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UNUSUAL CASE OF NON-ST ELEVATED MYOCARDIAL INFARCTION IN A MIIDDLE-AGED FEMALE; A CASE REPORT.

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Abstract

Fibromuscular dysplasia (FMD) is a non-atherosclerotic, non-inflammatory disease that affects any artery of the body.

We report a case of non-ST elevated myocardial infarction (N-STEMI) in a 60-year-old female who had a past history of hypertension, type 2 diabetes, and hyperlipidemia. She presented to the emergency department with dyspnea and chest pain of 1-day duration. The electrocardiogram showed inferior ischemia. A coronary angiogram showed spontaneous coronary artery dissection (SCAD). Magnetic resonance angiography (MRA) of the renal arteries and brain displayed distal stenosis in the right renal artery and middle cerebral artery aneurysms. The patient was treated conservatively and followed up.

Vascular, renovascular, and cerebrovascular manifestations are common in FMD. This case showcases both the common and uncommon presentations of the disease, most notably it highlights the systemic manifestations of FMD.

Introduction (explain how FMD differs from other vascular disases)

Fibromuscular dysplasia (FMD) is a nonatherosclerotic, non-inflammatory arteriopathy affecting the muscular layer of medium-sized arteries [1-2]. The prevalence of FMD is still unknown. Reviewing studies between 1963 & 2011 showed that the mean prevalence of FMD in healthy renal transplant donors was 3.3%. Another study showed that 3.8% of those undergoing renal artery angiograms had FMD [3].

It is primarily seen in young females but can be present in males. Lesions can be multifocal, giving it a characteristic (string of beads) appearance and focal stenotic lesions [4]. The most frequently affected arteries are the renal and cerebrovascular arteries [4]. More recently, the vascular phenotype of lesions associated with FMD has included arterial aneurysms, dissections, and tortuosity [4].

In this case report, we report a case of fibromuscular dysplasia uncommonly affecting the three major vascular territories (renal, coronary, and cerebrovascular).

Case Presentation

This patient is a 60-year-old female who presented to the emergency department with recurrent chest pain, dyspnea, and diaphoresis of one day duration. She had no associated symptoms or history of coronary artery disease or drug abuse.

The patient is a known case of type 2 diabetes, hypertension, and dyslipidemia. She is a chronic smoker and is on hormonal replacement therapy for hypothyroidism.

Five days prior to this presentation, she presented to the hospital complaining of mild chest pain. Physical examination was unremarkable; investigations included an electrocardiogram and cardiac biomarkers, which were normal, and the patient was discharged and advised for further work-up

During this presentation, the patient was vitally stable at room temperature. Auscultation revealed soft bilateral basilar crackles, no added sounds or murmurs, and the rest of the examination was insignificant.

On her latest visit. Investigations showed an elevation in troponin I, rising to 8.382Ng/L (normal<0.026 Ng/L). Electrocardiogram changes indicated inferior ischemia(Figure *1*). The Patient was admitted as a case of acute non-ST elevated myocardial infarction with mild heart failure. She was given three medications, aspirin, clopidogrel, and bisoprolol.

On the third day, the patient showed symptomatic improvement and no recurrent chest pain and underwent cardiac catheterization, revealing normal coronary anatomy but a dissection of the Left Circumflex Artery (LCA) with an intramural hematoma(Figure *2*). Diagnosis of spontaneous coronary artery dissection was made. Due to the long dissection segment and relatively small vessel size distally with the absence of active ischemic symptoms, it was decided to treat the patient conservatively.

Renal Magnetic Resonance Angiography (MRA) displayed stenosis of the distal segment of the right renal artery(Figure *3*). Moreover, a Magnetic resonance Angiography with a contrast of the

brain exhibited bilateral middle cerebral artery (MCA) trifurcation aneurysms with bilateral hypoplastic anterior and posterior communicating arteries(Figure *4*). With these findings, the suspicion of fibromuscular dysplasia was confirmed. The patient was regularly followed up in two-month intervals. The findings were unremarkable.

