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*Genitourinary and Gynecologic Imaging* | *Case report*

# A rare presentation of unilateral periureteral renal lymphangiomatosis

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## ABSTRACT

Renal lymphangiomatosis is a rare developmental malformation of the perirenal lymphatic system. We report a unique case with unilateral massive periureteral involvement in addition to intrarenal and peripelvic lymphangiomatosis. Although this is a rare entity, it should be considered in patients with peripelvic or periureteric cystic lesions as it may affect appropriate management and follow-up. This case report reviews the imaging features of this entity and a comprehensive literature review and discussion about the entity will be provided.

**Keywords:** Renal, Lymphangiomatosis, Peripelvic cyst

## INTRODUCTION

Renal lymphangiomatosis is a rare developmental malformation of the perirenal lymphatic system. We report a unique case with unilateral massive periureteral involvement in addition to intrarenal and peripelvic lymphangiomatosis.

## CASE REPORT

A 39-year-old female presented to her primary care physician with the right lower quadrant abdominal pain. She denied fever, chills, diarrhea, or weight loss. Her medical history was significant for only hyperlipidemia. Her surgical history was positive for remote cholecystectomy. The patient denied any significant family history of cancer.

Physical examination was unremarkable and vital signs were all within normal limits. Complete blood count, PT/INR, and PTT values were all within normal range. The patient underwent computed tomography (CT) abdomen/pelvis which demonstrated a 6.7 × 4.7 × 6.5 cm hypoattenuating lesion that extended caudally from the left renal pelvis. The patient was referred to urology for further management.

The patient received a follow-up CT urogram 4 months following the initial imaging. Her right lower quadrant abdominal pain resolved by now. There was enlargement of the cystic lesion which now measured 7.6 × 8.5 × 11.1 cm. Innumerable tiny cystic dilatation throughout the left renal parenchyma was more conspicuous on the CT urogram [Figures 1 and 2]. There was prompt excretion of contrast through the non-dilated left collecting system that was slightly compressed

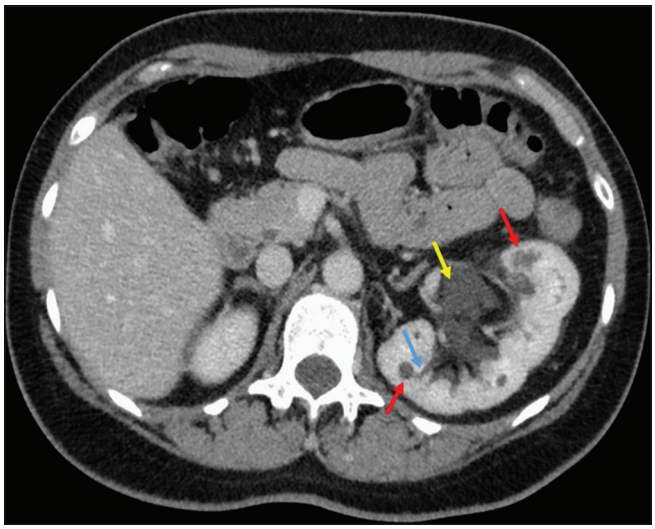

**Figure 1:** A 39-year-old female with renal lymphangiomatosis who presented with abdominal pain. Axial computed tomography urogram through the level of the left kidney in the nephrogenic phase demonstrates multiple foci of cystic dilation in the renal parenchyma (red arrows) some which communicate (blue arrow) with the peripelvic cystic lesion (yellow arrow). These cystic dilations most likely represent lymphangiomatosis within the renal parenchyma.

