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CASE REPORT

INTERMEDIATE

A Challenging Case of Extensive Spontaneous Coronary Artery Dissection

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ABSTRACT

The detection of spontaneous coronary artery dissection (SCAD) causing myocardial infarction is integral in pursuing the appropriate management. Our case posed a diagnostic challenge, with Takotsubo cardiomyopathy and coronary embolism among the potential differential diagnoses upon the initial presentation. Extensive propagation of spontaneous coronary artery dissection subsequently resulted in a significant challenge to management requiring surgical revascularization. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2020;2:1437–42) © 2020 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

HISTORY OF PRESENTATION

A 28-year-old female flight attendant without cardiac risk factors or a history of connective tissue disorders presented with substernal chest pain, shortness of breath, bilateral arm numbness, and emesis that started several hours after an international flight. En route to the hospital via ambulance, she was reportedly in atrial fibrillation. She was hemodynamically stable on arrival with a blood pressure of 107/71 mm Hg, a heart rate of 80 beats/min, and an oxygen saturation of 99% on room air. Cardiac, respiratory, and pulse examinations were unremarkable. Although she cited work-related stressors, there were no clear physical or emotional triggers preceding presentation. She additionally denied any antecedent infectious symptoms.

LEARNING OBJECTIVES

- SCAD is a common etiology of ACS in young women and can result in significant morbidity and mortality if not recognized in a timely manner.
- Repeat angiography should be pursued in individuals if clinical suspicion exists for worsening ischemia in the context of myocardial infarction with nonobstructive coronary arteries without clear etiology.
- Although there may be a potential association between SCAD and TTC, SCAD should be carefully excluded when the diagnosis of TTC is entertained because the management strategy may be different. CMR can be very valuable in characterizing the myocardial pathology.

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ABBREVIATIONS AND ACRONYMS

ACS = acute coronary syndrome

CABG = coronary artery bypass graft

CMR = cardiac magnetic resonance

ECG = electrocardiogram

LAD = left anterior descending artery

LGE = late gadolinium enhancement

LM = left main

MI = myocardial infarction

MINOCA = myocardial infarction with nonobstructive coronary arteries

SCAD = spontaneous coronary artery dissection

TTC = Takotsubo cardiomyopathy

PAST MEDICAL HISTORY

Before presentation, the patient reported good health with satisfactory employment-mandated routine physical examinations. She denied a history of smoking, alcohol, or illicit drug use. She had never been pregnant and was not on hormone supplementation.

DIFFERENTIAL DIAGNOSES

A wide differential was considered by the admitting team, including pulmonary embolism, acute coronary syndrome (ACS), and pericarditis/myocarditis.

