BANTAO Journal 2020; 18(2): 64-67

BJ
BANTAO Journal

*Case Report*

# Renal Lymphangiectasia: An Unusual Mimicker of Hydronephrosis – A Case Report

Pavlina Dzekova-Vidimliski, Vlatko Karanfilovski and Vesna Ristovska

University Hospital for Nephrology, Skopje, Republic of North Macedonia

**Abstract**

Renal lymphangiectasia is very rare benign lymphatic malformation of renal lymphatics, characterized by cystic dilatation of perirenal, peripelvic, or intrarenal lymphatic vessels. One of the rare mimickers of hydronephrosis and cystic renal lesions on the imaging findings. We report a case-patient with unilateral right renal lymphangiectasia with flank pain and arterial hypertension. The ultrasound examination revealed right kidney hydronephorosis. The renal lymphangiectasia was identified on contrast-enhanced computed tomography scan. The patient was managed conservatively with antihypertensive drugs. The report also contains a review of the literature on the pathophysiology of renal lymphangiectasia, clinical presentation, imaging findings, differentials, complications and treatment.

**Keywords:** renal, lymphatic system, lymphangiectasia, hydronephrosis, hypertension,

## Introduction

Renal lymphangiectasia (RLM) is a rare, benign, and developmental disorder of the renal lymphatic system characterized by cystic dilatation of perirenal, peripelvic, or intrarenal lymphatic vessels [1]. It is mostly asymptomatic or with nonspecific symptoms, unilateral or bilateral, and unrelated to age or gender [2]. The incidence of RLM accounts for approximately 1% of all lymphangiomas [3]. The identified cases with renal lymphangiectasia were with familial, developmental, and acquired causes for malformation of the renal lymphatic tissue leading to obstruction and accumulation of lymph [4]. The diagnosis of renal lymphangiectasia is usually done by imaging techniques, either ultrasound (US), computed tomography (CT) or magnetic resonance (MRI) [4,5]. It is commonly misdiagnosed with hydronephrosis, cystic renal lesions, lymphoma, nephroblastoma, and perirenal liposarcoma [1]. The evolution of renal lymphangiectasia is ambiguous, and both, spontaneous regression or rapid worsening were described in the literature [5].

## Case Report

A 30-year-old male presented with right flank pain, headache, and high blood pressure. The hiatal hernia, hydronephrosis of the right kidney, and well-controlled allergic asthma treated with an inhaled corticosteroid and oral antihistamines were conditions from his medical history. The hydronephrosis on the right kidney was diagnosed with an ultrasound examination before three years. Clinical examination of the patient showed increased blood pressure of 150/100 mmHg. All laboratory blood and urine analyzes were within normal limits. Ultrasound examination of the abdomen revealed increased right kidney with a dimension of 160 x 73.7 mm and parenchyma with granular appearance and thickness of 13.6 mm. Further US evaluation showed dilated anechoic pelvis and calyces of the right kidney, suspicious for a high grade of hydronephrosis. The left kidney was with normal size and parenchyma thickness of 19 mm. The patient underwent a contrast-enhanced CT scan of the abdomen. The CT scan demonstrated increased right kidney with multiple lobulated non-enhancing parenchymal and peripelvic fluid collections that were intimately associated with renal pelvis and calyces (Figure 1A). Fluid-filled lesions were also noted in the right perinephric space, surrounding the lower pole of the right kidney and adjacent to the abdominal aorta and inferior vena cava (Figure 1B). The delayed phase of the abdominal CT scan showed normal excretion of contrast with no evidence of leakage and dilatation of the pelvicalyceal system (Figure 2C and 2D). The CT angiography of renal vasculature was also performed because of suspicion for renovascular hypertension. CT angiography showed normal vascular anatomy of both kidneys, confirming the previous diagnosis of ectatic lymphatic vessels in peripelvic and retroperitoneal perivascular space. The unilateral right renal lymphangiectasia was diagnosed on a CT scan,

*Correspondence to:* Vidimliski-Dzekova Pavlina, University Hospital for Nephrology, "Mother Theresa" str 17, 1000 Skopje, R. N. Macedonia; Phone: +389 75 59 46 56; E-mail: pavlinadzekova@yahoo.com

and the other differential diagnoses were excluded. The patient was managed conservatively. The arterial hypertension was treated with a combination of an angiotensin-converting enzyme (ACE) inhibitor and a thiazide diuretic, with a good treatment response. No invasive interventions were considered necessary for further diagnosis or treatment of the patient. Regarding the reports for different clinical outcomes of RLM, the patient was advised for regular checkups to follow-up the course of the disease.

