American Journal of Case Reports

e-ISSN 1941-5923
© Am J Case Rep, 2021; 22: e933934
DOI: 10.12659/AJCR.933934

Received: 2021.07.09
Accepted: 2021.09.25
Available online: 2021.10.07
Published: 2021.11.08

Bilateral Renal Lymphangiectasia with No Significant Morbidity for Over 25 Years: A Case Report

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Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
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Financial support: None declared
Conflict of interest: None declared

Patient: Female, 39-year-old
Final Diagnosis: Lymphangiectasia
Symptoms: Pain
Medication: —
Clinical Procedure: —
Specialty: Urology

Objective: Unknown etiology

Background: Renal lymphangiectasia is a rare benign disorder with unknown pathophysiology. Renal lymphangiectasia can affect both males and females with no known age predilection. Its diagnosis can be accomplished with radiological images and confirmed by aspiration of lymphatic fluid in certain cases. However, there is no clear presentation to be relied on, apart from incidental findings or presentation of complications, such as hypertension due to mass effect on the kidney or renal failure from chronic compression and obstruction or renal vein thrombosis. Management is directed toward symptomatic relief and protection of the kidneys from failure or obstruction. The timing of possible complications and the duration of conservative therapy are undetermined.

Case Report: Here, we present a case of a healthy 39-year-old woman with bilateral renal lymphangiectasia. It was initially discovered and confirmed to be lymphangiectasia when she was 13 years old and underwent bilateral renal aspiration. She recently presented to the Emergency Department with abdominal symptoms that were found to be caused by diverticulitis. The radiological images showed the persistence of her previous diagnosis of bilateral renal lymphangiectasia. She has had the same condition for more than 25 years, with no related complications or further intervention beyond conservative management.

Conclusions: Renal lymphangiectasia has a benign long-term course with insignificant and minor effects in certain patients regardless of the considerable size of cysts and bilateral involvement of the kidneys. The findings of our case could reassure patients with a diagnosis of a similar condition.

Keywords: Abdominal Cystic Lymphangioma • Hydronephrosis • Polycystic Kidney Diseases

Full-text PDF: https://www.amjcaserep.com/abstract/index/idArt/933934

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Background

Benign, progressive lymphatic vessel proliferation and dilatation with or without associated obstruction involving the lymphatic system of the kidney constitute renal lymphangiectasia or lymphangioma. Based on the regions involved, renal lymphangiectasia can be subclassified as parapelvic, intrarenal, perirenal, or pararenal lymphangiectasia [1]. Primary retroperitoneal lymphatic malformation accounts for about 1% of lymphangiomatous lesions [2].

Renal lymphangiectasia affects both males and females and is not age specific. Additionally, it can occur as isolated unilateral renal involvement; however, bilateral cases have been reported with no significant distinctions from unilateral cases apart from the rarity of bilateral lymphangiectasia [3-5].

Presentations range from asymptomatic and incidental findings to severe flank pain, abdominal pain, hypertension, and renal failure. Overall, the engagement of adjacent spaces such as the pleura or peritoneum can be assumed [6,7]. Treatment is principally oriented to symptoms, and a conservative approach is the preferred management. Surgical interventions in the form of interventional aspiration with or without sclerotherapy injection, cyst marsupialization, and even nephrectomy have been reported [3,8,9]. We present a case of a 39-year-old woman with a history of bilateral renal lymphangiectasia for nearly 26 years with no significant consequences or regression.

Case Report

A healthy 39-year-old woman presented to our Emergency Department with progressive bilateral flank pain that started about 5 days earlier. The pain was dull in nature and alleviated by nonsteroidal anti-inflammatory drugs, and it had unknown aggravating factors. The patient had no fever, chills, or weight loss; however, she had experienced multiple episodes of bilious vomiting and watery diarrhea. The systemic review was unremarkable. The patient had a past surgical history of bilateral percutaneous renal cyst aspiration when she was 13 years old, when she presented with vague abdominal pain. The patient underwent a thorough investigation at that time, and it led to a diagnosis of bilateral renal lymphangiectasia. Multiple cysts of variable size were described as being located in the perirenal, intrarenal, and parapelvic areas. Additionally, the fluid aspirated was characteristically lymphatic (high lymphocyte counts and high levels of proteins and triglycerides). Unfortunately, the previous report did not have images attached, and no additional in-depth radiological details were included. However, the radiological description noted cysts of variable size, with significant fluid collection around both kidneys but no hydronephrosis. The patient's gynecological history was normal. She had 1 daughter, who was born via spontaneous vaginal delivery, and the patient reported no significant problems or complications during the pregnancy. A thorough physical examination revealed normal vital signs, and the cardiovascular and chest examinations were normal. The patient had a tender abdomen with no obvious, palpable masses. A digital rectal examination revealed internal hemorrhoids only.

