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Myocardial Infarction as the Initial Presentation for Fibromuscular Dysplasia

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Abstract **Background:** Fibromuscular dysplasia (FMD) is a non-atherosclerotic disease that affects medium-sized arteries and results in stenosis, dissection, aneurysm or occlusion. It is most commonly reported in the renal and carotid arteries. Involvement of coronary arteries is quite rare and and leads to serious consequences. **Case: A** 62-year-old African American woman with a history of mitral valve prolapse presented with chest discomfort associated with diaphoresis. Her EKG initially showed ST segment changes in leads II, III, and V2-V5 which resolved in approximately 30 minutes. Her troponin peaked to 20 ng/L during her hospital course. A bedside echocardiogram revealed an EF of 45% with mid, distal septal and apical hypokinesis. **Decision-making:** The patient was admitted to the Cardiac Care Unit for treatment of an NSTEMI. She underwent cardiac catheterization, which revealed single-vessel coronary disease with diffuse narrowing of the distal LAD, beyond the first diagonal branch down to the apex. CT angiography of her abdomen and pelvis showed mild narrowing of the mid-right renal artery with a small fusiform aneurysm measuring approximately 5 mm. Her carotid duplex showed tortuosity in the right internal carotid artery. Given the multiple vascular anomalies, a diagnosis of fibromuscular dysplasia was considered. **Conclusion:** Acute coronary syndrome in fibromuscular dysplasia requires an integrated approach to management, especially if there is associated malignant hypertension and/or dissection.

Keywords: *acute coronary syndrome, fibromuscular dysplasia, myocardial infarction*

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1. Introduction

Fibromuscular dysplasia (FMD) is a disease of blood vessels with neither atherosclerotic nor inflammatory components, commonly affecting middle age women. [1] With fibromuscular dysplasia, there are abnormal growths within the arterial wall resulting in a "string of beads" pattern commonly seen on imaging modalities. The vessels most commonly involved include the renal, carotid, and vertebral arteries; however, dysplasia can affect any artery and can occur in multiple arteries simultaneously. [1] The clinical presentation of FMD includes headaches, hypertension, dizziness, and pulsatile tinnitus, but adverse cardiovascular events can happen with a high frequency. [1] These events include aneurysm, dissection, and cerebrovascular accidents. [1]

Since the year 1964, FMD was thought to be specific to renal arteries; later studies identified FMD changes in the celiac and carotid arteries. [2] Since that time, several cases of extrarenal FMD have been published in the literature. Uncommonly seen, FMD can also affect the coronary arteries. A majority of patients with coronary FMD present with dissection of an epicardial artery or a major branch. [3] Small coronary artery FMD has been reported to be a cause of sudden cardiac death due to loss of the sinoatrial or atrioventricular nodal arteries [4]. It is important to administer intra-arterial vasodilators to rule out coronary spasm. [5] In young females with typical chest pain presenting with spontaneous coronary artery dissection (SCAD), FMD should be a kept as an index of suspicion.

