A Problem in the Main Street: Spontaneous Coronary Artery Dissection in The Left

Jawad Shabani[1], Vaibhav Shah [1], David Song[2], Abhigan Shrestha[3], Vikash Jaiswal[4], and Chadi M Alraies[5]

[1]Icahn School of Medicine at Mount Sinai
[2]Icahn School of Medicine at Mount Sinai Elmhurst Hospital
[3]M Abdur Rahim Medical Medical College Hospital
[4]AMA School of Medicine
[5]Detroit Medical Center

March 11, 2024

Introduction:

Spontaneous coronary artery dissection (SCAD) is a tear or separation within the coronary artery wall forming an intraluminal hematoma reducing downstream blood flow leading to myocardial ischemia (MI)**(Figure 1A-C)[1]** . Intraluminal hematomas may develop from "inside-out" due to endothelial-intimal injury or "outside-in" due to injury within the vasa vasorum leading to bleeding in the wall. The subsequent false lumen from either mechanism can extend and compress the true lumen to cause MI [2]. Unlike other luminal etiologies of MI, SCAD is not caused by atherosclerotic plaque rupture, coronary intervention, or trauma [3]. With significant MI from intraluminal hematoma, patients can present with signs and symptoms that mimic MI including chest pain with and without radiation, dyspnea, nausea and vomiting. It can also include elevated troponin with electrocardiogram (ECG) changes consistent with ST segment elevation or non-ST segment elevation MI. The diagnosis is made when other etiologies of acute coronary syndrome (ACS) are ruled out and there is an angiographic evidence of non-iatrogenic or non-atherosclerotic radiolucent intimal flap and contrast staining [1]. Left anterior descending (LAD) artery is the most commonly affected branch of the left main coronary [4]. Severe stenosis can lead to heart failure as well as cardiogenic shock amongst other post-MI complications including ventricular arrhythmias, ventricular free wall or septal wall rupture. If angiography is inconclusive, then intracoronary imaging with optical coherence tomography or intravascular ultrasound may be considered before repeat angiography [5].

The etiology of SCAD is thought to be multifactorial without a clear underlying cause. Risk factors associated include fibromuscular dysplasia (FMD), peri and postpartum period, hormonal fluctuations, and arteriopathies. SCAD is a rare cause of ACS, although when present, there is a higher prevalence in young females without cardiac risk factors [6]. Stable patients are treated conservatively with medication while percutaneous coronary intervention (PCI) and coronary artery bypass graft are reserved for severe stenosis and acute change in hemodynamics. Use of antiplatelet therapy remains controversial as medications such as acetylsalicylic acid (i.e. aspirin) and P2Y12 receptor blockers can worsen intraluminal hematoma. Dual antiplatelet therapy is recommended for those who undergo stent placement [5]. Similarly, guideline directed medical therapy is initiated for new onset of heart failure. Review of literature reveals 90% recovery within one month with coronary computed tomography angiography [5]. Many cases of SCAD have been reported in young females with ischemic symptoms and angiographic findings in the distal LAD but only few cases highlight proximal coronary involvement. We present a case of a young woman with extensive left main and proximal LAD SCAD requiring intervention.

Case History/examination:

A 29-year-old Colombian woman with a history of an unknown congenital cardiac condition, with a high suspicion for coarctation of the aorta that was surgically corrected for artery narrowing as a neonate in Colombia, presented to the emergency department with chest pain. The chest pain was sudden onset, dull and squeezing in nature that localized to the mid-sternum that radiates to the right side of her neck and right arm. It started while she was at rest in a seated position that lasted for 15 minutes before self-resolving associated with cramping, numbness and tingling. There were no exacerbating or relieving factors. She has never experienced the same or similar type of chest pain in the past. The patient denied fevers, cough, orthopnea, paroxysmal nocturnal dyspnea, nausea, vomiting, diaphoresis, swelling, abdominal pain, sick contacts, or significant emotional stressors. She was not taking any medications or supplements at home. She denied current or previous tobacco, alcohol, caffeine, energy drink and illicit drug use. There was no known family history of cardiac disease.

