Unique presentation of renovascular hypertension due to fibromuscular dysplasia

Mitali Doshi, MD,[a] Peter Layman, DO,[b] Henri Justino, MD,[c] *and* Mahmoud Malas, MD, MHS, RPVI, FACS,[b]
Worcester, MA; La Jolla and San Diego, CA

ABSTRACT

A 21-year-old female with a history of right nephrectomy due to trauma presented with several years of multidrug-resistant hypertension. Her workup included negative findings from autoimmune and vasculitides panels and urine catecholamine testing. Computed tomography showed an acute hairpin turn of her left renal artery. Intraoperatively, the artery demonstrated kinking with exhalation. She underwent excision of the diseased portion of the renal artery and an end-to-end anastomosis. Final pathologic examination demonstrated fibromuscular dysplasia. This is a unique case of mechanical artery kinking combined with fibromuscular dysplasia contributing to renovascular hypertension, for which open surgery was beneficial at improving the patient's hypertension. (J Vasc Surg Cases Innov Tech 2023;9:101257.)

Keywords: Fibromuscular dysplasia; Renal artery; Renal artery stenosis; Renovascular hypertension

Renovascular hypertension is predominantly due to atherosclerotic renal artery stenosis (90%).[1] Fibromuscular dysplasia (FMD) is the second most common cause of renovascular hypertension (10%) and the most common cause in the younger population.[1] FMD is an idiopathic, nonatherosclerotic, and noninflammatory arterial disease with a female gender predominance, with ~80% to 90% of patients being women. The disease primarily manifests as beaded lesions in medium or small-size arteries and most commonly affects the renal and extracranial carotid and vertebral arteries. We describe the case of a 21-year-old woman with renovascular hypertension who had no radiographic evidence of FMD, including no "beads on a string" and the absence of vasculitis or autoimmune laboratory test results, with a prominent area of kinking in the renal artery related to respiration. The patient provided written informed consent for the report of her case details and imaging studies.

From the University of Massachusetts, Chan Medical School, Worcester[a]; the Division of Vascular and Endovascular Surgery, University of California, San Diego, La Jolla[b]; and the Division of Pediatric Cardiology, Rady Children's Hospital, San Diego.[c]

Author conflict of interest: none.

Additional material for this article may be found online at www.jvscit.org.

Correspondence: Mahmoud B. Malas, MD, MHS, RPVI, FACS, Division of Vascular and Endovascular Surgery, University of California, San Diego, La Jolla, CA 92093 (e-mail: mmalas@health.ucsd.edu).

The editors and reviewers of this article have no relevant financial relationships to disclose per the Journal policy that requires reviewers to decline review of any manuscript for which they may have a conflict of interest.

2468-4287

https://doi.org/10.1016/j.jvscit.2023.101257

CASE REPORT

A 21-year-old, otherwise healthy, female patient presented to the hospital after a seizure and hypertensive emergency. She had had multiple prior admissions with similar presentations. Eleven years before the current admission, she had undergone right nephrectomy due to a traumatic motor vehicle accident. In recent years, she was taking amlodipine, metoprolol, and lisinopril daily for hypertension. An outpatient evaluation by her nephrologist included a urine catecholamine test with negative findings. A renal angiogram demonstrated left renal artery stenosis at the site of an acute hairpin turn in the vessel, with kinking of the area of stenosis with exhalation, followed by straightening during inhalation (Supplementary Video, online only and Fig 1). The mean arterial pressure of 75 mm Hg in the proximal renal artery decreased across the stenosis to 23 mm Hg and was not affected by the respiratory cycle. Intravascular ultrasound confirmed the presence of high-grade stenosis at the area of the hairpin turn of the artery (Fig 2). Renal artery duplex ultrasound demonstrated >60% stenosis of the proximal left renal artery (Fig 3). Computed tomography angiography showed an acute bend of her proximal left renal artery with a caliber change in the diameter and no intraluminal thrombus. At the site of maximal stenosis, the residual lumen was ~1 mm. None of the imaging studies showed a "beads on a string" appearance of the renal artery, carotid arteries, or other arteries to suggest FMD.