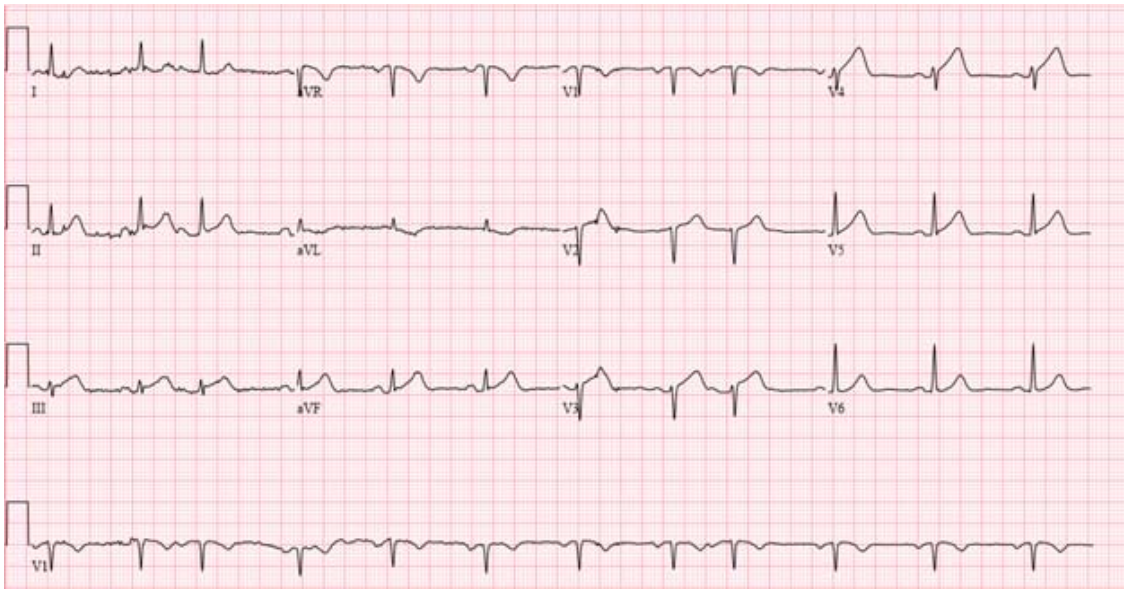

Figure 1A. EKG showed sinus rhythm with ST segment changes in II, III, V2-5

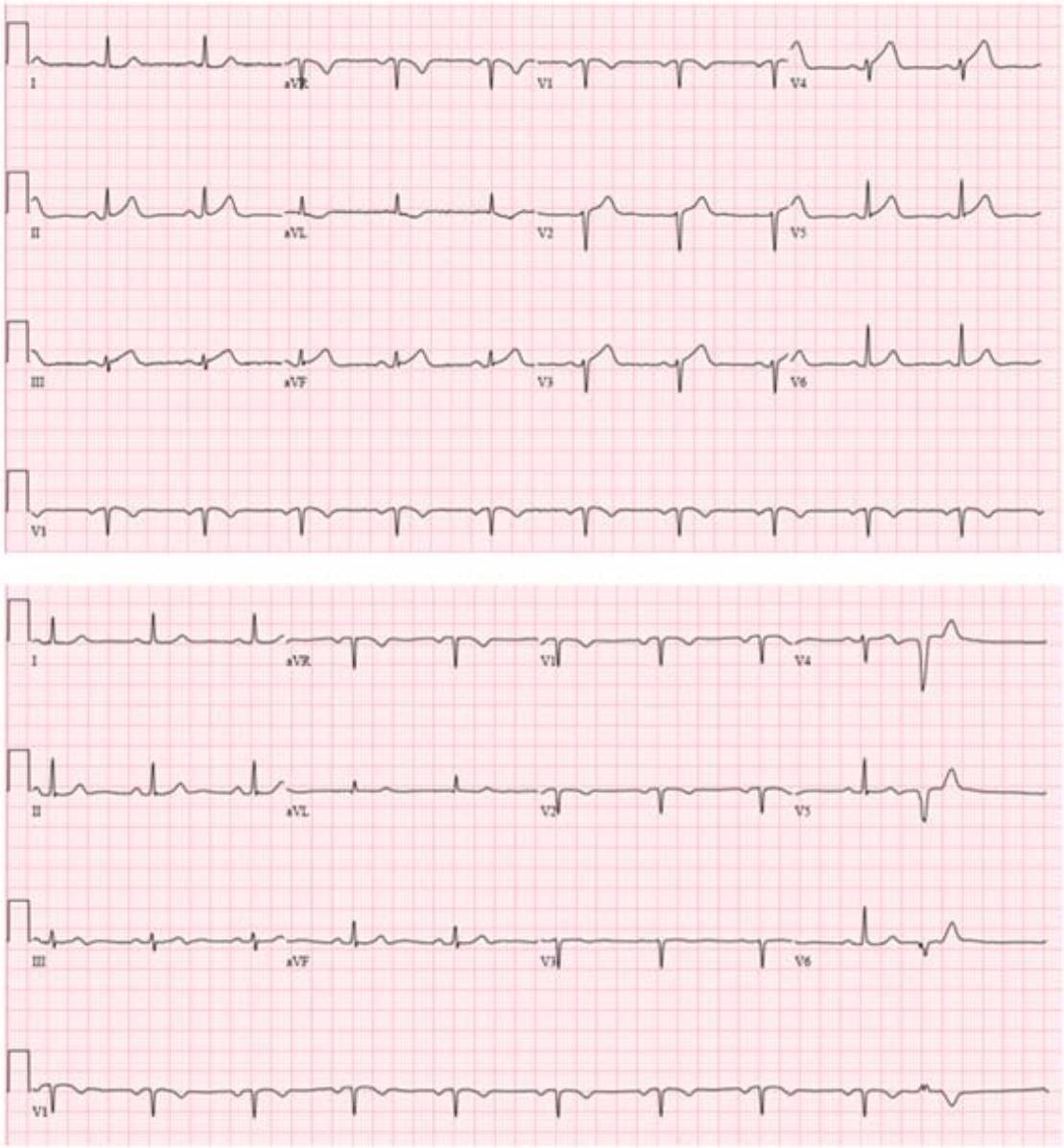

Figure 1B. EKG showed sinus rhythm