

A Normotensive Patient with Fibromuscular Dysplasia Presenting as Unilateral Renal Infarction

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Fibromuscular dysplasia is a non-inflammatory non-atherosclerotic vascular disease that affects the entire arterial bed, but most frequently the renal and internal carotid vasculature [1]. Clinical manifestations can range from asymptomatic disease to a multisystem disease resembling necrotizing vasculitis [1]; most commonly however, FMD presents as poorly controlled hypertension, typically in younger women.

We describe the case of a normotensive woman with renal artery FMD who presented with unilateral renal infarction as a complication that has been sparsely described in the past [2].

PATIENT DESCRIPTION

A 56 year old woman presented with a 2 day history of abdominal pain and vomiting. On presentation she denied fever, changes in bowel movements, and urinary complaints. Her medical history was remarkable for hyperlipidemia, hypothyroidism and fibromyalgia. The patient did not smoke. She was treated with venlafaxine (75 mg twice daily) and levothyroxine (75 µg/day).

Her vital signs were normal, as were blood pressure values (124/75 mmHg). Physical examination revealed slight abdominal tenderness but was otherwise unremarkable. Laboratory evaluation yielded an elevated white blood cell count

of 17.25 K/µl with neutrophilia, and elevated lactate dehydrogenase levels of 863 IU/L (normal 100–260 IU/L). Other blood chemistry assays were within normal limits. Plasma renin activity and aldosterone levels were not obtained. Chest and abdominal X-rays were normal. A kidney triple-phase protocol computed tomography scan with contrast demonstrated a peripheral wedge-shaped area of low attenuation in her right kidney, suggesting the diagnosis of renal infarction. Although the CT was not performed in a dedicated angiographic phase, irregularities were demonstrated in both renal arteries and further dedicated workup was recommended. For further evaluation a magnetic resonance angiogram was performed, which demonstrated a peripheral wedge-shaped non-enhancing area, correlating with a renal lower pole infarction. Both renal arteries demonstrated segments with alternating stenosis and dilatation [Figure]. This configuration is commonly referred to as the “chain of beads.” The aorta, celiac trunk and superior mesenteric artery were normal. These imaging findings support the diagnosis of bilateral FMD of the renal arteries. In addition, workups for a thromboembolic source, genetic or acquired hypercoagulable state, and arrhythmia yielded no abnormal findings.

During her hospitalization the patient was normotensive (blood pressure levels 100–125/65–85 mmHg) and completely asymptomatic. The abnormal laboratory findings, including LDH levels, were normalized within a few days and the patient was discharged with the addition of

aspirin. Since then, she has undergone an MR angiogram of the carotid and cranial vasculature, which ruled out pathological processes in other vascular areas.

COMMENT

We present a female patient with renal infarct due to FMD. Patients with renal infarction usually have risk factors for thromboembolic events, such as atrial fibrillation, valvular heart disease, or intrarenal arterial thrombosis, yet some are reported to be of idiopathic origin. When the workup reveals no source for emboli or intrarenal arterial thrombosis, it is reasonable to proceed with either CTA or MRA scans. Such studies should be tailored to the renal arteries in order to search for arterial pathologies such as FMD, even in normotensive patients. Our patient demonstrated no source of emboli and the MRA was diagnostic for FMD.

Fibromuscular dysplasia is a non-inflammatory non-atherosclerotic vascular disorder that leads to arterial stenosis [1]. The disease involves the renal arteries (60–75%) followed by the internal carotids (25–30%), but it may affect the entire arterial bed. The most common presentation is resistant hypertension, followed by transient or sustained cerebral ischemia, headaches, dizziness, and tinnitus [1]. Our patient was normotensive and the first presentation of FMD was renal infarction. Several case reports of renal infarct in patients with FMD have been reported, but almost all were described in hypertensive subjects [2]. To our knowledge this is the first report of renal infarction due

