Oncology

Refractory hypertension due to unilateral renal lymphangiectasia: An uncommon case with a surgical solution

Robert N. Uzzo a, Evan Bloom a, Andrew Peters b, Meena Parab c, Selma Masic a, Alexander Kutikov a,c,*

a From the Department of Surgery, Division of Urology, Fox Chase Cancer Center – Temple University Health System, USA
b From the Department of Cardiology, Heart and Vascular Institute – Temple University Health System, USA
c From the Department of Anatomic Pathology, Fox Chase Cancer Center – Temple University Health System, USA

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ABSTRACT

We describe a case of unilateral renal lymphangiectasia (RLM) in a 30-year-old male with severe, refractory hypertension (HTN) and end-organ effects despite five anti-hypertensives. After diagnostic testing, the patient ultimately underwent a successful right laparoscopic nephrectomy with significant improvement of HTN. We review the literature regarding the pathophysiology and management strategies of HTN in patients with renal lymphangiectasia.

1. Introduction

While primary hypertension (HTN) accounts for most cases of sustained HTN, secondary HTN, due to a remediable cause, accounts for 5-10% of additional cases. Other than sleep apnea, secondary HTN is most commonly due to renovascular pathophysiology. We present a unique case of secondary HTN and end-organ effects due to unilateral renal lymphangiectasia (RLM). We review the presentation, evaluation, treatment with laparoscopic nephrectomy, and subsequent course. To our knowledge, this represents the first case of severe, refractory HTN due to unilateral RLM, and improvement following nephrectomy.

2. Case presentation

A 30-year-old male presented with severe, refractory HTN (>160/110 mmHg) despite five anti-hypertensive agents (lisinopril, amlodipine, clonidine, carvedilol, hydrochlorothiazide which was replaced with chlorthalidone). He was first diagnosed with HTN at age eight and started on lisinopril at age 19. He developed hypertensive retinopathy in his twenties with retinal hemorrhage and partial vision loss requiring multiple bilateral eye surgeries. His past medical history was remarkable for mild diabetes mellitus (NIDDM), chronic kidney disease (CKD) stage 2 (eGFR = 86), and obesity (BMI 31.4). Physical exam was unremarkable. Serum creatinine was 1.14 mg/dl, hemoglobin 13.5 g/dl, resting peripheral renin 21.7 ng/dl (ref = 0.167–5.38) on lisinopril, aldosterone 4.9 ng/ml/hr (ref = 0.30), and aldosterone/renin ratio 0.23 ng/ml/hr.

Renal artery duplex ultrasound revealed no arterial stenosis and normal resistive indices. Abdominal computerized tomography noted multiple right parapelvic cysts with pelvic calyceal system distortion and perinephric fluid suggestive of intrarenal and retroperitoneal lymphangiectasia (Fig. 1).

The patient underwent aspiration of the paranephric fluid to assess for lymphatic content given clinical suspicion for lymphangiectasia. The fluid noted a neutrophil-predominant (74%) leukocytosis with elevated white cell count (31 cells/mm3), absent chylomicrons, total protein content (<3.0 g/dl), and normal creatinine (0.9 mg/dl). These findings are consistent with prior descriptions of renal lymphangiectasia with the exception of his leukocytosis, which was neutrophil-predominant compared to previous small published series, which were lymphocyte predominant.

Following aspiration of all the paranephric fluid, the patient’s blood pressure (BP) remained unchanged. Further review of imaging revealed a diminutive versus absent main renal vein, and dilated perirenal and pararenal lymphatics. The lymphangiectasia appeared predominantly intrarenal and not amenable to additional percutaneous drainage or sclerosis. Given the patient’s refractory HTN with hypertensive retinopathy and adverse sequela, a right laparoscopic nephrectomy was recommended and subsequently performed (Fig. 2). Microscopic

* Corresponding author. Division of Urologic Oncology, Fox Chase Cancer Center, 333 Cottman Avenue, Philadelphia, PA, 19111, USA.
E-mail address: Alexander.Kutikov@fccc.edu (A. Kutikov).

