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Case Report

Development of unilateral renal peripelvic lymphangiectasia after renal vein thrombosis ☆

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ABSTRACT

Renal peripelvic lymphangiectasia is a rare entity that can mimic hydronephrosis on routine contrast-enhanced computer tomography (CT). While it may remain asymptomatic, symptomatic cases can exhibit refractory hypertension (HTN) and recurrent abdominal pain. Diagnostic challenges stem from its nonspecific symptoms and imaging characteristics, which can overlap with other renal disorders. Thereby, adequate protocolling of CT or magnetic resonance (MR) imaging is important for accurate diagnosis. In this report, we present a case of renal lymphangiectasia that developed in a medically complex patient following renal vein thrombosis.

Introduction

Renal lymphangiectasia (RLT) is a rare benign retroperitoneal lymphatic disorder characterized by ectasia of the perirenal and peripelvic lymphatic channels [1]. Given its rarity, there is a scarcity of literature describing this condition. The existing reports emphasize the pivotal role of radiologists and their expertise in accurately reporting findings and aiding in the diagnosis of RLT, as it is frequently encountered incidentally and thus misdiagnosed. In this context, we present a case of unilateral RLT developed after subsequent episodes of renal vein thrombosis in a patient with a complicated medical history encompassing renal and autoimmune manifestations, initially misdiagnosed as hydronephrosis at an outside hospital.

Case report

A 37-year-old female presented to the emergency department complaining of severe bilateral flank pain for the past 2 weeks. Her past medical history was significant for systemic lupus

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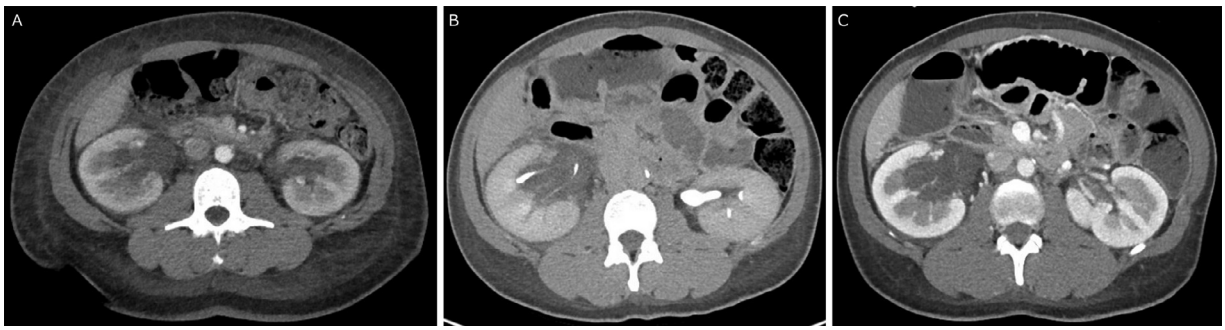

Fig. 1 – (A) Axial view of venous phase contrast-enhanced CT obtained approximately 4 months after initial episode of renal vein thrombosis; Imaging reveals expanded fluid density structure throughout the renal hilum mimicking hydronephrosis. (B) Excretory (delayed) phase showcasing enlarged fluid density structure encasing the calyces, with excreted contrast in normal caliber calyces; images were collected a month prior to current admission. (C) Venous phase contrast-enhanced CT obtained during the current admission, displaying no evident interval changes from delayed phase collected a month prior.

erythematosus (SLE), hypertension (HTN), and membranous nephropathy (MN) complicated by left and right renal vein thromboses respectively 9 and 5 months prior to presentation.

Upon presentation, the patient was hemodynamically stable with marked HTN (167/85). The physical examination revealed tenderness in the left costovertebral angle but was otherwise unremarkable. In light of the patient's worsening flank pain and complex rheumatologic and nephrologic history, a decision was made to admit the patient for comprehensive evaluation and management.

During the patient's hospitalization, an extensive laboratory workup was conducted. Noteworthy laboratory findings included a decreased creatinine of 0.48 mg/dL (normal range:0.6-1.3 mg/dL), proteinuria, and otherwise unremarkable renal function. Surprisingly, a comprehensive autoimmune panel encompassing antidouble stranded DNA (anti-dsDNA), antineutrophil cytoplasmic antibody (ANCA), antihistone, antiphospholipid panel, as well as inflammatory markers (CRP and ESR) and complement, was unrevealing.

