, and there were no other physical or psychological stressors identified in the history. However, there was a family history of early cardiac death from myocardial infarction affecting her mother at 55 years of age. Her other past medical history included right nephrectomy for renal cell carcinoma.

Initial physical assessment was unremarkable; she was normotensive with no clinical signs of cardiac failure. Her electrocardiogram demonstrated high-lateral T-wave changes with normal corrected QT interval (Figure 1A). High-sensitivity cardiac troponin I was elevated (peak 6807 ng/L; normal range <10 ng/L), and a diagnosis of non-ST elevation myocardial infarction was made. Anticoagulation with unfractionated heparin and dual antiplatelet therapy with aspirin and clopidogrel were initiated. Transthoracic echocardiography demonstrated severe LV dysfunction with hyperkinetic basal segments and circumferential akinesia of the mid to apical segments (Figure 2, Video 1). Akinesia of the mid to apical right ventricular segments were also noted suggestive of biventricular stress cardiomyopathy (Video 1). The patient proceeded to selective coronary angiography (Figure 3A) that identified severe stenosis of the distal first diagonal branch, with an appearance typical for intramural haematoma and type 2A SCAD. The remaining coronary arteries were angiographically normal. Left ventriculography demonstrated extensive apical and mid-wall regional wall motion abnormalities extending beyond the vascular territory of the affected coronary artery territory.

Learning points

- Whilst the association between spontaneous coronary artery dissection (SCAD) and fibromuscular dysplasia (FMD) is well documented, stress cardiomyopathy may also occur concomitantly and shares a similar risk factor profile.
- Cardiac magnetic resonance imaging is useful for confirming the diagnosis of stress cardiomyopathy and may identify high-risk features (i.e. left ventricular thrombus, right ventricular involvement or outflow-tract obstruction).
- Extended vascular investigation in patients with SCAD with contrast enhanced magnetic resonance angiography or computed tomography angiography may identify other vascular abnormalities such as FMD or cerebral aneurysms.
- Use of anticoagulation (i.e. for left ventricular thrombus) remains controversial in cases of SCAD due to theoretical concerns for extension of intramural haematoma.
artery (Figure 3B, Video 2). Serial electrocardiography demonstrated progressive wide-spread deep T-wave inversion, suggestive of a stress cardiomyopathy (Figure 1B).

Cardiac magnetic resonance imaging (MRI) demonstrated severe LV dysfunction (ejection fraction [EF] 29%) with circumferential akinesis of the mid to apical segments. Right ventricular EF was normal at 49%, with akinesis of the apical segments. Tissue characterization with native T1 mapping and T2-weighted imaging demonstrated circumferential myocardial oedema of the mid-wall and apical segments consistent with myocardial oedema (Figure 4A). Near transmural late Gadolinium enhancement (LGE) in a subendocardial pattern was noted on post-contrast imaging in the apical lateral and apical inferior segments consistent with myocardial infarction (Figure 4B). The degree of myocardial oedema was disproportionate to the territory of infarction, supporting a concomitant diagnosis of stress cardiomyopathy. A non-enhancing filling defect in the LV apex was noted consistent with intracardiac thrombus.

A computed tomography (CT) renal angiogram was arranged to screen for systemic arteriopathy and demonstrated FMD involving both external iliac arteries and the left renal artery (Figure 5A). Subsequent CT angiography of the head and neck vessels confirmed FMD of the internal carotid arteries (Figure 5B) with otherwise unaffected intracranial vessels and excluded intracerebral aneurysms.

On patient preference, rivaroxaban 15 mg was initiated in combination with clopidogrel 75 mg daily. Candesartan and metoprolol succinate were progressively up-titrated to maximally tolerated doses. Following discharge, the patient represented with headache and hypertension, but no significant abnormalities such as intracranial bleeding or vascular abnormalities (i.e. intracranial aneurysms) were detected on CT brain imaging.

Repeat 2D (Video 3) and contrast enhanced transthoracic echocardiography was performed at 6 weeks, demonstrating recovery of the stress cardiomyopathy (LV EF 56% with normal right ventricular size and function). No residual LV thrombus was identified. There were no adverse bleeding events on therapy, no thrombo-embolic events, and no clinical features to suggest reinfarction related to propagation of intramural haematoma.

Figure 1 Electrocardiography. Isolated high-lateral T-wave inversion on presentation (A). Diffuse T-wave inversion and QTc prolongation at 48 h consistent with stress cardiomyopathy (B).
Figure 2. Echocardiography. Akinesis of the mid-wall and apical segments with hyperdynamic basal function in the apical four-chamber (A) and apical two-chamber (B) views at end-diastole (left) and end-systole (right). Global longitudinal strain is reduced (7.6%) with apical akinesis extending to the mid-wall segments (C).

Video 1. Echocardiography. Apical four-chamber view demonstrating akinesis of the mid-wall and apical left ventricular segments with hyperdynamic basal function. Right ventricular involvement is also seen.
The identification of SCAD as a non-atherosclerotic cause of acute coronary syndrome is increasingly common, accounting for up to 4% of presentations and features prominently in current international position statements. Early identification of intramural haematoma is important as selective angiography and percutaneous coronary intervention confer a greater risk of iatrogenic complications in patients with SCAD. As such, a conservative approach is often preferred due to the risk of haematoma propagation with coronary instrumentation, the typical self-limiting course, and expected resolution of coronary dissection.

