

MINI-FOCUS ISSUE: CARDIOMYOPATHIES

INTERMEDIATE

CASE REPORT: CLINICAL CASE SERIES

Dynamic Takotsubo Syndrome

When SCAD Hides in a Pot



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ABSTRACT

Both Takotsubo cardiomyopathy and spontaneous coronary artery dissection (SCAD) of the distal portion of the left anterior descending artery affect the apical myocardium. It is important to distinguish between both diseases, because therapy and follow-up differ. Revascularization may be lifesaving in SCAD, whereas heart failure management is vital in Takotsubo cardiomyopathy. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2020;2:1923-31)
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PATIENT #1

PRESENTATION. A 50-year old woman was transferred to the hospital with acute onset of chest pain. At presentation, she was hemodynamically stable. Electrocardiography (ECG) showed nonspecific ST-T alterations in the inferolateral leads.

MEDICAL HISTORY. The patient had a medical history of Sudeck atrophy leading to amputation of her left forearm.

DIFFERENTIAL DIAGNOSIS. Differential diagnosis included acute coronary syndrome and pulmonary embolism (PE).

INVESTIGATIONS. Due to subtle ECG changes, an urgent transthoracic echocardiography (TTE) was performed, which showed akinesia of the apicolateral segment with suspicion of an apical thrombus (**Video 1A**). This structure, however, was not visualized in other fragments (**Video 1B**). Coronary angiography revealed a moderate stenosis in the distal portion of the left circumflex artery (LCX) (**Figure 1**). The right coronary artery (RCA) (**Figure 2**) and the left anterior descending artery (LAD) (**Figure 3**) did not show any apparent stenosis. Ventriculograms showed akinesia of the apex without evidence of an apical thrombus (**Video 2**). ECGs obtained on day 3, showed a typical evolution with negative T waves in the precordial leads. Takotsubo cardiomyopathy (TC) was diagnosed. The patient was treated with angiotensin-

LEARNING OBJECTIVES

- To examine the distal location of SCAD in middle-aged women with nonobstructive, nonatherosclerotic coronary arteries and wall motion abnormalities consistent with TC.
- To be aware of the dynamic nature of coronary pathology seen in patients with SCAD who have recurrent chest pain and have had an initial diagnosis of TC.
- To understand that treatment and prognosis of SCAD and TC differ: single antiplatelet therapy and beta blockers are used in SCAD, whereas ACE inhibitors are the treatment of choice in TC.

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**ABBREVIATIONS
AND ACRONYMS**

ACE = angiotensin-converting enzyme

ECG = electrocardiography

FMD = fibromuscular dysplasia

LAD = left anterior descending artery

LCX = left circumflex artery

PE = pulmonary embolism

RCA = right coronary artery

SCAD = spontaneous coronary artery dissection

TC = Takotsubo cardiomyopathy

TTE = transthoracic echocardiography

converting enzyme (ACE) inhibitors and a beta-blocker.

During admission, the patient suffered repeated mild attacks of angina, despite declining troponin levels. TTE on the third day showed dyskinesia in the basal and mid portions of the inferolateral and inferior walls. On day 5, Q-waves formed in the inferior leads, suggesting ischemia in the LCX or RCA territory. Due to these evolving signs of ischemia on ECG and TTE, angiography was repeated on the seventh day, revealing type I spontaneous coronary artery dissection (SCAD) of the LCX (Figure 4) with subsequent occlusion and type IIB SCAD of the RCA (Figure 5) with partial occlusion of the distal portion. The caliber of the distal portion of the LAD (Figure 6) was normal. Optical coherence tomography and intravascular ultrasonography were not attempted because of the risk that dissection would worsen with subsequent occlusion. Computed tomography angiography of the cerebral blood vessels showed no signs of fibromuscular dysplasia (FMD). Computed tomography angiography of the renal blood vessels was not performed.