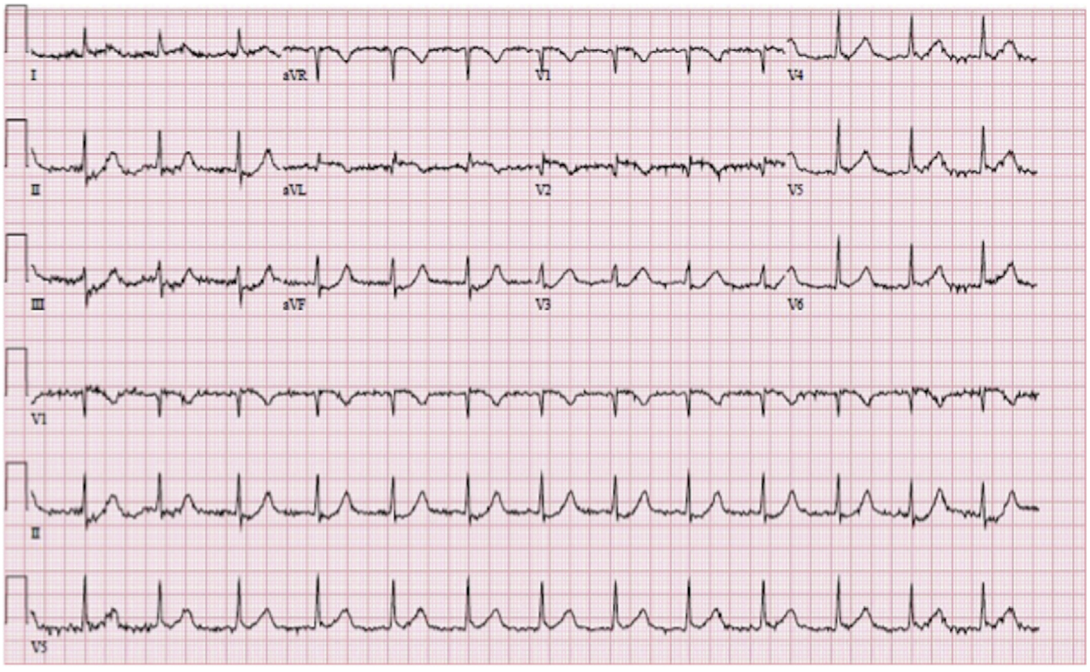
INVESTIGATIONS

The initial laboratory values demonstrated a normal complete blood count, normal metabolic panel, and troponin I. D-dimer was positive (228 ng/ml). A computed tomographic chest scan excluded pulmonary embolism. An electrocardiogram (ECG) revealed anterolateral injury and reciprocal changes in the inferior leads (**Figure 1**). Troponin I peaked at 48.52 ng/ml. Emergent cardiac catheterization revealed mild luminal irregularity of a small diagonal branch and otherwise angiographically normal coronary arteries (**Figure 2**). A ventriculogram demonstrated a severely reduced left ventricular ejection fraction (LVEF) of 30% to 35% with akinesis of the anterior, apical, and inferior walls and relative preservation of the basal territories consistent with Takotsubo cardiomyopathy (TTC) or a resolved myocardial infarction (MI) in the proximal wrap-around left anterior descending (LAD) artery distribution. An echocardiogram confirmed reduced LVEF with severe septal and apical hypokinesis.

Given the degree of troponin rise that seemed out of proportion to that typically observed in TTC (1) or expected from the abnormality associated with a small diagonal branch, cardiac magnetic resonance (CMR) was pursued. This demonstrated wall motion abnormalities in the LAD artery territory with late gadolinium enhancement (LGE), strongly suggesting MI of the mid to distal anterior wall, anteroseptum, and apex (**Figure 3**, Video 1). Her coronary angiogram was re-reviewed by multiple interventional cardiologists without noted evidence of stenosis, dissection, or occlusion involving the LAD (**Figure 2**).

At this stage, her presentation was suspicious for myocardial infarction with nonobstructive coronary arteries with coronary thrombosis/emboli highest on the differential.

FIGURE 1 Initial Electrocardiogram at Presentation



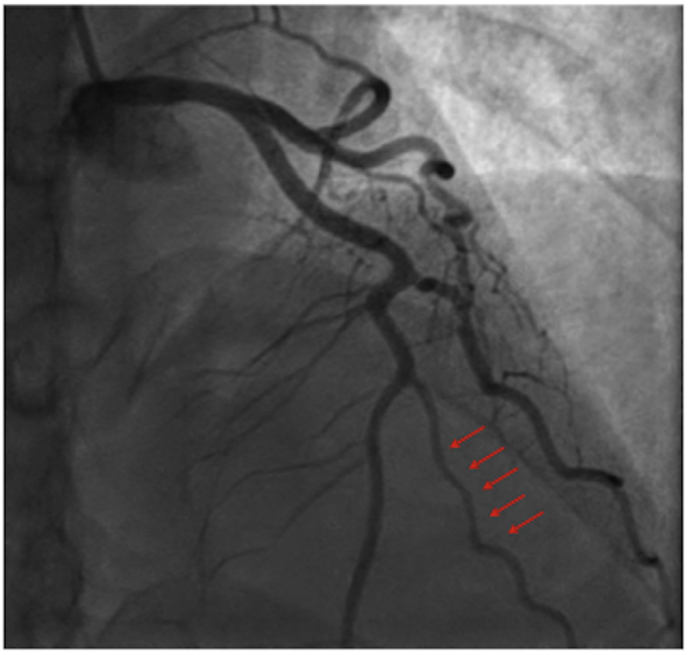
The electrocardiogram revealed evidence of repolarization abnormalities consistent with anterolateral injury.

