Recurrent Idiopathic Spontaneous Coronary Artery Dissection: A Case Report and a Review

1. Introduction:

Spontaneous coronary artery dissection (SCAD) is a rare cause of acute coronary syndrome (ACS).[7] Traditionally, SCAD has been observed in two groups: those with coronary atherosclerosis and women in the peripartum period.[1, 9, 14] Other less common associations were prescribed, such as vigorous exercise, vasoactive drug use, inflammatory conditions, and connective tissue diseases.[16-19, 27] SCAD is considered idiopathic when all of the previously described associations are excluded.[6] We report a case of recurrent idiopathic SCAD, and propose a systematic approach for identifying the etiology behind SCAD.

2. Case:

A 52-year-old woman presented to the emergency department (ED) with unstable angina. The electrocardiogram (ECG) was unremarkable and Troponin I was mildly elevated at 0.477ng/mL. The patient had no significant risk factors for coronary artery disease (CAD), was not on hormone replacement therapy and was under no physical dynamic stress. Coronary angiography revealed a spontaneous dissection of the right posterior descending artery (PDA) (Image 1). The lesion was deemed unsuitable for intervention due to its small lumen size. For the same reason intravascular ultrasound (IVUS) was not performed. The patient was subsequently discharged and treated medically with aspirin, clopidogrel, beta-blocker and simvastatin.

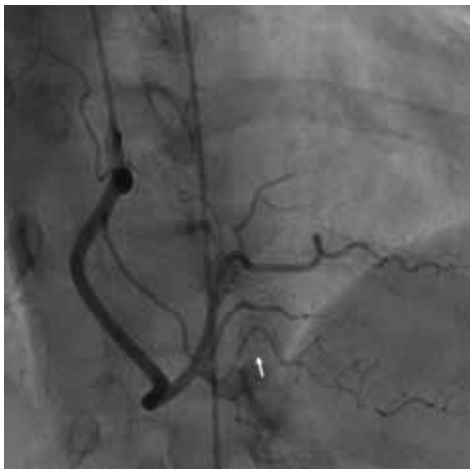


Image 1. Spontaneous coronary artery dissection of right posterior descending artery.

Two-and-a-half months following the inciting event, the patient again presented to the ED with unstable angina. ECG showed ischemic ST-T wave changes (Figure1) with elevated CPK of 203U/L, CK-MB 16.6ng/mL and Troponin I of 6.210ng/mL. A repeat coronary angiography showed complete resolution of the previous PDA dissection (Image 2), however a new spontaneous dissection in the second diagonal branch of the LAD (Image 3), not previously present on angiogram (Image 4), was noted. Again, secondary to the small lumen size, medical management was determined to be the best form of treatment.

