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CASE REPORT

INTERMEDIATE

CLINICAL CASE

Acute De Novo Multivessel Spontaneous Coronary Artery Dissection

A Case Report and Review of Literature

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ABSTRACT

Spontaneous coronary artery dissection (SCAD) is an important cause of acute coronary syndrome, especially in young women. Recurrent SCAD can be due to extension of dissection or de novo dissection of unrelated vessel. The authors present a case of acute de novo recurrent SCAD treated conservatively and with coronary intervention. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2019;1:32–5) © 2019 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/).

Spontaneous coronary artery dissection (SCAD) is an important cause of acute coronary syndrome (ACS). Recurrent SCAD can be due to extension of SCAD of the index vessel or de novo dissection of an unrelated vessel. Recurrence can occur acutely within hours or it might occur several years after the first SCAD episode. The acute phase de novo SCAD involving multiple vessels is uncommon. Here, we present an interesting case of de novo recurrent SCAD involving multiple vessels successfully treated with both conservative therapy and percutaneous coronary intervention (PCI).

LEARNING OBJECTIVES

- To know the clinical presentations and risk factors associated with SCAD.
- To recognize the difference angiographic appearances of SCAD on coronary angiograms.
- To recognize the role of intravascular imaging in the diagnosis of SCAD.
- To recognize the management options and the indications for an invasive approach in such cases.

HISTORY OF PRESENTATION

Our case is a 49-year-old woman, who presented to our facility with worsening anginal chest pain for the past few weeks. Her blood pressure on arrival was 202/100 mm Hg, and her heart rate was 102 beats/min. Physical exam was otherwise unremarkable.

PAST MEDICAL HISTORY

The patient had been diagnosed with hypertension and ACS, with stenting of the obtuse marginal coronary artery (where SCAD was misdiagnosed as atherosclerotic coronary artery disease [CAD]) in 2016.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis was ACS secondary to plaque rupture, very late stent thrombosis, embolic phenomenon to coronary vessels, or SCAD.

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INVESTIGATIONS

The electrocardiogram (ECG) was normal, and troponin was 0.93 ng/ml. Her blood work-up, including cell count and comprehensive metabolic profile, was unremarkable.

MANAGEMENT

ACS therapy with heparin drip, aspirin, clopidogrel, statin, beta-blocker, and angiotensin-converting enzyme inhibitor was initiated. The following day, the coronary angiogram revealed a patent obtuse marginal coronary artery stent and new SCAD of the distal right coronary artery (RCA) (**Figure 1**). Given the Thrombolysis In Myocardial Infarction (TIMI) flow grade III and resolution of the symptoms, SCAD of the distal RCA was treated conservatively.

On day 3, the patient experienced recurrent severe chest with the troponin level rising to 25.50 ng/ml. The ECG showed T-wave inversions in lead I and AVL. Echocardiogram showed mild anterolateral wall hypokinesis with a left ventricular ejection fraction of 60%. Repeat coronary angiogram revealed slight progressive SCAD of the distal RCA into the posterior descending coronary artery (PDA) and posterolateral vessel with TIMI flow grade III and new SCAD of the septal perforator and bifurcating diagonal coronary artery with TIMI flow grade 0 (**Figure 2**). Intravascular ultrasound (IVUS) confirmed the dissection and a large intramural hematoma in the diagonal vessel wall (**Figure 3**).

Given TIMI flow grade 0, ongoing chest pain despite medical treatment, the ECG and echocardiogram findings, the decision was made to proceed with PCI, and a total of 4 stents were deployed to the diagonal artery with restoration of TIMI flow grade III (**Figure 4**, Videos 1 and 2). Computed tomography angiography of head, neck, chest, abdomen, and pelvis did not show evidence of fibromuscular dysplasia. Patient's symptoms resolved, and she was discharged on hospital day 5.

FOLLOW-UP

Patient was doing well at 1-month follow-up in the cardiology clinic.

