Radiological Sciences Proceedings

UCLA Health

Renal Lymphangiectasia: A Case Report

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UCLA Radiol Sci Proc. 2025;5(4):47-51

Abstract: Renal lymphangiectasia is a very rare disorder that results from developmental malformations affecting the renal lymphatic system. Patients of any age may present with a range of symptoms, such as abdominal pain, flank pain, a palpable mass, and hypertension. Radiologic features include lymphatic cysts forming a rind-like appearance along the outer edge of the renal cortex. We report a case of renal lymphangiectasia in a 49-year-old man presenting with headache and back pain, highlighting the characteristic imaging features and key diagnostic considerations of this rare condition.

Keywords: *abdominal imaging, renal lymphangiectasia, magnetic resonance imaging, ultrasound, computed tomography*

Introduction

Renal lymphangiectasia—also referred to as renal lymphangiomatosis, parapelvic lymphangiectasia, or hygroma renale—is a rare disorder believed to originate from developmental malformations in the renal lymphatic system. The condition has been documented in a total of 104 well-characterized cases globally.[1] It accounts for approximately 1% of all detected lymphangiomas and has been associated with somatic mutations in the *PIK3CA* gene but may also result from inherited mutations in genes involved in lymphatic development.[2]

Renal lymphangiectasia presents in both pediatric and adult populations. It is most often a congenital condition caused by a developmental malformation of the renal lymphatic system.[3] However, previously reported cases have also suggested hormonal influences during pregnancy and immune-related mechanisms triggered in transplant patients.[4] This case report is presented to educate radiologists and trainees about the imaging and clinical findings in a patient with renal lymphangiectasia and was prepared following the CARE guidelines.[5]

Key Points

- Renal lymphangiectasia's clinical and radiologic presentations closely resemble those of several other conditions.
- A diagnosis of renal lymphangiectasia can only be confirmed with pathologic evidence, but clinicians typically rely on imaging due to the risks associated with tissue sampling.
- On computed tomography, magnetic resonance, and ultrasound images, renal lymphangiectasia appears as collections of cysts and fluid in the perinephric and parapelvic spaces.

Abbreviations

ADPKD: autosomal dominant polycystic kidney disease
ALP: alkaline phosphatase
ALT: alanine aminotransferase
AST: aspartate aminotransferase
CT: computed tomography
MRI: magnetic resonance imaging
SSFSE: Single-shot fast spin echo

Case Presentation

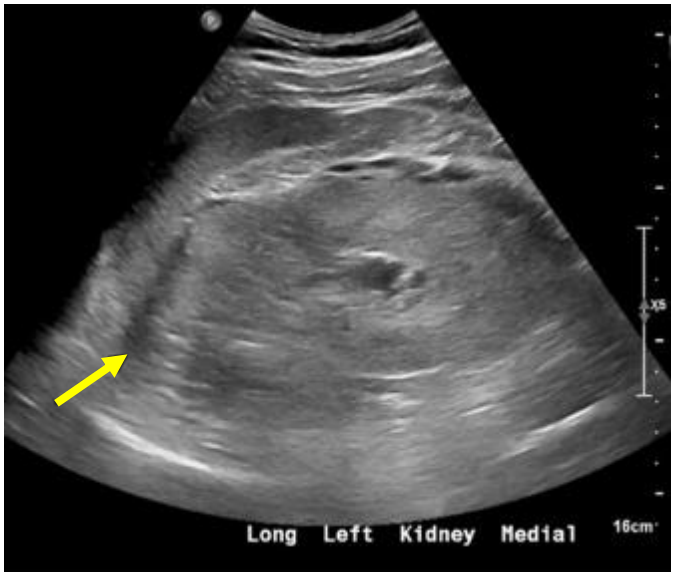
A 49-year-old man with a history of hyperlipidemia and medically refractory hypertension presented to a primary care clinic with headaches and back pain. Laboratory tests ordered by his physician revealed that his alanine aminotransferase (ALT) level was outside of the reference range. His liver enzyme values included an ALT level of 115 U/L (reference range, 8–70 U/L) (to convert U/L to μkat/L, multiply by 0.0167), an aspartate aminotransferase (AST) level of 57 U/L (reference range, 13–62 U/L), and an alkaline phosphatase level of 91 U/L (reference range, 37–113 U/L).

