

Clinical Image

An Unusual Presentation of Fibromuscular Dysplasia

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CLINICAL IMAGE

Case

A 47-year-old man developed sudden, severe left flank pain while driving, and went to a local emergency room. He had a past medical history significant for kidney stones. Family history was significant for aneurysm on the mother's side. On presentation, physical examination revealed a blood pressure (BP) of 139/88 mm Hg and regular pulse 62/min, with no radio-radial or radio-femoral delay. Heart and lung examination was unremarkable. There was no carotid, renal or femoral artery bruits. Initial work-up included serum creatinine 1.06 mg/dL, and urinalysis negative for blood, protein, and white blood cells. The erythrocyte sedimentation rate (ESR) was 36 mm/hr. The remaining laboratory values including plasma biochemistry, inflammatory markers, and complements were within normal range. Renal ultrasound was obtained and which showed left renal infarction. Echocardiogram did not show any thrombus or vegetation. Computed tomography (CT) angiography revealed beading consistent with fibromuscular dysplasia (FMD) at the distal main renal artery at the junction of the bifurcating branches (Figure 1) and tapered narrowing and occlusion in the more proximal artery (Figure 2) most likely caused by spontaneous dissection with intramural hematoma, with resulting substantial upper pole infarct. The vascular surgery consult was obtained and with normal BP and renal function no angioplasty was pursued. Renal infarct was managed conservatively with hydration and non-steroidal anti-inflammatory medications.

Diagnosis

Left renal infarction in the setting of fibromuscular dysplasia.

DISCUSSION

FMD is a non-inflammatory, non-atherosclerotic disorder that leads to arterial stenosis, occlusion, aneurysm, and dissection. FMD typically presents in middle aged women, often with hypertension [1]. Depending on which arteries are involved, patients may present with transient ischemic attack, headache, pulsatile tinnitus, flank pain, or stroke [2]. CT angiography,

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Submitted: 17 January 2016

Accepted: 08 February 2017

Published: 09 February 2017

ISSN: 2379-0652

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Figure 1 Computed tomography (CT) angiography revealed beading consistent with fibromuscular dysplasia (FMD) at the distal main renal artery at the junction of the bifurcating branches.



Figure 2 Tapered narrowing and occlusion in the more proximal renal artery.

magnetic resonance angiography, duplex ultrasound and catheter based digital subtraction angiography can be ordered to confirm the diagnosis. Differential diagnosis includes atherosclerosis, polyarteritis nodosa, takayasu arteritis, ehlers-danlos syndrome, marfan's syndrome, and primary and secondary hypertension. Treatment strategies for FMD includes BP control, and angioplasty and surgery is pursued in some cases. Renal infarct in the setting of FMD is managed by supportive therapy, including analgesics, anti-coagulation and adequate control of BP.

CONCLUSION

FMD is a non atherosclerotic disease of medium-sized arteries that affects mainly women. The hypertension is the most common clinical presentation. Renal infarction is seen in only 0.9% of the patients with FMD [3]. This case is unique as we reported FMD in male who presented with renal infarction. Our patient had no inflammatory characteristics or clinical symptoms such as fever, night sweats, malaise, weight loss, arthralgia, or myalgia thus

distinguishing the diagnosis from vasculitis disorders such as takayasu arteritis. There was no prior history of renal disease, ehlers-danlos syndrome, or marfan's syndrome. Our patient has done well with no anticoagulation or invasive intervention and return of renal function to his prior baseline.

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Cite this article

Al Nimri O, Malhotra R, Mazer M, Salani M, Burks ML, et al. (2017) An Unusual Presentation of Fibromuscular Dysplasia. *J Clin Nephrol Res* 4(1): 1057.