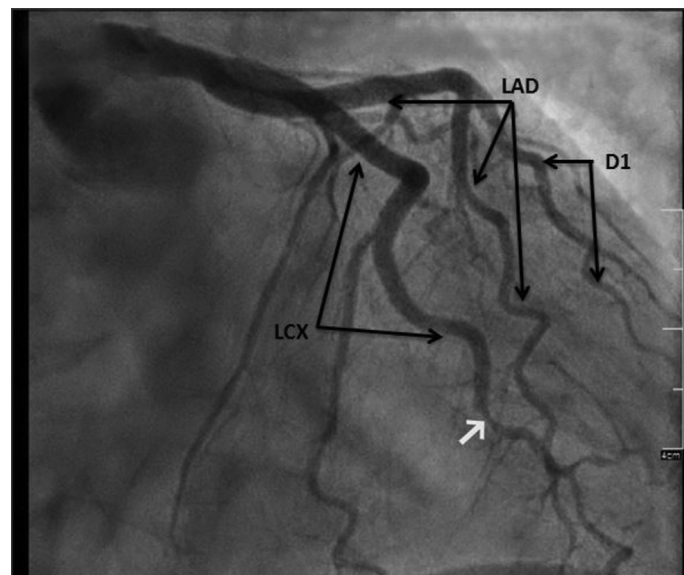
MANAGEMENT. The patient was treated medically with low-dose aspirin, an ACE inhibitor, spironolactone, and beta-blockers. TTE on the day of discharge (day 18) showed a nearly full recovery of the global and regional left ventricular functions. The patient was discharged in good health.

FOLLOW-UP. TTE 6 weeks after discharge showed persistent full recovery of left ventricular function. Angiography at 2 months after discharge showed complete resolution of the lesions (Figures 7 to 9). Medical treatment with ACE inhibitors, beta-blockers, and low-dose aspirin was continued. Because of this unusual clinical picture, all angiographies were reviewed, and expert opinion was sought. On review, the first angiography showed evidence of coronary dissection of the distal portion of the LCX (Figure 1) and the LAD (Figure 3). This explains why wall motion abnormalities differed between the echocardiography and ventriculography on day 1 and also explains the negative T waves in the precordial leads on day 3.

PATIENT 2

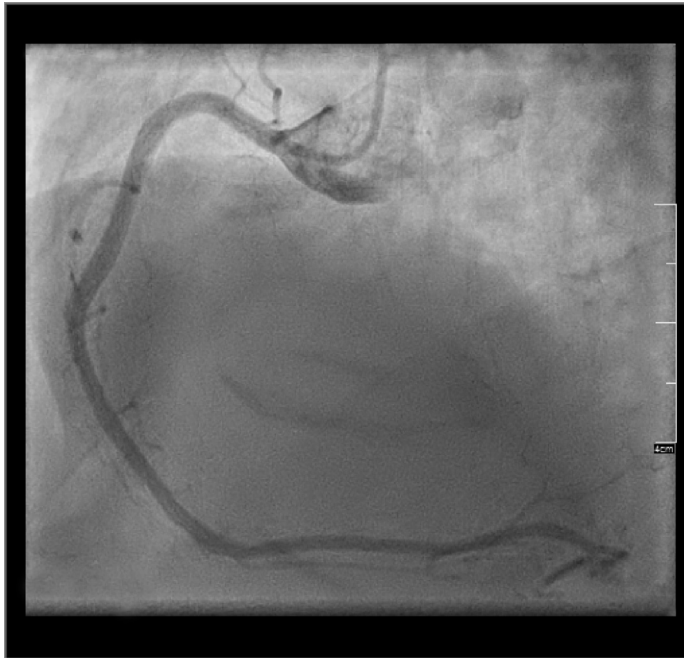
PRESENTATION. A 59-year-old woman presented to the emergency ward with symptoms of angina and dyspnea. At presentation, she was pain free and