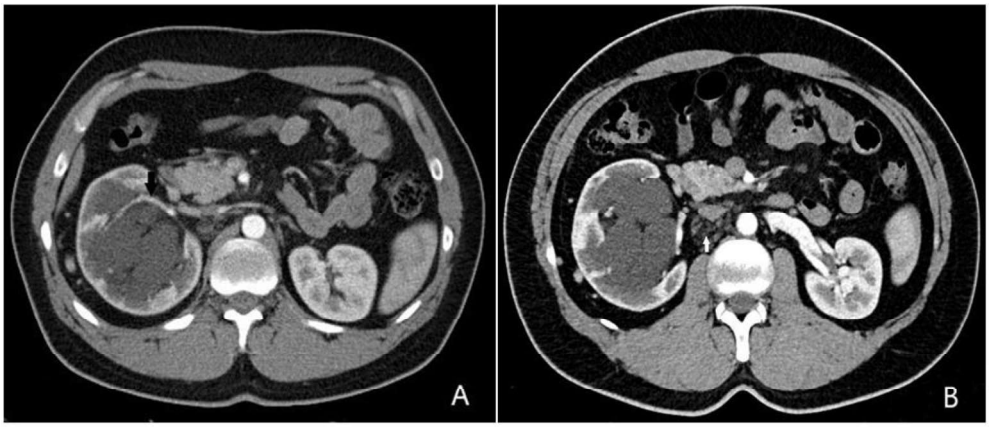


**Fig. 1.** (A) Axial contrast-enhanced CT scan at the level of right renal hilum showed thin renal parenchyma compressed by multiple lobulated non-enhancing intraparenchymal and peripelvic fluid collections with secondary displacement of hilar vessels (black arrow); (B) Retroperitoneal peri-aorto-caval cysts (white arrow).

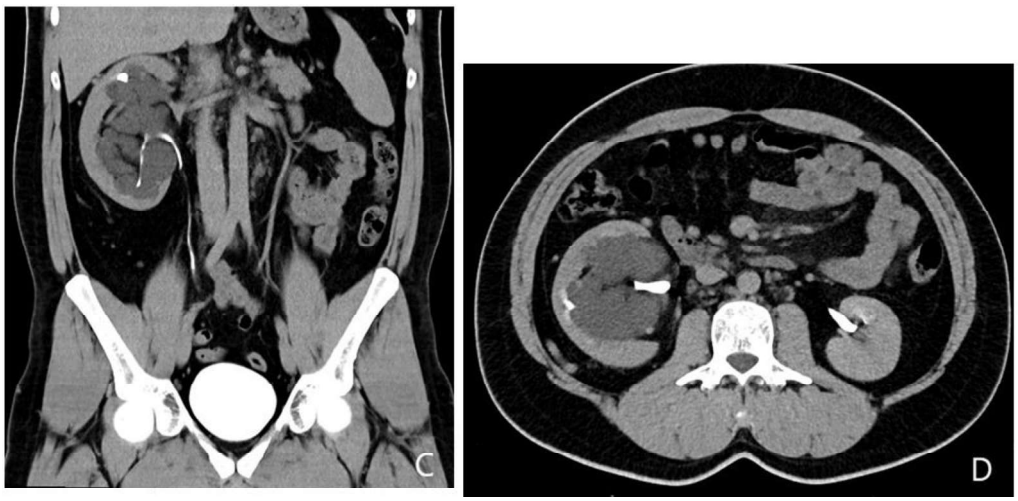


**Fig. 2.** Coronal (C) and axial (D) CT scan of the abdomen showed normal excretion of contrast and non-dilated excreting collecting system