Although right hydronephrosis was diagnosed on a single-venous-phase contrast-enhanced computed tomography (CT) scan at an external institution (Fig. 1A), a multiphasic contrast-enhanced CT scan performed near the time of her admission showed a fluid density structure encasing the calyces on excretory (delayed) phase, rather than true hydronephrosis (Fig. 1B). A CT conducted during the current admission showed no interval changes compared to her imaging 1 month prior (Fig. 1C). Remote prior imaging before the renal vein thromboses showed normal kidneys.

Nuclear medicine mercaptoacetyltriglycine-3 (MAG -3) renal function study (Fig. 2) was performed and demonstrated symmetric normal flow and function, confirming no evidence of obstruction or hydronephrosis thus concordant with the multiphasic CT findings.

Magnetic resonance imaging (MRI) was performed to further characterize the right renal peripelvic process, showing fluid signal in the peripelvic and perinephric structure with minimal delayed enhancement on the 10-minute post-contrast images (Fig. 3). It is worth noting that imaging at an outside hospital had raised concerns regarding retroperitoneal fibrosis because of the perceived hydronephrosis and retroperitoneal soft tissue on single phase CT, however, multiphasic CT and MR showed these findings to be right perinephric, peripelvic, and perihilar lymphangiectasia with cavernous transformation of the right renal vein.

The patient was eventually discharged and advised to follow up with both her nephrologist and rheumatologist.

Discussion

Renal Lymphangiectasia (RLT), also referred to as renal lymphangiomatosis, represents a distinct subset of retroperitoneal lymphangiomas characterized by ectasia of perirenal, peripelvic, and in some cases intrarenal lymphatic channels [1,2]. RLT does not exhibit a gender predilection and affects all ages equivalently [2].

By nature of its rarity, most of the documented information on RLT derives from isolated case reports, posing challenges in understanding its pathophysiology and clinical course. The renal lymphatic drainage ultimately converges into retroperitoneal lymph nodes situated in the para-aortic, para-caval, and inter-aorto-caval lymph nodes [3]. Although no consensus exists on the underlaying mechanism leading to lymphatic ectasia, it is presumed that in adults an acquired etiology is more prevalent. Various reports have associated renal vein thrombosis with the development of RLT [4–6]. One report documented the case of a young male that developed bilateral renal vein thrombosis secondary to antiphospholipid syndrome [4]. Subsequent longitudinal monitoring spanning 14 years revealed the progressive development of RLT in this patient. Another case reported a middle-aged male with a history of chronic renal vein thrombosis with multiple perirenal collaterals that was incidentally noted to have RLT after an evaluation for abdominal pain [7]. Some authors have proposed that an elevation in starling hydrostatic pressure within the renal capillaries owing to thromboembolic events in the renal vessels augments trans-endothelial filtration, resulting in increased lymphatic drainage and the formation of ecstatic channels [4].

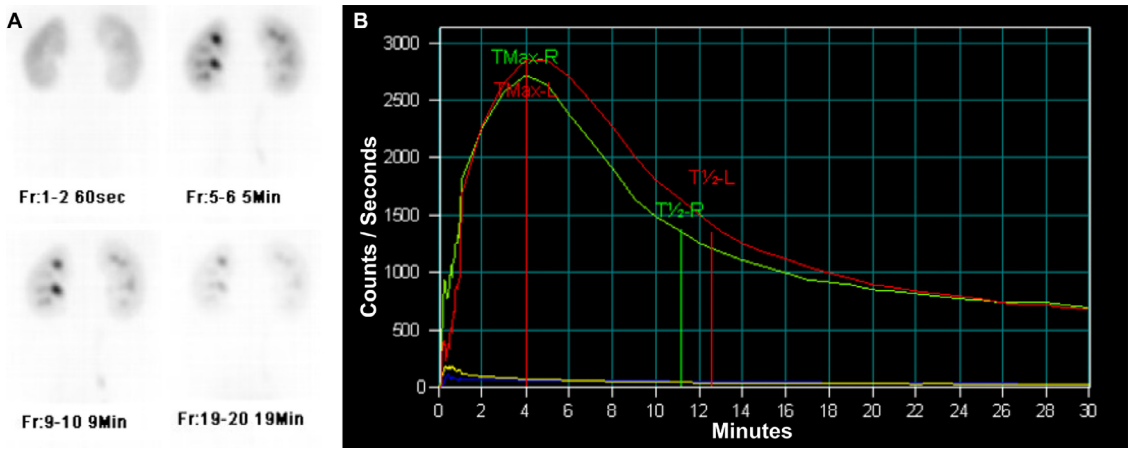

Fig. 2 – (A and B) MAG-3 renal function study demonstrating symmetric normal flow and function. On the renogram curve, red color represents the left kidney and green color represents the right kidney.