Patient's vital signs on admission were blood pressure of 106/73 mm Hg, pulse of 76 beats per minute, afebrile, and breathing comfortably on room air. Physical examination was unrevealing.

Methods:

Chest plain radiograph did not reveal any acute cardiopulmonary abnormalities. The initial ECG showed T-wave inversion in lead V1 and biphasic T-wave in lead V2 **(Figure 2A)** , while the subsequent ECG one hour later showed persistent T-wave inversion in lead V1, V2, but a new biphasic T-wave in lead V3 **(Figure 2B)** . Cardiology was consulted who recommended serial cardiac troponin which continued to increase to 0.241 ng/mL, 0.650 ng/mL, and 0.728 ng/mL. The patient was given an aspirin load of 324 mg and urgent echocardiography revealed a mildly reduced ejection fraction of 45% with akinetic apical anterior, apical septal, apical inferior, apical lateral and apex left ventricular wall segments **(Figure 3A-B)** . She was started on a heparin, eptifibatide, and nitroglycerin. A cardiac catheterization was performed and revealed coronary dissection in the left main coronary artery from the ostium to the distal in addition to a second dissection in the proximal and mid LAD **(Figure 4A-B)** . Three drug-eluting stents, one in the left main coronary and two in the proximal and mid LAD, were placed without complications. The patient was started on aspirin 81 mg daily, clopidogrel 75 mg daily, atorvastatin 40 mg daily, lisinopril 5 mg daily, and metoprolol succinate 50 mg daily. Initial FMD workup including computed tomography angiography (CTA) of the head, neck and chest was unremarkable. The patient was safely discharged home with plans to enroll in the SCAD registry and repeat echocardiogram to assess for left ventricle recovery. Unfortunately, the patient was lost to follow up despite multiple attempts to establish communication with the patient by the cardiology team.

Conclusion and results:

SCAD is becoming an entity that is more recognized, especially in young women without prior risk factors. Distal LAD is the most affected vessel, mostly managed conservatively, but proximal SCAD is rare and requires invasive treatment such as PCI, increasing the risk of possible complications. Therefore, the location of the lesion is critical to risk stratify the patient and requires close follow up with enrollment in the national SCAD registry to help researchers better understand this condition.

Discussion

SCAD mainly affects middle-aged women comprising 87% to 95% of all cases with a mean age ranging between 44 to 53 years. There also have been few cases reported in teens and elderly as well. Out of all patients who present with ACS, SCAD's prevalence is estimated to be 4%. In addition, SCAD accounts for up to 35% of ACS in women under 50 years of age, a number that may be underestimated given that SCAD is often both underdiagnosed and misdiagnosed. According to multiple studies, SCAD often co-exists with other conditions and is initiated by various triggers. In a descending order of prevalence, such co-existing conditions may include FMD, hypertension, connective tissue disorders, migraine, genetic susceptibility, and pregnancy. Some of the triggers may include hormone imbalance, significant emotional distress, intense

exercise, medications and illicit substances [8, 10-11].

One of the developing and well-supported explanations for the pathophysiologic emergence of SCAD is the "outside-in" hypothesis. It attributes the initiating event to a primary hemorrhage of a vaso vasorum within the medial coronary layer. As a sequelae, the new-born hematoma may either self-dissolve or further expand in a longitudinal and circumferential fashion, creating a false lumen that eventually accumulates to compress and narrow the true lumen causing ischemic symptoms. SCAD is classified into 4 types based on angiographic appearance. Type 1 is pathognomonic arterial wall staining by contrast dye with multiple radiolucent lumens. Type 2 is diffuse, long, and smooth stenosis of variable degrees. Type 3 often mimics atherosclerotic lesions and is the most often to be misdiagnosed requiring further characterization and specialized imaging. Type 4 is complete occlusion of distal coronaries mimicking coronary embolism **(Figure 5)**[9] . The majority of SCAD cases present as type 2, which is mainly seen in the mid-to-distal left anterior descending coronary artery lesions [10].