with occasional premature ventricular complexes

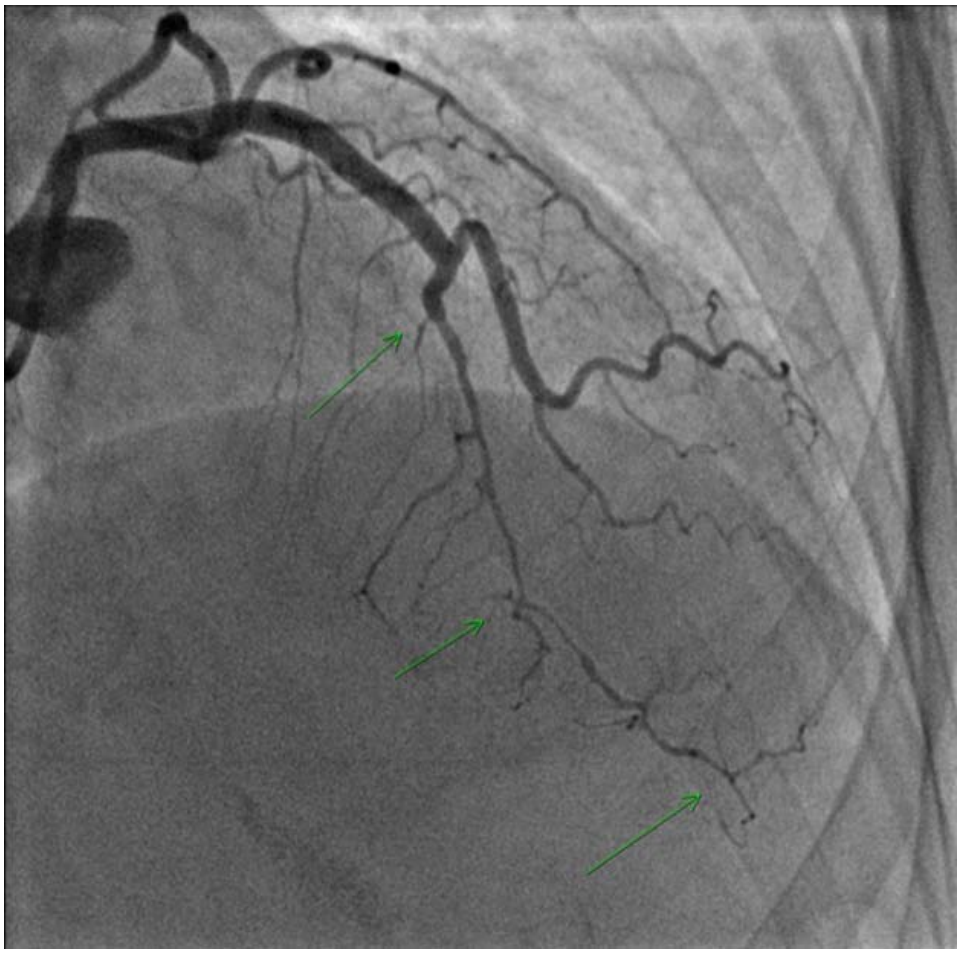

Figure 1C. EKG showed sinus rhythm with occasional premature ventricular complexes and Septal infarct