Moreover, the patient had no lower limb edema, and neurological examinations were normal. Laboratory examinations showed leukocytosis (14×10^3 white blood cells/mm^3), normal renal and liver profiles, and a negative urinalysis. The initial impression was food poisoning, but computed tomography (CT) imaging with intravenous contrast led to a diagnosis of rectosigmoid diverticulitis with ascending and descending colitis. The radiological images incidentally showed bilateral renal lymphangiectasia (**Figures 1, 2**). The patient was admitted and treated uneventfully for the diverticulitis and colitis with antibiotics, hydration, and septic workup for a few days, and antibiotics were continued for a few weeks after her discharge from the hospital. For bilateral renal lymphangiectasia, the patient was treated with conservative management and no intervention was needed. She provided informed consent for publication of her clinical details including radiological images that protected her privacy.

Discussion

The kidney is surrounded by sophisticated lymphatic drainage that traverses through renal tissue and merges into a more prominent channel at the renal sinuses and further drains into regional lymph nodes (paraaortic, interaortocaval, and paracaval). Disruption of that network by congenital malformations or acquired causes can lead to lymphangiectasia [10].

The dilatation of lymphatics produces cysts that vary in size and location, causing a diagnostic dilemma. The top differential diagnosis is hydronephrosis, polycystic kidney disease, and urinoma. However, information is still lacking regarding prevalence and the precise pathophysiology, and most knowledge predominantly originates from case reports. A thorough, careful history and physical examination will certainly provide clues about the possible diagnosis. The presentation is essentially asymptomatic, and lymphangiectasia is usually found incidentally through radiological investigations for another disorder. However, hypertension, hematuria, abdominal pain, ascites, pleural effusion, and renal failure have been reported as probable presentations of lymphangiectasia [3,5,9,11,12]. An adjunct imaging study with or without aspiration of the cyst will lead to the diagnosis.

Ordinary renal ultrasound examination can raise the suspicion of lymphangiectasia, but it cannot verify the lymphangiectasia diagnosis. The ultrasound will show multiple noncommunicating

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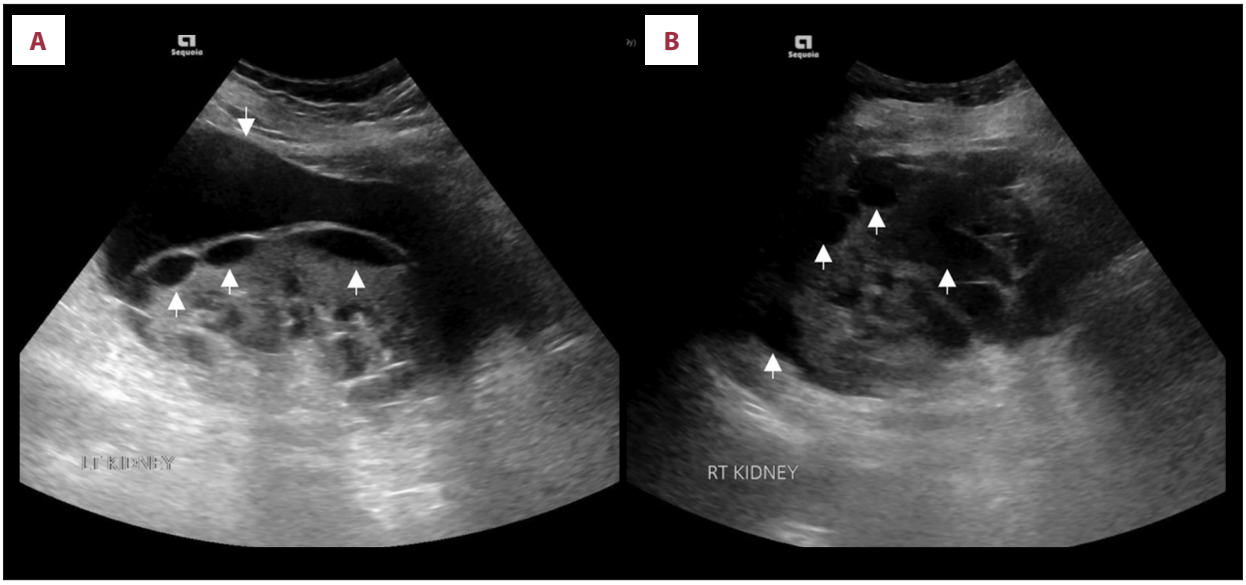

Figure 1. Ultrasounds of the (**A**) left and (**B**) right kidneys showing perirenal, intrarenal, and parapelvic anechoic cysts (white arrows) of inconstant size and site with thin walls.