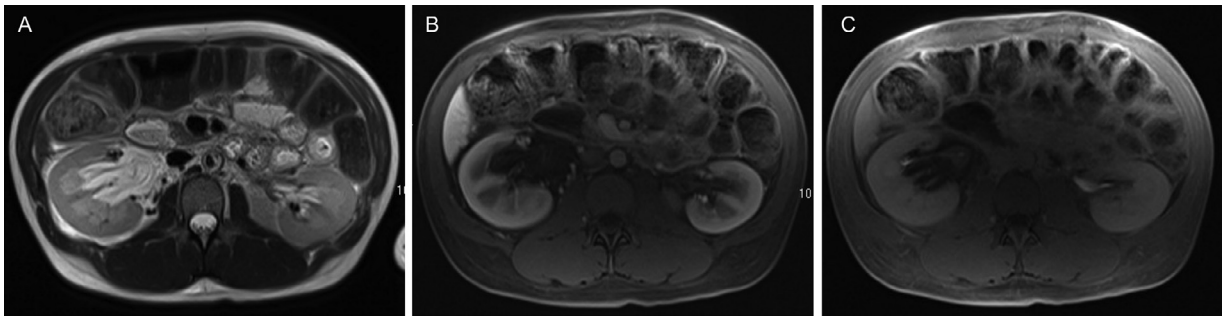

Fig. 3 – (A) Axial T2-weigthed MRI revealing a right kidney peripelvic fluid signal structure encasing the calyces, consistent with peripelvic lymphangiectasia. There is also a small perinephric fluid signal structure along the renal capsule. (B) Postcontrast T1-weighted fat-saturated images show no arterial enhancement of the peripelvic lymphangiectasia. (C) Postcontrast 10-minute delayed phase shows excreted contrast in the calyces and minimal enhancement of the peripelvic lymphangiectasia.

RLT is typically diagnosed incidentally, as most cases are asymptomatic [8]. Nevertheless, uncomplicated RLT can present with bilateral or unilateral abdominal pain, abdominal distention, hematuria, and proteinuria, among others [9,10]. Surprisingly, renal function is largely preserved in most cases [11]. Several reports have documented instances of complicated RLT encompassing refractory HTN, pleural effusion, ascites, rupture after abdominal blunt trauma, and renal insufficiency and anemia [8,12–14]. It is noteworthy that certain reports identify renal vein thrombosis as a complication and a potential etiology [4–6].

Accurate diagnosis of RLT heavily relies on the expertise of radiologists, given that cases are frequently detected incidentally and can be easily confused with other cystic etiologies [7,15]. In the vast majority of cases ultrasonography (US), contrast computed tomography (CT) and magnetic resonance imaging (MRI) are employed to characterize these lesions. US commonly shows anechoic perinephric or parapelvic fluid collection, encompassed by well-defined thin cystic walls that may exhibit multiloculated or unilocular configurations, with cystic spaces insinuating into the renal sinus [9,16]. Contrast-enhanced CT demonstrates cystic lesions with fluid attenuation (0-10 HU) in the parapelvic or perinephric region with or without septations [7,15]. On MRI, the cystic lesions appear hyperintense on T2-weighted sequences and hypointense on T1-weighted images [16]. In some cases, retroperitoneal thin dilated lymphatic channels can be visualized [7]. Although radiological diagnosis is a mainstay, percutaneous aspiration of the renal lymphatic fluid can confirm a diagnosis by showing a predominant lymphocytic infiltrate with high relative levels of renin and low levels of fat and protein. These features can aid clinicians in distinguishing typical lymphatic fluid (lacks renin and lymphocytes) from RLT-derived fluid [11,13,17]. Histopathological tissue analysis can also be used for diagnostic purposes, but it is rarely employed [1].