These laboratory results prompted the physician to order an abdominal ultrasound (Figure 1), which revealed hepatomegaly with hepatic steatosis, as well as increased left renal echogenicity suggestive of chronic renal disease with mild left hydronephrosis or parapelvic cysts. A subsequent computed tomography (CT) urogram revealed a rind-like, intermediate density infiltration surrounding the left kidney with equivocal enhancement, as well as several parapelvic left renal cysts without hydronephrosis (Figure 2). Magnetic resonance imaging (MRI) of the abdomen showed a thin, rind-like complex cystic process surrounding the left kidney (Figure 3), which was believed to be benign due to its stability compared to a lumbar MRI conducted 4 years prior to evaluate the patient's chronic low back pain.

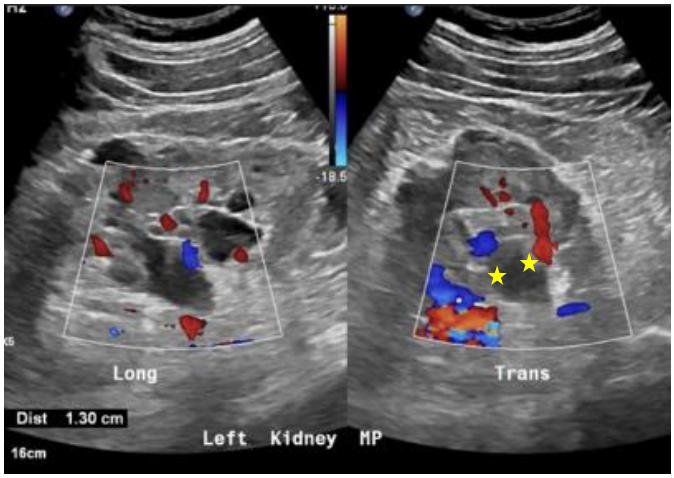
The primary consideration following imaging workup was renal lymphangiectasia, with other potential diagnoses, including perinephric hematoma, Erdheim-Chester disease, IgG4 disease, or lymphoproliferative disease, considered less likely. The patient was referred to a urologist, who agreed with the suggested diagnosis of renal lymphangiectasia and suggested a short-term repeat MRI to confirm the stability of the findings over time.

Figure 1. Doppler Ultrasound Images of the Left Kidney of a 49-Year-Old Man with Renal Lymphangiectasia.

A Longitudinal grayscale



B Longitudinal and transverse color



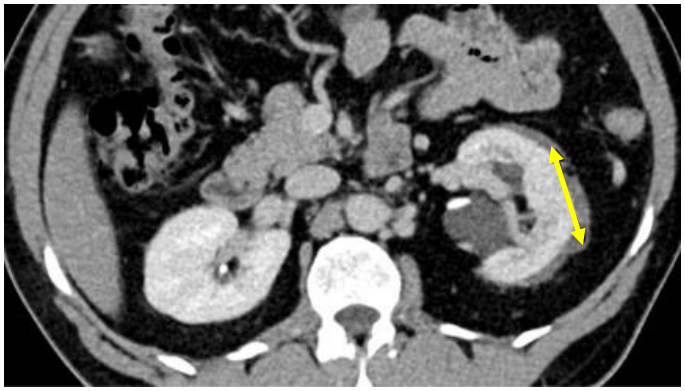
(A) Longitudinal grayscale and (B) longitudinal and transverse color Doppler images of the left kidney show a perinephric rind of anechoic cystic spaces (A, arrow) and parapelvic cysts (B, stars).