FIGURE 1 First Angiogram of Patient 1: SCAD of LCX



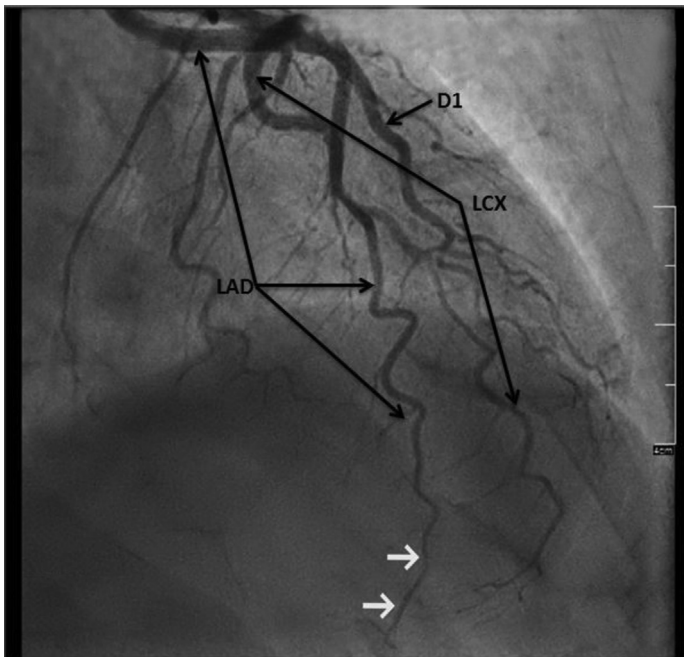
The first angiogram of Patient 1 shows moderate stenosis of the distal LCX (white arrow), suspected for SCAD. D1 = first diagonal branch; LAD = left anterior descending artery; LCX = circumflex artery; SCAD = spontaneous coronary artery dissection.

FIGURE 2 First Angiogram of Patient 1: No SCAD of RCA

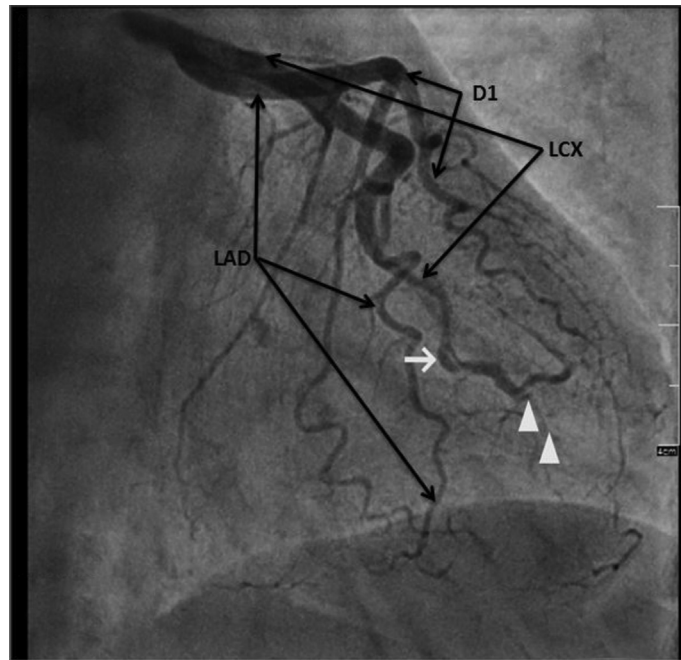


The first angiogram in Patient 1 shows right coronary artery without stenosis. RCA = right coronary artery; SCAD = spontaneous coronary artery dissection.

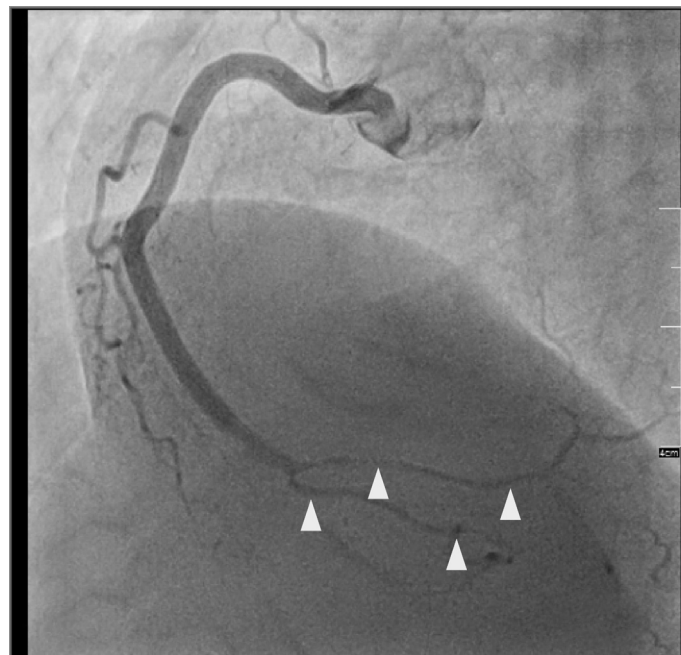
FIGURE 3 First Angiogram of Patient 1: SCAD of LAD



The first angiogram of Patient 1 shows a narrow lumen in the distal part of the LAD (**white arrows**), suspected for spontaneous coronary artery dissection after review. D1 = first diagonal branch; other abbreviations as in [Figure 1](#).