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evaluation demonstrated endothelial-lined cysts containing lymph fluid and surrounding focal inflammation (Fig. 3).

After surgery, the patient’s BP regimen was reduced to two medications (lisinopril and carvedilol). At his 3-week follow-up, he reported systolic BPs between 140 and 150 mmHg at home. Two months postoperatively, his BP was 140/90 mmHg, and his carvedilol was increased slightly with a BP goal of <130/80 mmHg. Four months postoperatively, his BP was 135/100 mmHg on the same regimen. The patient recovered well surgically with a stable creatinine of 1.4–1.5 mg/dl. He was referred to nephrology given his complex comorbidity profile and solitary kidney.

3. Discussion

Renal lymphangiectasia is a rare clinical entity. Diagnosis is based primarily on radiological features of dilated perinephric and/or peri-pelvic lymphatic channels. It is believed to be a benign developmental malformation that results from noncommunication of perirenal and peri-pelvic lymphatic channels with the main lymphatic system. The resulting cystic lesions may be intrarenal and involving the renal sinus or parenchyma, or extrarenal and involving the perirenal fascia. Familial associations have been made in some cases arguing for a congenital etiology.

The nomenclature for the disorder is varied in the literature and includes renal lymphangiomatosis, renal lymphangioma, and renal peri-pelvic multi-cystic lymphangiectasia. Clinically the condition is most often asymptomatic but may present with variable manifestations including flank pain, hematuria, ascites, weight loss, HTN, or other vague constitutional symptoms. While overall renal function is usually preserved, cases of renal insufficiency with anemia or polycythemia have been reported, particularly when bilateral. Radiographically, the lesions may present as diffuse renal enlargement and/or dilated cystic lesions affecting various parts of the renal parenchyma and collecting system. Pathology can be unilateral or bilateral as well as focal or diffuse. Occasionally, patients present with retroperitoneal fluid in the subcapsular, perirenal or pararenal spaces.

In a series by Schwarz et al. examining 42 cases of symptomatic RLM, nearly 60% were associated with HTN. Of these, nearly 2/3 reported that HTN was reversible or markedly improved by drainage or resection of the cysts or nephrectomy. Several authors presume the mechanism of HTN is related to a Goldblatt-like effect on the renal parenchyma causing pressure-related ischemia and renin release. This compression effect is known as Page Kidney and has been described under a number of clinical conditions. The diagnosis is usually based on clinical suspicion and radiographic findings on ultrasound and/or CT. In typical cases, renal compression leads to ischemia and triggers release of vasoactive hormones by the renin-angiotensin-aldosterone system (RAAS). This in turn results in arterial HTN. Multiple pharmacological therapies targeting RAAS may improve BP control but fail to completely overcome the effects of renal compression on systemic BP. Interventions to reduce parenchymal compression such as cyst decortication or paraneaphric fluid aspiration may diminish the compressive effects on the kidney and improve BP. Unfortunately, in cases of predominantly intrarenal RLM as described in our patient, the compressive effects on renal parenchyma are intrarenal and diffuse and therefore less amenable to such interventions. While RLM may respond to medical therapy or even spontaneously regress in rare patients, the condition usually persists and necessitates escalation of therapy as was the case for our patient.

4. Conclusion

We present an unusual case of unilateral intrarenal RLM in a young man with refractory HTN and end-organ disease. Despite aggressive pharmacologic therapy with five antihypertensives, the HTN persisted. Given the diffuse and predominantly intrarenal distribution of lymphangiectasia, aspiration did not improve BP control and the patient ultimately underwent laparoscopic nephrectomy. He recovered well postoperatively and had significant BP improvement with a reduced antihypertensive regimen.

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Declaration of competing interest

The authors have no conflicts of interest to disclose.

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Fig. 3. Microscopic examination with hematoxylin and eosin (H&E) stains. (a) 2x magnification showing lymphangioma (center) in relation to renal parenchyma (top) and sinus fat (bottom). (b) 10x magnification showing cystic spaces lined by endothelial cells with focal inflammation as demonstrated by a lymphocytic infiltrate.