FMD = fibromuscular dysplasia

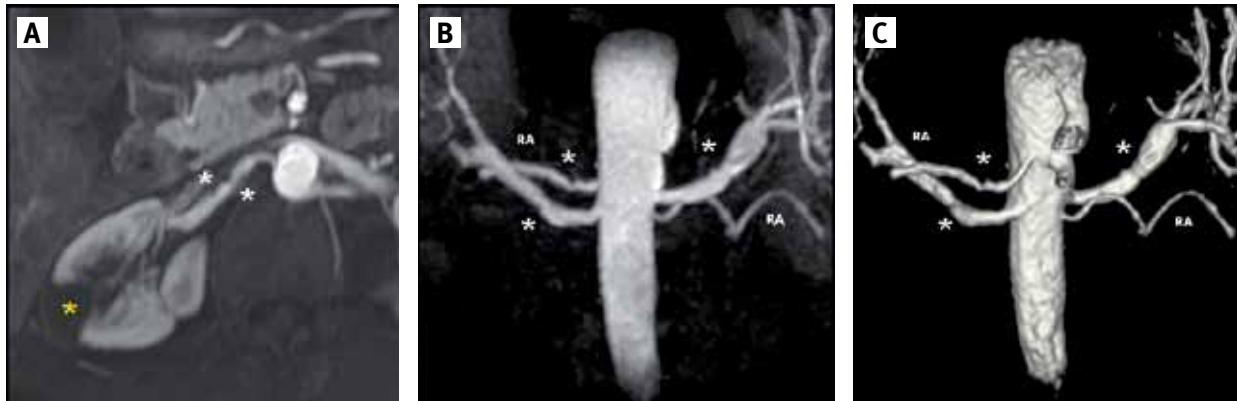
LDH = lactate dehydrogenase

CTA = computed tomography angiography
MRA = magnetic resonance angiography

Scan performed using a 1.5T scanner (GE)

[A] Axial three-dimensional spoiled gradient echo sequence with volume acceleration (LAVA) is a technique designed to acquire images with high definition, coverage and speed, following gadolinium administration (1.5 Tesla, Signa HDx Ver 15, GE Healthcare, Milwaukee, WI, USA). A wedged-shaped hypointense segment in the right kidney (yellow asterisk) represents a renal infarct. In the right renal artery the adjacent segments of stenosis and dilatation (white asterisks)

resemble “chain of beads.” **[B]** Non-contrast MRA (Inhance, GE Medical Systems). Maximal intensity projection demonstrates two accessory renal arteries (RA) bilaterally. Multiple segments of stenosis and dilatation are shown (white asterisks). **[C]** Non-contrast MRA (Inhance, GE Medical Systems). Volume rendering demonstrates two accessory renal arteries (RA) bilaterally. Multiple segments of stenosis and dilatation are shown (white asterisks)



to FMD in a normotensive subject. The normotensive state in our patient may be related to non-obstructive lesion in the renal artery which does not activate the renin angiotensin system. Unfortunately, we do not have data on plasma renin activity to confirm this explanation.

In the past the gold standard for diagnosis of renal artery FMD was digital subtraction angiography [1]. There are several non-invasive imaging modalities that are also useful for diagnosis of FMD, such as Doppler sonography, CTA and MRA [1]. In our patient the diagnosis was made by renal CTA and MRA.

The primary goal in treating patients with renal artery FMD is to control blood pressure and prevent the sequelae of long-standing hypertension. Revascularization should be considered in patients with recent-onset hypertension, drug-resistant hypertension, intolerance of hypertensive medications, and loss of renal volume secondary to ischemic nephropathy [1]. Our patient was normotensive and presented with renal infarction due to bilateral FMD. It is not clear whether this presentation justifies revascularization. Notably, a recent meta-analysis has demonstrated that both percutaneous transluminal renal artery

angioplasty and surgery have yielded only moderate benefits in patients with renal artery FMD, with substantial variations across various studies [3]. Although the loss of renal mass occurs in up to 63% of renal artery FMD patients, renal failure is rare. The benefit of revascularization in atherosclerotic renal artery stenosis is doubtful. Wolak et al. [4] showed that PTRAs improved blood pressure control but was ineffective in restoring renal function [4], whereas Kobo et al. [5] showed that PTRAs in symptomatic patients with renal artery stenosis improves blood pressure control and renal function. In light of the accumulated data in the literature we decided not to perform revascularization in our patient.

Since the underlying mechanism for the renal infarct in FMD is not entirely clear, there is a dilemma whether anticoagulation should be initiated to prevent another renal infarct and subsequent renal failure. Anticoagulation is clearly indicated when warranted by the underlying disease (e.g., atrial fibrillation, left ventricular thrombus, hypercoagulable state) but not in other settings, such as

PTRA = percutaneous transluminal renal artery angioplasty

tumor or fat embolization or aortic dissection. Evidence is lacking regarding the benefit of anticoagulation for patients with infarction secondary to FMD.

In conclusion, the present case suggests that FMD should be suspected as a cause of renal infarction even in a normotensive subject.

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