Given the patient's multiple hospital admissions with uncontrolled hypertension, intervention was deemed warranted. The initial plan was for excision of the redundant portion of her renal artery and an aortorenal bypass with a segment of the internal iliac artery. A left retroperitoneal approach was performed, and the aorta, left internal iliac artery, and left renal artery were exposed carefully. Clear kinking of the renal artery occurred with diaphragm movement during respiration (Fig 4). The patient became anuric before clamping of the vessel; thus, the plan was changed immediately to excision of the redundant

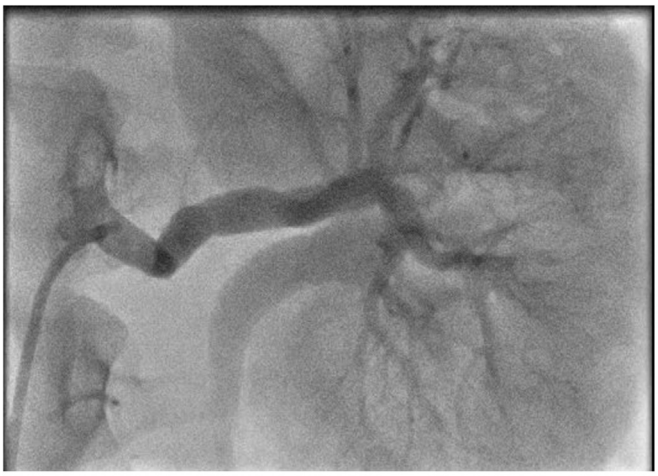

Fig 1. Still image of renal angiogram demonstrating maximal left renal artery mechanical kinking during exhalation.

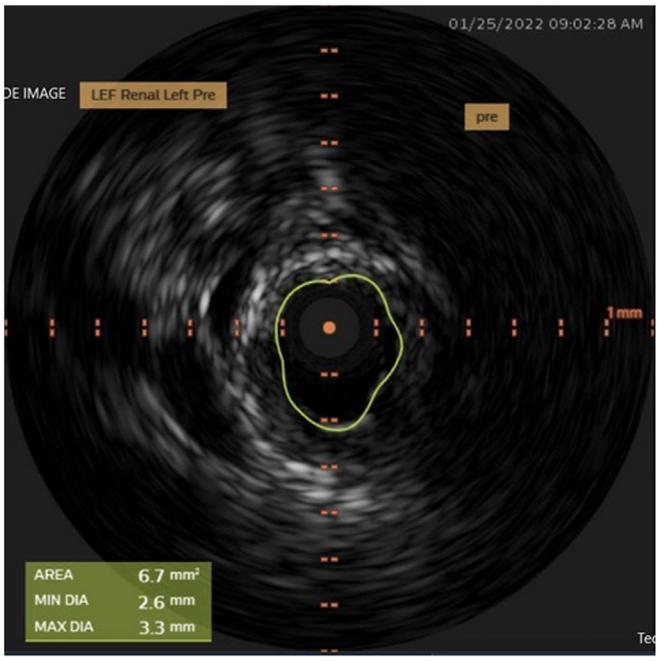

Fig 2. Intravascular ultrasound of left renal artery confirming stenosis. *DIA,* Diameter; *LEF,* left; *MAX,* maximum; *MIN,* minimum.

kinked segment of the artery and an end-to-end anastomosis (Fig 5). The anastomosis was technically difficult due to the location deep in the retroperitoneal space. The total renal ischemia time was 24 minutes. Intraoperative renal artery duplex ultrasound demonstrated no residual stenosis. However, the patient remained anuric for 1 hour after the anastomosis. The decision was made to close the abdomen and transfer the patient to the intensive care unit. The patient started to produce urine at 3 hours postoperatively, with 2 L of urine output within 2 hours. She recovered well, with a return of creatinine to the baseline value. Her average blood pressure before surgery was 165/108 mm Hg on her antihypertensive treatment. Postoperatively, it was 125/71 mm Hg on no antihypertensive medication. She was discharged home on postoperative day 4 with instructions to take 81 mg of aspirin daily.