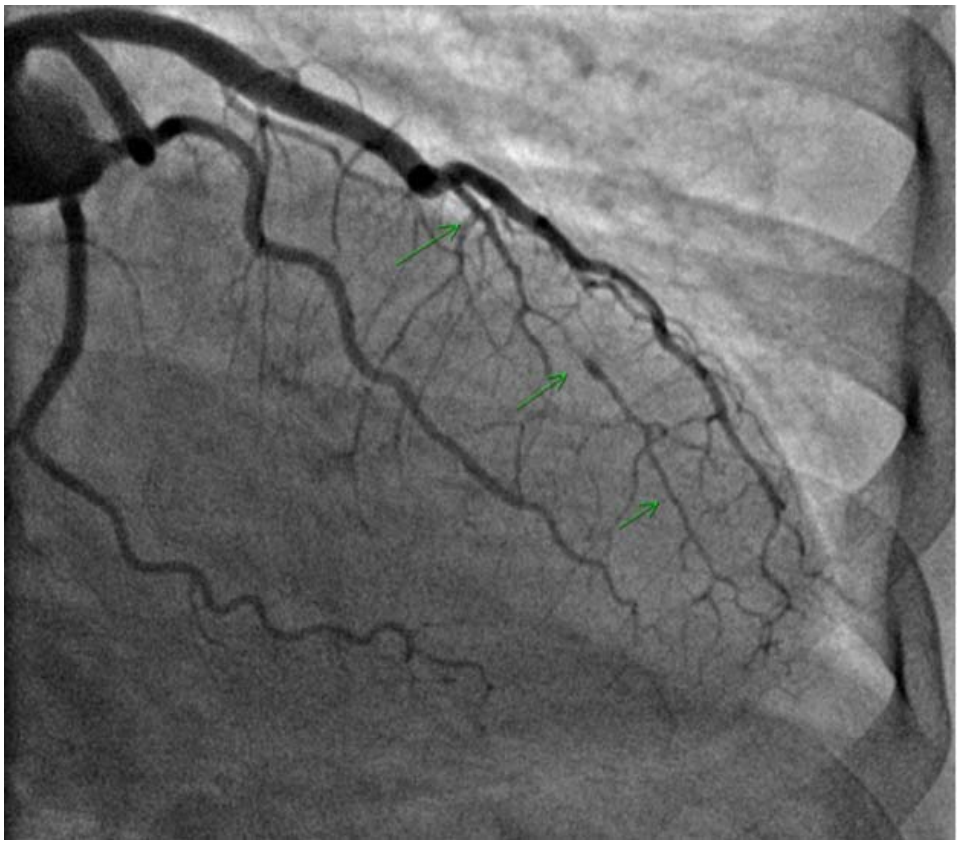

Figure 1D. LCA in RAO Cranial view; the arrows located the diffusely narrowed distal LAD