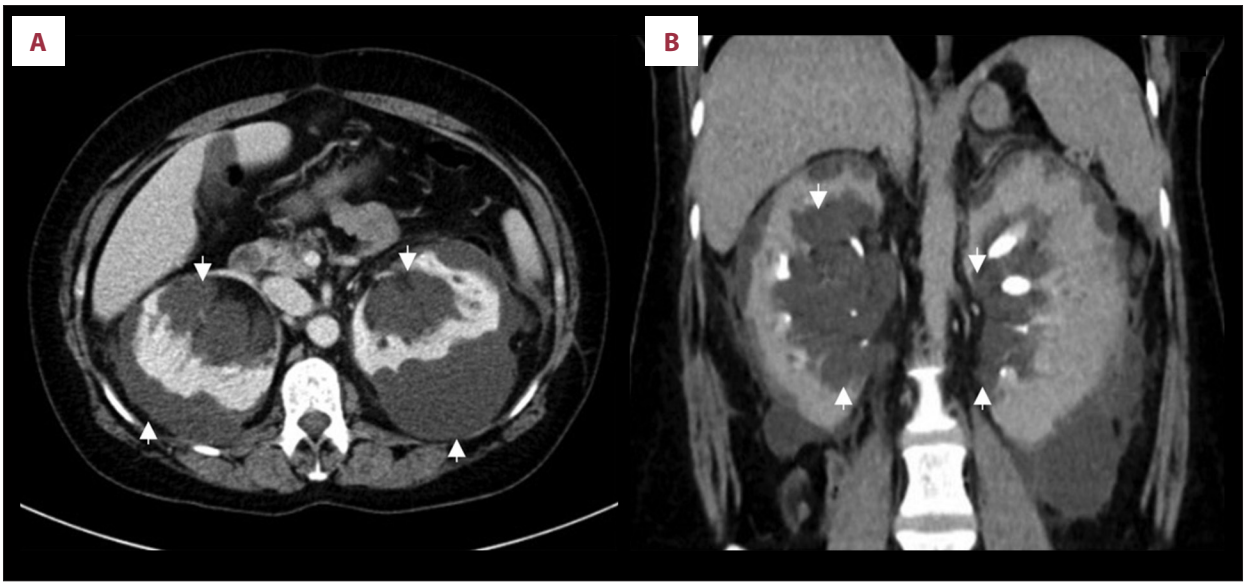

Figure 2. Abdominal computed tomography scan with contrast. (**A**) Portal venous phase, axial section showed large nonenhanced bilateral renal cysts (white arrows) in the perirenal and parapelvic areas. (**B**) Delayed phase, coronal section showed the delayed secretion within the parapelvic lymphangiectasia (white arrows).

septated anechoic cysts with thin walls at inconstant locations in the renal parenchyma or pararenal or parapelvic areas. It is challenging to differentiate such findings from polycystic kidney disease, hydronephrosis, or even urinoma due to perirenal fluid collection [13,14].

A CT scan has more diagnostic value. The renal lymphangiectasia appears as "a well-defined low-attenuation (0-20 HU density) multiseptated collections in perinephric or peri-pelvic regions with normal renal parenchymal enhancement and contrast excretion," as stated by Pandya et al [15]. Three-phase modalities of CT scan have more radiological implications; the lymphatics have low-Hounsfield, nonenhanced cysts during the contrast phase; and the delayed phase helps to track the drainage and allows the exclusion of hydronephrosis [15,16]. Magnetic resonance imaging strengthens the CT findings and will show the lymphatic cysts as hypodense or hyperdense on T1 and T2, respectively. Again, there is no enhancement or opacification of lymphatic cysts during the magnetic resonance excretory phase of T1 [13].

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The management options are broad and depend on the presentation. Percutaneous aspiration with or without sclerotherapy has a reasonable success rate in cases in which it is feasible, but the recurrence rate is high. Marsupialization has an excellent effect in decompression of the cysts, but in cases of refractory hypertension caused by renal lymphangiectasia, nephrectomy can be done as a last resort [7-9].

Conservative treatment is the principal means of management, including watchful waiting and medical treatment of pain and hypertension. This form of therapy depends on the natural history of the disease, which is poorly understood. Therefore, the duration of management is poorly defined. Many theories and reported cases note possible regression, progression, and complications, including renal vein thrombosis, renal failure, and refractory hypertension. Our patient has had renal lymphangiectasia for more than 25 years, with no significant morbidity on conservative therapy. To the best of our knowledge, this is the first reported case of such long-term conservative therapy for renal lymphangiectasia. This case report should indicate a favorable prognosis of renal lymphangiectasia in some patients.

Conclusions

For unknown reasons, renal lymphangiectasia has nonthreatening progression with negligible effects in some patients, regardless of sizable cysts and involvement of both kidneys. Thereby, this case can reassure patients with a diagnosis of this disorder.

Acknowledgments

We wish to acknowledge the Radiology Department at our hospital for their help and technical assistance.

Institution Where Work Was Done

This work was done at King Fahd Hospital of the University, Dammam, Saudi Arabia.

Declaration of Figures' Authenticity

All figures submitted have been created by the authors who confirm that the images are original with no duplication and have not been previously published in whole or in part.

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