Both SCAD and stress cardiomyopathy share several risk factors. They most commonly affect women and are often precipitated by emotional or physical stress. Both have a predilection for involvement of the left anterior descending artery, and selective angiography is often required to delineate between acute coronary syndrome associated with SCAD, plaque rupture, and stress cardiomyopathy. The concomitant diagnosis of SCAD and stress cardiomyopathy has previously been described; however, evidence-based management is limited, particularly in the setting of intracardiac thrombus. A retrospective survey where stress cardiomyopathy was the provisional diagnosis found that angiographically proven SCAD co-existed in 2.5% of cases, with a further 9% being indeterminate. In this case, stress cardiomyopathy was temporally related to myocardial infarction secondary to SCAD. In the absence of other precipitants, the infarct was the presumed trigger for stress cardiomyopathy.

The detection of SCAD by angiography can be challenging; though intracoronary imaging techniques are particularly useful, they may not be technically possible in small branch vessels and are not widely available. Angiography in this case confirmed type 2A SCAD of the first diagonal branch artery, and the extent of the regional wall motion abnormalities on ventriculography and echocardiography was disproportionate to the vascular territory of the limited coronary dissection. Multimodality imaging was particularly useful in this case. Due to its high spatial resolution and parametric mapping techniques, cardiac MRI helped confirm the concomitant diagnoses of SCAD with associated focal infarct, and stress cardiomyopathy and with right

**Discussion**

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**Figure 3** Coronary angiography. Type 2A spontaneous coronary artery dissection of the first diagonal artery (A). Left ventriculogram demonstrating wall motion abnormalities consistent with Takotsubo cardiomyopathy (B).

**Video 2** Left ventriculogram. Findings consistent with stress cardiomyopathy.
ventricular involvement, which may suggest worse prognosis. The pattern of LGE identified the region of myocardial infarction—a technique that may improve SCAD detection rates when correlated to angiographic findings. Finally, cardiac MRI is more sensitive than echocardiography for detection of LV thrombus, which is important as thrombus may occur in 1.3–5.3% of stress cardiomyopathies even in those on anticoagulation therapy. Extended vascular screening with CT angiography identified features of FMD. Importantly, this did not demonstrate evidence of intracerebral aneurysms that can affect as many as 14% of patients with FMD.

Treatment of intracardiac thrombus requires special consideration in patients with LV thrombus and SCAD. Empiric anticoagulation is often initiated in patients with SCAD or stress cardiomyopathy at the time of initial presentation as management for presumed atherosclerotic acute coronary syndrome. However, cessation of anticoagulation following diagnosis is often recommended in SCAD due to theoretical risks of intramural haematoma extension. The management of cases requiring anticoagulation is not well supported by current guidelines. This case of SCAD and stress cardiomyopathy complicated by LV thrombus required extended duration anticoagulation, which was combined with antiplatelet therapy. This poses several theoretical risks for potential propagation of the intramural haematoma associated with SCAD and increased bleeding risks in those with FMD who have associated vascular abnormalities such as intracerebral aneurysms or carotid dissection. Though warfarin was offered, a direct oral anticoagulant was prescribed on patient request. Though data are limited, non-vitamin K-dependent anticoagulants appear safe and effective in the management of LV thrombus. A total of 3 months of anticoagulation was planned with a total of 12 months of clopidogrel therapy.

Figure 4 Cardiac magnetic resonance imaging. Four-chamber post-contrast imaging showing a non-enhancing filling defect in the left ventricular apex consistent with apical thrombus and near transmural late gadolinium enhancement in a subendocardial pattern involving the apical lateral segment consistent with myocardial infarction (A). Two-chamber image showing near transmural late Gadolinium enhancement of the apical inferior segment consistent with myocardial infarction (B). Native T1 (C) and T2 mapping (D) showing elevated values in the mid and apical segments extending beyond the zone of infarction consistent with myocardial oedema, a pattern seen in stress cardiomyopathy.
Figure 5  Computed tomography angiography. Bilateral beading of the external iliac arteries with mild fusiform dilation of the mid to distal left renal artery (A). Mild wall irregularity of the cervical internal carotid arteries (B). Findings consistent with fibromuscular dysplasia.

Video 3  Echocardiography. Apical four-chamber view demonstrating resolution of stress cardiomyopathy.
Follow-up contrast enhanced echocardiography at 6 weeks demonstrated resolution of stress cardiomyopathy and LV thrombus with no adverse clinical events on this therapy.

**Lead author biography**

Dr Penni Blazak graduated medicine with honours from The University of Queensland, Australia. She is a final year physician trainee planning to undertake specialist cardiology training. She is trained in Nuclear Medicine and has ongoing interest in cardiac imaging and heart failure.

**Supplementary material**

*Supplementary material* is available at the *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:** The authors confirm that written consent for submission and publication of this case report including imaging and associated text has been obtained from the patient in line with COPE guidance.

**Acknowledgements:** None declared.

**Funding:** None declared.

**Conflict of interest:** None declared.

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