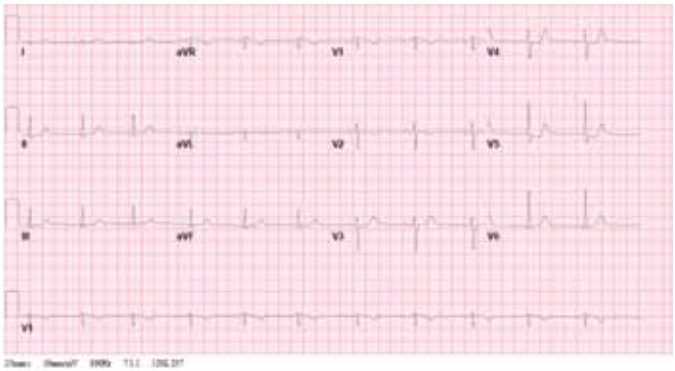


Figure 1

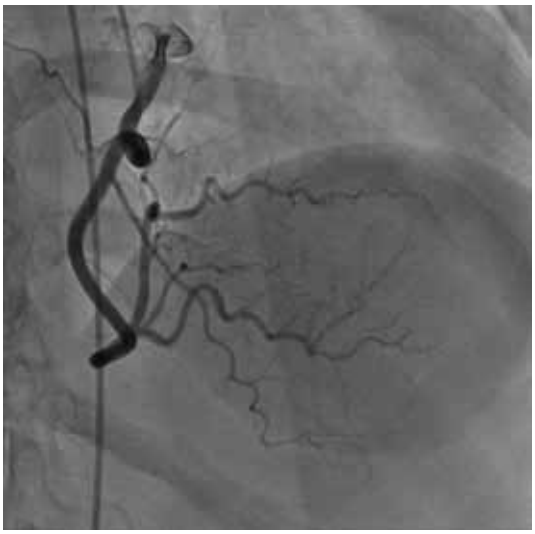


Image 2. Complete healing of the previous spontaneous coronary artery dissection of right posterior descending artery.

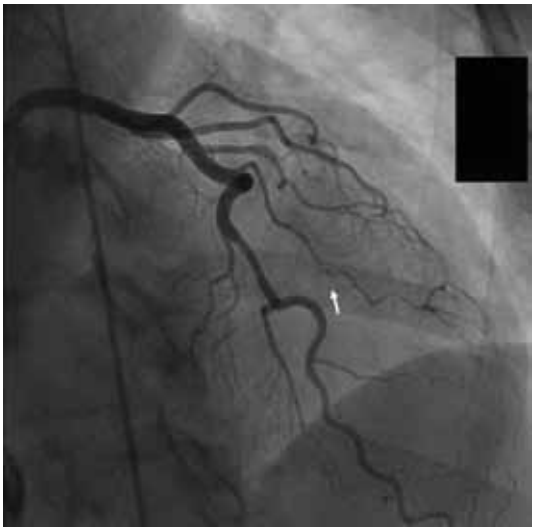


Image 3. New spontaneous coronary artery dissection of the second diagonal branch of the left anterior descending artery.

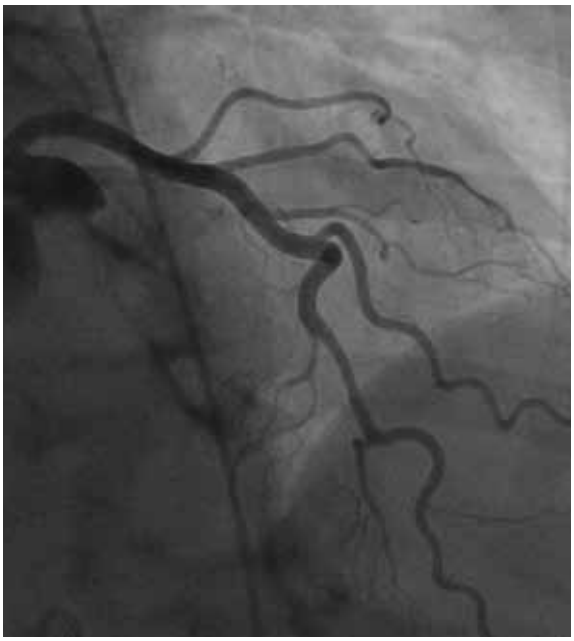


Image 4. Absence of coronary artery dissection of the second diagonal branch of the left anterior descending artery in first angiography.

In an attempt to reveal the etiology of this patient's recurrent SCADs a comprehensive work-up was done, including: proteinase 3 antibody test, myeloperoxidase antibody test, antinuclear antibody titers, erythrocyte sedimentation rate, C-reactive protein, C3 and C4 complement levels. The findings of these tests were within normal limits. Lipid panel revealed no significant dyslipidemia. Genetic testing for hereditary connective tissue disorders was not conducted because no aortic root dilation was noted on echocardiogram. Urine analysis was negative for toxicology. Following this work-up and patient history, a diagnosis of recurrent idiopathic spontaneous coronary artery dissection (SCAD) was made.

