

## Spontaneous coronary artery dissection, a commonly overlooked etiology of acute coronary syndrome

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### ABSTRACT

Spontaneous coronary artery dissection (SCAD) is a type of non-atherosclerotic coronary artery disease, initially thought to be uncommon but is now being increasingly recognized as a cause of acute coronary syndrome in females. The exact incidence of this remains unknown and most of these cases undergo emergent percutaneous intervention (PCI) due to concern for acute coronary syndrome (ACS). Prior studies have shown that PCI can be detrimental in these cases. It is important to recognize the possibility of SCAD in young female patients so that potentially harmful interventions, such as starting these patients immediately on heparin, use of thrombolytic therapy, and emergent PCI that can lead to worse outcomes, are avoided.

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### 1. Introduction

SCAD is a type of non-atherosclerotic coronary artery disease. The risk factors for SCAD include female sex, young age, extreme emotional stress, exertion, pregnancy, fibromuscular dysplasia and it is also associated with inherited connective tissue diseases such as Marfan syndrome, Ehlers-Danlos, and Loeys-Dietz syndrome. There are limited data to determine the ideal management for SCAD and PCI is reported to cause higher rate of complication and emergency coronary artery bypass grafting. Thrombolytic therapy and glycoprotein IIb/IIIa inhibitors and anticoagulants are not recommended in SCAD. In this case report, we discuss a female patient presenting with typical symptoms and angiographic findings of SCAD. We also have provided a brief review of literature of prior reported cases.


### 2. Case presentation

A 66 year old female with past medical history of hyperlipidemia, gastroesophageal reflux disease (GERD), long-term tobacco use, and obstructive sleep apnea was admitted with chest pain. On presentation, the patient reported acute chest pain, located in the center of the chest, 10/10 in intensity, radiating to both arms and neck. Pain was associated with diaphoresis and nausea. She reported taking four baby aspirin prior to calling for help. The patient was given nitroglycerin in the emergency department

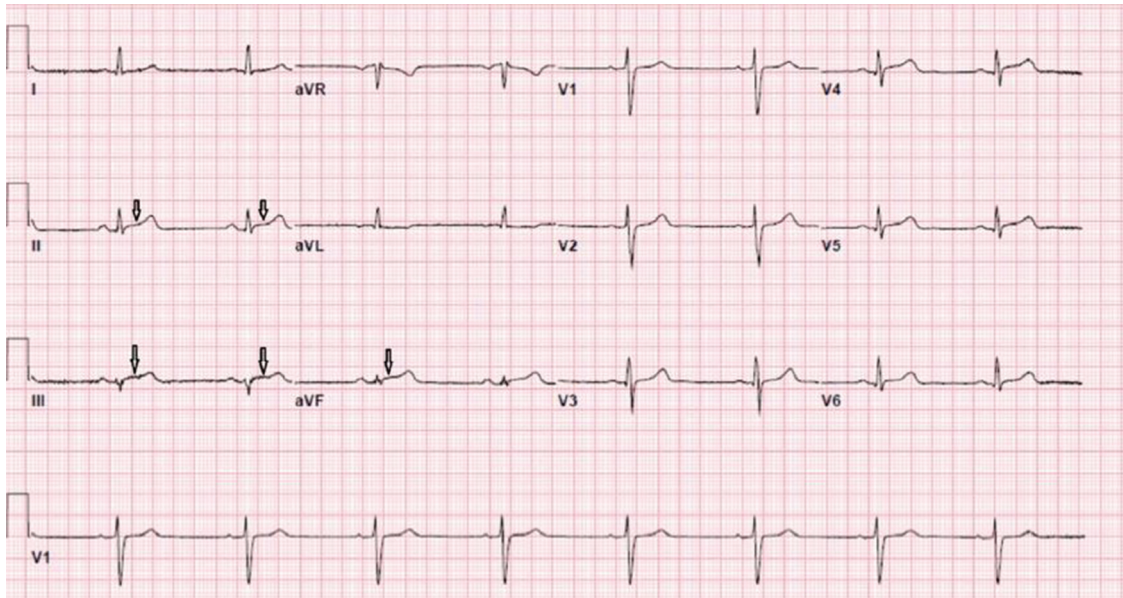
with resolution of her pain. Her family history was significant for myocardial infarction in father at age 48.