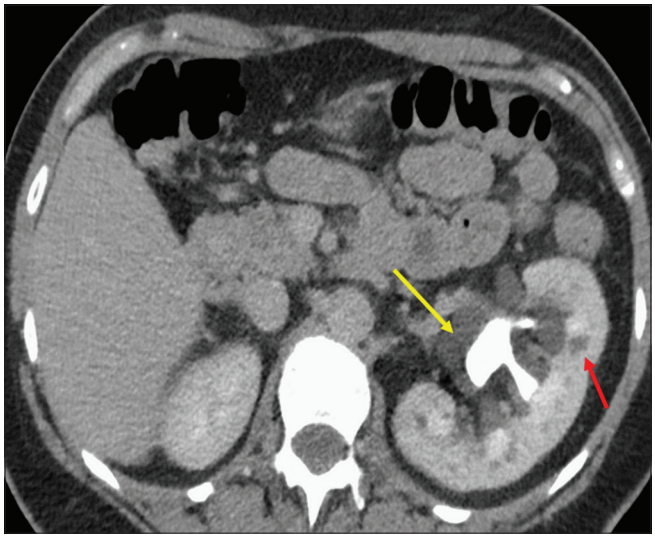

**Figure 2:** A 39-year-old female with renal lymphangiomatosis who presented with abdominal pain. Axial computed tomography urogram through the level of the left kidney in the excretory phase again demonstrates multiple foci of cystic dilation in the renal parenchyma (red arrows). The peripelvic cystic lesion (yellow arrow) does not demonstrate opacification on the delayed excretory images indicating that it is separate from the renal collecting system.

at the proximal and mid ureter level [Figure 3]. The ureter coursed through the center of the fluid density lesion located inferior to the kidney. No lymphadenopathy or free fluid was noted. A follow-up ultrasound (US) demonstrated a 7.7 cm × 4.5 cm cystic lesion with few thin internal septations. There

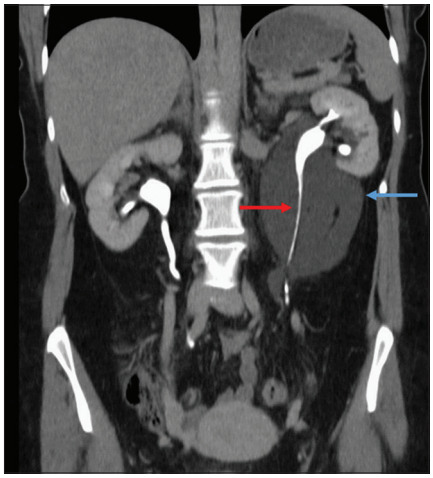

**Figure 3:** A 39-year-old female with renal lymphangiomatosis who presented with abdominal pain. Coronal reformats of the computed tomography urogram during the excretory phase demonstrates the large left periureteric cystic lesion (blue arrow). The ureter is coursing through the center of the lesion and there is mild compression of the proximal and mid ureter (red arrow).

was a communication of the periureteral cystic lesion with the peripelvic cysts [Figure 4].

Our patient underwent an US-guided aspiration of the left retroperitoneal cystic lesion under conscious sedation. 30 cc of clear, yellow fluid was aspirated. Pathology from the fluid showed benign appearing epithelial cells and chronic inflammation with no evidence of malignancy. The fluid creatinine was 0.7 mg/dL. The fluid serous profile showed specific gravity of 1.023 and white blood cell of 39/uL. The fluid differential count showed 84% lymphocyte and 16% monocyte/macrophage. The fluid triglyceride level was 16 mg/dL and fluid total protein level was 3.3 g/dL, ruling out chylous fluid, which is white and has elevated protein and triglyceride concentrations. Given the CT findings and lack of signs of obstruction, the most likely diagnosis was renal lymphangiomatosis which was confirmed by analysis of the aspirated fluid. The patient was scheduled for regular follow-up and to return if symptoms recur.

## DISCUSSION

Renal lymphangiomatosis is a rare developmental malformation of the perirenal lymphatic system. It is caused by dilatation of the lymphatic ducts and cystic collections in the perinephric space due to failure of the perirenal branch to communicate with the rest of the lymphatic system.[1] Clinically, the condition may be asymptomatic, when symptomatic, flank pain is the most frequent

complaint.[2] Other reported symptoms and signs include palpable abdominal mass, gross or microscopic hematuria, ascites, weight loss, and hypertension. Renal function is usually preserved; however, a case with renal insufficiency has been reported.[3] Other isolated associations reported include a case of renal insufficiency with anemia and a case of bilateral renal vein thrombosis.[3,4] These lesions are often incidentally found. Parapelvic areas and the renal sinus are the commonly affected anatomical areas.[5] In cases when they are located within the renal parenchyma, they can mimic multilocular cystic disease of the kidney.[5] Lymphangiomatosis may sometimes extend from the peripelvic area into the periureteric area. Renal lymphangiomatosis is usually bilateral with only few reported cases being unilateral.[6]