Although advancements in the understanding of SCAD's pathophysiology have been made, the predilection to certain sides and segments of the coronary arteries remains unclear. SCAD tends to present in the distal coronary arteries and its smaller branches. Specifically, it has the propensity to affect the mid-to-distal portions of the LAD. [8,10]. In a study by Jackson et al that focused on the pathophysiologic mechanisms through optical coherence tomography of 65 SCAD cases, the percentage affecting the left main stem and the proximal LAD coronary artery were 1% and 4%, respectively. In comparison, the percentage found within the distal portions of the LAD coronary artery was 45% [12]. Studies have yet to explore this regional favoritism of SCAD, but it is thought to be influenced by physical mechanics of blood flow, native structural differences within certain areas of the tunica media, or a combination of both. Thus far, the literature has not revealed such explanations to any epidemiological or genetic basis for a high prevalence within the aforementioned coronary segments.

Although guideline-supported treatment of SCAD, once confirmed by coronary angiography, is stent; a growing number of clinicians and centers opt to manage conservatively without PCI, especially if coronary flow is preserved and the lesion is distal. As high as 95% of such cases commonly undergo spontaneous resolution within 30 days [13]. Conservative management is favored because of the unpredictable outcomes of PCI as well as the myriad of complications including PCI failure due to unwanted false lumen interactions with the wire or the stent itself, or iatrogenic expansion of the dissection which happens in up to one third of cases.In a study by Tweet et al, proximal and ostial left main coronary arteries in SCAD had the lowest rates of conservative treatment compared to lesions in the mid-to-distal portions.[14]. Therefore, patients with proximal SCAD lesions, such as ours, are at a propagated disadvantage given the innate risks associated with intra-angiographic and post-PCI outcomes.

References:

1. Hayes, Sharonne N et al. "Spontaneous Coronary Artery Dissection: Current State of the Science: A Scientific Statement From the American Heart Association." *Circulation* vol. 137,19 (2018): e523-e557. doi:10.1161/CIR.0000000000000564
2. Garcia-Guimarães, M., Bastante, T., Antuña, P., Jimenez, C., de la Cuerda, F., Cuesta, J., Rivero, F., Premawardhana, D., Adlam, D., & Alfonso, F. (2020). Spontaneous Coronary Artery Dissection: Mechanisms, Diagnosis and Management. *European cardiology* ,*15* , 1–8. https://doi.org/10.15420/ecr.2019.01
3. Yip, Amelia, and Jacqueline Saw. "Spontaneous coronary artery dissection-A review." *Cardiovascular diagnosis and therapy*vol. 5,1 (2015): 37-48. doi:10.3978/j.issn.2223-3652.2015.01.08
4. Tweet, Marysia S et al. "Clinical features, management, and prognosis of spontaneous coronary artery dissection." *Circulation* vol. 126,5 (2012): 579-88. doi:10.1161/CIRCULATIONAHA.112.105718
5. Pristera, Nicole et al. "Spontaneous coronary artery dissection: Principles of management." *Cleveland Clinic journal of medicine* vol. 88,11 623-630. 2 Nov. 2021, doi:10.3949/ccjm.88a.20162
6. Nakashima, Takahiro et al. "Prognostic impact of spontaneous coronary artery dissection in young female patients with acute myocardial infarction: A report from the Angina Pectoris-Myocardial In-

farction Multicenter Investigators in Japan." *International journal of cardiology* vol. 207 (2016): 341-8. doi:10.1016/j.ijcard.2016.01.188

