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Case Report

Intracoronary Imaging Isolates Type 3 Spontaneous Coronary Artery Dissection of the Left Main -

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KEYWORDS: Spontaneous Coronary Artery Dissection (SCAD); Intramural Hematoma; Intravascular ultrasound (IVUS); Optical Coherence Tomography (OCT); Post-Partum; Fibromuscular Dysplasia (FMD)

INTRODUCTION

Spontaneous Coronary Artery Dissection (SCAD) is an often under diagnosed condition given its spectrum of clinical presentation, thus posing a significant diagnostic dilemma for clinicians. Pathologically, it occurs as a consequence of non-traumatic, non-iatrogenic separation of arterial walls, creating a false lumen with intramural hematoma (IMH) formation, which may compromise anterograde blood flow and cause ischemia [1]. First defined in 1931 via a post-mortem study, and arbitrarily labeled as a predominantly idiopathic condition, the recent advent and utilization of intracoronary imaging has increased the diagnostic yield of SCAD, highlighted multiple associated predisposing risk factors, and defined the disease subtypes.

CASE SERIES

A 37-year-old woman 2 weeks postpartum, complicated by preeclampsia and gestational diabetes, who initially complained of substernal chest pain and dizziness, was found by the emergency medical services immediately after losing consciousness. On cardiac monitoring, she was in Ventricular Fibrillation (VF), and was successfully defibrillated with 200 Joules.

In the emergency room, she was hemodynamically stable with complaints of persistent substernal chest pain. Her initial electrocardiogram (ECG) showed sinus tachycardia without active ischemic changes. She developed progressive shortness of breath, with coarse crackles at bilateral lower lung fields, and significant jugular venous distension. She was subsequently intubated for acute hypoxemic respiratory failure due to cardiogenic pulmonary edema. A repeat ECG now showed ST segment elevations in leads V1 and aVR, with extensive ST depression in the inferior and anterolateral leads (Figure 1). Emergent coronary angiogram showed a right dominant circulation, with 40% stenosis of the mid left main coronary artery (LM). Remaining coronary circulation shows no significant atherosclerotic disease and an Ejection Fraction (EF) was estimated at 25%, with a left ventricular end diastolic pressure of 32mmHg (Figure 2).

She was transferred to the cardiac ICU for further management. An ECG within an hour after catheterization showed resolution of the ST segment deviations. She was started on intravenous diuretics with improvement in hypoxemia and eventually extubated. Initial troponin I was 0.019 with a peak of 68 (normal range 0–0.05 ug/l) and peak creatinine phosphokinase of 313U/l (normal range 24–170 U/l). A follow up trans-thoracic echocardiogram within 24 hours showed an EF of 49%, moderate diffuse hypokinesia, with mildly increased wall thickness. Her clinical status improved significantly with guideline-directed heart failure therapy.

A Cardiac MRI revealed normalization of her EF to 65%, however with antero-septal hyper-enhancement consistent with a small, subendocardial infarct. With a newfound suspicion for a true ischemic event as the etiology of her cardiac arrest, a repeat coronary angiogram coupled with IVUS was performed, which revealed a plaque with IMH compromising the lumen of the mid segment of the LM, with a minimal luminal area of 6mm² (Figure 3). Interestingly, no dissection plane was identified and the coronary

circulation continued to show non-significant atherosclerotic disease. Based on these findings, a diagnosis of LM Type 3 SCAD with IMH formation was made. A consideration for Implantable Cardioverter Defibrillator (ICD) implantation for secondary prevention of Sudden Cardiac Death (SCD) was made, given her unique presentation with VF. She was discharged with a life vest as she deferred subcutaneous ICD placement. On post subsequent post-hospitalization follow

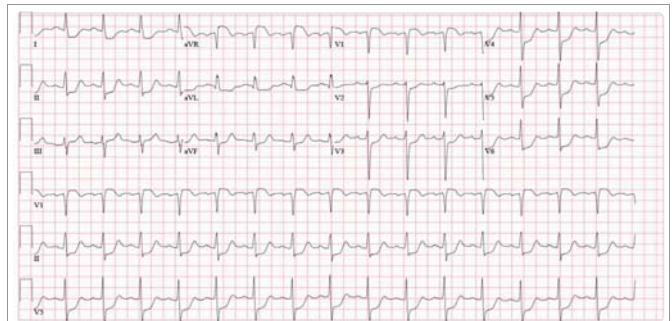


Figure 1: 12-Lead ECG: Sinus Tachycardia, ST segment elevation in leads aVR and V1, with diffuse ST segment depressions.