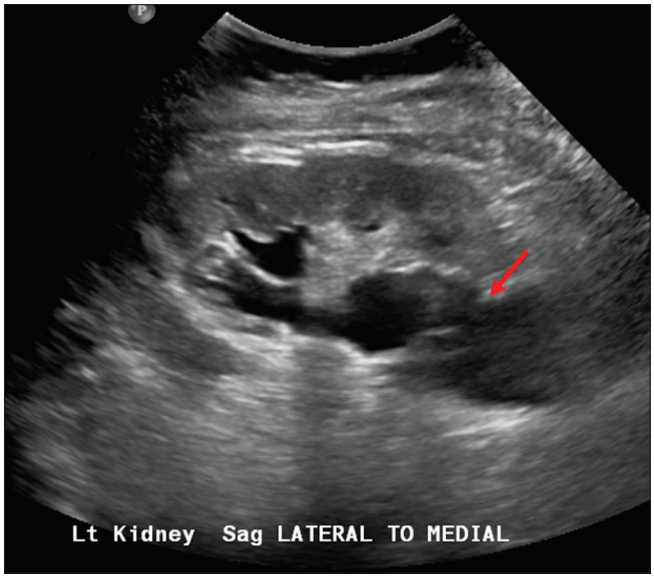


**Figure 4:** A 39-year-old female with renal lymphangiomatosis who presented with abdominal pain. Sagittal gray scale ultrasound demonstrates communication of the periureteral cystic lesion with the peripelvic cysts (red arrow).

Retroperitoneal lymphangiomatosis accounts for approximately 1% of all lymphangiomatosis.[7] A unique finding of our case is unilateral massive periureteral involvement in addition to intrarenal and peripelvic lymphangiomatosis. Anechoic and non-complex appearing cystic structures with no associated solid components are the typical sonographic findings, which may be misdiagnosed as hydronephrosis. There are a few additional entities that may also be confused for renal lymphangiomatosis but it has some distinguishing features [Table 1]. Minimal septal enhancement may be detected on CT images. Displacement of surrounding structures without invasion is common. As compared to US, CT and magnetic resonance (MR) imaging provide better delineation of the anatomic boundaries as well as the relationship between the lesion and the neighboring anatomic structures.[5] On contrast-enhanced CT, there is no enhancement in the early phases and there is no opacification of cystic lesions in the excretory phase. The cystic lesion appears hypointense on T1-weighted images and hyperintense on T2-weighted images. On contrast-enhanced T1-weighted images, there is no enhancement on the early phases. There is no opacification of cystic lesions in the post-contrast T1-weighted MR excretory urography images.[2,5,8] Although the imaging features are typical, in a few cases, the confirmation is done by percutaneous aspiration of the cyst and by assessing the aspirated fluid. The aspirated fluid of renal lymph differs from that of thoracic duct contents because renal lymphatic vessels are out of the pathway of lymphatic drainage of the mesentery. Thoracic duct contents are highly rich in fat and protein, whereas renal lymph contains mostly lymphocytes and small amounts of fat and protein.[9]

The treatment of asymptomatic patients is conservative. Percutaneous aspiration of the collection is required in symptomatic cases and also in cases presenting with pain on account of compression by the collection.[10] However,

**Table 1:** Key features of renal lymphangiomatosis mimics.

| Entity | Description | Important points |
|---|---|---|
| Hydronephrosis | Dilatation of the renal collecting system | The dilated renal collecting system opacifies on delayed post-contrast imaging |
| Vesicoureteric reflux | Typically seen in young children and refers to abnormal reflux of urine from the bladder into the upper urinary tract | Primary diagnostic procedure for evaluation of the vesicoureteric reflux is a voiding cystourethrogram |
| Parapelvic cyst | Originate from the renal parenchyma and protrude into the renal sinus | Low attenuation lesions originating from the renal parenchyma that do not show enhancement/opacification on early or delayed post-contrast images |
| Renal Lymphangiomatosis | Rare entity where there is dilation of the perirenal, parapelvic, periureteric or intrarenal lymphatics | Appear as low attenuation lesions on computed tomography scans and show no contrast enhancement/opacification on early or delayed post contrast images given their lack of communication with real collecting system. Diagnosis can be confirmed with percutaneous aspiration and analysis of the fluid |

the success rate is less in multiseptated larger lesions and leads to more recurrences. Surgery, which consists of cyst marsupialization, is considered as a last therapeutic measure as it may result in nephrectomy.[11,12]

## CONCLUSION

Although renal lymphangiomatosis is rare, this diagnosis should be considered in patients with peripelvic or periureteric area cystic lesions as it may affect appropriate management and follow-up.

### Declaration of patient consent

Patient's consent not required as patients identity is not disclosed or compromised.

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Nil.

### Conflicts of interest

There are no conflicts of interest.

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