Clinical Case Report

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# Unique imaging findings in fibromuscular dysplasia of renal arteries

## A case report

Weiying Kong, MD, Zhangxue Hu, MD*

**Abstract**

**Rationale:** Fibromuscular dysplasia (FMD) is a rare vascular disorder that causes abnormal cell growth in arterial walls. The classic "string of beads" sign has been reported in many cases, whereas the appearance of tubular stenosis and distal tapering of renal arteries with multiple renal infarctions, as well as left kidney atrophy occurring in one patient, has not been precisely described.

**Patient concerns:** A 19-year-old woman presented to us with a history of elevated blood pressure without any symptoms for the past 1 month. Routine laboratory tests indicated a moderately impaired renal function, and ultrasound examination demonstrated a small-sized left kidney and seriously decreased blood flow of the left renal artery and its branches.

**Diagnosis:** Subsequent contrast-enhanced computed tomographic angiography (CTA) demonstrated multiple ischemic infarctions in the bilateral kidneys, and FMD was suggested at that time. Thereafter, we performed selective reno-angiography, which confirmed that the all left renal arteries had tubular stenosis and that right renal arterial branches presented distal tapering.

**Intervention:** Antihypertensive drugs were prescribed conservatively, including nifedipine 60 mg/d and prazosin 4 mg/d, to lower the patient's blood pressure.

**Outcomes:** The patient had a well-controlled blood pressure and an improved renal function at her 6-month follow-up.

**Lessons:** We should take the diagnosis of FMD into account if young women develop asymptomatic hypertension. To our knowledge, this is the first case that exhibited renal artery FMD manifesting as tubular stenosis and distal tapering, especially followed by bilateral renal infarctions and significant atrophy of the left kidney. In addition, CTA combined with digital subtraction angiography (DSA) may be more sensitive than other tests with respect to the detection of intrarenal infarctions and arterial variants of FMD.

**Abbreviations:** 3D = three-dimensional, CRP = C-reactive protein, CTA = computed tomographic angiography, DSA = digital subtraction angiography, FMD = fibromuscular dysplasia, GFR = glomerular filtration rate, LDH = lactate dehydrogenase, SCr = serum creatinine.

**Keywords:** distal tapering, fibromuscular dysplasia, renovascular hypertension, tubular stenosis

### 1. Introduction

Fibromuscular dysplasia (FMD) is a noninflammatory, non-atherosclerotic disease of the arterial wall musculature that commonly involves the renal, extracranial carotid, and vertebral arteries.[1–3] Areas involving the arteries can thicken, enlarge, narrow, and even can manifest as tapering, making it difficult for blood to flow through. The classic "string of beads" appearance has been described in many cases; however, the appearance of tubular stenosis and distal tapering with bilateral renal infarctions has not been precisely described, to the best of our knowledge. Here we report a rare case of a young female with a history of hypertension whose renal arteries manifested as typical tubular stenosis and distal tapering with multiple ischemic infarctions of the bilateral kidney as well as an atrophic left kidney on imaging. The patient was treated conservatively with antihypertensive drugs and found to have a well-controlled blood pressure as well as improved renal function at her 6-month follow-up.

### 2. Case report

A 19-year-old female presented to us with a history of elevated blood pressure for 1 month. She had been feeling well until a routine physical examination performed 1 month before her presentation showed that her blood pressure was 197/138 mmHg. She denied experiencing fever, arthralgia, dizziness, nausea, vomiting, or gross hematuria. On admission, her blood pressure was 183/131 mmHg with a heart rate of 83 beats/min. No abdominal bruits were auscultated. Routine laboratory tests showed that her serum creatinine (SCr) level was 1.5 mg/dL, the levels of serum C-reactive protein (CRP) and lactate dehydrogenase (LDH) were in the normal

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*Informed Consent: Written informed consent for publication of clinical details and images were obtained from the patient. A copy of the consent form is available for review upon request*