Figure 2: Coronary Angiography: Left main coronary with 40% stenosis, EF = 25 % and severely elevated LVEDP = 32 mmHg, TIMI 3 anterograde flow.

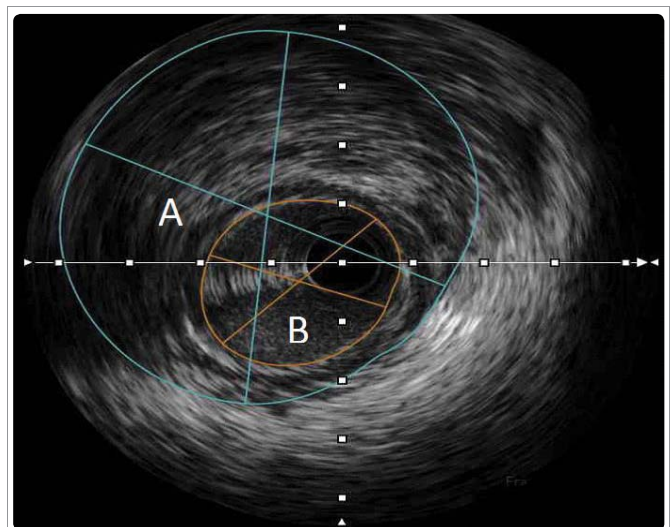


Figure 3: IVUS: Intramural hematoma (A) within the media of the arterial wall resulted in compression of the real lumen (B). An intimal tear was not observed.

up, she has remained asymptomatic and clinically euvoletic. A repeat TTE six months after presentation showed normal left and right ventricular function, no regional wall motion abnormalities, normal diastolic function, and no valvular disease. She is scheduled to undergo a nuclear stress test for surveillance of LM disease.

DISCUSSION

SCAD could mimic an acute myocardial infarction with similar presenting symptoms, electrocardiographic findings, and serological elevation of cardiac biomarkers. In contrast to ACS, SCAD is notably a non-atherosclerotic entity, induced either by a tear in the intimal layer of the arterial wall or by rupture of the supporting vasa vasorum, with a subsequent separation between intima and media, or media and adventitia [2]. An IMH forms as blood accumulates extraluminally, compressing the vessel, and consequentially reducing anterograde blood flow. As a result, patients could develop myocardial ischemia, infarction, ventricular arrhythmias, or sudden cardiac death [3]. In the University of British Columbia (UBC) series, the largest SCAD registry to date, all cases presented with an elevation of troponin, 26% with STEMI, and a smaller percentage (3.6%) with ventricular arrhythmias [4].

Previously defined as an idiopathic condition of young women and identified primarily on post-mortem studies, advances in intracoronary imaging has enabled better understanding of the disease pathophysiology. The reported prevalence of SCAD may be as high as 8.7% in women presenting with ACS and 10.8% in a subgroup presenting with ST segment elevation [5]. In the UBC series, the average age was 52.1 +/- 9.2 years, with 58% presenting at age 50 years or older. In the same study, angiographic review identified SCAD in as high as 24% of women < 50 years of age who had an MI [4].

Postulated risk factors include fibromuscular dysplasia (FMD), connective tissue disorders, systemic inflammatory diseases, as well as pregnancy [6]. Of the 168 patients with SCAD in the UBC series, 72% were found to have FMD [4]. Given the infrequency of both conditions, a causal implication was suggested as the pathophysiological changes associated with FMD could cause weakened arterial walls, predisposing coronary arterial segments to dissection. Similarly, a retrospective study conducted at the Mayo Clinic that involved 200 patients, also found iliac FMD in 50% of femoral angiograms performed on SCAD patients [7].