Discussion

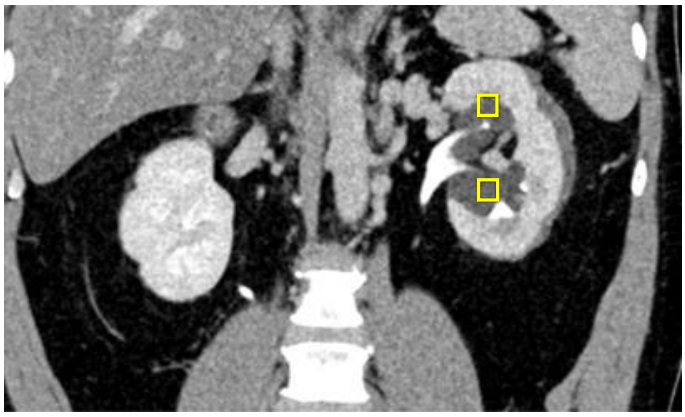
Renal lymphangiectasia has a variety of clinical presentations. In a large portion of cases, patients are asymptomatic, and the condition is detected incidentally.[3,6] However, some patients may present with symptoms such as flank or back pain, along with associated findings including hypertension, ascites, a palpable abdominal mass, and edema.[2,7] In pediatric populations, a palpable mass is more common, and adult populations more often report flank pain.[6] Patients with severe renal lymphangiectasia may present with elevated creatinine levels, hematuria, and renal vein thrombosis, in which case immediate

Figure 2. Excretory-Phase CT Urogram of a 49-Year-Old Man with Renal Lymphangiectasia.

A Axial view



B Coronal view



(A) Axial and (B) coronal excretory phase CT urogram images demonstrate a low-attenuation perinephric rind of tissue (A, arrow) and numerous parapelvic cysts (B, squares) in the left kidney. No hydronephrosis is present.

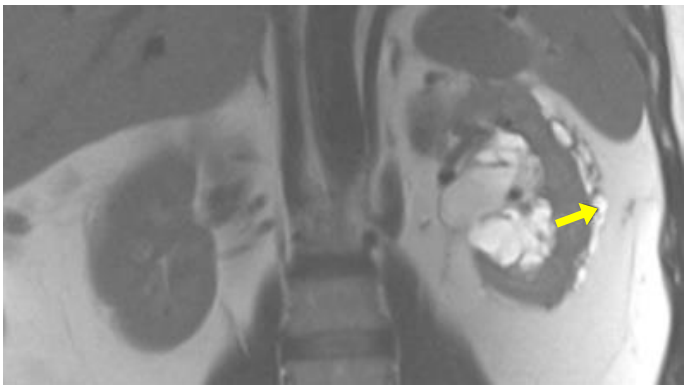
nephroprotective treatment is required to address impairment of renal function.[2,6,7]

Renal lymphangiectasia is most often a congenital condition caused by a developmental malformation of the renal lymphatic system that impairs lymphatic drainage. However, renal lymphangiectasia may also develop due to secondary lymphatic obstruction caused by infection, inflammation, or malignant infiltration.[3,6,7] This dysfunction in drainage results in lymphatic vessel dilation and fluid accumulation in the kidney,[3,6] particularly in the perirenal, parapelvic, and intrarenal spaces.[2,7,8] Progressive fluid accumulation leads to cystic expansion that compresses the renal parenchyma, which can potentially result in renal insufficiency or failure in severe cases.[2,8] Compression of the renal structures may also induce the activation of the renin-angiotensin system, which contributes to hypertension.[2,8]