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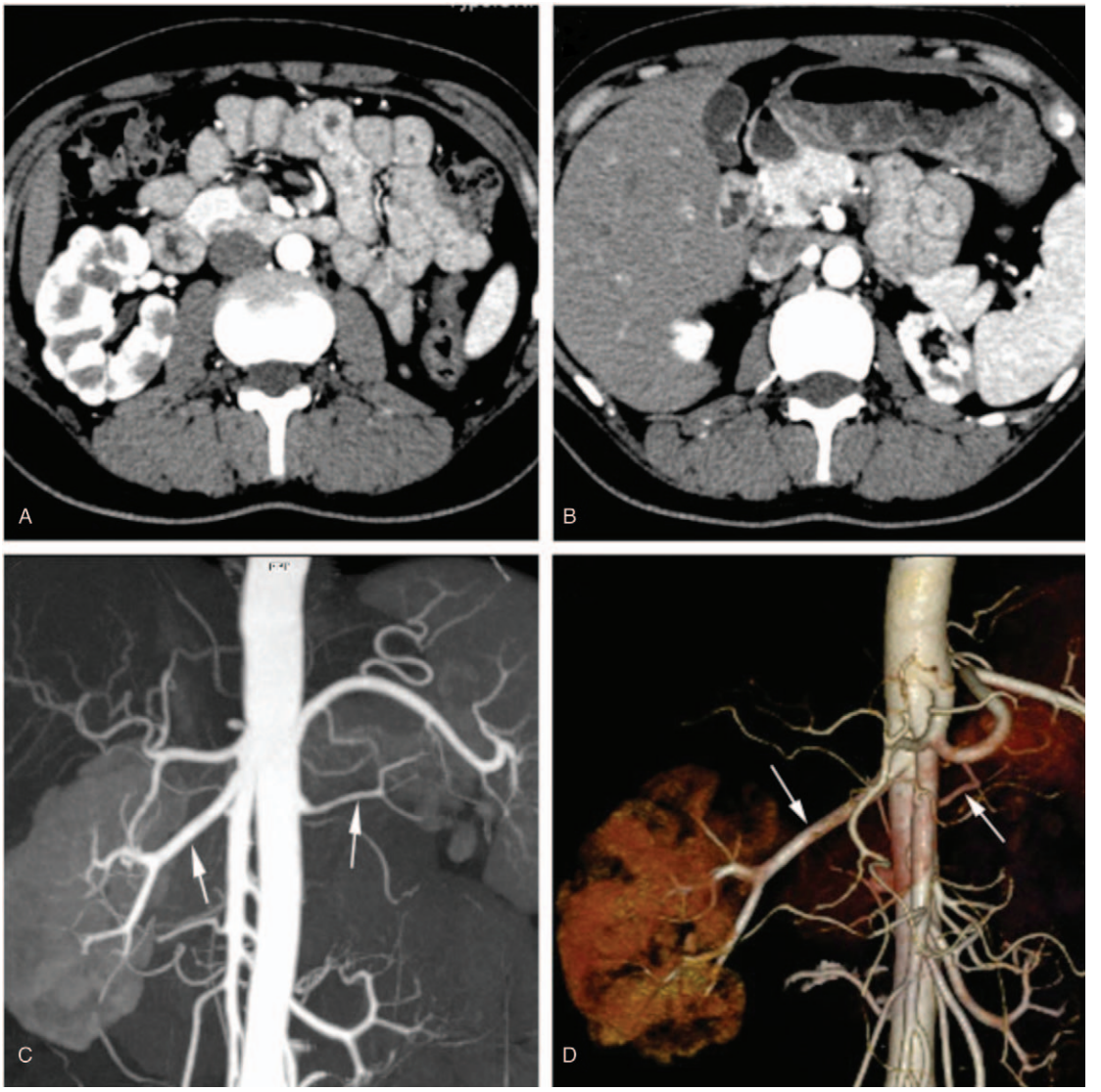


**Figure 1.** (A–B) CTA showed that multiple ischemic infarctions, partial cortical thinning, and irregular jagged edges in the bilateral kidney. (C–D) 3D reconstruction showed that the left renal artery was much thinner and smaller than the right on imaging (arrows).

range, and the aldosterone-to-renin ratio (ARR) was 5.16 ng/dL per ng/(mL·h). Her urinalysis was normal, and the urinary protein excretion was 590 mg/24 h. Other tests such as evaluation of leukocyte, blood lipid, and hepatitis marker levels and immunological evaluations revealed negative results. Echocardiography showed that the left ventricle was hypertrophic, and duplex ultrasonography of the carotid arteries revealed the presence of a thickened intima-media at the origin of the right subclavian artery. Further renal ultrasonography indicated that the left kidney was significantly atrophic ($6.3 \times 3.5 \times 3.7$ cm) and that the blood flow to the kidney from the left renal artery and its branches was severely impaired, with the peak systolic velocity of the left renal artery being 46.6 cm/s and the peak velocity of the segmental arteries being 29.6 cm/s. In addition, both kidneys were determined to have a decreasing glomerular filtration rate (GFR: left kidney, 12.6 mL/min; right kidney, 30.7 mL/min). Computed tomographic angiography (CTA) was performed subsequently, which revealed multiple ischemic infarctions, partial cortical thinning, and irregular jagged edges in the bilateral kidneys (Fig. 1A, B). Meanwhile, three-dimensional (3D) reconstruction revealed that the left renal artery was much thinner and smaller than the right (Fig. 1C–D, arrows). FMD was suggested at that time, and selective reno-angiography was performed, which confirmed that tubular stenosis of the left renal artery extended into the branches (Fig. 2A). Although there was no obvious stenosis in the right renal artery, distal tapering was clearly seen at its branches (Fig. 2B). In addition, thrombus formations at the branches of the right artery were observed on imaging (Fig. 2B). The patient was therefore diagnosed with "tubular and tapering" FMD. Considering that the left renal artery and its branches all had severe tubular stenosis, nifedipine at 60 mg/d and prazosin at 4 mg/d were prescribed to lower the patient's blood pressure. Her blood pressure was controlled at 138/89 mmHg and SCr level was 1.3 mg/L when she was discharged. During the subsequent 6-month follow-up, her blood pressure remained well controlled and SCr level was stable.