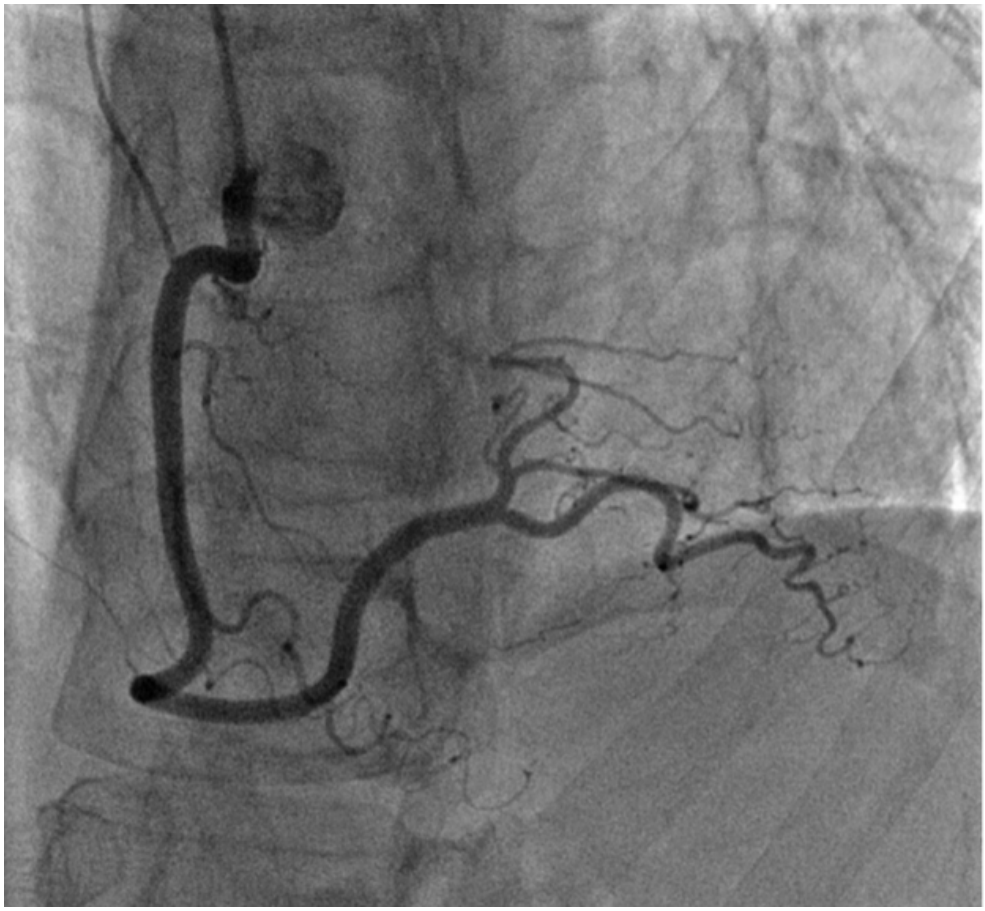

Figure 1E. Dominant normal RCA

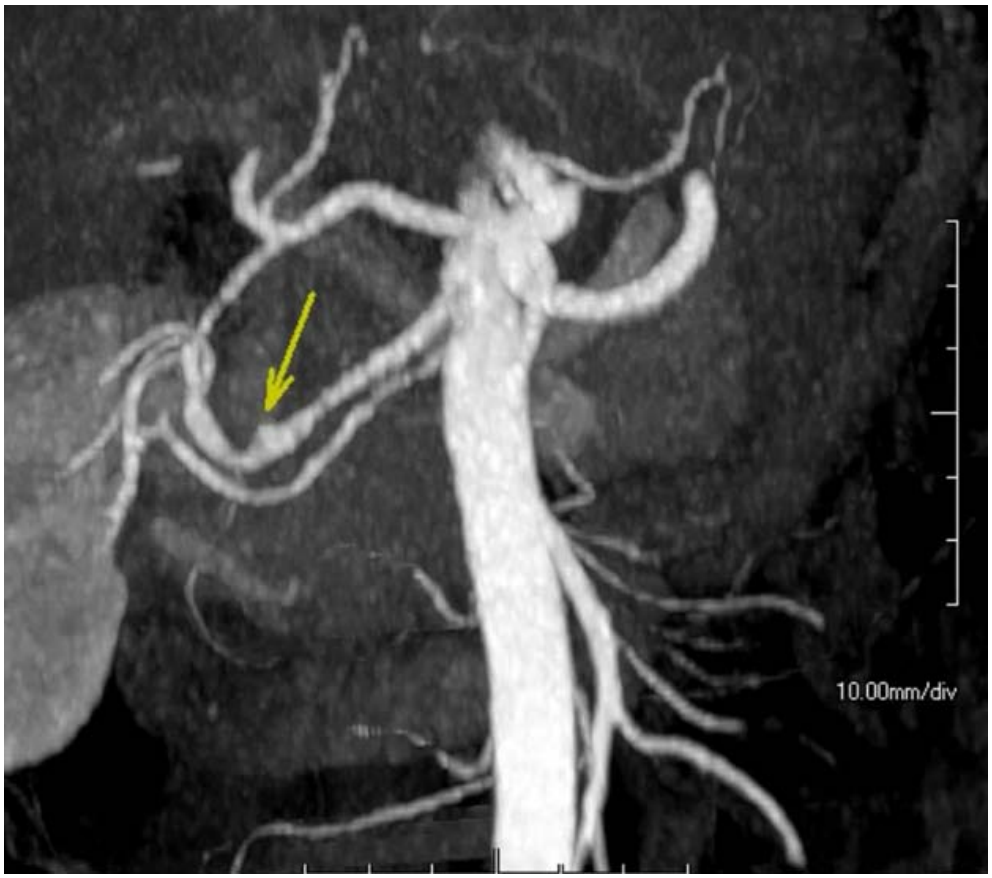

Figure 1F. CTA of abdomen shows mild narrowing of the mid right renal artery with a small fusiform aneurysm measuring approximately 5 mm. No intramural hematoma or dissecting intimal flap