3. Discussion:

3.1. Pathology:

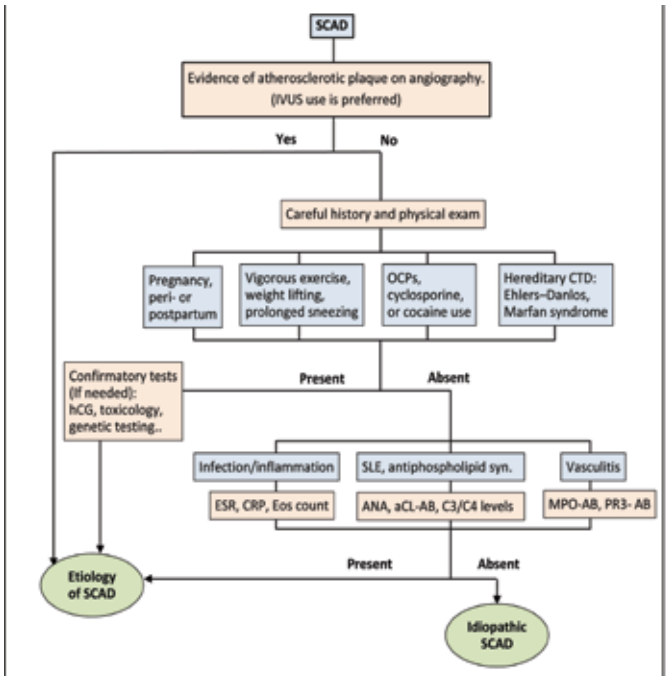
By definition, SCAD is coronary artery dissection that does not extend from the aortic root and is not caused by cardiac surgery, trauma, coronary angiography or percutaneous intervention.[1] SCAD is described much like aortic dissection, with a disruption to the vascular endothelium causing accumulation of blood or thrombus that leads to the formation of a false lumen.[2] The false lumen can compress the true lumen and create an ischemic event that is detrimental to the myocardium distal to the lesion.[3] The hematoma or thrombus can be due to an intimal tear or a medial dissection leading to bleeding from the vasa vasorum causing a hematoma with no associated intimal tear; the latter is generally more common.[4, 5, 6]

3.2. Epidemiology and Etiology:

SCAD has an incidence of 0.1 to 1.1 % on angiography, but most cases go undiscovered until post mortem assessment, or are asymptomatic and go undiagnosed.[9-12] In cases of SCAD the left anterior descending artery (LAD) is the more commonly involved, in about 80 % of cases.[3, 8] The right coronary artery (RCA) is implicated more commonly in men and the LAD in women.[13] SCAD has a predilection for younger females in the peri- and postpartum periods, but is still a rare cause of acute myocardial infarction, affecting only 1 in 100,000 pregnancies.[14] In SCAD cases with underlying CAD, ruptured plaque was found in the majority.[9] It has also been associated with instances of increased shear stress such as intense exercise, hereditary connective tissue disorders (Marfan's Syndrome), inflammatory disorders including vasculitis, and vasoactive drugs like cocaine, cyclosporine and ergotamine.[27]

The pathophysiology of SCAD has not been fully understood, but it has been found that changes in the vascular architecture in conjunction with increased shear forces can lead to the lesion. In the gravid or postpartum patient, hormonal fluctuations affect collagen, smooth muscle cells and proteoglycan matrix production.[14, 15] These changes weaken the vessel wall, increasing the likelihood of SCAD. This phenomenon has occurred with the use of oral contraceptives and menstruation, supporting the idea that hormonal involvement is associated with the pathophysiology.[16, 17, 18] Inflammatory causes have also been proposed based on eosinophilic involvement in up to 43 % of SCAD cases.[19] Eosinophils contain cytotoxic enzymes, released by degranulation, that may damage the vasculature. Their presence may also merely represent an inflammatory reaction to vascular damage already present.

A systematic approach to identifying the etiology of SCAD is lacking. Based on previous SCAD reports and its common etiologies, we propose a work-up algorithm to identify the cause of SCAD (Figure 2).



Algorithm for working-up etiology of spontaneous coronary artery dissection.

SCAD = spontaneous coronary artery dissection; IVUS = intravascular ultrasound; OCPs = Oral contraceptives pill; CTD = Connective tissue disorder; hCG = human chorionic gonadotrophin; SLE = Systemic lupus erythematosus; ESR = erythrocyte sedimentation rate; CRP = C-reactive protein; Eos = eosinophils; ANA = antinuclear antibody; aCL-AB = anticardiolipin antibody; MPO-AB = myeloperoxidase antibody; PR3-AB = Proteinase 3 antibody.