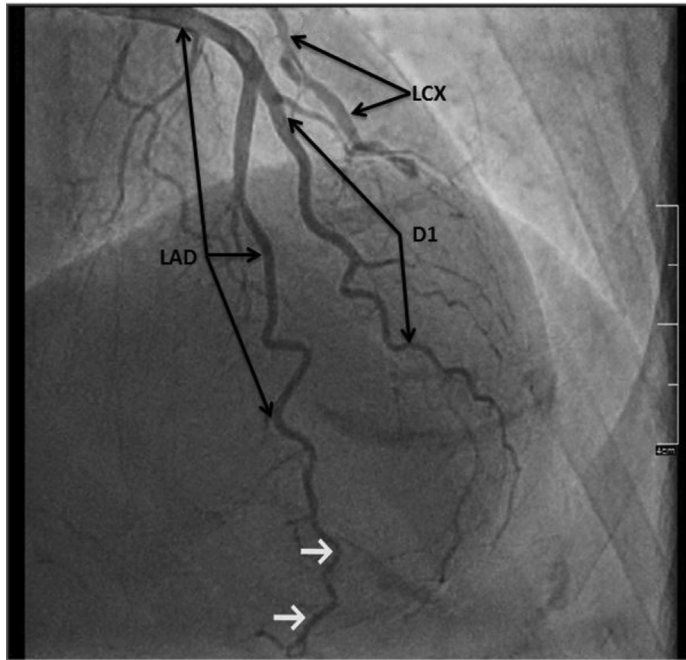
FIGURE 4 Second Angiogram of Patient 1: Type I SCAD of LCX

The second angiogram of Patient 1 shows type I SCAD of the distal portion of the LCX (**white arrow**) with subsequent occlusion (**white arrowheads**). Abbreviations as in [Figure 1](#).

FIGURE 5 Second Angiogram of Patient 1: Type IIb SCAD of RCA

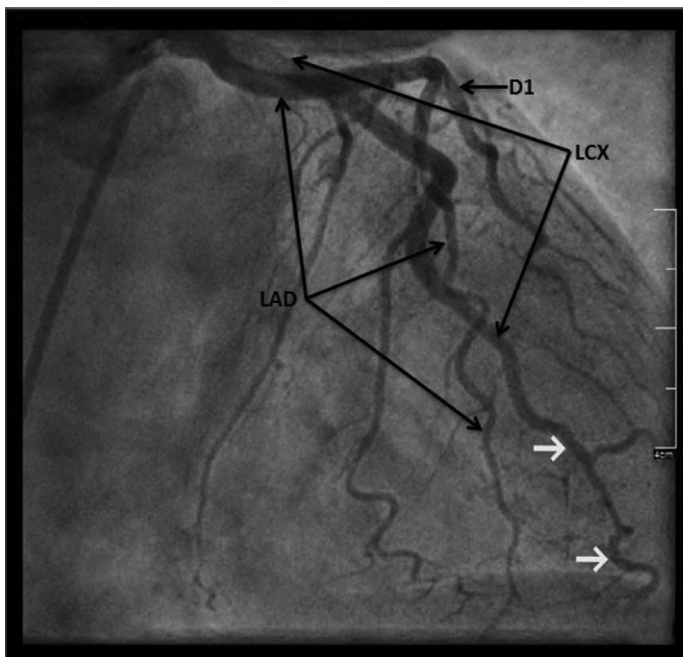
The second angiogram of Patient 1 shows type IIb SCAD of the distal portion of the RCA with partial occlusion. (**white arrowheads**) Abbreviations as in [Figures 1 and 2](#).

FIGURE 6 Second Angiogram of Patient 1: No SCAD of LAD



The second angiogram of Patient 1 shows normal caliber of the distal part of the LAD (**white arrows**). Abbreviations as in [Figure 1](#).