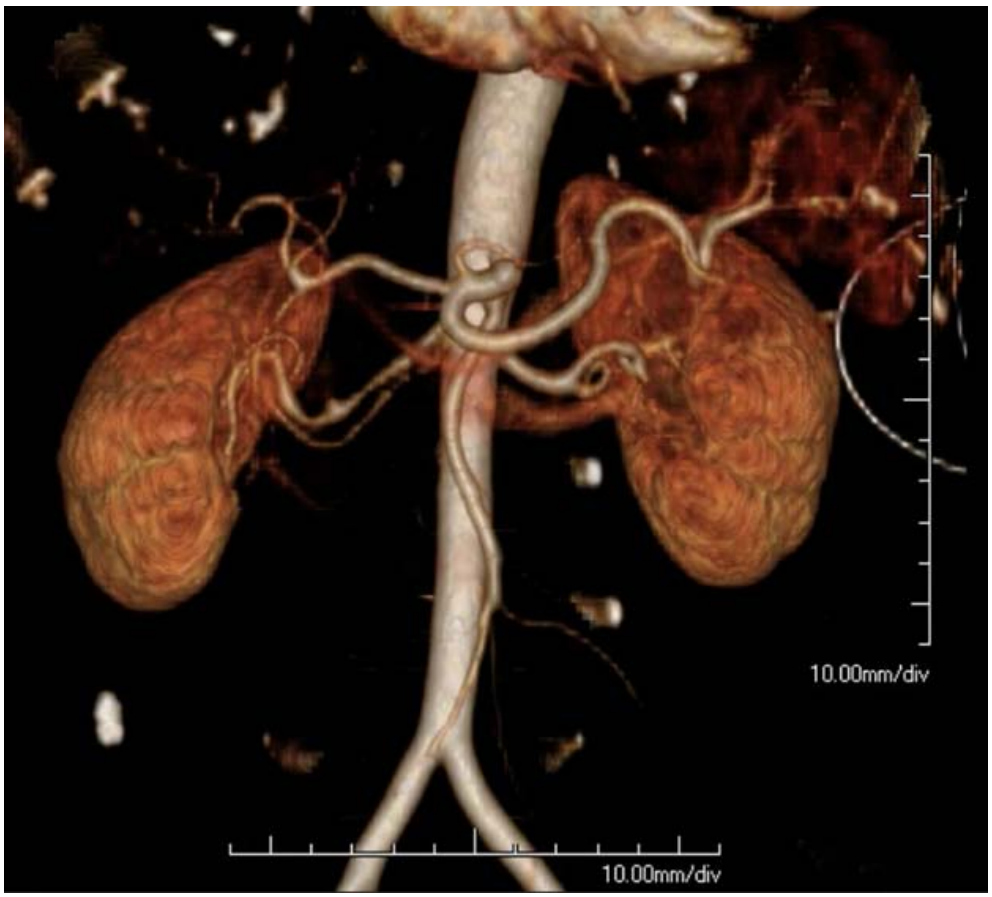

Figure 1G. CTA abdomen with 3D reconstruction shows right renal artery aneurysm

2. Case Presentation

A 62-year-old African-American female with a past medical history of mitral valve prolapse presented to our hospital with chest discomfort associated with diaphoresis. She reported palpitations accompanied by weakness and sweating while driving. She pulled over and called Emergency Medical Services. In the Emergency Department (ED), physical examination revealed a well-developed female, anxious, in mild distress with the appearance of pectus excavatum of her chest. She was afebrile with a blood pressure of 117/50 mmHg and a heart rate of 55 bpm. Her electrocardiogram initially showed ST segment changes in leads II, III, and V2-V5 (Figure 1A), which resolved within 30 minutes of arrival (Figure 1B). Her troponin I was initially 0.06 ng/L [Normal <0.04 ng/L] but peaked to 20 ng/L in 12 hours. She was admitted to the cardiac critical care unit for acute non-ST segment elevation myocardial infarction (NSTEMI). She was started on aspirin, clopidogrel, and a therapeutic dose of enoxaparin. The symptoms were alleviated in a few hours and her troponin trended downward. Within 48 hours, she was taken for cardiac catheterization, which revealed single-vessel disease involving the left anterior descending artery (LAD) extending from the area distal to the last diagonal artery to the apex of the heart. The diseased segment had the appearance of fibromuscular dysplasia with possible areas of spontaneous coronary artery dissection (SCAD). There were no changes in vessel caliber with nitroglycerin injection (Figure 1C, D, E). Post-cardiac catheterization transthoracic echocardiography revealed mid and distal septal and apical hypokinesis with ejection fraction (EF) 45%. CT angiography for abdomen and pelvis showed mild narrowing of the mid right renal artery with a small fusiform aneurysm measuring approximately 5 mm (Figure 1F, G). Carotid duplex studies showed abnormal velocities in the distal left internal carotid artery, concerning for a possible aneurysm. The provisional diagnosis was fibromuscular dysplasia; the possibility for medium-vessel vasculitis was less likely given normal erythrocyte sedimentation rate (ESR), C-reactive protein (CRP), and absence of anemia or thrombocytopenia. Connective