

Spontaneous Coronary Artery Disease in a Healthy Fibromuscular Dysplasia Patient: A Case Report

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How to cite this paper: Shunnar, M., Alshaikh, O., Alzubaidi, M. and Choi, H.-R. (2024) Spontaneous Coronary Artery Disease in a Healthy Fibromuscular Dysplasia Patient: A Case Report. *Case Reports in Clinical Medicine*, 13, 375-381.

<https://doi.org/10.4236/crcm.2024.139046>

Received: August 8, 2024

Accepted: September 10, 2024

Published: September 13, 2024

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Abstract

Our case is a 72-year-old female with Fibromuscular Dysplasia (FMD) presented to the Emergency Department (ED) with acute coronary syndrome (ACS), more specifically NSTEMI. In ED, troponin levels were elevated and non-ST segment elevation myocardial infarction (NSTEMI) was detected on electrocardiogram (ECG). Computed tomography (CT) scan showed Internal Carotid artery (ICA) pseudoaneurysm. Cardiac catheterization (CATH) was performed and revealed occlusion of the Left anterior descending (LAD) artery. Spontaneous coronary artery dissection (SCAD) was suspected due to the history of FMD without a history of hypertension, smoking or substance use.

Keywords

SCAD, NSTEMI, STEMI, ACS, Eliquis, Pseudoaneurysm, Fibromuscular Dysplasia, Anticoagulant, Antiplatelet, Coronary Dissection, Neuroendovascular, Cardiology, Warfarin, Cath Lab, Apixaban, Clopidogrel

1. Introduction

Spontaneous coronary artery dissection is a tear in the intimal layer of epicardial coronary arteries causing an intramural hematoma that can partially or completely occlude the blood flow in the vessels. It is an underrecognized condition that can cause myocardial ischemia or infarction, arrhythmia, or sudden cardiac death. There are multiple risk factors that can precipitate SCAD including hypertension, hyperlipidemia, connective tissue diseases, fibromuscular dysplasia, emotional stress and hypothyroidism. SCAD is diagnosed primarily by cardiac catheterization and coronary angiogram, but intravascular imaging can also be done

[1]. FMD is a rare arteriopathy that causes narrowing of blood vessels commonly renal, carotid and vertebral vessels, but can impact any blood vessel in the body [2]. The etiology of FMD is accounted for by an interplay among genetic predisposition, estrogen hormone exposure especially in middle-aged females, and environmental factors [2]. There is an increased prevalence of FMD in patients with SCAD presenting with symptoms of acute coronary syndrome [3]. Research is sparse regarding how ACS manifests in FMD patients and how it can be investigated. ACS itself entails NSTEMI, STEMI, or unstable angina depending on the ECG findings and troponin levels. In this case, the patient elevated enzymes with normal ECG strongly correlated with NSTEMI. While more common causes of NSTEMI exist [4], we take the opportunity here to warn against overlooking SCAD-related NSTEMI especially in patients who denied smoking, alcohol, drug use, and had a generally healthier active lifestyle.

2. Case Presentation

A 72-year-old woman with a history of paroxysmal atrial fibrillation, deep vein thrombosis, pulmonary embolism diagnosed a year prior, fibromuscular dysplasia and hyperlipidemia presented to the Emergency Department by EMS on the night of April 30th of 2024. She reported experiencing chest pain throughout the day, prompting an initial visit to the ED earlier that day. However, her symptoms subsided, and she was discharged home. Later that evening, while standing at her kitchen counter, she lost consciousness and fell backward, striking her head.

Upon arrival at the ED, the patient reported chest pain and the fall. Although the patient had a history of paroxysmal atrial fibrillation, her ECG was normal when she presented. Arrhythmias including atrial fibrillation itself can be due to many underlying causes, one of these causes is SCAD [5]. The medication on file was Eliquis (apixaban) 5 mg twice daily. Vitals signs were stable, and the physical exam confirmed non reproducible chest pain at presentation. Labs showed an initial troponin level of 279 ng/dL, suggestive of potential heart damage. A potassium level of 3.0 mEq/L was noted. A CT scan raised concerns about a possible nondisplaced cervical spine fracture at C2. Repeat troponin levels showed an increase to 364 ng/dL. Due to the elevated troponin levels suggestive of NSTEMI and the potential cervical spine fracture, the decision was made to place the patient in a cervical collar and initiate a Heparin drip. The patient was transferred to a level 1 trauma center for further evaluation and management.