7. Clare R., Duan L., Phan D., et al. "Characteristics and clinical outcomes of patients with spontaneous coronary artery dissection". J Am Heart Assoc 2019;8:e012570.
8. Saw J. Coronary angiogram classification of spontaneous coronary artery dissection. *Catheterization and cardiovascular interventions* . 2014;84(7):1115-1122. doi:10.1002/ccd.25293
9. Saw, J.; Humphries, K.; Aymong, E.; Sedlak, T.; Prakash, R.; Starovoytov, A.; Mancini, G.J. Spontaneous Coronary Artery Dissection: Clinical Outcomes and Risk of Recurrence. *J. Am. Coll. Cardiol.***2017** , *70* , 1148–1158.
10. Hayes SN, Tweet MS, Adlam D, et al. Spontaneous Coronary Artery Dissection: JACC State-of-the-Art Review. *Journal of the American College of Cardiology* . 2020;76(8):961-984. doi:10.1016/j.jacc.2020.05.084
11. Saw J, Aymong E, Sedlak T, Buller CE, Starovoytov A, Ricci D, Robinson S, Vuurmans T, Gao M, Humphries K, Mancini GB. Spontaneous coronary artery dissection: association with predisposing arteriopathies and precipitating stressors and cardiovascular outcomes. Circ Cardiovasc Interv. 2014 Oct;7(5):645-55. doi: 10.1161/CIRCINTERVENTIONS.114.001760. Epub 2014 Oct 7. PMID: 25294399.
12. Jackson R, Al-Hussaini A, Joseph S, et al. Spontaneous Coronary Artery Dissection Pathophysiological Insights From Optical Coherence Tomography. *JACC Cardiovascular imaging* . 2019;12(12):2475-2488. doi:10.1016/j.jcmg.2019.01.015
13. Hassan S, Prakash R, Starovoytov A, Saw J. Natural History of Spontaneous Coronary Artery Dissection With Spontaneous Angiographic Healing. *JACC Cardiovascular interventions* . 2019;12(6):518-527. doi:10.1016/j.jcin.2018.12.011
14. Tweet MS, Eleid MF, Best PJ., et al. Spontaneous Coronary Artery Dissection: Revascularization Versus Conservative Therapy.*Circulation Cardiovascular interventions* . 2014;7(6):777-786. doi:10.1161/CIRCINTERVENTIONS.114.001659

Images:

Figure 1A-C: Hayes et al [1] illustration of coronary artery intramural hematoma formation. A: Normal coronary artery. B: Coronary artery with intramural hematoma. C: Coronary artery with intimal tear. Spontaneous coronary artery dissection is characterized by the spontaneous formation of an intramural hematoma, which can lead to compression of the true lumen and myocardial infarction. An intimal tear may be present.

Figure 2A-B: A, initial electrocardiogram: V1 T-wave inversions and V2 biphasic T-waves. B, repeat electrocardiogram: V1 persistent T-wave inversions, V2 T-wave inversions, and V3 biphasic T-waves.

Figure 3A-B: Echocardiographic 4 chamber view during ventricular diastole (A, left) followed by ventricular systole (B, right) showing mildly reduced ejection fraction (45%) with akinetic apical left ventricular wall segments: anterior, septal, inferior, lateral including the apex.

Figure 4 A-B: A (left) : Coronary angiography showing luminal narrowing of left main coronary artery (red arrow) and radiolucent intraluminal hematoma (blue circle) in the proximal LAD with long narrowing (blue arrows) in the middle LAD. **B (right)** : Post PCI coronary angiography with restoration of normal caliber in the left main coronary (red arrow) as well as the proximal and middle LAD (blue arrows).

Figure 5: Spontaneous coronary artery dissection Type 1-4 [8]

Hosted file

`fig1.docx` available at https://authorea.com/users/566196/articles/724010-a-problem-in-the-main-street-spontaneous-coronary-artery-dissection-in-the-left

Hosted file

fig2.docx available at https://authorea.com/users/566196/articles/724010-a-problem-in-the-main-street-spontaneous-coronary-artery-dissection-in-the-left

Hosted file

fig3.docx available at https://authorea.com/users/566196/articles/724010-a-problem-in-the-main-street-spontaneous-coronary-artery-dissection-in-the-left

Hosted file

fig4.docx available at https://authorea.com/users/566196/articles/724010-a-problem-in-the-main-street-spontaneous-coronary-artery-dissection-in-the-left

Hosted file

fig5.docx available at https://authorea.com/users/566196/articles/724010-a-problem-in-the-main-street-spontaneous-coronary-artery-dissection-in-the-left