### 3. Methods and procedures

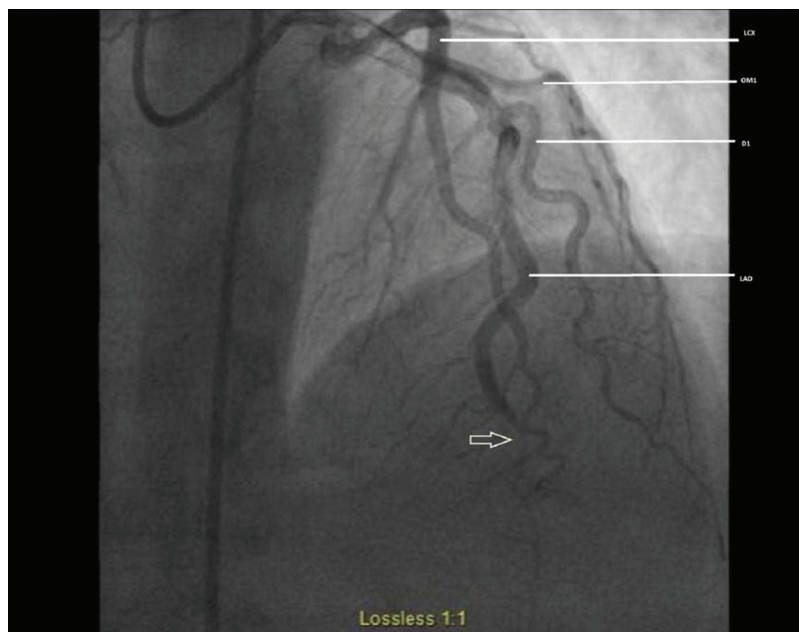
Electrocardiography (EKG) showed inferior ST segment elevation myocardial infarction (STEMI) and patient was taken for emergent coronary angiography (Figure 1). Emergent cardiac catheterization showed the mid segment of left anterior descending (LAD) artery with 90% rapid tapering followed by distal pruning (Figure 2). The LAD finding on coronary angiography was consistent with spontaneous coronary artery dissection (SCAD). During the procedure, patient was having active chest pain; therefore, percutaneous coronary intervention with ballooning under low inflation pressure was done for 1 min with advancement of guidewire into distal LAD, but no change in vessel size or flow was appreciated with the intervention or retraction of guidewire (Figures 3,4). Left circumflex artery (LCX) was noted to be a large codominant vessel with 20% stenosis. Right coronary artery (RCA) with no evidence of stenosis was noted. Due to cardiac catheterization findings suggestive of SCAD, a stent was not placed. Left ventricular ejection fraction on cardiac catheterization was noted to be low at 40% and a follow up echocardiogram was done. Transthoracic echocardiogram

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**Figure 1.** EKG on admission with ST elevation in inferior leads. (Downward arrow showing ST elevation).



**Figure 2.** Right anterior oblique cranial view on angiogram showing mid-segment rapid tapering of left anterior descending artery with distal pruning. (Left arrow – showing LAD taper, coronary arteries marked as, LCX – left circumflex, OM1 – obtuse marginal 1, D1 – diagonal 1, LAD – left anterior descending).

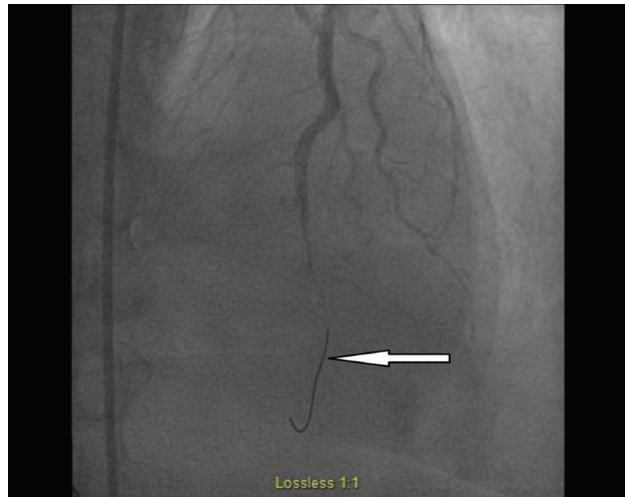
confirmed a left ventricular ejection fraction of 40%, akinesia of the mid to apical segments and inferior hypokinesia (Figure 5).

With new diagnosis of systolic heart failure, the patient was started on beta blocker and angiotensin-converting-enzyme inhibitor. Our patient was initially loaded with ticagrelor and after the procedure was switched to dual antiplatelet therapy with aspirin and clopidogrel. Statin treatment was started for hyperlipidemia. The patient was educated on lifestyle modifications and smoking cessation. She was followed up at the cardiology outpatient clinic within 2 weeks and then every 3 months. The