## Discussion

**Pathophysiology:** the lymphatic system of the kidneys begins in the cortex with a intralobular lymphatics which connect to the larger arcuate and interlobar lymphatic vessels that drain into a hilar lymphatics in renal sinus and hilum [6]. Via larger lymphatic trunks, lymph from both kidneys drains into the paraaortic, pericaval and interaortocaval lymph nodes [6,7]. In renal lymphangiectasia, there is impairment in the drainage of larger renal sinus lymphatic trunks with resultant dilatation of peripelvic, perinephric and intrarenal lymphatics [7,8]. The accumulation of lymph in renal lymphatic vessels causes subsequent dilatation and formation of localized or generalized cystic masses [7]. This condition can be either congenital or acquired. Familial predilection is seen in very few reported cases. Meredith *et al*. supported familial predilection of the disease, describing exacerbation of renal lymphangiectasia during pregnancy in two sisters [9]. No family association was found in any of the eight patients with RLM presented in the case study of Pandya VK *et al*. [4]. Blockage of lymphatic vessels due to inflammation or other obstruction like neoplasm, may cause acquired renal lymphangiectasia [1].

The lesions are usually asymmetric, bilateral, and may involve renal sinus, renal parenchyma or perirenal regions [7,10]. In case of unilateral disease, the left kidney is more frequently affected [4]. Our case patient had unilateral right kidney lymphangiectasia with

peripelvic and peri-aorto-caval fluid collections with no familial association. Out of eight patients in a case study of Pandya VK *et al*., six (75%) had peripelvic lymphangiectasia, and only two (25%) had peri-nephric lymphangiectasia. Four (50%) patients had bilateral lesions and rest four (50%) patients had unilateral lesions [4]. The retroperitoneal involvement with formation of peri-vascular cysts is the rarest form of the disease [7].

**Clinical presentation:** more often it is an asymptomatic condition and it is diagnosed incidentally. However, it may be presented with symptoms like flank pain, hematuria, proteinuria, abdominal mass, ascites, lower extremity edema, and hypertension. [11]. A few cases have also been reported with renal insufficiency and renal vein thrombosis [12,13]. Schwarz A et al. reported that hypertension was present in 59% of the cases with peripelvic or perirenal cysts and it was reversible or markedly improved after drainage or resection of the cysts [14]. The same authors also noted that hypertension was associated with elevated levels of circulating renin and aldosterone, and plasma levels of these factors dropped significantly postoperatively. These findings strongly support the assumption of renin-dependent hypertension secondary to renal ischemia caused by parenchyma compression from surrounding cysts. The flank pain was the most common complaint of the patients in the case study of Pandya VK *et al*. [4]. Five patients out of eight patients (62.5%) presented with flank pain and only one patient had associated hypertension. Rarely, formation of a junction between cyst and peritoneum or pelvi-calyceal system can result with ascites or chyluria, respectively [10]. Less commonly reported symptoms are rupture of the cyst with hemorrhage and hematuria [7,8,13,14]. Flank pain and hypertension were the main symptoms in our case patient with peripelvic lymphangiectasia.

**Imaging techniques** are essential in the diagnosis and further management of this condition. The dilated lymphatics appear as cystic lesions in the perirenal, peripelvic, and intrarenal locations. Ultrasound frequently reveals anechoic, multi-spectated, sharply defined cysts with thin walls located in peripelvic and/or perirenal regions [2,8,15]. Sometimes, cystic lesions are seen in the renal parenchyma, extending from there into the renal sinus and it can appear as a focal hyperechoic lesion in the renal cortex. US examination could misdiagnose this condition with cystic renal lesions, hydronephrosis, urinoma, or other renal cystic masses like Wilm's tumor or lymphoma [4,7]. Our case-patient was misdiagnosed with right kidney hydronephrosis with the US examination. Almost all case-patients (6 out of 8 patients) from the study of Pandya VK *et al.* were misdiagnosed with hydronephrosis with the US examination. One of the eight patients was misdiagnosed with polycystic kidney disease, and the other one was misdiagnosed with urinoma on ultrasound [4].