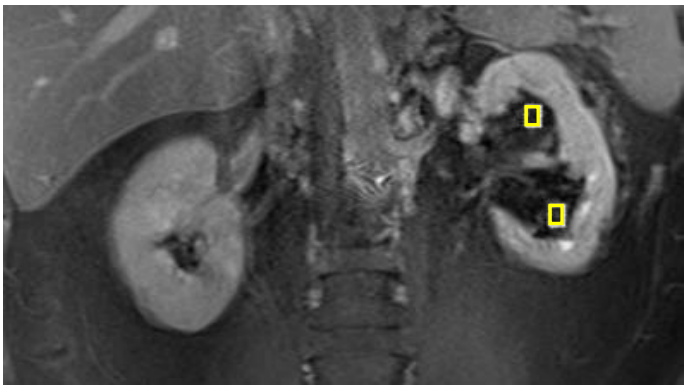
Characteristic imaging features of renal lymphangiectasia on CT, MRI, and ultrasonography are the presence of renal cystic and fluid collections, characteristically in the perinephric and parapelvic spaces. The cysts may demonstrate equivocal enhancement or septations and may extend to the renal hilum and paraaortic space. Additionally, retroperitoneal fluid collections have been reported, which represent dilated lymphatic vessels. Renal cortical thinning may be present in the affected kidney. This process can be seen unilaterally or bilaterally.

Figure 3. Coronal View T2-Weighted Magnetic Resonance Images of the Abdomen of a 49-Year-Old Man with Renal Lymphangiectasia.

A Single-shot fast spin echo (SSFSE)

