CLINICAL VIGNETTE

Coronary Computed Tomography Angiography in Fibromuscular Dysplasia Patients with Chest Pain

Abdelmalek Habel[1], Emily Melik Aslanian[2], Roya Mojarrad, MD[3], Renata Stankovic, MD[3] and Peyman N. Azadani, MD[3]

[1]School of Medicine, University of Algiers, Algiers, Algeria
[2]Postbac MEDPREP Program, University of Michigan Medical School, Ann Arbor, Michigan, United States
[3]Department of Medicine, University of California Los Angeles (UCLA), Los Angeles, California, United States

Background

Involvement of coronary arteries is not well known in stable patients with Fibromuscular Dysplasia (FMD). We report a case with coronary artery involvement of FMD documented on coronary CT angiogram (CTA), obtained in absence of acute coronary syndrome. Two other cases with FMD did now show involvement of the coronary arteries.

Case 1: A 51-year old female with no significant past medical history was admitted to the Emergency Department with sudden onset of severe unilateral headache and neck pain. Carotid angiogram identified Internal Carotid dissection consistent with fibromuscular dysplasia (Figure.1). She later reported atypical chest pain and was evaluated for coronary artery disease with a coronary ECG-gated computed tomography angiography. This showed beaded appearance of proximal left anterior descending artery (Figure 2) most consistent with coronary artery involvement of her FMD.

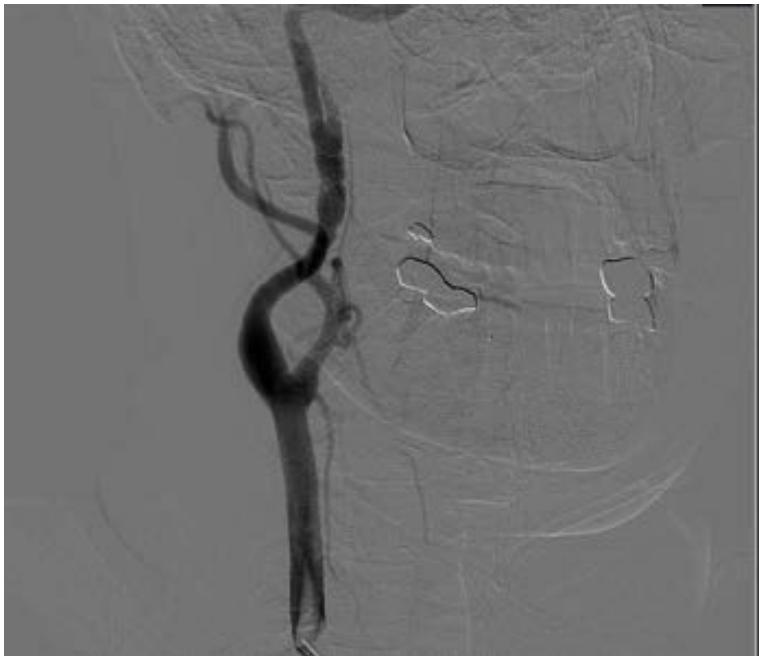

Figure 1: Left internal carotid artery angiography consistent with dissection extending from the C2 vertebral body level to the vertical petrous segment of the ICA. There is also a beaded appearance throughout the entire abnormality consistent with fibromuscular dysplasia.

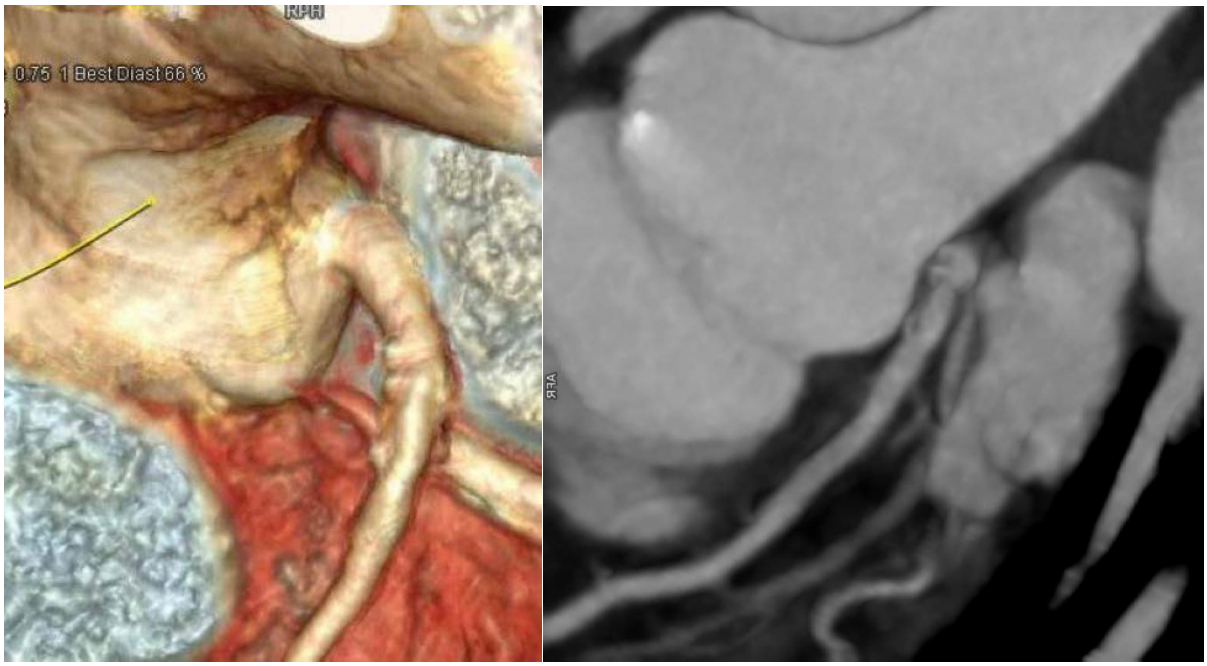

Figure 2. Coronary CTA of left anterior descending artery with old dissection/beaded appearance suggestive for FMD involving the coronary arteries.