FIGURE 7 Third Angiogram of Patient 1: No SCAD of LCX



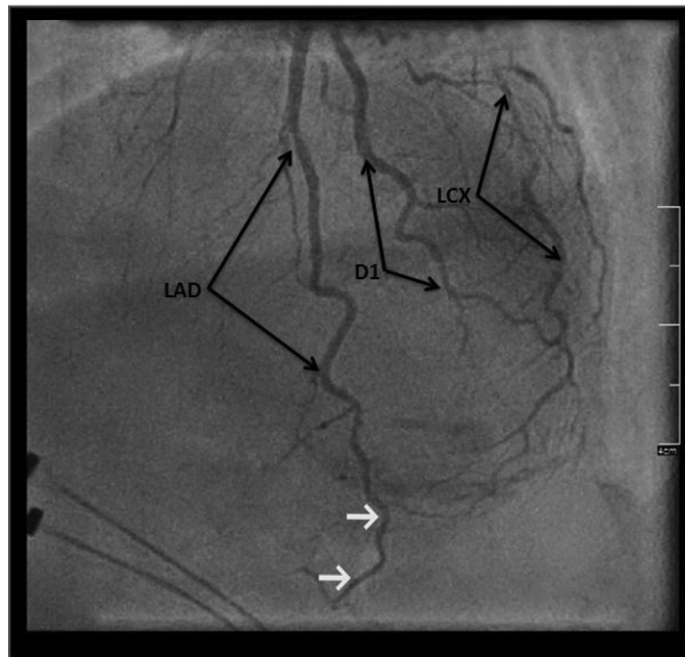
The third angiogram of Patient 1 shows complete resolution of lesion in the LCX (**white arrows**). Abbreviations as in [Figure 1](#).

FIGURE 8 Third Angiogram of Patient 1: No SCAD of RCA



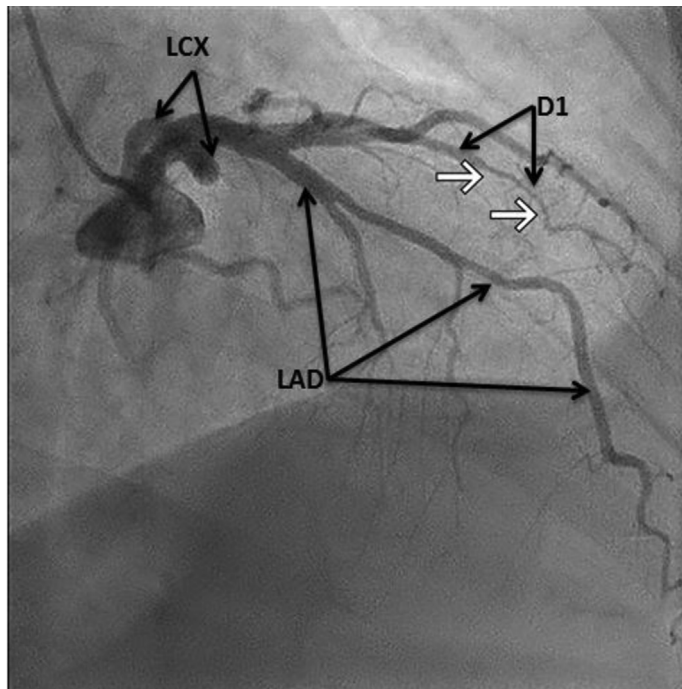
The third angiogram of Patient 1 shows complete resolution of the lesion in the RCA. Abbreviations as in [Figures 1 and 2](#).

FIGURE 9 Third Angiogram of Patient 1: No SCAD of LAD



The third angiogram of Patient 1 shows normal caliber of the distal part of the LAD (**white arrows**). Abbreviations as in [Figure 1](#).

FIGURE 10 Second Angiogram of Patient 2



The second angiogram of Patient 2 shows type IIb SCAD of the first diagonal branch of the LAD (white arrows). Abbreviations as in Figure 1.

hemodynamically stable. ECG showed nonspecific ST-T alterations in the inferolateral leads.

MEDICAL HISTORY. The patient had a medical history of arterial hypertension, hypercholesterolemia, and TC diagnosed 3 months earlier, based on akinesia of the apex seen on ventriculography.

DIFFERENTIAL DIAGNOSIS. Differential diagnosis included acute coronary syndrome, TC, SCAD, and pulmonary embolism.