Peripartum SCAD has gained significant interest in the recent years. The UBC series reported a rate of 3-8% of pregnancy related SCAD, with the highest frequency noted during the first post-partum month, with a peak in the second week [5]. The hormonal and hemodynamic changes that occur during pregnancy have been implicated in creating a weakened media due to impaired collagen synthesis, generating a hypercoagulable or prothrombic milieu, increasing the shear stress from augmented cardiac output and increased circulatory volume, with a cumulative risk of false lumen creation and thrombosis [8,9].

SCAD may involve any coronary artery, however it has been shown that the proximal, mid, and distal LAD are more frequently affected. In the Mayo series, the frequency of LM involvement as compared to other coronary branches was only 1.2% [7]. With the emergence of intracoronary imaging, three distinct angiographic patterns of SCAD have been described. Type 1 illustrates the pathognomonic angiographic appearance of SCAD with contrast staining of both the arterial wall and false lumen of the dissected lesion (Figure 4). Type 2 involves stenosis of varying severity and usually affects the mid to

distal segments of coronary arteries (Figure 5). Type 3, also known as angiographically silent SCAD, is notoriously the most challenging to differentiate from atherosclerotic disease and requires intracoronary imaging for definitive diagnosis. Angiographic features include the lack of atherosclerotic changes in other coronary arteries, longer lesions between 11-20 mm, hazy, and linear stenosis [6]. Interestingly, the UBC series reported that only 3.9% had Type 3 SCAD whereas an overwhelming majority (67%) had Type 2 [4]. It was also noted in this series that recurrent dissection occurred in 13.1% of cases, compared to the Mayo Clinic study which observed a rate of 17% [6].

Requiring a high index of suspicion for SCAD, angiographers must consider intracoronary imaging for definitive diagnosis. Conventional, gold standard angiography is a 2-dimensional luminogram that can effectively depict luminal narrowing in Type 1 disease, but has been shown to be inferior for assessing arterial wall structure pathologies that predominates SCAD Types 2 and 3. Intracoronary imaging modalities include intravascular ultrasound (IVUS) and optical coherence tomography (OCT), which differ in their spatial resolution, and depth of penetration. IVUS has a lower spatial resolution but penetrates deeper, thus allowing for full vessel visualization. With this imaging modality, IMH appears as a homogenous collection behind the intima-media membrane. OCT, on the contrary, has a higher resolution with the capability to

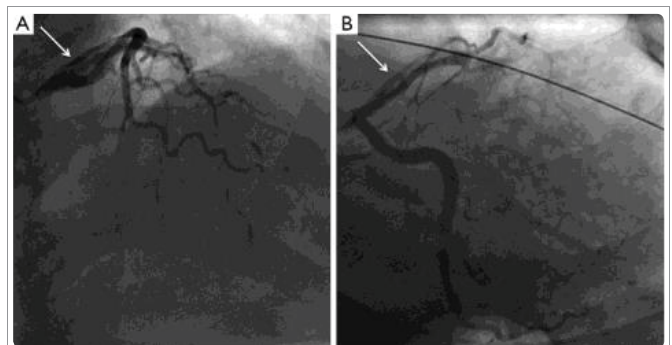


Figure 4: Type 1 SCAD. Illustrates the pathognomonic angiographic appearance of SCAD with contrast staining of both the arterial wall and false lumen of the dissected lesion [6].

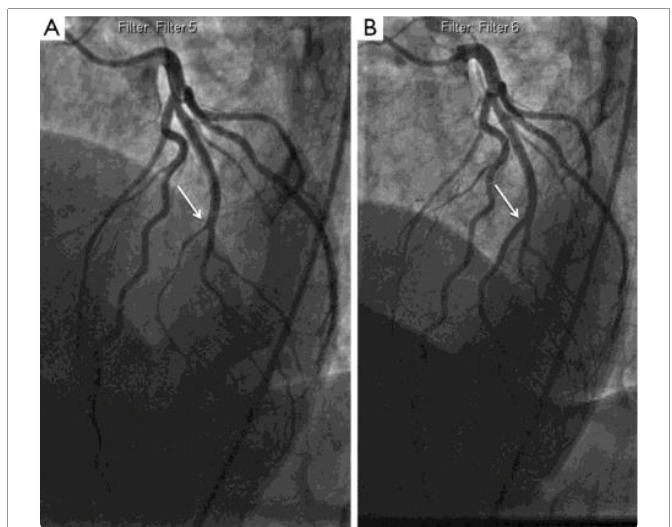


Figure 5: Type 2 SCAD. Stenosis of varying severity and usually affects the mid to distal segments of coronary arteries. [6].

visualize true and false lumens, and intimal tears, but at the expense of poor penetration. In comparative studies, OCT has been shown to be more sensitive and better at detecting SCAD than IVUS, though both are able to identify IMH equally [10,11].