MANAGEMENT

She was started on metoprolol. Her blood pressure did not tolerate further neurohormonal blockade. Unfractionated heparin was started on arrival given the reported atrial fibrillation in the ambulance and was continued because of the concern for potential embolic MI. Ultimately, these ECG strips were obtained, and no atrial fibrillation was noted. A transesophageal echocardiogram did not reveal an embolic source or patent foramen ovale, and unfractionated heparin was promptly discontinued.

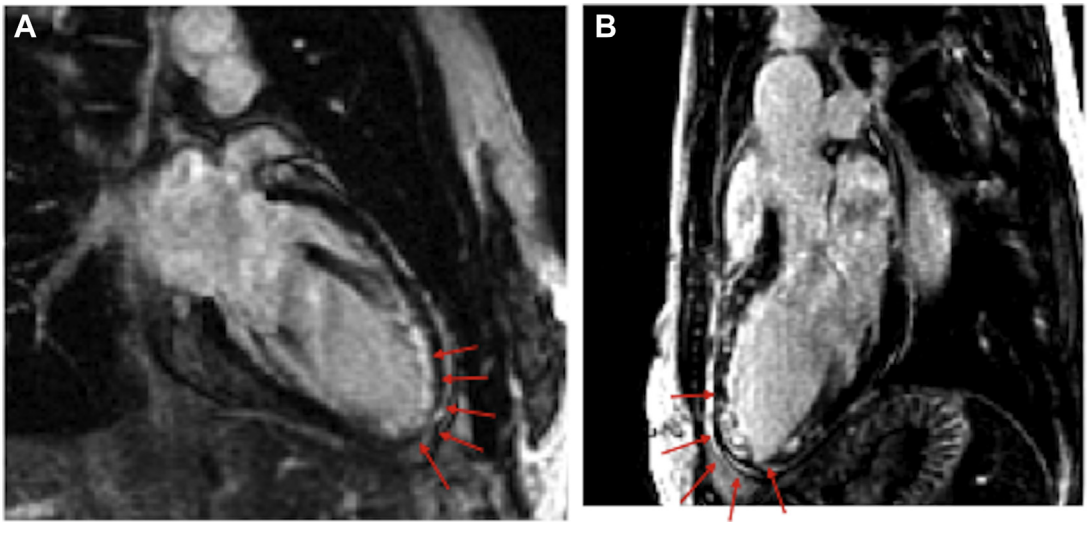
Four days into hospitalization, she developed arm numbness, mild chest discomfort, and emesis. An ECG showed more prominent ST-segment depressions in the inferior leads, which had otherwise improved during hospitalization, and <1-mm ST-segment elevations in the anterior leads (Figure 4). Given the recurrent symptoms, ECG findings, and the unclear explanation for her LAD territory MI, a repeat angiogram was cautiously pursued with low-pressure contrast injection not directly engaging the coronary arteries as the presence of coronary artery dissection was considered and the operators wanted to reduce the risk of further coronary artery injury or dissection propagation. The angiogram demonstrated evidence of extensive spontaneous coronary artery dissection (SCAD) involving the left main (LM),

FIGURE 2 Initial Coronary Angiogram



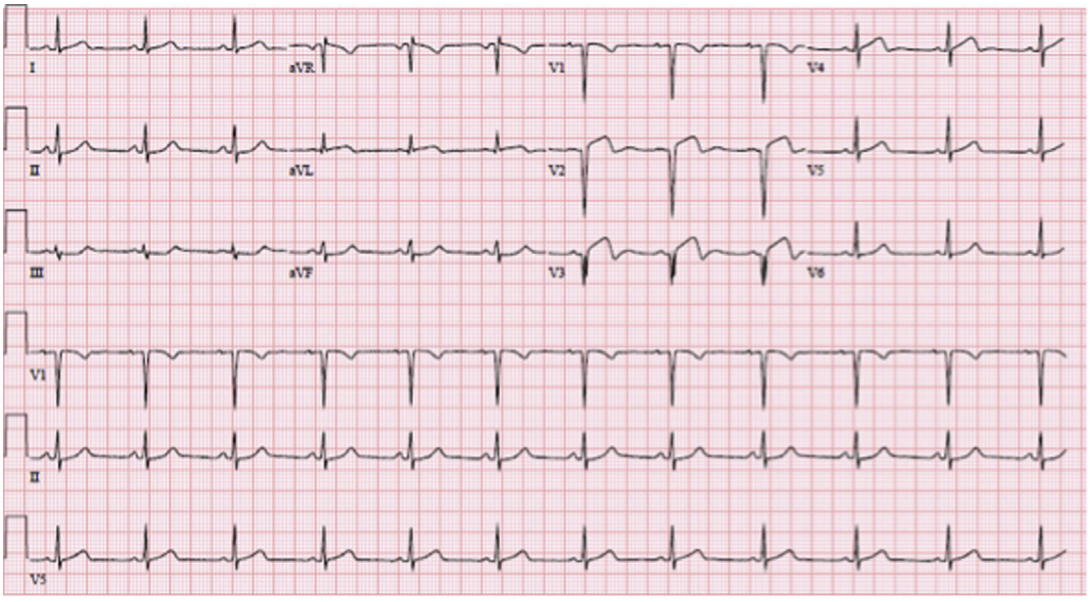
The initial angiogram revealed mild luminal irregularity involving the second diagonal branch. The **arrows** show the second diagonal branch. The other coronary arteries were angiographically normal.