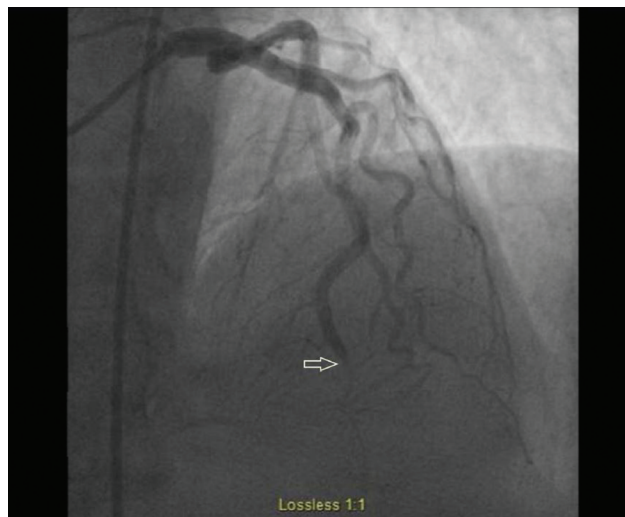
patient did not report any exertional chest pain on follow up.

#### 4. Discussion

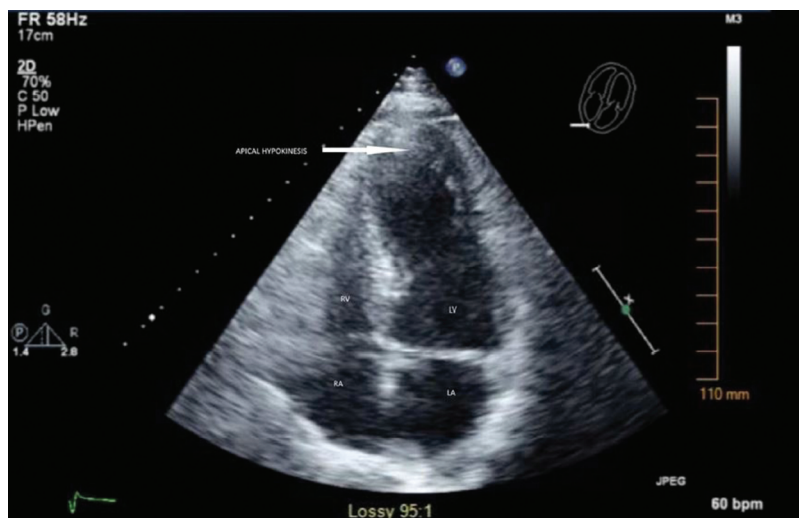
Spontaneous coronary artery dissection (SCAD) is an acute coronary event of uncertain origin. It is a type of non-atherosclerotic coronary artery disease. As the name suggests, it is a spontaneous dissection in the coronary arterial wall, leading to intimal tearing and compression of true lumen. Clinical features and prognosis of the disease remain insufficiently



**Figure 3.** Right anterior oblique cranial view on cardiac angiogram shows guidewire in distal LAD. (Left arrow – showing guidewire).



**Figure 4.** Right anterior oblique cranial view on cardiac angiogram after guidewire retraction, with no improvement in flow in distal LAD noted. (Right arrow – showing no improvement in flow).



**Figure 5.** Four chamber apical view on transthoracic echocardiogram showing reduced left ventricular ejection fraction and apical akinesis. (Right arrow showing apical akinesis, RV – right ventricle, LV – left ventricle, RA – right atrium, LA – left atrium).



characterized at this time. Pretty reported the first case of SCAD in 1931 [1]. With increasing focus on women's cardiovascular health, over the last decade, several case reports with SCAD have been published [2–12].

Atherosclerotic disease with plaque rupture was considered to be the most common pathophysiology behind ACS, it is now recognized that women may have other underlying pathophysiology, such as plaque erosion and SCAD [13,14]. The exact prevalence of SCAD remains unclear and reported prevalence ranges from 0.1% to 25% [15,16]. Studies in Europe reported the incidence of SCAD being three times more in women as compared to men but the underlying cause of this difference still remains unidentified [17,18]. Pregnancy-associated spontaneous coronary artery dissection is considered a common cause of ACS in young pregnant females [13]. More cases of SCAD in non-pregnant women are being recognized and remains an area of further research.

The risk factors of SCAD have been poorly elaborated till date. Initially, SCAD was thought to be only associated with hormonal and hemodynamic changes due to pregnancy and postpartum state but risk factors now include female sex, young age, extreme emotional stress, exertion, and fibromuscular dysplasia [2,3,13]. With increasing recognition of the disease in non-pregnant healthy women, other risk factors highlighted include systemic connective tissue disorders such as Marfan's syndrome, Ehlers-Danlos and Loeys-Dietz syndrome. Underlying connective tissue disorders in general are known risk factors of intimal wall rupture in any arterial vessel, including coronary arteries. Fibromuscular dysplasia (FMD) is now known to be associated with this condition. In one study, noncoronary FMD was seen in 86% of patient [3]. Several other authors have reported association between FMD and SCAD [19–24]. Our patient did not have a known history of FMD, or history such as uncontrolled hypertension suggestive of it. No clear recommendation to evaluate for underlying FMD in young females diagnosed with SCAD exist. Our patient therefore did not undergo further imaging study to evaluate for possible underlying FMD.