INVESTIGATIONS. Because of the atypical presentation, mild ECG changes and troponin concentrations of 267 ng/l (<14 ng/l), urgent angiography was not performed. On the second day, however, troponin concentration had risen 4-fold. Coronary angiography revealed type IIb SCAD of the first diagonal branch of the LAD (Figure 10). Reviewing the first angiography, performed in a nearby heart center, showed evidence of coronary dissection of the distal portion of the LAD

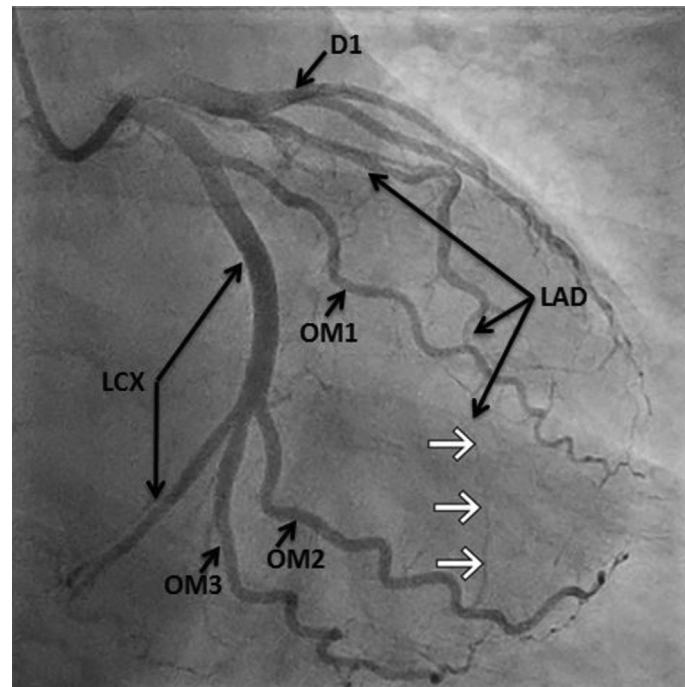
and a diagonal branch (Figure 11). TC should not have been diagnosed exclusively by ventriculography.

MANAGEMENT. The patient was treated medically with low-dose aspirin. She was already taking ACE inhibitors, spironolactone, and beta-blockers.

DISCUSSION

SCAD is a noniatrogenic separation of the coronary arterial wall. The underlying mechanism has yet to be elucidated, but an intimal tear or bleeding of the vasa vasorum could be the driving force (1), leading to a false lumen with intramural hematoma (2).

Until recently, SCAD was considered a rare disease, primarily seen in young women during the peripartum period and in patients with connective tissue disorders. The emergence of high sensitivity biomarkers and the use of intracoronary imaging have

FIGURE 11 First Angiogram of Patient 2

The first angiogram of Patient 2 shows type IIb SCAD of the distal LAD (**white arrows**) and a diagonal branch. OM1-OM3 = obtuse marginal artery 1-3; other abbreviations as in [Figure 1](#).

changed our mindset. A recent multicenter prospective observational study (3) showed that SCAD predominantly affects young to middle-aged women (mean age: 52 years old). FMD was seen in one-third of the patients, and peripartum SCAD or association with other connective diseases was rare. Emotional stress was reported in 50.3% and physical stress in 28.9% of patients. The association between SCAD (4) and FMD has recently been confirmed at a genetic level (4,5). A common variant of chromosome 6 at the PHACTR1 locus could be a risk locus for SCAD and FMD.

Because shear stress may be the driving force for SCAD, medical therapy with beta-blockers and single-antiplatelet therapy is the treatment of choice. Revascularization is needed in some cases of SCAD patients who have ongoing ischemia (6), in contrast to TC, where treatment is based on the acute presentation of the disease, management of heart failure or in some cases shock. The role of long-term therapy with ACE inhibitors and beta-blockers is not well established.

CONCLUSIONS

This paper describes 2 cases of SCAD which were initially misdiagnosed as TC. These cases underscore the need for interventionists to familiarize themselves with the often subtle and sometimes hard to recognize angiographic appearance of SCAD. These cases also emphasize the fact that one should at all times be prepared to challenge an initial diagnosis and use additional imaging if deemed necessary. We clinicians should be aware of dynamic coronary pathology in SCAD.

AUTHOR RELATIONSHIP WITH INDUSTRY

All authors have reported that they have no relationships relevant to the contents of this paper to disclose.

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KEY WORDS acute coronary syndrome, case series, coronary angiography, spontaneous coronary artery dissection, takotsubo cardiomyopathy

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