Pathologic examination of the removed diseased portion of her renal artery demonstrated intimal hyperplasia, a medial myxoid change, elastic layer fragmentation, and medial fibrosis. Although these changes are consistent with a diagnosis of FMD, in the present patient, these pathologic changes were likely a result of the angulation and motion of the redundant renal artery. The laxity of the artery and kinking during the respiratory

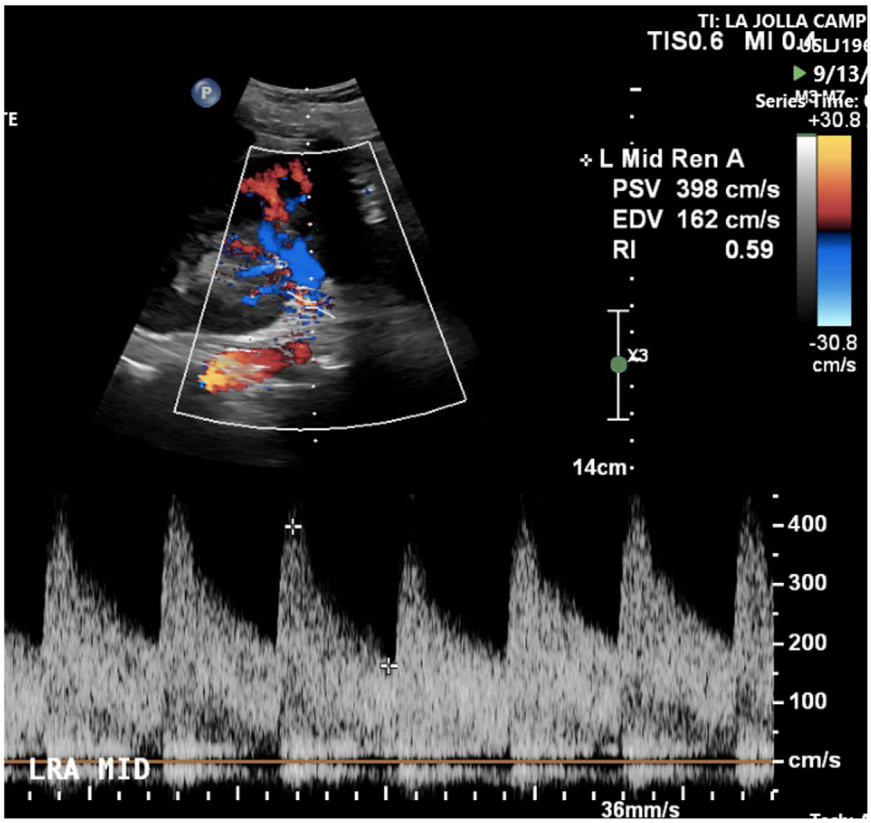

Fig 3. Preoperative renal artery duplex ultrasound demonstrating elevated velocities. *A,* Artery; *ESV,* end-systolic velocity; *L,* left; *LRA,* left renal artery; *MID,* middle; *Mid,* middle, *PSV,* peak systolic velocity; *Ren,* renal; *RI,* resistive index.

cycle must have led to the intimal hyperplasia and stenosis. This was probably an induced phenomenon, although underlying congenital FMD could have also contributed.

The patient was seen in the clinic at 1 and 6 months postoperatively. Duplex ultrasound of her left renal artery at 6 months after surgery demonstrated a peak systolic velocity of 144 cm/s, end-diastolic velocity of 38 cm/s, and a resistive index of 0.74.

