Introduction

Spontaneous coronary artery dissection (SCAD) is an extremely rare cause of acute coronary syndrome (ACS), with a reported angiographic incidence of 0.1–1.1% (1,2). Patients may present with a broad spectrum of clinical scenarios, ranging from angina pectoris to myocardial infarction, cardiogenic shock, and sudden death (3). It occurs predominantly in women, with a female: male ratio of 2:1. The majority of patients are young adults, with an average age of 46 years for men and 38 years for women (1). In > 70% of cases, the diagnosis is made on postmortem examination (4,5). SCAD can present with sudden death and, unless an autopsy is performed, the true number of cases will remain underestimated. Other factors leading to underestimation of its prevalence are false-negative results on coronary angiography and spontaneous closure of the dissection (6).

We report a case of a 39-year-old male who presented with chest pain for 4 days and electrocardiographic findings of anterior myocardial infarction. He was found on angiography to have SCAD of the distal left anterior descending (LAD) coronary artery.

Case

A 39-year-old Caucasian male with past medical history of hypertension, diabetes mellitus type I and tobacco use presented with multiple complaints including non-exertional substernal chest pain, epigastric abdominal
pain and emesis ongoing for 3 days. Chest pain had been preceded by dyspnea on exertion over a period of one week. Physical examination showed mild epigastric tenderness. Blood pressure was 119/79 mm Hg, with a pulse rate of 111 beats per minute and respiratory rate of 13 breaths per minute. His temperature was 36.7°C (98 °F). Oxygen saturation was 100 % on room air. Electrocardiography (ECG) showed sinus tachycardia, left posterior fascicular block, ST elevation in the anterior and inferior leads(Figure 1). Transthoracic Lab work revealed elevated cardiac troponin 2.95 ng/mL, blood glucose of 984 mg/dL, HbA1c=10.7 %, creatinine 1.73 mg/dL, hemoglobin of 8.8 g/dL, Sodium level 124 mEq/L and Potassium level of 6.7 mEq/L. Arterial blood gas showed pH of 7.20, bicarbonate level 9 mEq/L, PCO2 14 mm Hg. He was admitted to intensive care unit for diabetic ketoacidosis and a stat cardiology consult was placed. Stat bedside echocardiography showed absence of thickening of the apex and periapical region, gigantoid, multilobar, complex, separate pendular mobile thrombi (Figure 2a to 2e). Urgent coronary angiography showed total occlusion of distal portion of LAD artery with spiral dissection of LAD (figure 3) thought to be subacute or chronic, given collateral branches from diagonal branch supplying cardiac apex.

**Figure Legend 1**

*Electrocardiogram (ECG) showing a sinus tachycardia, acute anterior and inferior infarct, left posterior fascicular block (LPFB) and prolonged QT interval*

He received loading dose of aspirin 325 mg then 81 mg daily, loading dose of clopidogrel 600 mg once and then 75 mg daily, unfractionated heparin intravenous bolus of 60 units per kilogram then 12 units per kilogram intravenous infusion, atorvastatin 80 mg nightly and metoprolol tartrate 25 mg twice daily. He was placed on Warfarin with INR target of 2-3 given his left ventricular thrombus. He was also placed on intravenous insulin per diabetic ketoacidosis protocol. Afterwards hospital course was stable. He was discharged home with plan for outpatient cardiology follow up.
Unusual Distal LAD Spiral Dissection in Young Male

Figure Legend 2

Parasternal long axis

Figure Legend 3

Apical 4 chambers
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Figure Legend 4

3 dimensional apical 4 chambers

Figure Legend 5

Definity study
DISCUSSION

Primary SCAD is a rare cause of acute myocardial infarction and is associated with a high mortality rate of about 50%. It has been observed in four groups of patients: those with coronary atherosclerosis, women in peripartum and early postpartum period, women taking oral contraceptives, and lastly in an inhomogeneous group labelled “idiopathic” as illustrated by this case due to there being no obvious associated factors. (1).

SCAD is defined as a spontaneous separation of the coronary artery wall that is not iatrogenic or related to trauma. As such, dissections due to blunt trauma, surgical instruments, or those that are catheter-induced are not deemed to be SCAD. Furthermore, contemporary usage of the term SCAD is typically reserved for the nonatherosclerotic variant, and most modern series exclude SCAD due to atherosclerotic coronary artery disease.

SCAD was previously, incorrectly believed to be very rare and to be frequently associated with pregnancy. Unfortunately, the true incidence and prevalence of SCAD in the general population is unknown due to significant under-diagnosis of this condition. SCAD affects women in >90% of cases with rate of 9 to 1 ratio (female:male). In contemporary series that excluded patients with atherosclerotic causes, women accounted for 92% to 95% of the population with SCAD (12,15,16,18–20). The reported mean age ranged from 44 to 55 years in contemporary series, reflecting a relatively young to middle-age population (12,15,16,18–21).