FIGURE 3 Follow-Up Cardiac Magnetic Resonance Imaging



Cardiac magnetic resonance showed late gadolinium enhancement of the mid-distal anterior wall extending around the apex **(A, red arrows)** and the mid-distal septum and apex **(B, red arrows)**.

FIGURE 4 Evolving Electrocardiographic Changes



Evolving electrocardiogram changes revealed anteroseptal myocardial infarction and repolarization abnormalities consistent with anteroseptal injury or ventricular aneurysm.

proximal to distal LAD, left circumflex, and first and second diagonal (D1 and D2) coronary arteries (**Figure 5**, Video 2). An intra-aortic balloon pump was placed because of persistent ischemic symptoms. Although surgical intervention is rarely pursued for SCAD given the technical difficulty in differentiating true and false lumens, cardiothoracic surgery consultation was obtained because of the ongoing ischemic symptoms, hypotension, and extent of dissection with LM involvement. After multidisciplinary discussion, the decision was made to pursue surgical intervention, and a 4-vessel coronary artery bypass graft surgery (CABG) was performed with good graft anastomoses in the unaffected portions of the target vessels.

DISCUSSION

HOW COMMON IS SCAD? SCAD is an underrecognized cause of ACS in young women, despite accounting for over 25% of ACS in those ≤50 years, and can result in significant morbidity and mortality (2).

HOW IS THE DIAGNOSIS OF SCAD SUSPECTED AND CONFIRMED? A high index of suspicion should exist in young individuals, primarily of female sex, with a paucity of coronary artery disease risk factors presenting with MI. In these cases, coronary angiography should be pursued expeditiously for definitive diagnosis with the use of supplemental intracoronary imaging such as intravascular ultrasonography and optical coherence tomography if the diagnosis remains unclear (3).

WHY WAS THE DIAGNOSIS OF SCAD IN THE CURRENT CASE CHALLENGING? The case was challenging in light of her fairly unremarkable coronary angiogram relative to her dramatic presentation. After her initial angiogram, which did not demonstrate an obstructive coronary lesion, concern remained high for TTC or missed SCAD. Given the small caliber of the abnormal diagonal branch, intravascular imaging was not feasible for further investigation of the abnormal coronary segment. Repeat angiography, although initially considered, has associated risk if SCAD is on the differential diagnosis, given the possibility of propagating dissection with coronary instrumentation. Cardiac imaging in this case, namely with CMR, helped to narrow the differential diagnosis (see later), and recurrence of ischemia drove the decision for repeat angiography, which ultimately confirmed the diagnosis.

DO SCAD AND TTC COEXIST, AND, IF SO, HOW DO WE DIFFERENTIATE BETWEEN THE 2 ETIOLOGIES? HOW DOES CMR HELP? The potential concurrence of