Upon arrival at the trauma center on May 1st, additional CT scans were performed to assess other potential injuries from the fall. The Heparin drip resumed. Consultations were made with cardiology, neurosurgery and trauma surgery for comprehensive evaluation and treatment planning. Repeat troponin levels showed a further increase to 544 ng/dL. The patient was admitted under the care of the trauma team. CT angiography revealed a possible pseudoaneurysm on the left internal carotid artery (**Figure 1**), prompting consultation with neuroendovascular specialists. Neurosurgeons cleared the cervical spine, allowing for the removal of the cervical collar.

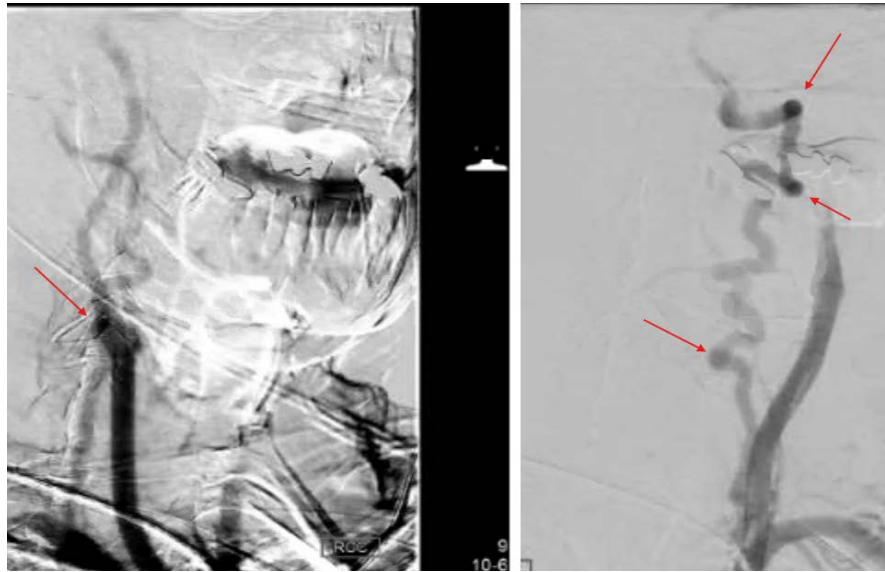


Figure 1. Arrows pointing to the pseudoaneurysmal malformation.

Cardiology evaluated the patient on May 2nd and planned for a left heart catheterization later that day. They also recommended a cath lab and initiating medications including intravenous heparin drip, aspirin, a statin, and a beta-blocker. The patient underwent a left heart cath in the afternoon. The procedure revealed a 100% blockage in the distal-apical left anterior descending artery. This blockage could be due to either embolic phenomena or SCAD. Other coronary arteries showed mild disease, and the left ventricle maintained good systolic function. A transesophageal echocardiogram (TEE) was performed during the cath, which revealed mild to moderate tricuspid regurgitation and a slightly enlarged left atrium. The ejection fraction was 55%. Cardiology recommended continued medical management with Eliquis and lifestyle modifications to control risk factors.

Later that night, the patient developed severe left-sided chest pain radiating to the neck and back improved with sublingual nitroglycerin. An ECG and chest X-ray were normal. Troponin levels continued to rise, reaching 901 ng/dL. Cardiology placed the patient on triple therapy with aspirin, Plavix (generic name Clopidogrel) and Eliquis. Hematology and oncology were consulted to investigate a possible hypercoagulable state. They suggested anticoagulation failure due to the recurrence of presumed thromboembolic phenomenon while on anticoagulant therapy. This also raised the possibility of antiphospholipid syndrome. However, testing for antiphospholipid antibodies came back negative. Hematology and oncology recommended switching the blood thinner to Warfarin.

On May 5th, the patient complained of chest discomfort described as indigestion. The ECG was normal, but troponin levels continued to climb, reaching 1120 ng/dL. Sublingual nitroglycerin was administered again. Morphine and Zofran were given intravenously. A nitroglycerin drip was started, and the patient was transferred to the cardiac ICU due to worrying symptoms and rising troponin levels.