Figure 2

3.3. Clinical Presentation and Diagnosis:
SCAD can present as ACS or sudden cardiac death (SCD).[7] Diagnosis has thus far depended on coronary angiography or autopsy. Angiography may depict a double radiopaque lumen and a radiolucent intimal flap or trapping of contrast in the false lumen with slow clearance.[20] The aforementioned presentation will only occur when an intimal tear is the cause of the dissection, but as previously discussed this may not be the most likely pathology. A hematoma between the media and adventitia may easily be missed on angiography or be misdiagnosed as atherosclerotic stenosis. In such cases intravascular ultrasound (IVUS) can provide a superior diagnostic tool,[4] and can also reveal angiographically "missed" ruptured plaque causing the dissection.[9] However, the availability of this technology and the small lumen of the artery are frequent challenges for its routine use.[4,9] Contrast tomography (CT) angiography has also been shown to have a role in SCAD diagnosis. CT angiography is a non-invasive method that can accurately image the intimal flap or intramural hematoma and can be an alternative to coronary angiography when following the progression of SCAD in a medically managed patient.[21-23]

3.4. Management:
Many of the treatment modalities have been derived from case reports. The mainstay of treatment includes medical management, percutaneous intervention and surgical revascularisation. The treatment of choice is dependent on the presentation, location and extent of the dissection. Medical management includes antiplatelet therapy, beta-blockers, and heparin or enoxaparin, and has been shown to be successful, allowing for spontaneous healing of the dissection.[24] Antiplatelet agents may reduce the contents within the false lumen and decrease compression.[25] Beta-blockers reduce stress on the vessels by decreasing force, and it has been theorised that they can accomplish the same effect on coronary vessels.[27] Anticoagulation therapy has been debated due to some evidence that it may cause the dissection to propagate by increasing hemorrhage into the dissection.[27] Conservative follow-up is recommended to monitor the progression of the lesion. Some patients, like ours, have shown complete resolution; others have experienced recurrence and acute deterioration and propagation.

Percutaneous coronary intervention (PCI) has shown mixed outcomes. It has been recommended that if the dissection does not lead to compression of the true lumen, stenting should be avoided and medical management considered.[27] PCI can cause extension of the false lumen and dissection by contrast and guide wire.[27] Nevertheless, stent placement should strengthen the vessel wall and successfully seal the dissection.
Percutaneous dilation without stent deployment should be avoided in the treatment of SCAD because it has a higher likelihood of extending the dissection.[8] Petronio *et al*.[26] described an approach that deploys the stent only at the proximal edge of the dissection to resolve the acute ischemia, which may reduce the incidence of dissection progression and the need for extensive stenting.

A total occlusion of the lumen may call for surgical revascularisation to a viable distal target. Other instances that lead to surgical intervention include: SCAD presenting with severe ischemia, cardiogenic shock, three-vessel involvement, left main dissection, or patients who do not respond to alternate forms of management.[13, 5, 8]

Due to the variability in presentation of SCAD, a "one size fits all" treatment option does not exist. No treatment is currently considered superior, and management approach should be individually tailored. Careful consideration of the location, extent, severity, and stability of the lesion should be undertaken.

4. Conclusion:
Little is known about SCAD, due to its low incidence and "devious" presentation. It mainly occurs in patients with CAD, peri- and postpartum young females and less frequently with other conditions that may increase shear stress on arterial wall, or affect its architecture. Medical management is typically the default as PCI and surgical revascularisation have their limitations. Our proposed systemic approach should help the clinician in identifying the etiology of SCAD, which in some rare instances may not be revealed. The idiopathic SCAD case that we reported is one example. Therefore, more research is needed to further investigate the mechanism by which SCAD occurs so as to tailor future therapies. As it was once said: "If you know your enemies you will not be imperiled in a hundred battles, but if you do not know your enemies you will win one and lose one."[28]

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