Case 2: A 54-year-old female with history of hypertension and cardiac murmur presented to Cardiology Clinic to establish care. Her echocardiogram showed bicuspid aortic valve with dilated proximal part of ascending aorta. As a result, she underwent chest and abdominal CT angiogram for further assessment of her aorta which showed dilated mid ascending aorta at 4.6 cm along with beaded appearance of the right renal artery and bilateral external iliac arteries consistent with FMD. Later she presented for preop cardiovascular exam with atypical chest pain. Her coronary CT angiogram showed normal coronary arteries with stable aortic aneurysm.

Case 3: A 43-year-old female with history of hypertension presented to outside hospital Emergency Department with acute chest pain due to ST elevation MI (STEMI). She underwent PCI to left anterior descending artery. She established care with UCLA cardiology clinic. Since coronary dissection could be a manifestation of FMD, a renal ultrasound was ordered for the patient which showed elevated velocities. Abdominal MR angiogram multifocal areas of ectasia consistent with fibromuscular dysplasia also involving her iliac arteries. Later she presented with chest pain and coronary CT angiogram showed patent stent with no involvement of FMD in the other coronary arteries.

Discussion

There is increasing evidence of higher prevalence of FMD in patients presenting with spontaneous coronary artery dissection.[1] Coronary artery involvement of FMD is not common but is an important condition that can manifest as acute coronary syndrome with potential cardiac death. Involvement of the left anterior descending artery has been reported.[2] However, prevalence and other risk factors are still unknown.[1-4] The beaded appearance suggestive for FMD involving the coronary arteries has been previously reported.[3] Involvement of mid to distal segments of coronary arteries has been shown in patients with FMD[1] versus proximal artery involvement as shown in our first case. If coronary artery involvement in FMD in not a recognized risk of developing acute coronary syndrome, coronary artery dissection, left ventricular dysfunction, and mortality, may not be appreciated.[3]

Coronary CTA is a useful imaging modality in patients with known FMD. Although limited evidence exists to guide treatment, coronary CTA may help with the decision to imitate antiplatelet therapy and tighter blood pressure control. It may further assess risk of dissection in patients with coronary involvement. The first patient had coronary CT angiographic manifestations of coronary FMD and was treated with baby aspirin and anti-hypertensive agents. The second patient was reassured regarding her chest pain and CT angiogram prevented an invasive procedure. Tighter blood pressure control was also obtained. Coronary CT angiogram prevented a repeat higher risk angiogram in the third patient given her FMD involving her iliac artery.

In conclusion, coronary CTA is useful to assess coronary artery involvement in FMD patients. CTA can rule out obstructive disease and prevent invasive coronary angiography. We believe the first patient's strong of beads appearance on the proximal left anterior descending coronary artery is the first demonstration of FMD involvement on coronary CT angiogram in the absence of acute coronary syndrome. Future studies should prospectively review findings and outcomes in patients with known FMD involving coronary arteries to better guide the treatment.

REFERENCES

1. **Michelis KC, Olin JW, Kadian-Dodov D, d'Escamard V, Kovacic JC**. Coronary artery manifestations of fibromuscular dysplasia. *J Am Coll Cardiol*. 2014 Sep 9;64(10):1033-46. doi: 10.1016/j.jacc.2014.07.014. PMID: 25190240; PMCID: PMC4370367.
2. **Alfonso F, García-Guimaraes M, Bastante T, de la Cuerda F, Antuña P, Cuesta J, Rivero F**. Spontaneous coronary artery dissection: from expert consensus statements to evidence-based medicine. *J Thorac Dis*. 2018 Jul;10(7):4602-4608. doi: 10.21037/jtd.2018.07.10. Erratum in: *J Thorac Dis*. 2019 Jan;11(1):E19. PMID: 30174912; PMCID: PMC6105997.
3. **Saw J, Bezerra H, Gornik HL, Machan L, Mancini GB**. Angiographic and Intracoronary Manifestations of Coronary Fibromuscular Dysplasia. *Circulation*. 2016 Apr 19;133(16):1548-59. doi: 10.1161/CIRCULATIONAHA. 115.020282. Epub 2016 Mar 8. PMID: 26957531.
4. **Gornik HL, Persu A, Adlam D, Aparicio LS, Azizi M, Boulanger M, Bruno RM, de Leeuw P, Fendrikova-Mahlay N, Froehlich J, Ganesh SK, Gray BH, Jamison C, Januszewicz A, Jeunemaitre X, Kadian-Dodov D, Kim ES, Kovacic JC, Mace P, Morganti A, Sharma A, Southerland AM, Touzé E, van der Niepen P, Wang J, Weinberg I, Wilson S, Olin JW, Plouin PF**. First International Consensus on the diagnosis and management of fibromuscular dysplasia. *Vasc Med*. 2019 Apr;24(2):164-189. doi: 10.1177/1358863X18821816. Epub 2019 Jan 16. Erratum in*: Vasc Med*. 2019 Oct;24(5):475. Erratum in: *Vasc Med*. 2021 Aug;26(4):NP1. PMID: 30648921.