The differential diagnosis of RLT encompasses several conditions, including hydronephrosis, cystic diseases, perinephric urinoma, nephroblastomatosis, and lymphomas

[7]. Hydronephrosis and polycystic kidney disease are the 2 most commonly misdiagnosed conditions [10,18]. In adults, hydronephrosis, characterized by luminal dilation of the collecting system, is a common misdiagnosis for RLT on nonenhanced or early-phase-contrast-enhanced CT. A critical CT sign for distinguishing between these 2 entities is the absence of enhancement during the excretory (delayed) phase, as RLT lacks communication with the collecting system [19]. Because renal function is preserved in RLT, there is normal excretion of contrast into the collecting system, interdigitating between the lymphangiectasia. Conversely, hydronephrosis often fills with excreted contrast in the excretory phase [20].

Conservative management is typically used for asymptomatic patients [16]. However, patients with abdominal pain and compressive symptoms may be treated with percutaneous aspiration, although the presence of multiseptated lesions can lead to recurrence [7,17]. Laparoscopic ablation, marsupialization, and nephrectomy are reserved for recurrent cases or complex presentations, such as refractory hypertension or kidney failure [7,10,17]. Overall, the management approach is highly influenced by the clinical presentation and the likelihood of recurrence.

To return to our patient, we present a case of a 37-year-old female with a history of refractory hypertension (HTN), systemic lupus erythematous (SLE), and membranous nephropathy (MN) complicated by bilateral renal vein thrombosis, who subsequently developed unilateral renal lymphangiectasia (RLT). To the best of our knowledge, no previous association exists between RLT and MN or SLE. MN, the leading cause of nephrotic syndrome in adults, is strongly linked to SLE and renal vein thrombosis [21]. While the literature on RLT is scant, a few case reports have described an association with renal vein thrombosis [4–6]. However, given the intricate profile of our patient, it remains uncertain whether an autoimmune factor played a role in the accelerated development of RLT. Moreover, the underlying cause of refractory hypertension in our patient remains ambiguous considering his autoimmune profile and confounding RLT. Furthermore, our patient was misdiagnosed with hydronephrosis and retroperitoneal fibrosis at an outside hospital, but appropriately protocoled CT and MRI ruled out these diagnoses.

Conclusion

Renal lymphangiectasia is an elusive condition with uncertain etiology susceptible to misdiagnosis. Some reports have speculated an association with renal vein thrombosis similar to our case, although this connection lacks concrete evidence. Although a significant number of patients remain asymptomatic, a subset may experience recurring abdominal pain. Medically complex patients with multiple comorbidities can pose a challenge to accurately diagnose, as RLT is predominantly detected incidentally. Delayed diagnosis can adversely impact patient care and result in unnecessary interventions. Radiologists play a critical role in the diagnosis of this condition through appropriate CT and MR protocol creation, and by remaining vigilant to this uncommon condition which can mimic hydronephrosis and other renal cystic entities.

Patient consent

I hereby confirm that patient reported in the manuscript signed the informed consent/authorization for participation in research which includes the permission to use data collected in future research projects including presented case details and images used in this manuscript. No protected health information is shown in this case report.

REFERENCES

[1] Rajasekaran S, Parkhi M, Kanojia R, Sekar A, Nada R. Renal lymphangiectasia. Autop Case Rep 2022;12. doi:10.4322/ACR.2021.399/PDF/AUTOPSY-12-E2021399.PDF.

[2] Hauser H, Mischinger HJ, Beham A, Berger A, Cerwenka H, Razmara J, et al. Cystic retroperitoneal lymphangiomas in adults. Eur J Surg Oncol (EJSO) 1997;23(4):322–6. doi:10.1016/S0748-7983(97)90777-0.

[3] Russell PS, Hong J, Windsor JA, Itkin M, Phillips ARJ. Renal lymphatics: anatomy, physiology, and clinical implications. Front Physiol 2019;10 MAR. doi:10.3389/FPHYS.2019.00251.

[4] Villard N, Meuwly JY, Righini M, Alberti N, Habre C. Renal peripelvic lymphangiectasia after bilateral renal vein thrombosis. BMJ Case Rep CP 2022;15(7):e245666. doi:10.1136/BCR-2021-245666.

[5] Riehl J, Schmitt H, Schäfer L, Schneider B, Sieberth HG. Retroperitoneal lymphangiectasia associated with bilateral renal vein thrombosis. Nephrol Dial Transplant 1997;12(8):1701–3. doi:10.1093/NDT/12.8.1701.