SCAD and TTC has been previously described (4). It is hypothesized that an acute adrenergic rise in TTC may potentiate coronary artery sheer stress and SCAD or that stress associated with SCAD may theoretically manifest as TTC (3,5). Interestingly, in 2 series of patients originally diagnosed with TTC, re-review of angiograms revealed the presence of previously unrecognized SCAD in up to 9% (6,7). The persistent clinical syndrome, discordantly elevated troponin peak, ECG evolution, and presence of LGE in a single coronary territory played a critical role in differentiating our case from TTC and prompting the team to evaluate for alternative diagnoses. Furthermore, in TTC, the wall motion abnormalities typically extend beyond territories supplied directly by the LAD, including the inferior, posterior, and lateral territories, which were not noted in this case. Although routine follow-up angiography is not often pursued, there is a role for repeat testing in cases with diagnostic uncertainty or ongoing ischemia. Therefore, this case underscores the importance of avoiding committing to a single diagnosis when discordant clinical and investigational findings develop. Our case additionally highlights the utility of CMR in delineating myocardial pathology and the various etiologies of myocardial infarction with nonobstructive coronary arteries (MINOCA) through the identification of LGE, myocardial edema, and wall motion abnormalities (8).

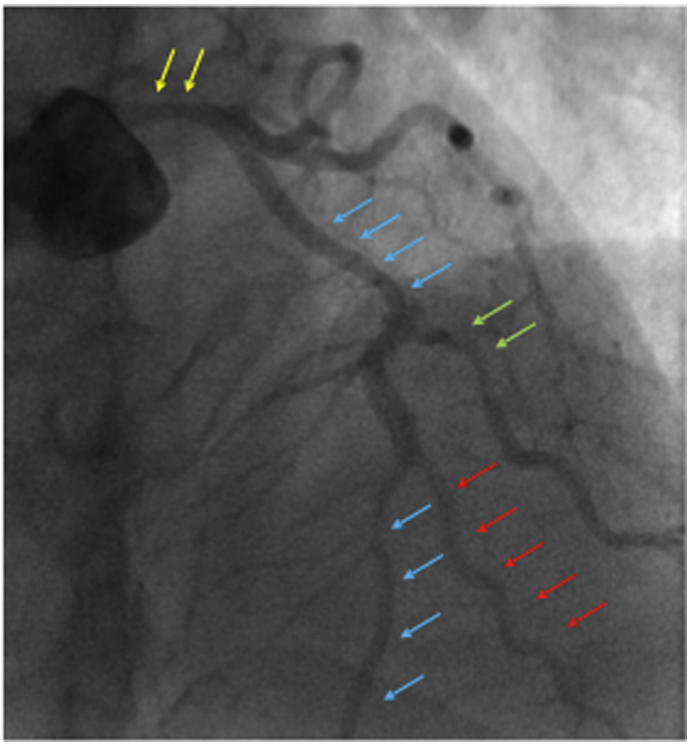
HOW COMMONLY DOES SCAD INVOLVE MULTIPLE TERRITORIES? The LAD is the most commonly affected coronary artery, with multivessel involvement in less than one-fourth of cases (3,9).

WHAT IS THE MAINSTAY OF THERAPY IN SCAD, AND WHY IS REVASCULARIZATION UNCOMMONLY PURSUED? The mainstay of management for most SCAD is medical therapy due to spontaneous healing over time in the majority of cases, the risk of propagating dissection with repeat invasive evaluation, and poor long-term patency of grafts; however, revascularization with CABG is considered in very select situations after comprehensive clinical decision making, including in cases with LM or proximal coronary involvement, ongoing ischemia despite conservative measures, or cardiogenic shock (9,10). In these scenarios, it is critical for the surgeon to differentiate a false versus a true lumen to provide the highest likelihood of graft patency.

FOLLOW-UP

The patient recovered well post-CABG and will be returning to her hometown (internationally) where she will undergo evaluation for fibromuscular dysplasia and connective tissue disorders.

FIGURE 5 Repeat Coronary Angiogram



Evidence of spontaneous coronary artery dissection Involving the left main **(yellow arrows)**, proximal to distal left anterior descending artery **(blue arrows)**, and first **(green arrows)** and second diagonal **(red arrows)** arteries.

CONCLUSIONS

SCAD can have a challenging clinical and angiographic presentation. As such, clinical familiarity with this entity is integral to prevent associated morbidity and mortality. Although there may be an association or overlap between SCAD and TTC, careful consideration with various imaging modalities, including intravascular or magnetic resonance imaging as well as a detailed review of coronary angiography, may be integral in delineating etiology and guiding further management.

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KEY WORDS spontaneous coronary artery dissection, Takotsubo cardiomyopathy, women's health

APPENDIX For supplemental videos, please see the online version of this paper.

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