DISCUSSION

The present case represents a unique case of renal artery stenosis. The phenomenon of renal artery redundancy causing stenosis has been described in renal transplant patients as a rare, but important, cause of early graft dysfunction for which balloon angioplasty is ineffective.[2,3] The present patient also had renal artery redundancy that was, however, due to unique laxity and angulation associated with the respiratory cycle. Few case reports have described such an induced phenomenon.

Additionally, our patient had had a solitary kidney for the previous 11 years before the current presentation. Research has demonstrated that in children with an acquired solitary functioning kidney, the risk of developing hypertension, albuminuria, and chronic kidney disease in later life is increased.[4] These compensatory mechanisms such as glomerular hyperfiltration of the solitary kidney and the physiology of the renin-angiotensin-aldosterone system likely also contributed in small part to her renal injury and hypertension.

Generally, endovascular therapy via renal artery angioplasty is a safe option for treating renal FMD, with favorable patency rates.[5] However, the outcomes after stenting and angioplasty vary and do not always cure the hypertension.[6] A literature review demonstrated that stent kinking and fracture have been reported in the setting of renal FMD.[7] Also, positional changes of the kidneys during respiration causes both bending and a change in angulation of the renal arteries, which can adversely affect the renal arteries after stenting owing to the risk of stent fractures from cyclical bending.[8] A case report of a female patient with FMD after renal artery stenting demonstrated similar renal artery intermittent kinking with respiratory and cardiac motion, leading to recurrent hypertension.[9]

To avoid these potential complications in our young patient with a long life expectancy, surgical repair was deemed the best option. The options discussed for

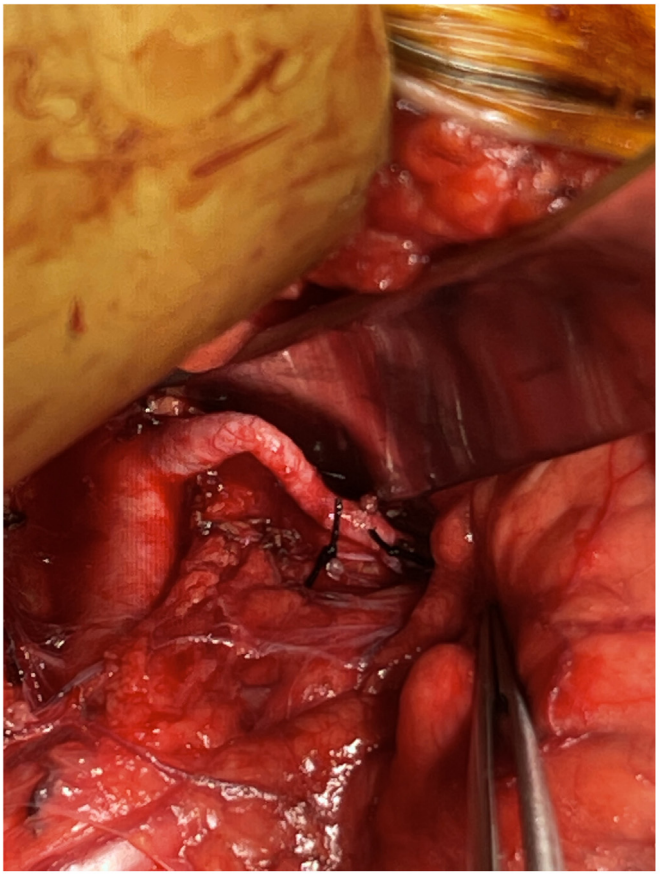

Fig 4. Intraoperative photograph showing left renal artery kinking with forced expiration during Valsalva maneuver.