Due to the persistently elevated troponin levels suggestive of a type 1 NSTEMI. Cardiology decided to perform coronary angiography on May 7th to investigate potential distal coronary embolization or thrombotic phenomena. The coronary angiogram confirmed the very distal-apical LAD is very tortuous with 100% occlusion in the apical area and is small in caliber, left to left collaterals seen, possible embolic phenomena vs SCAD, the artery is small around a very tortuous segment and will treat medically. Giving the findings of the angiogram, conservative management was more appropriate in this case, taking into consideration the consequences on any procedural intervention [6]. A TEE with bubble study was performed to assess for a patent foramen ovale. Agitated saline was used to create bubbles and see if they passed through the PFO. The TEE with color flow showed a normal left ventricular function. The interatrial septum had a Grade I positive bubble study where 1 to 9 bubbles were seen passing through the PFO during the test which is considered insignificant, and 100 - 300 bubbles is the most significant or Grade IV.

After the patient stabilized, she was discharged home on Warfarin and Clopidogrel. Eliquis and Aspirin were discontinued at discharge with instructions to follow up with cardiology, neuroendovascular, and hematology/oncology for continued management of the hypercoagulable state.

3. Discussion

FMD is a non-atherosclerotic, non-inflammatory vascular disease that predominantly affects medium-sized arteries, causing arterial stenosis, occlusion, aneurysm, and dissection [7]. It has a strong association with SCAD, particularly in women, and is recognized as a significant underlying condition in many SCAD cases. In this patient, the diagnosis of FMD was reinforced by neuroendovascular findings of multivessel involvement, including a pseudoaneurysm in the left internal carotid artery (ICA), highlighting the systemic nature of the disease.

The exact mechanism linking FMD and SCAD is not very clear, but FMD's inherent structural arterial abnormalities likely predispose to dissection under stress or minor trauma. FMD causes fibroplasia of the media with alternating areas of smooth muscle loss in the vascular walls eventually leading to dilatation and formation of fibromuscular ridges [8]. The alternating areas of dilatation and constriction lead to the beaded appearance of the vessels like the pattern depicted in this patients' imaging in **Figure 2** and **Figure 3**. Those weakened vessel walls are at higher risk of damage by sheer force of blood leading to intimal tear and dissection [9]. The tear can trigger thrombus formation leading to complete or partial blockage of the blood flow. In cases that occur in the coronary arteries, it manifests as one of the diseases under the umbrella of ACS such as NSTEMI in this patient.

The pathophysiology highlighted above provides a reasonable explanation for this case disease progression. The patient's SCAD presented as a 100% occlusion of the distal-apical LAD artery, detected during a coronary angiogram. The

absence of significant atherosclerotic disease elsewhere in the coronary circulation supports the SCAD diagnosis over atherosclerotic plaque rupture, which typically underlies the NSTEMI. The patient's recurrent episodes of chest pain and rising troponin levels with normal ECG further corroborate the diagnosis of SCAD-related NSTEMI.



Figure 2. Carotid angiogram with arrows pointing to the FMD showing the beaded appearance of the vessel. There is a fusiform dilatation near the bifurcation representing the pseudoaneurysm.



Figure 3. Similar Finding to **Figure 1.**

Management of SCAD in the context of FMD is primarily conservative, focusing on symptom relief and prevention of further dissection or embolization compared to interventional approach with cath lab with or without stents if required

[10]. In this patient, initial inpatient treatment included Heparin drip and Nitroglycerin followed by antiplatelet therapy (ASA, Plavix) and anticoagulation with Warfarin. We here encourage prophylaxis against thromboembolic events with multiple medications in patients who showed recurrence of symptoms while on one therapy (for example, Eliquis in this case) or switch to more potent alternative treatment options. Given the systemic involvement of FMD and associated complications such as pseudoaneurysms, a multidisciplinary team approach involving cardiology, neurosurgery, trauma surgery, and neuroendovascular specialists is also crucial in long-term management [11].

4. Conclusion

This case report details a complex interplay between FMD, a known predisposing factor, and SCAD, leading to a NSTEMI in a 72-year-old female patient. We highlight the findings suggesting FMD as a plausible underlying cause of SCAD. Patients who have recurrent episodes of NSTEMI without underlying risk factors should investigate the not-so-common causes. To ensure better control of these episodes, a more adamant plan with anticoagulant and antiplatelet should be considered in addition to a rigorous follow-up. Interventional procedures might not be necessary since conservative management proved to be as effective with less adverse events.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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