The arterial dissection with SCAD can occur within or between any of the 3 layers (intima, media, or adventitia) of the coronary artery wall. Two potential mechanisms for the initiation of arterial wall separation have been proposed (23). The first is the intimal tear hypothesis, in which a primary disruption in the intimal-luminal interface creates an entry point for intramural hematoma (IMH) accumulation inside the false lumen, leading to separation of the arterial wall. The second is the medial hemorrhage hypothesis, in which a hemorrhage into the
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arterial wall is the primary mechanism, perhaps due to spontaneous rupture from the increased density of the vasa vasorum. This creates a false lumen in the vessel, which is further expanded by bleeding, accumulation, and enlargement of the hematoma. This causes further separation of the dissected layers, leading to compression of the true lumen. Subsequently, this results in myocardial ischemia or infarction (7).

Figure Legend 7

Transthoracic echocardiogram 4 chambers apical view showing LV thrombus

Figure Legend 8

Transthoracic echocardiogram parasternal short axis view showing LV thrombus
Coronary angiogram showing intimal dissection flap with preserved flow in distal LAD (Arrows)

The underlying etiology of SCAD appears to be multifactorial. There is often an associated underlying predisposing arteriopathy, which may be compounded by a precipitating stressor (Intense exercise, emotional stress, hormonal therapy, Valsava-type activities and recreational drugs) culminating in the phenotypic expression of SCAD. Many potential predisposing nonatherosclerotic arteriopathies for SCAD have been reported. The most dominant association reported is fibromuscular dysplasia (FMD), in addition to, pregnancy, connective tissue...
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disease, systemic inflammatory disease, coronary vasospasm or idiopathic, cyclosporine therapy, and heavy exercise (12). A hereditary factor has been discussed previously (17).

There is a wide spectrum of clinical presentations and severity of SCAD. Almost all patients with SCAD present with ACS and elevation of cardiac enzymes (15,17,18,21). The proportion who presented with STEMI varied widely in different series, ranging from 24% to 87% (15–21). A small proportion can be complicated with ventricular arrhythmias (3% to 10%) (15–17), cardiogenic shock (<3%) (15,17), or sudden cardiac death (<1%), although this presentation may be underestimated (22).

The right coronary artery (RCA) is most commonly involved in men (67%), whereas the LAD artery and the left circumflex artery are more commonly involved in women (87%). However, irrespective of gender, overall, the LAD is affected in 75–80% of the cases, and the RCA is affected in only 20% of cases (4). The usual location of the dissection is in the proximal portion of a single coronary artery, but involvement of multiple arteries has been described (8, 9). Concerning the prognostic correlations, left main or LAD disease, and multi-vessel dissection tend to indicate a poor outcome, commonly resulting in sudden death or extensive myocardial infarction (6).

Our case is uncommon presentation as SCAD occurred in the distal portion of the LAD, and caused ST-segment elevation seen on the inferolateral leads on ECG.

Per previous reported literature, in rare instances, acute inferior myocardial infarction may result from occlusion of the recurrent LAD branch, which is the terminal portion of a “wraparound” LAD. Consequently, besides a wraparound LAD, a more distally occluded LAD is also thought to be a prerequisite for an isoelectric or even elevated ST segment inferiorly (10).

Accurate and early diagnosis of SCAD is important because the management and investigation of SCAD is different from atherosclerotic disease. Coronary angiography is widely available and is the first-line imaging for patients presenting with ACS. However, coronary angiography has significant limitations in diagnosing SCAD because it is a 2-dimensional luminogram that does not image the arterial wall. Dedicated intracoronary imaging (optical coherence tomography [OCT] and intravascular ultrasound [IVUS]) that images the arterial wall layers improves SCAD diagnosis, but it is not as widely available and is associated with additional risks and costs. Thus, coronary angiography remains instrumental in SCAD diagnosis, and angiographers should gain familiarity with the angiographic variants of SCAD. Both OCT and IVUS provide complementary details to diagnose SCAD, which requires the presence of IMH or a double lumen. OCT has a superior spatial resolution of 10 to 20 mm versus IVUS, with lower resolution (~150 mm), but better penetration.