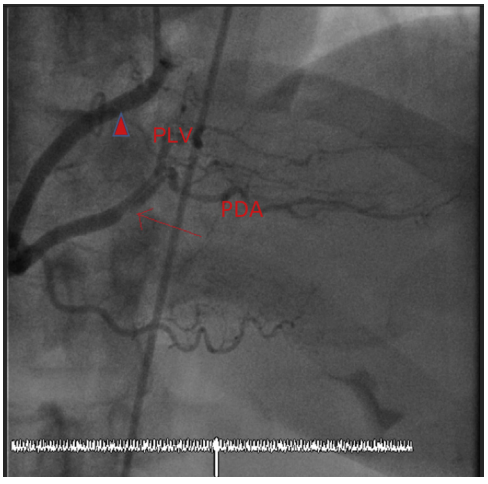
DISCUSSION

SCAD has emerged as an important cause of ACS, comprising 1% to 4% of all ACS cases overall, and up to 35% of ACS in young women <50 years of age (1). Our understanding of the pathophysiology of SCAD has evolved in the past few years with 2 main hypotheses on how SCAD occurs; the first is intramural hematoma caused by intimal tear, and the second is intramural hematoma caused by spontaneous hemorrhage in the vasa vasorum. The intramural hematoma compresses the true vessel lumen, leading to decreased coronary perfusion and myocardial infarction (1–3).

SCAD has been associated with several risk factors. It has a strong association with fibromuscular dysplasia, in up to 86% in some studies; that is why many experts recommend screening for fibromuscular dysplasia in all patients with SCAD (1). Other important associated risk factors for SCAD include pregnancy, multiparity, external hormone use, systemic inflammatory diseases, emotional stress, and history of migraine (1,4,5).

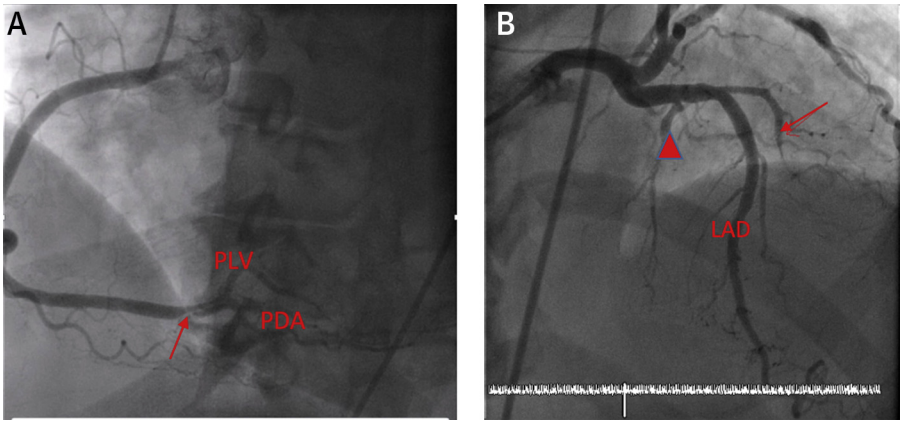
The left anterior descending coronary artery territory and its associated branches, the diagonal and septal branches, is the most commonly involved territory, with the left circumflex coronary artery territory being the second most common territory involved, then the right coronary system, with the left main vessel being the least commonly involved vessel in SCAD (**Table 1**) (1). Multivessel involvement in SCAD occurs in 9% to 23% of cases, with noncontiguous vessel involvement in 5% to 10% (1).

FIGURE 1 SCAD Involving the Distal RCA



The figure shows the angiographic appearance of type 1 with double lumen in the distal RCA extending to the PDA and PLV. The **arrow** indicates the distal RCA SCAD. The **arrowhead** indicates the proximal RCA. PDA = posterior descending coronary artery; PLV = posterolateral vessel; RCA = right coronary artery; SCAD = spontaneous coronary artery dissection.

FIGURE 2 SCAD Involving the Distal RCA and Diagonal Coronary Artery

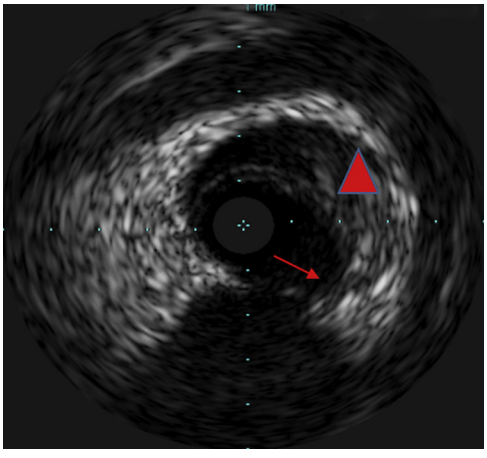


(A) SCAD involving distal RCA. There is slight progression in dissection in the distal RCA into the PDA and PLV as shown by the **arrow**. **(B)** SCAD involving the diagonal coronary artery with TIMI flow grade 0. This figure shows SCAD with angiographic appearance of type 2, as it illustrates the long-segment narrowing (>20 mm) of the vessel. The **arrow** shows diagonal SCAD. The **arrowhead** shows septal perforator SCAD, which was treated conservatively. TIMI = Thrombolysis In Myocardial Infarction; other abbreviations as in Figure 1.