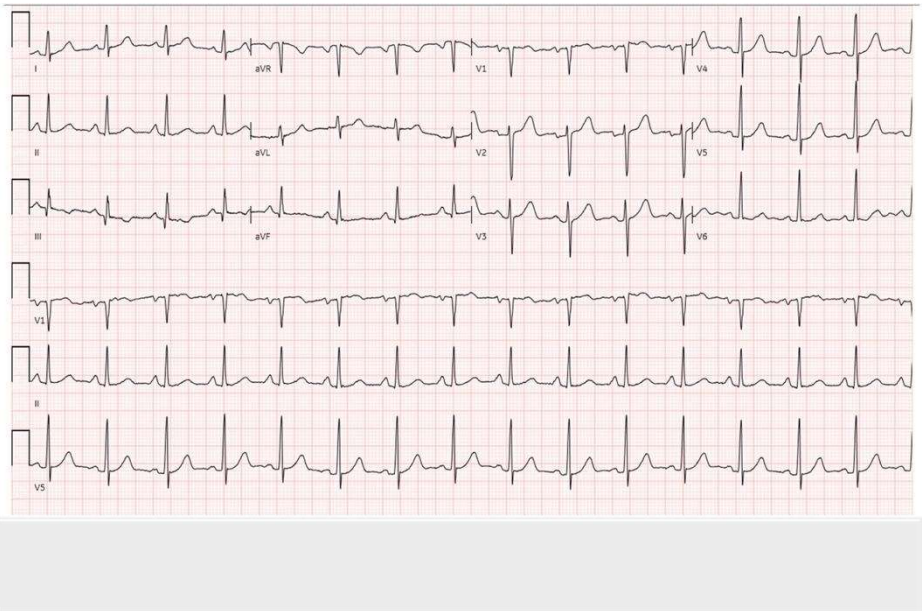

FIGURE 1: inferior ischemia

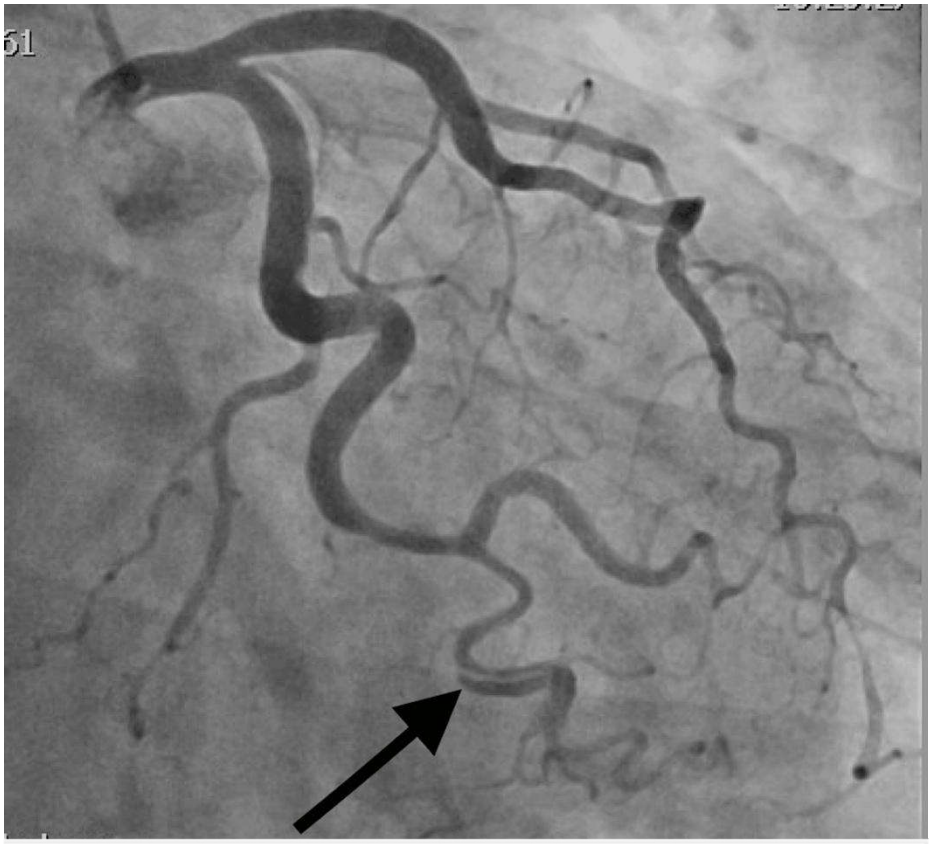

FIGURE 2: dissection of the mid left circumflex artery with intramural hematoma

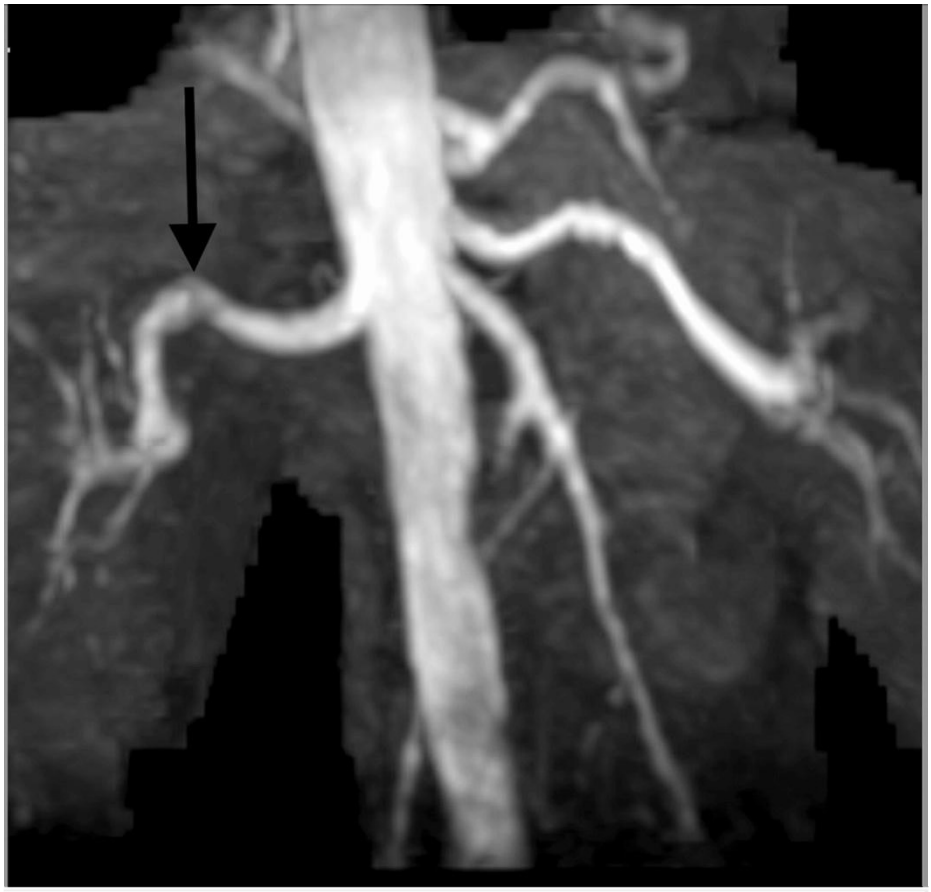

FIGURE 3: stenosis of the distal segment of the right renal artery

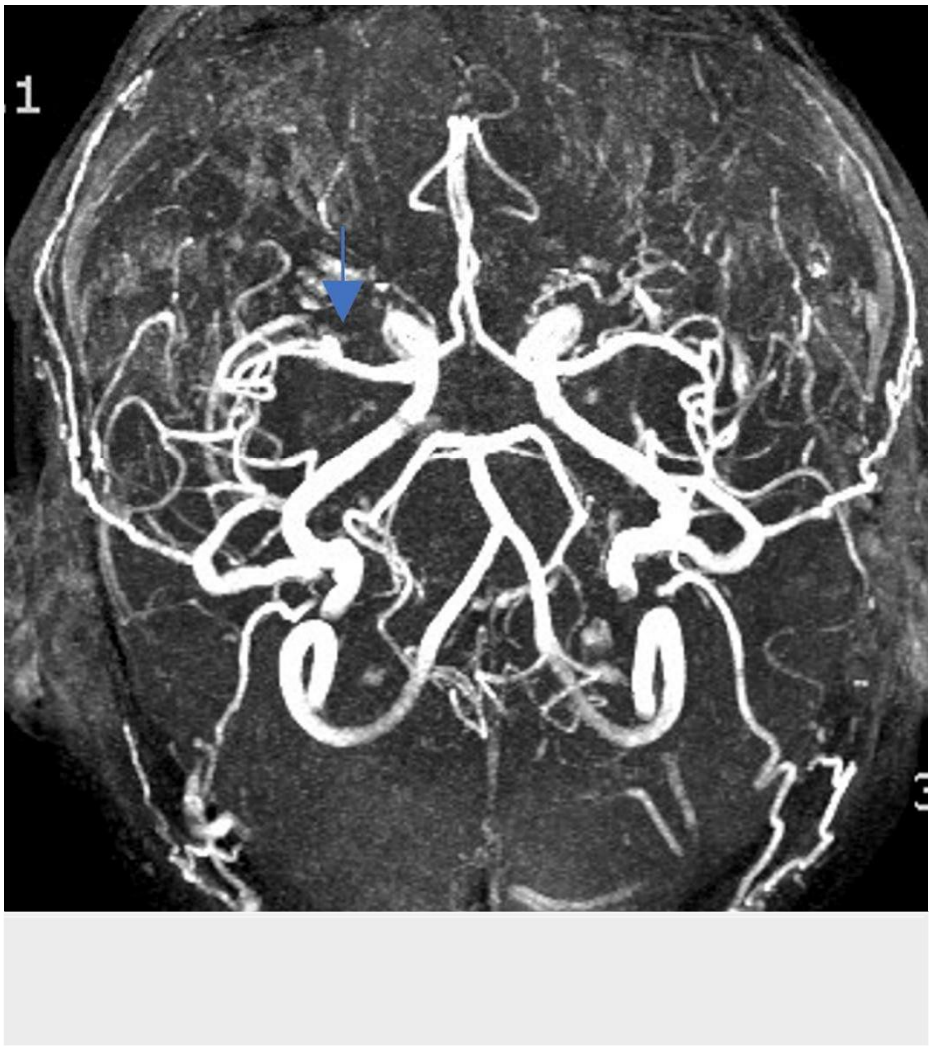

FIGURE 4: bilateral middle cerebral artery trifurcation aneurysm with bilateral hypoplastic anterior and posterior communicating arteries

Discussion

Fibromuscular Dysplasia is a non-inflammatory, non-atherosclerotic disease primarily affecting middle-aged women. The clinical picture somewhat differs between males and females[4]. Males may present with manifestations that include Hypertension, Aneurysms, and Arterial dissections. Manifestations of cerebrovascular origin are prominent in females[4], which include Carotid bruit and Headaches.

In our case, the patient presented with controlled Blood Pressure and no history of neurological manifestations. No carotid bruit was heard on examination. Suspicion of non-ST elevated Myocardial Infarction was raised as the patient presented with recurrent chest pain, and the electrocardiogram indicated inferior ischemia with elevated cardiac markers. Suspicion for Fibromuscular Dysplasia was not present initially.

Regarding imaging, it is important to appreciate the diverse presentations on angiograms, which are classified into two types: Multifocal Fibromuscular Dysplasia, which resembles the "string of beads" appearance due to varying areas of stenosis and dilation. It is seen in the middle and distal portion of the artery. Focal Fibromuscular Dysplasia is less common and occurs in any artery segment.