Another interesting association that was recently highlighted by Madias JE is the relationship of SCAD with Takotsubo cardiomyopathy [25,26]. Both these entities are more common in females and emotional stress is recognized as risk factor for both. It is still unclear if there exists a causal relationship between the two. Of interest, our patient did have akinesis of mid to apical segment noted on echocardiography (Figure 5).

In reported cases, left anterior descending (LAD) artery and its branches are more commonly involved in SCAD [6–11]. Our patient had LAD involvement with rapid tapering and distal pruning

(Figure 2). An angiographic series of 246 SCAD patients by Eleid et al. found significantly increased occurrence of coronary tortuosity in SCAD patients as compared to 313 matched controls and suggested increased tortuosity as potential mechanism for SCAD [27]. Saw J characterized angiographic appearance in SCAD into three categories [28]. These subtypes included Type 1 (contrast staining of arterial wall delineating multiple lumens), Type 2 (diffuse smooth narrowing of arterial lumen usually involving mid to distal arterial segment with abrupt change in caliber), and Type 3 (similar to atherosclerosis). Saw and Yip in another review noted that type 3 dissection is commonly misdiagnosed due to similarity with atherosclerosis [29]. They stated that long lesions with hazy, linear stenosis, and lack of atherosclerotic changes in other coronary arteries favor SCAD diagnosis over atheroma. Our patient had findings consistent with type 2 SCAD where acute change in caliber of LAD was noted, involving a long distal segment of the artery [6]. Intravascular ultrasound and optical coherence tomography may be used to visualize arterial wall disruption and intramural thrombus in cases where SCAD is missed on angiography [30–35].

As was seen in our patient, STEMI has been reported to be the most common finding on EKG in cases with SCAD. This raises the clinical dilemma of proceeding with emergent percutaneous coronary intervention as suggested by American heart association in cases with acute myocardial infarction. There are no clear guidelines based on randomized control trials for management of these cases. One study compared PCI with conservative management and showed that PCI in these cases is associated with higher rate of complication and need of emergency coronary artery bypass grafting [7]. Stent placement in these cases may lead to progression of intramural hematoma. Most of these cases are treated with aspirin. When no stent is placed, use of clopidogrel in addition to aspirin is questionable, but theoretically can be beneficial in prothrombotic state with intimal tear and mural thrombus. Choi et al. reported a case of multivessel coronary dissection treated with dual antiplatelet therapy [36]. Our patient was started on dual antiplatelet therapy based on literature review and recommendations. There are limited data to determine the ideal conservative management for SCAD.

Thrombolytic therapy and glycoprotein IIb/IIIa inhibitors are avoided in known cases of SCAD due to risk of thrombus extension [37,38]. Likewise, treatment with anticoagulants such as warfarin and heparin should be avoided. Therefore, it is important to recognize the possibility of SCAD in young female patients so that potentially harmful interventions such as starting these patients immediately on

anticoagulants, use of thrombolytic therapy and emergent PCI that can lead to worse outcomes and unnecessary stent placement are avoided.

Long-term beta-blocker and angiotensin converting enzyme inhibitors are added when left ventricular dysfunction, hypertension or angina are present based on American heart association guidelines. Statins are not routinely administered unless indicated by primary prevention guidelines. In our patient with reduced ejection fraction and history of hyperlipidemia, beta blocker, angiotensin receptor blocker, and statin was started.

In-hospital prognosis of SCAD has been reported to be favorable and most cases have shown resolution with vessel healing. In a retrospective 87 patient series, the estimated 10-year rate of death, heart failure, myocardial infarction, or dissection recurrence was 47% [4]. Early recurrence is usually in the form of dissection extension whereas late recurrence invariably occurs in new coronary distribution. With more focus on women's cardiovascular health, SCAD remains an area of interest, with need for larger studies to determine the correct management protocol.

## 5. Learning points

- (1) Spontaneous coronary dissection remains a commonly overlooked cause of ACS with increasing number of cases now being recognized due to greater availability of invasive procedures and imaging modalities.
- (2) SCAD is commonly associated with FMD in young females.
- (3) Angiographic imaging with distal pruning and no improvement of blood flow noted with ballooning is consistent with SCAD.
- (4) Treatment for SCAD may differ from atherosclerotic causes of ACS. Thrombolytic therapy, anticoagulant and coronary stent placement may be harmful in these cases.
- (5) Differentiating SCAD from atherosclerotic disease is prudent to prevent unnecessary intervention that may be rather harmful for the patient.

## Disclosure statement

No potential conflict of interest was reported by the authors.

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