Diagnosis of SCAD is challenging because it might have different angiographic appearances on coronary angiography. SCAD is classified into 3 types on the basis of the appearance on the coronary angiogram: type 1 shows multiple radiolucent vessel lumens, type 2 shows long-segment narrowing of the vessel (>20 mm), and type 3 shows a focal narrowing of the vessel lumen mimicking atherosclerotic CAD. In cases where SCAD is suspected, intravascular imaging using IVUS or computed tomography might be helpful in confirming the diagnosis (1,5).

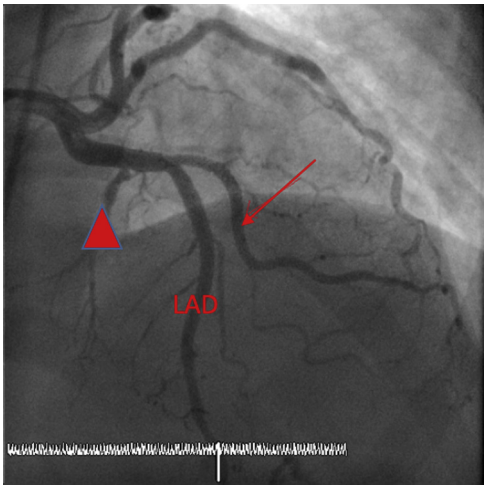
Most cases of SCAD are managed conservatively; as most dissections heal spontaneously in up to 97% of

FIGURE 3 IVUS of the Diagonal Vessel



The figure shows the intimal flap **(arrow)** and a large intramural hematoma **(arrowhead)** compressing the true lumen. IVUS = intravascular ultrasound.

FIGURE 4 Coronary Angiogram Showing the Diagonal Coronary Artery Post-PCI



Diagonal SCAD status post-successful PCI with deployment of 4 drug-eluting stents. The **arrow** indicates the diagonal coronary artery. The **arrowhead** indicates the septal perforator SCAD. Also see Videos 1 and 2. LAD = left anterior descending coronary artery; PCI = percutaneous coronary intervention; SCAD = spontaneous coronary artery dissection.

TABLE 1 The Incidence of Vessel Territory Involved in SCAD

Vessel Territory	Incidence
LAD, diagonal, septal	45%–61%
Left circumflex, obtuse marginal, ramus	15%–45%
Right coronary system	10%–39%
Left main	Up to 4%

LAD = left anterior descending coronary artery; SCAD = spontaneous coronary artery dissection.

cases. It is estimated that there is 1:6 hazard ratio of serious deterioration by day 6 in SCAD patients treated conservatively. Thus, it is of paramount importance to monitor these patients closely (6). Patients with impaired blood flow and persistent symptoms despite medical treatment need invasive treatment with either PCI or coronary artery bypass graft (CABG) (1). PCI is performed in 5% to 10% of all cases of SCAD. PCI is associated with higher complication and lower success rates in SCAD compared with atherosclerotic CAD, mainly due to the possibility of extension of the dissection with PCI and its potential catastrophic consequences. Studies have shown that SCAD patients treated with PCI needed emergent CABG in 30% to 53% of cases and/or emergent PCI in 4% of cases; this highlights the importance of selecting appropriate patients for intervention. CABG is usually performed for patients with left main SCAD (1,3,5).

CONCLUSIONS

SCAD is an important cause of ACS, especially in young women with minimal risk factors for atherosclerosis. Diagnosis is key because the management approach is different from atherosclerotic CAD. Cardiologists should be aware of the diagnostic and therapeutic challenges of this important disease entity.

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KEY WORDS de novo SCAD, multivessel SCAD, recurrent SCAD, SCAD, spontaneous coronary artery dissection

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