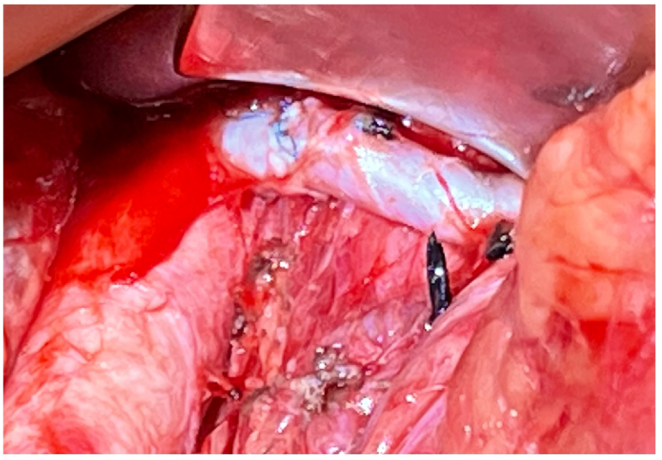

Fig 5. Intraoperative photograph showing final anastomosis of end-to-end renal artery transposition.

surgery included using an autogenous vein, an ipsilateral internal iliac artery as a conduit for the bypass graft, prosthetic material such as polytetrafluoroethylene for an aortorenal artery bypass, or an end-to-end anastomosis of the renal artery. Lower extremity vein mapping before surgery did not reveal adequate vein diameters. The left internal iliac artery was dissected before renal artery clamping to use as a possible conduit, and a polytetrafluoroethylene graft was also available. Because the patient became anuric during surgery, we chose to perform an end-to-end renal artery anastomosis because it would require the least renal ischemia time. The patient did well postoperatively.

CONCLUSIONS

We present the case of a 21-year-old female with elongation of her renal artery with prominent kinking seen during exhalation that caused multidrug-resistant hypertension and multiple admissions to the hospital. The patient's hypertension quickly resolved after open surgery. Management of renal artery FMD remains controversial. Open surgical revascularization can be beneficial in certain circumstances, especially for cases of cyclical kinking of the renal artery related to respiration. We recommend performing dynamic angiography and intravascular ultrasound for patients with FMD. If mechanically induced stenosis of the renal artery is documented, the patient should be offered open surgical repair because angioplasty has a high likelihood of recurrence.

REFERENCES

1. Gornik HL, Persu A, Adlam D, et al. First international consensus on the diagnosis and management of fibromuscular dysplasia. Vasc Med 2019;24:164-89.
2. DeVries BL, Wechsler B, Yim D. Case report of transplant renal artery stenosis secondary to mechanical renal artery kinking: balloon angioplasty as a supportive diagnostic tool? Int J Surg Case Rep 2021;83:106052.
3. Miah M, Madaan S, Kessel DJ, Newstead CG, Guleria S. Transplant renal artery kinking: a rare cause of early graft dysfunction. Nephrol Dial Transplant 2004;19:1930-1.
4. Westland R, Schreuder MF, Bökenkamp A, Spreeuwenberg MD, van Wijk JA. Renal injury in children with a solitary functioning kidney–the KIMONO study. Nephrol Dial Transplant 2011;26:1533-41.
5. Mousa AY, Campbell JE, Stone PA, Broce M, Bates MC, AbuRahma AF. Short- and long-term outcomes of percutaneous transluminal angioplasty/stenting of renal fibromuscular dysplasia over a ten-year period. J Vasc Surg 2012;55:421-7.
6. Birrer M, Do DD, Mahler F, Triller J, Baumgartner I. Treatment of renal artery fibromuscular dysplasia with balloon angioplasty: a prospective follow-up study. Eur J Vasc Endovasc Surg 2002;23:146-52.
7. Draney MT, Zarins CK, Taylor CA. Three-dimensional analysis of renal artery bending motion during respiration. J Endovasc Ther 2005;12:380-6.
8. Raju MG, Bajzer CT, Clair DG, Kim ESH, Gornik HL. Renal artery stent fracture in patients with fibromuscular dysplasia: a cautionary tale. Circ Cardiovasc Interv 2013;6:e30-31.
9. Wang LC, Scott DJ, Clemens MS, Hislop SJ, Arthurs ZM. Mechanism of stent failure in a patient with fibromuscular dysplasia following renal artery stenting. Ann Vasc Surg 2015;29:123.e19-21.

Submitted Apr 8, 2023; accepted Jun 2, 2023.