tissue disorders, such as Ehlers-Danlos syndrome vascular-type, were less likely given absence of uterine prolapse, bruising, family history, or hypermobility. Moreover, inflammatory vasculitides were excluded given negative results in complement 3, complement 4, antinuclear antibody panel (ANA), antineutrophil cytoplasmic antibodies (ANCA), Rheumatoid Factor (RF), Cryoglobulin, Hepatitis B and C, and urinalysis looking for red blood cells and active sediments. She was discharged in stable condition with follow-up with cardiology and rheumatology services.

3. Discussion

3.1. Introduction

Fibromuscular dysplasia (FMD) can affect any number of blood vessels in the body simultaneously. When it affects the coronary arteries, it can manifest with multiple cardiovascular effects, such as dissection, aneurysm, and

CVA. [1] Traditionally, it was thought to affect only the renal arteries, however in 1964, an initial case of extrarenal FMD was reported. [2] Since that time, several cases of extrarenal FMD have been published in the literature. Also, FMD can present with spontaneous coronary artery dissection (SCAD), especially in young women with typical chest pain. [6] However, SCAD can occur independently of FMD.

3.2. Epidemiology

The prevalence of FMD in the general population is not known, and it is exceedingly rare according to some researchers. [7] Hence, diagnosis and treatment are not always done in a timely fashion, causing potentially devastating complications. [7] However, other investigators claim that FMD not as uncommon as previously thought. [7] Arteriograms of the renal vasculature in 1862 prospective renal donors were evaluated, and FMD was identified in 3.8% of these patients. [7]

3.3. Diagnosis: Methods and Confounders

Patients who present with acute coronary syndrome (ACS) or left ventricular dysfunction in the absence of cardiac risk factors, may have FMD. [8] This possibility becomes likely in middle-aged women with isolated hypertrophy of a coronary artery in its mid-distal portion without involvement of other coronary arteries. [8] FMD is considered as a diagnosis whether there is coronary or noncoronary involvement. Specific symptoms can provide a clue to diagnosis. [8] For example, renal artery involvement can lead to headache, and FMD of a cervical vessel can cause pulsatile tinnitus. [8]