The Best initial study to confirm or exclude Fibromuscular dysplasia is computed tomographic angiography (CTA). If it is contraindicated, magnetic resonance angiography is preferred [4]. DUPLEX ultrasound is considered the initial imaging modality in qualified centers [4]. Invasive Digital subtraction angiography is regarded as the "gold standard." It excludes background bone and soft tissue and provides higher resolution [4]. In this case, the symptoms and elevated biomarkers of the patient required an initial coronary angiogram, revealing a spontaneous coronary artery dissection (SCAD). The association of spontaneous coronary artery dissection with Fibromuscular Dysplasia has become more apparent over the years. The "DISCO" study, conducted over a two-year span, showed that in 373 patients with spontaneous coronary artery dissection,340 had concomitant Fibromuscular Dysplasia. Spontaneous coronary artery dissection is a non-atherosclerotic, non-iatrogenic separation of the coronary wall with accumulation of blood. Coronary insufficiency occurs when the false lumen compresses the true lumen externally. The cause is not known [4]. Magnetic resonance angiography was performed, and renal artery stenosis was present in the classical "string of beads" appearance, as multi-focal Fibromuscular dysplasia is found in 91.5% of patients with renal involvement [6]. Catheter-based angiography still stands as the gold standard when detecting renal artery stenosis. In our case, a renal magnetic resonance angiogram confirmed the presence of Fibromuscular Dysplasia. Although the patient did not experience any neurological symptoms, computed tomography angiography of the brain was performed to rule out cerebrovascular involvement. Multiple Intra-cranial aneurysms were present. Patients are recommended for screening for intracranial aneurysms as it is prevalent in this population [7]. The risk of rupture depends on factors related to the aneurysm (size, location, number) and patient features (co-morbidities, smoking history)[6].

Management: Hypertension in this population group should be addressed as most patients have Hypertension. The Initial drug class to treat renovascular Hypertension is an Angiotensin-converting enzyme inhibitor(ACEI) or angiotensin receptor blocker (ARB). Revascularization is an option. However, clinical trials assessing the outcomes of renal artery revascularization occurred in atherosclerotic renal artery stenosis. Difference in disease pathophysiology and progression compared to Fibromuscular Dysplasia makes these studies un-applicable in this patient population [4]. Options for revascularization include percutaneous transluminal angioplasty or surgery. Extensive Comparative trials between the two have not been performed. However, observational studies have shown that percutaneous transluminal angioplasty can achieve similar success with less risk of complications[1-8]. Surgical intervention is warranted in the occurrence of a renal artery aneurysm or multifocal Fibromuscular Dysplasia in children[1].

Clinical Trials evaluating the efficacy of medical therapy are scarce. Nevertheless, the use of anti-platelet medication is acceptable due to the risk of thrombotic and thrombo-embolic complications Fibromuscular Dysplasia patients face, even in the absenteeism of aneurysms or dissections [10]. In the US registry, 72.9% of patients were given anti-platelet therapy, aspirin being the most prescribed [11].

Performance of percutaneous coronary intervention (PCI) is linked with less favorable results compared to patients with atherosclerotic disease due to the possibility of dissection extension with the false lumen along with it [12]. Moreover, patients who have undergone coronary artery bypass grafting (CABG) showed high long-term graft failures [12]. Thus, medical management is preferred. However, the optimal length and regimen of anti-platelet therapy are still unclear [4].

An intracranial aneurysm can be managed by surgical clipping or coil embolization. Factors such as patient preference, expert opinion, and anatomical variations should be considered [13].

In cases of an unruptured aneurysm, follow-up imaging is advised to assess the growth of the aneurysm. The frequency of follow-up imaging is yet unclear [7]. In our case, the patient was put on dual anti-platelet aspirin and clopidogrel therapy. The patient's spontaneous coronary artery dissection and intracranial aneurysms are monitored and have not deteriorated.

Conclusions

Our case report emphasizes the importance of exploring all vascular territories potentially implicated in this disease. Multiple imaging modalities can showcase typical radiological characteristics. Data interpreting long-term outcomes is scarce. However, association with cranial nerves shows to have a poorer prognosis.

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