The optimal management of SCAD remains undetermined because no randomized trials have compared medical therapies or revascularization strategies, unlike atherosclerotic disease. Standard guideline indicated medical therapies administered for ACS have not been specifically studied for SCAD, and it is unclear if they are beneficial in this unique population (25). Current recommendations on management are largely used on the basis of expert opinions from observational series (11,24,26). Medical therapy includes Beta-Blockade, dual antiplatelet therapy (DAPT) could be empirically beneficial, the role of novel P2Y12 antagonists (Ticagrelor and Prasugrel) for SCAD management is undefined. Glycoprotein IIb/IIIa inhibitors have also not been evaluated for SCAD; however, they are not recommended because of their greater potency, higher bleeding risk, and a potential risk of extending the dissection (11). The role of anticoagulation for SCAD is controversial and has not been studied. Heparin agents are routinely administered for ACS management in hospital, but the clinical benefit has not been established for SCAD. There is a potential risk of extending the dissection with anticoagulation, which is balanced by the potential benefit of resolving overlying thrombus and improving true lumen patency. Heparin...
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should likely be discontinued once the SCAD diagnosis is made; however, this practice remains controversial and lacks supportive data. Thrombolytic therapy should be avoided in SCAD because there have been reports of harm and clinical deterioration due to extension of IMH and dissection (28, 29). The use of statins for SCAD is controversial. A small retrospective study demonstrated potentially higher SCAD recurrence with statins (13); however, the bulk of data for ACS demonstrates significant benefit with lipid lowering, and statins are routinely recommended post-MI (27). Because of the uncertainty and the general lack of atherosclerosis in SCAD patients, statins tend to only be administered to patients with pre-existing dyslipidemia. One series reported a high rate of statin use without apparent safety concerns (18).

Following diagnosis of SCAD, it has become routine practice in several centers specializing in SCAD to screen for predisposing arteriopathies associated with SCAD.

An overall conservative approach is preferred on the basis of expert opinions derived from observational data (11, 14–17). This recommendation relies on observations that SCAD arteries heal spontaneously in most cases, and that revascularization is associated with high failure rates. Nevertheless, a small proportion of patients should be considered for revascularization, including those with ongoing or recurrent ischemia, hemodynamic instability, ventricular arrhythmias, or left main dissection.

PCI should be performed in these cases if the anatomy is suitable; otherwise, CABG should be considered. Several series have reported poor technical success with PCI for SCAD related to iatrogenic dissection and propagation to distal portion (Not suitable for stenting, longer stents), difficulty advancing coronary wire into distal true lumen and risk of stent malapposition after resorption of IMH, with risk of late stent thrombosis.

Patients are advised to avoid lifting weights >20 pounds, and to have a low target exercise heart rate and systolic blood pressure with this program. SCAD patients who participated in the dedicated rehabilitation program had lower long-term MACEs compared with those who did not participate (31).

The in-hospital outcomes of SCAD patients are reasonably good in contemporary prospective series. Acute in-hospital mortality was <5% in modern series, and in-hospital recurrent MI, need for urgent revascularization in conservatively managed patients, or other MACEs were 5% to 10% (13, 15, 17). However, following hospital discharge, a significant proportion of patients can have recurrent chest pains and MACEs. Subacute MACEs were reported in 10% to 20% of patients at 2-year follow-up, with recurrent SCAD occurring in 15% (15). Longer-term recurrent SCAD rates at 4 to 5 years were reported at ~27% (16, 21). Although overall long-term survival is good in this cohort (>95%), long-term MACE rates can be high, and were reported to be 15% to 37% at 5 to 7 years and estimated at ~50% at 10 years (13, 15, 17, 21). In the updated prospectively followed cohort of 280 patients, MACEs were 20.4% at a median 2.3-year follow-up; recurrent MI was 19.0%, with a recurrent SCAD rate of 12.2% (30). Such high long-term MACE rates emphasize the importance of close follow-up of SCAD survivors by cardiovascular specialists. Of note, patients with post-partum SCAD may have worse prognoses than other SCAD cohorts. In a small retrospective series, post-partum patients had larger infarcts, lower mean left ventricular ejection fractions, and tended to have more proximal artery dissections (32).

We report an unusual case of distal LAD spiral spontaneous dissection in young male without obvious triggers, however, it is unclear whether his acute diabetic ketoacidosis and tobacco use may contribute to his presentation. With the exception of smoking, hypertension and DM, no other risk factors were present in our patient. It is noteworthy that the distal portion of the LAD is an unusual location for dissection in male like our case. PCI was not performed due to distal location of the dissection, vessel diameter <2.5 mm and the risk of propagation.
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We did not use a GP IIb/IIIa inhibitor in the present patient because of clinical success with dual antiplatelet therapy and heparin. Warfarin was initiated with INR goal of 2-3. The prognosis was favorable in our case and he was discharged home in stable condition with outpatient follow up plan.

CONCLUSION

SCAD is a rare entity, often underdiagnosed, however, it is increasingly being recognized as a significant cause of ACS. It is frequently associated with predisposing and precipitating factors. Coronary angiography with IVUS or OCT remains instrumental in SCAD diagnosis. Conservative therapy is preferred, except for patients with unstable symptoms, hemodynamic instability or left main dissection. Overall survival is good; however, long-term MACEs are frequent, including recurrent SCAD, therefore, SCAD patients should be closely followed for cardiac events.

REFERENCES


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