[6] Chang MS, Yang DM, Yoon MH, Kim HS, Jin W, Kim HS, et al. Peripelvic and perirenal lymphangiectasia and bilateral renal vein thrombosis: a case report. J Korean Radiol Soc 2000;43(1):91–4. doi:10.3348/JKRS.2000.43.1.91.

[7] Umapathy S, Alavandar E, Renganathan R, S T, Kasi Arunachalam V. Renal lymphangiectasia: an unusual mimicker of cystic renal disease - a case series and literature review. Cureus 2020;12(10). doi:10.7759/CUREUS.10849.

[8] Bouardi N EL, Haloua M, Alami B, Youssef AL, Boubbou M, Maaroufi M. A ruptured renal lymphangiectasia in the retroperitoneum: a rare complication of a rare condition. Radiol Case Rep 2022;18(1):271–4. doi:10.1016/J.RADCR.2022.10.029.

[9] Davidson AJ, Hartman DS. Lymphangioma of the retroperitoneum: CT and sonographic characteristic. Radiology 1990;175(2):507–10. doi:10.1148/RADIOLOGY.175.2.2183287.

[10] Li Y, Wang Q, Kan G, Gong H, Zhang H, Tao X, et al. Renal lymphangiomatosis: literature analysis on research progress and presentation of four cases. Quantitat Imag Med Surg 2023;13(1):518–28. doi:10.21037/QIMS-22-314/COIF.

[11] Uzzo RN, Bloom E, Peters A, Parab M, Masic S, Kutikov A. Refractory hypertension due to unilateral renal lymphangiectasia: an uncommon case with a surgical solution. Urol Case Rep 2020;32:101177. doi:10.1016/J.EUCR.2020.101177.

[12] Pulaski JE. Perirenal lymphangioma causing hypertension. Ann Int Med 1950;33(1):234–43. doi:10.7326/0003-4819-33-1-234.

[13] SAA Al-Dofri. Renal lymphangiectasia presented by pleural effusion and ascites. Radiol Case Rep 2009;3(10):5–10. doi:10.3941/JRCR.V3I10.317.
[14] Cadnapaphornchai MA, Ford DM, Tyson RW, Lum GM. Cystic renal lymphangiectasia presenting as renal insufficiency in childhood. Pediatr Nephrol 2000;15(1-2):129–31. doi:10.1007/S004670000413/METRICS.
[15] Kashgari AA, Ozair N, Al Zahrani A, Al Otibi MO, Al Fakeeh K. Renal lymphangiomatosis, a rare differential diagnosis for autosomal recessive polycystic kidney disease in pediatric patients. Radiol Case Rep 2017;12(1):70–2. doi:10.1016/J.RADCR.2016.11.016.
[16] Gupta R, Sharma R, Gamanagatti S, Dogra PN, Kumar A. Unilateral renal lymphangiectasia: imaging appearance on sonography, CT and MRI. Int Urol Nephrol 2007;39(2):361–4. doi:10.1007/S11255-006-9039-Z.
[17] Schwarz A, Lenz T, Klaen R, Offermann G, Fiedler U, Nussberger J. Hygroma renal: 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):953–7. doi:10.1016/S0022-5347(17)35660-4.
[18] Sarikaya B, Akturk Y, Bekar U, Topaloglu S. Bilateral renal lymphangiomatosis mimicking hydronephrosis: multidetector CT urographic findings. Abdom Imaging 2006;31(6):732–4. doi:10.1007/S00261-005-8014-Y.
[19] Chua N, Wolfe K, Mehta S, Lodge RN, Liyanage SH. Triparametric ultrasound in differentiating multicystic renal masses: a rare presentation of unilateral focal renal lymphangioma. Radiol Case Rep 2017;12(4):731–7. doi:10.1016/J.RADCR.2017.07.005.
[20] Robins SA, Fischmann J. Hydronephrosis; a radiologic classification based on anatomical variations. Radiology 1948;50(5):632–8. doi:10.1148/50.5.632.
[21] Ronco P, Beck L, Debiec H, Fervenza FC, Hou FF, Jha V, et al. Membranous nephropathy. Nat Rev Dis Prim 2021;7(1):69. doi:10.1038/s41572-021-00303-z.