## 3. Discussion

FMD, initially described in 1938 by Leadbetter and Burkland, is a rare noninflammatory, nonatherosclerotic vascular disorder that causes abnormal cell growth in arterial walls. As a result of

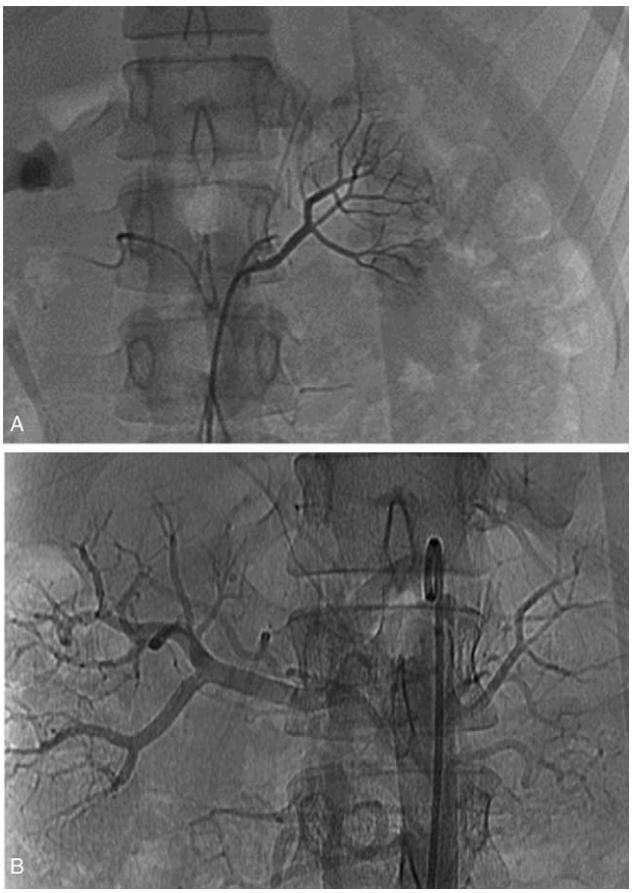


**Figure 2.** (A) Selective reno-angiogram showing tubular stenosis of the left renal artery extending into its branches. (B) Distal tapering and thrombi were observed at the branches of the right renal artery.

abnormal cell growth in the arterial walls, areas of the arteries can thicken, enlarge, narrow, and even can manifest as tapering, making it difficult for blood to flow through. It can affect every artery in the body, but usually affects the renal, extracranial carotid, and vertebral arteries.[1–3] It is important to note that FMD is a significant cause of renovascular hypertension, especially in young women. The histologic descriptions of medial, intimal, and adventitial FMD by Harrison and MCcormack in 1971[4] have been replaced with an angiographic classification system.[1,5] On the basis of pathological–angiographic correlations, Kincaid et al proposed the following 3 types of renal artery stenosis of FMD: multifocal (string-of-beads appearance), unifocal (solitary stenosis $<1$ cm in length), and tubular (stenosis $\geq 1$ cm in length).[6] Specifically, FMD may also manifest as aneurysm, dissection, occlusion, arterial tortuosity as well as distal tapering on angiographic imaging.[7] Tubular stenosis is characterized by the presence of an elongated, smooth, concentric narrowing of the renal artery caused mainly by medial hyperplasia and intimal fibroplasia.[6] This Chinese patient presented with renovascular hypertension, and comprehensive examinations showed significant tubular stenosis of the left renal arteries and distal tapering of the branches of the right artery with marked atrophy of the left kidney, as well as bilateral renal infarctions, but normal levels of CRP and LDH. There was no evidence of extrarenal artery stenosis, nor was there evidence of an immune disorder. All of the above findings confirmed the diagnosis of FMD.

Interestingly, CTA and 3D reconstruction demonstrated the presence of heterogeneously enhancing lesions, multiple filling defects, partial cortical thinning, and irregular jagged edges. The above findings proved that there were multiple ischemic infarctions in both kidneys, which is a rare complication of FMD that results from tubular stenosis, distal tapering, as well as thrombi.[8–13] It should be noted that bilateral renal infarctions are much less common than unilateral renal infarctions.[8–10,14,15] Acute large renal infarctions usually present with abdominal and flank pain, as well as fever, nausea, vomiting, hypertension, elevated LDH, and leukocytosis. Microscopic hematuria and mild proteinuria may be noted on urinalysis. SCr levels may increase if the infarctions are bilateral and involve important branches. Although this patient had suffered multiple renal infarctions and had developed severe left renal arteries stenosis, the right main renal artery was patent, which may explain why she was able to maintain moderate renal function despite her disease. The patient was virtually asymptomatic, which suggests that her infarctions happened over a long period of time. It is notable that, although invasive angiography is the gold standard for diagnosing FMD, the infarcted area seemed to be more obvious on 3D-CTA than on invasive angiography. The combination of CTA and DSA may be more sensitive than other tests with respect to the detection of intrarenal infarctions, as well as arterial "tubular and tapering" variants of FMD.

## Author contributions

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