Far better assessment of this condition could be achieved by computed tomography (CT) scan and magnetic resonance imaging (MRI). On CT scan, renal lymphangiectasia is presented as well-delineated, multiseptated, non-enhancing fluid collections in perirenal or peripelvic regions, which could compress the kidney parenchyma and distort the calyceal system [2,8,15]. Less commonly, as in our case patient, there are dilated retroperitoneal lymphatics around the aorta and inferior vena cava with similar CT characteristics [2,4]. CT examination reveals cystic lesions showing fluid attenuation in the renal sinus (peripelvic or perirenal location) with or without septations. In contrast-enhanced CT, there is no opacification of cystic lesions on delayed scans in the excretory phase, which is an important feature that differentiates RLM from the dilated pelvicalyceal system [15,16]. Administration of iodinated contrast agents should be avoided in patients with impaired kidney function. In those cases, MRI with excretory urography could be used as an alternative imaging technique for the diagnosis of renal lymphangiectasia. The cystic lesion appears hypointense on T1-weighted images and hyperintense on T2-weighted images. In contrast-enhanced T1-weighted images, there is no enhancement in the early phases. There is no opacification of cystic lesions in the post-contrast T1-weighted MR excretory urography images [17]. Lymphoscintigraphy could be also used for the detection of renal lymphangiectasia [2]. In uncertain cases, percutaneous fluid aspiration with a subsequent cytological evaluation of the sample could confirm the diagnosis [4,15].

**Differential:** most common differential diagnoses are hydronephrosis and polycystic kidney disease [10,11, 17]. In polycystic kidneys, there are massively enlarged kidneys with multiple welldefined cysts of varying sizes in the cortex, replacing the normal renal parenchyma. There are no cysts in the renal cortex in renal lymphangiectasia, only enlarged kidney with raised renal cortical echoes and loss of corticomedullary differenttiation. The pelvicalyceal system appears normal in renal lymphangiectasia, in contrast to hydronephrosis.

**Evolution and complications:** lymphangiectasia may show sudden appearance and rapid growth or cessation of growth and spontaneous regression of symptoms [13]. Pickering SP *et al.* reported partial regression of this condition in neonatal patient, 13 months after initial diagnosis [18]. Battaglia M *et al*. observed no progression of the lesions in 10 patients with peripelvic multicystic lymphangiectasia during 8-years of follow-up [19]. In case described by Liorente JG *et al*., perirenal collections completely resolved after 6 years, but intrarenal lesions had progressive course and resulted with increasing nephromegaly [5]. The complications of RLM include renal vein thrombosis, renin-dependent arterial hypertension, obstructive uropathy features due to compression of the collecting system

by the larger cysts, intracystic hemorrhage, ascites, and superimposed infection [7,9,17,20].

**Treatment:** it is a benign entity. Asymptomatic cases usually receive conservative management, but due to potential complications, periodic follow-up is necessary [1,4,7]. Hypertension and secondary infections of the lesions are managed conservatively with antihypertensive drugs and antibiotics [4]. Ultrasound-guided percutaneous aspiration of the fluid is reserved for symptomatic patients, presenting with pain on the account of compression by the collection [12]. However, the success rate of percutaneous aspiration was very low in multi-septated larger lesions and led to a high rate of recurrences. Laparoscopic ablation and nephrectomy is reserved for complicated cases with severe renal disease, renal vein thrombosis, and cases with multiple recurrences. However, nephrectomy is not considered as a choice treatment in the case of bilateral renal involvement, because the cyst formations in the contralateral kidney may increase in size [7].

## Conclusion

Renal lymphangiectasia is benign disorder that should be differentiated from other causes with intrarenal or perinephric cystic masses. Radiological modalities are important for early and proper diagnosis, determining extension, and further management of the disease. During the disease, the size of dilated lymphatics may remain unchanged or may increase causing renin-dependent hypertension, ascites, renal vein thrombosis, or nephromegaly. Therefore the patients with this condition need regular follow-up with guided treatment by the extensity and evolution of the disease.

