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Case Report

Bilateral Renal Lymphangiomatosis: Case Report

Kamal Hlioui, Fatma El Mabrouk*, Karima Sif Nasr, Ouijdane Zamani, Tarik Salaheddine, Rachida Saouab, Jamal El Fenni and Meriem Boui

Department of Radiology, Mohammed V Military Hospital, Rabat, Morocco

***Corresponding Author:** Fatma El Mabrouk, Department of Radiology, Mohammed V Military Hospital, Rabat, Morocco.

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Abstract

Renal lymphangiomatosis is a very rare disorder, characterized by the abnormal development of lymphatic channels in the kidney, resulting in cystic dilatations, with only a few reported cases the misdiagnosis rate of renal lymphangioma is high, as it is easily confused with other renal cystic diseases [1]. We present a case of bilateral renal lymphangiomatosis, manifested by bilateral flank pain, that was falsely diagnosed as hydronephrosis. MRI findings are described.

Keywords: *Renal Lymphangioma; Typical MRI Feature*

Introduction

Renal lymphangiomas also called benign cystic lymphangioma is a rare disorder [2], where there is malformation with insufficient drainage of the intra- or peri-renal lymphatic tissue [3]. The diagnosis can be suggested by imaging, and aspiration of chylous fluid is usually confirmatory.

We present imaging findings of a patient who was admitted with bilateral lower back pain. The patient was diagnosed as having bilateral renal lymphangiomatosis based on MRI imaging.

Case Report

We present the case of 64-years-old woman with a chronic intermittent lower back pain, who's treated for a mild arterial hypertension. She had never showed symptoms attributable to renal disease such as hematuria or urinary tract infections. She had never taken any nephrotoxic medication or illicit drugs and there is no known renal diseases in the family.

The physical examination and blood tests was unremarkable, in particularly the renal function was normal, serum creatinine concentration was 0.7 mg/dl (61.9 µmol/l), urea 22 mg/dl (3.66 mmol/l).

The patient underwent an MRI scan that revealed enlarged kidneys with numerous tubular cystic structures in the cortical region, but preserved parenchyma (Figure 1).

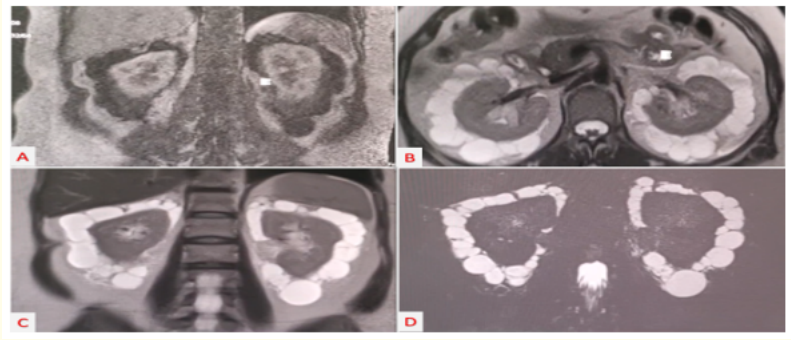

Figure 1: *MRI imaging: (A) T1 coronal, (B) T2 axial (C) T2 coronal (D) T2 HASTE coronal, showing multiple cystic lesions involving the perinephric region. These lesions demonstrate low signal intensity on T1-weighted images and high signal intensity on T2-weighted images, consistent with fluid content with thin internal septations.*

Discussion

Renal lymphangiomatosis is a rare, benign, developmental malformation of renal lymphatics that has been documented in the literature under various names, such as renal lymphangiomatosis, renal lymphangioma, renal lymphatic malformation, peri-pelvic lymphangiectasia, renal sinus polycystic disease, and renal hygroma [4].

Lymphangiectasia is usually found in children, most commonly involving the head and neck regions (approximately 70%), followed by the chest and axillary region (20 - 25%) Only about 5% of cases are located in internal organs, with kidneys accounting for a rare 1% [5].

Due to the relative rarity of the disease, its understanding is based on a few case reports and case series reports. The misdiagnosis rate of renal lymphangioma is high, as it is easily confused with other renal cystic diseases, imaging plays an important role in its diagnosis and differential diagnosis.

Most patients are asymptomatic and are diagnosed incidentally on imaging. In symptomatic cases, clinical manifestations vary, with lumbar pain and hypertension being the most common that can be explained by compressing adjacent structures; Our patient presented a chronic lumber pain and was diagnosed with bilateral RL based on characteristic radiological findings observed in MRI imaging.

Imaging is the gold standard for diagnosis, statistics show that ultrasound and CT can be used as the primary means for the diagnosis of the disease because they have no side effects and are easy to operate. However, CEUS or MRI can provide further information regarding tumor size and extension [5].

Ultrasound may be used as an initial screening modality, demonstrating echogenic cystic lesions with septations in the perinephric region or renal sinus [6]. Computed tomography (CT) is the modality of choice, typically showing multiple well-defined, thin-walled hypodense cystic lesions in the perinephric, pararenal, or intrarenal spaces [7]. Contrast-enhanced CT is essential for differential diagnosis, as renal lymphangiomatosis characteristically shows no enhancement of the cyst walls or contents on delayed images. On magnetic resonance imaging (MRI), the renal cortex typically appears normal, lesions demonstrate low signal intensity on T1-weighted images and high signal intensity on T2-weighted images, with improved visualization of cystic septations.

Differential diagnosis

Polycystic kidney disease and hydronephrosis are the main differential diagnoses for renal lymphangiomatosis.

Polycystic kidney disease is characterized by multiple cysts of varying sizes within the renal parenchyma, and these cysts do not communicate with the renal pelvis or calyces.

Hydronephrosis manifests as dilatation of the collecting system, with contrast enhancement visible in the collecting system on contrast-enhanced CT.

Conclusion

Bilateral renal lymphangiomatosis is a rare benign developmental disorder that may closely mimic more common renal conditions such as hydronephrosis or polycystic kidney disease. Recognition of its characteristic imaging features, particularly on MRI, is essential for establishing the correct diagnosis and avoiding unnecessary invasive investigations or inappropriate treatment. This case highlights the pivotal role of cross-sectional imaging in differentiating renal lymphangiomatosis from other cystic renal diseases and emphasizes the importance of including this uncommon entity in the differential diagnosis of bilateral perirenal cystic lesions.

Informed Consent

Written informed consent was obtained from the parents of patients for the publication of this case report.

Ethical Approval

Our institution does not require ethical approval for reporting individual cases or case series.

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