The differential diagnosis of coronary artery FMD includes ACS due to atheromatous plaque rupture, Ehlers Danlos syndrome type IV, Takayasu's arteritis, vasculitis, cocaine use, Prinzmetal's angina, and myocardial bridging. This bridging involves embedding of an epicardial coronary artery into the myocardium, i.e. mid LAD tunneled artery. [8]

Currently, angiographic catheterization is the only reliable modality for diagnosing FMD of the coronary arteries. [8] Coronary artery involvement in fibromuscular dysplasia has the following angiographic features: smooth narrowing, dissection, tortuosity, and intramural hematoma. [8] Spasm can also occur and resemble smooth narrowing, but spasm can be differentiated by intracoronary vasodilator administration. [8] Noninvasive imaging techniques have not been useful for diagnosing coronary artery involvement in FMD. [8]

These features can occur either individually or concomitantly, and multiple features can be seen in the same coronary artery or in the same patient. [8] The most common manifestation of FMD in coronary arteries is SCAD. [8] Coronary FMD commonly involves only one epicardial vessel. [8] Visualization of FMD is commonly done with Intravascular Ultrasound (IVUS), which is a supplemental method to angiography. [8] The appearance of FMD has been characterized as a "string of beads," which looks different from the beading found in noncoronary FMD, suggesting the possibility that coronary FMD beading may represent a particular manifestation of dissection. [3,9] The demarcation between healthy and diseased coronary artery is usually well-defined on angiography. [10] The use of both IVUS and angiography simultaneously allows for detailed characterization of the arterial wall, where FMD can be distinguished from other etiologies of coronary artery disease. [11] FMD tends to involve the mid or distal coronary artery segments, but atherosclerosis usually affects the proximal arterial portion. [12]

3.4. Treatment

The treatment strategy for FMD of coronary arteries is not standardized. [8] Guidelines are limited regarding treatment due to lack of trials and observational studies. [8] Current therapy is guided by a limited number of case reports and series with treatment strategies extracted from articles that present nonrandomized data. [8]

In patients with FMD of coronary arteries without active myocardial ischemia, conservative medical management is preferred over invasive intervention. [8] The reason is that SCAD usually heals by itself. [13] If percutaneous intervention is done, there is a tendency for the dissection flap to expand in either direction away from the stented segment, potentially leading to intramural hematoma. [8] Hence, the recommendation is to perform medical management, but if the patient has persistent ischemia, percutaneous or surgical intervention is favored. [8] One method of medical management is to use dual antiplatelet therapy with daily aspirin and clopidogrel for up to 1 year, so long as bleeding risk is minimized. [13] After that year passes, monotherapy with aspirin can be continued indefinitely. [8] Regarding coronary artery dissection, a beta-blocker can provide benefit by slowing the heart rate and reducing the blood pressure, thereby lowering the risk of continued dissection. [13] Furthermore, hypertension in the presence or absence of renal artery FMD should be managed appropriately. [15] In symptomatic patients experiencing ongoing ischemia, coronary intervention may be necessary. [8] Compared to the typical angioplasty done for renal artery FMD, placement of an intracoronary stent placement is normally needed in patients diagnosed with SCAD. [8] The stent should be carefully placed such that excessive vasodilation is avoided, in order to minimize the spread of dissection. [8] Coronary artery bypass grafting (CABG) should be considered in patients with multiple diseased vessels who also have good distal perfusion. [8] Also, no evidence exists to support the use of statin therapy in these patients. [15,17]

Coronary artery FMD can also be managed by cardiac rehabilitation, especially for young patients with anxiety. [16] Also, patients should avoid strenuous physical activity due to risk of SCAD. [16]

4. Conclusion

Clinicians should have a high index of suspicion of FMD as a cause of ACS. Even though it is rare, we should suspect isolated coronary FMD as a cause of ACS, especially in females with SCAD. A proper diagnosis of FMD is needed in order to optimize management.

Appropriate treatment will help to achieve favorable patient outcomes.

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