*Conflict of interest statement.* None declared.

## References

1. Ramseyer LT. Case 34: renal lymphangiectasia. *Radiology* 2001; 219(2): 442-444.
2. Elbanna KY, Almutairi BM, Zidan AT. Bilateral renal lymphangiectasia: radiological findings by ultrasound, computed tomography, and magnetic resonance imaging. J Clin Imaging Sci 2015; 30: 5-6.
3. Hauser H, Mischinger HJ, Beham A, *et al*. Cystic retroperitoneal lymphangiomas in adults. *Eur J Surg Oncol* 1997; 23: 322-326.
4. Pandya VK, Sutariya HC, Gandhi SP, *et al*. Role of CT scan in diagnosis of renal lymphangiectasia: our single-center experience. *Ren Fail* 2017; 39(1): 533-539.
5. Llorente JG, Garcia AD, Sacristan JS, Chicharro GN. Renal lymphangiectasia: radiologic diagnosis and evolution. *Abdom Imaging* 2002; 27(6): 637-639.
6. Russell PS, Hong J, Windsor JA, *et al*. Renal Lymphatics: Anatomy, Physiology, and Clinical Implications. *Front Physiol* 2019; 10: 251.
7. Upreti L, Dev A, Kumar Puri S. Imaging in renal lymphangiectasia: report of two cases and review of literature. *Clin Radiol* 2008; 63(9): 1057-1062.
8. Gorantla R, Yalapati A, Dev B, Joseph S. Case report: Perinephric lymphangiomatosis. Indian *J Radiol Imaging* 2010; 20(3): 224-226.
9. Meredith WT, Levine E, Ahlstrom NG, *et al*. Exacerbation of familial renal lymphangiomatosis during pregnancy. *AJR Am J Roentgenol* 1988; 151: 965-966.
10. Leite AF, Venturieri B, de Araujo RG, *et al*. Renal lymphangiectasia: know it in order to diagnose it. *Radiol Bras* 2016; 49(6): 408-409.
11. Blanc M, Schmutz G, Belzile F, Sabbagh R. Renal lymphangiectasia presenting with hypertension and polycythemia. *Can Urol Assoc J* 2014; 8: 160-166.
12. Cadnapaphornchai MA, Ford DM, Tyson RW, *et al*. Cystic renal lymphangiectasia presenting as renal insufficiency in childhood. *Pediatr Nephrol* 2000; 15: 129-131.
13. Riehl J, Schmitt H, Schafer L, *et al*. Retroperitoneal lymphangiectasia associated with bilateral renal vein thrombosis. *Nephrol Dial Transplant* 1997; 12(8): 1701-1703.
14. Schwarz A, Lenz T, Klaen R, *et al*. Hygroma renale: pararenal lymphatic cysts associated with renin-dependent hypertension (Page kidney). Case report on bilateral cysts and successful therapy by marsupialization. *J Urol* 1993; 150(3): 953-957.
15. Murray KK, McLellan GL. Renal peripelvic lymphangiectasia: appearance at CT. *Radiology* 1991; 180(2): 455-456.
16. Pandya VK, Shah MK, Gandhi SP, Patel HV. Bilateral Renal Lymphangiectasia. *J Clin Diagn Res* 2016; 10(9): TD01-TD02.
17. Kumar K, Khan S, Tanveer S, Khan AS. Case Report: Renal Lyphangectesia with Nephritic Syndrome. *J Nephrol Ther* 2018; 8: 318.
18. Pickering SP, Fletcher BD, Bryan PJ, Abramowsky CR. Renal lymphangioma: a cause of neonatal nephromegaly. *Pediatr Radiol* 1984; 14: 445-448.
19. Battaglia M, Ditonno P, Mancini V, *et al*. Long-term follow- up of peripelvic renal multicystic lymphangiectasia. *Arch Ital Urol Androl* 2002; 74(4): 200-205.
20. Wadhwa P, Kumar A, Sharma S, *et al*. Renal lymphangiomatosis: imaging and management of a rare renal anomaly. *Int Urol Nephrol* 2007; 39(2): 365-368.