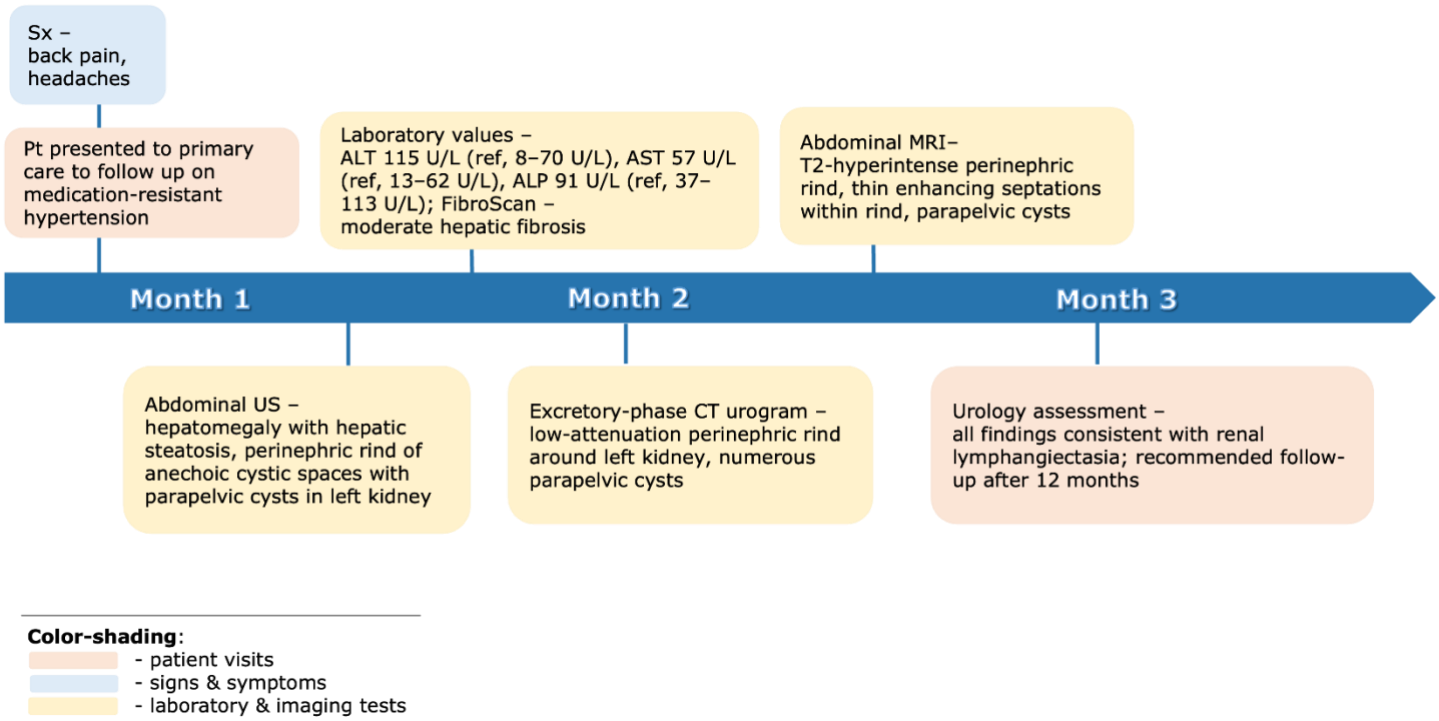


B Post-contrast MRI



(A) Coronal single-shot fast spin echo (SSFSE) T2 and (B) coronal fat-saturated postcontrast MRI images show a T2-hyperintense rind of perinephric tissue with thin enhancing septations (A, arrow) and parapelvic cysts (B, squares).

Case report timeline



Abbreviations: ALP, alkaline phosphatase; ALT, alanine aminotransferase; AST, aspartate aminotransferase; CT, computed tomography; MRI, magnetic resonance imaging; Pt, patient; US, ultrasound; Sx, symptoms

Renal lymphangiectasia can closely resemble several other conditions both clinically and radiologically, making accurate recognition and differentiation essential during evaluation.

Autosomal dominant polycystic kidney disease (ADPKD) presents with bilateral renal cysts. However, in contrast to renal lymphangiectasia, it is typically associated with a positive family history and shows progressive renal enlargement with cortical replacement by cysts on imaging. Renal lymphoma may also mimic renal lymphangiectasia due to lymphomatous infiltration of the renal and perirenal lymphatics that produces overlapping imaging features, such as perinephric fluid or mass-like appearance. However, renal lymphoma usually demonstrates solid and infiltrative lesions on imaging and is often accompanied by systemic symptoms such as fever, weight loss, and multistation lymphadenopathy. Erdheim-Chester disease may present with perirenal soft tissue infiltration that resembles the perirenal fluid collections in renal lymphangiectasia. However this entity is commonly associated with symmetric skeletal involvement and systemic fibrosis, which helps differentiate it from renal lymphangiectasia.

A diagnosis of renal lymphangiectasia can only be definitively confirmed through pathologic examination, but because of the risks associated with tissue sampling and the strength of radiologic evidence, clinicians typically rely on imaging when evaluating and managing this entity.

Conclusion

Renal lymphangiectasia is a rare disorder that accounts for approximately 1% of all lymphangiomas. Clinical presentations can vary widely in both pediatric and adult populations, ranging from asymptomatic cases to symptomatic presentations with flank pain, hypertension, ascites, and renal insufficiency. Diagnosis is best established by imaging via ultrasound, CT, or MRI. Unilateral or bilateral thin-walled cystic fluid collections in perirenal and parapelvic spaces, as well as possible septations, renal cortical thinning, or retroperitoneal extension, are characteristic imaging findings. Differential considerations for renal lymphangiectasia include ADPKD, renal lymphoma, and Erdheim-Chester disease.

Because of the rarity of the disease, it is crucial that radiologists be able to recognize the characteristic imaging features of renal lymphangiectasia to ensure accurate diagnosis and appropriate management.

Author Contributions

Conceptualization, M.P.; Acquisition, analysis, and interpretation of data, J.C., T.K.; Writing – original draft preparation, T.K.; Review and editing, J.C., T.S., M.P.; Supervision, T.S., M.P. All listed authors agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. All authors had full access to all the data in the study and take responsibility for the integrity of the data and accuracy of the data analysis.

Disclosures

None to report.

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