The proposed approaches to the management of SCAD have been met with significant controversy, given the scarcity of supportive data to serve as precedence. The choice between medical versus revascularization therapies depends on the clinical status of the patient and affected coronary vessel. In the Mayo series, all 79 patients were treated conservatively and demonstrated spontaneous angiographic healing at > 4 weeks following their event [7]. In the UBC study, an excellent clinical outcome was obtained after a series of 50 patients were treated with conservative management. There was no demonstrable in-hospital mortality and only 4.8% of patients suffered a recurrent MI. Theoretically, reducing prothrombotic and shear stress burden on coronary vessels should confer a protective effect. Beta-blockers have been routinely administered for both acute and long-term management of SCAD. Extrapolating from the established benefits of dual anti-platelet agents in secondary prevention of CAD, aspirin and clopidogrel have been promoted in the acute phase of SCAD, with aspirin encouraged for life-long therapy. As the pathophysiology of SCAD entails dissection and IMH formation, the role of anti-coagulation and anti-thrombotic therapy has been debated for its risk of dissection extension. A retrospective study by Shamloo et al (2010) [12], looked at 440 cases of SCAD and reported that of the patients who received thrombolytic therapy, 60% consequently required percutaneous coronary intervention or Coronary Artery Bypass Grafting (CABG) to prevent progression of dissection [12]. PCI is preferred for patients with significant hemodynamic instability, dissected left main or proximal LAD segments.

For the smaller subset of patients presenting with cardiac arrest due to ventricular fibrillation or ventricular tachycardia, the main controversy remains the indication for ICD implantation for secondary prevention of SCD. Based on the 2012 ACCF/AHA/HRS guidelines for device-based therapy of cardiac rhythm abnormalities, in patients resuscitated from cardiac arrest, ICD is associated with clinically and statistically significant reductions in SCD and total mortality, with Class I and Level A evidence of support based on various large clinical trials. However, these accredited recommendations apply to patients with irreversible coronary artery disease, underlying structural heart disease, or severe systolic dysfunction [13]. To date, there is no level of evidence to support any class recommendation for ICD implantation for secondary prevention in patients with SCAD presenting with VF arrest, especially in patients with normalized cardiac function. With an overwhelming majority of patients showing a positive response to conservative medical therapy, Physicians are left to individualize device implantation therapy based on clinical judgment. Without much clinical evidence, the benefit of ICD placement for secondary prevention in VF SCAD remains unknown, and continues to be a topic of significant debate.

CONCLUSION

Spontaneous coronary artery dissection is an under diagnosed condition with a variable clinical presentation. An increase in disease prevalence is largely attributed to the coupling of conventional coronary angiography with intracoronary imaging including IVUS and OCT.

Retrospective angiographic studies have identified left main coronary disease in only a small percentage of cases. Amongst the subtypes, Type 3 is the rarest and most challenging of all to identify with conventional angiography alone. Intracoronary imaging has alleviated this diagnostic dilemma with a significant uptrend in detection rates. Treatment approaches continue to be heavily debated in the cardiovascular arena. There has been a trend towards conservative pharmacological therapy with dual anti-platelet therapy and beta-blockers, with PCI or CABG reserved only for hemodynamically unstable patients or subsets with left main or proximal LAD involvement. The benefit of ICD implantation remains unanswered.

Our case not only illustrates the unusual presentation of SCAD in a post-partum woman, it also highlights the infrequency of LM involvement, and the rarity of Type 3 disease presenting with left main STEMI, with spontaneous resolution on conservative pharmacological therapy.

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