

## Primary Renal Lymphoma: Diagnostic Approach of an Atypical Presentation

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### ARTICLE INFO

#### Article history:

Received: 21 November 2017

Accepted: 01 December 2017

Published: 06 December 2017

#### Keywords:

Kidney neoplasms;

Lymphoma, Non-Hodgkin;

Urology

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G. Urol Res Ther J

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**Citation this article:** Sarmiento-Sarmiento G, Mantilla-Rey DJ, Rueda-Quijano SM. Primary Renal Lymphoma: Diagnostic Approach of an Atypical Presentation. Urol Res Ther J. 2017; 1(2):116.

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### ABSTRACT

**Background:** Primary Renal Lymphoma (PRL) is an infrequent pathology which existence has been debated due to the extranodal nature of the kidney and the absence of lymphocytes at this level. PRL is defined as the presence of a non-Hodgkin's lymphoma in the kidney without a primary lymphatic extrarenal involvement, representing 0.68% of all extranodal lymphomas in North America. In Colombia there is no epidemiological data about PRL.

**Methodology:** We report the clinical presentation and diagnostic approach of a patient with PRL in a health institution in Colombia.

**Case report:** A 64-year-old female patient centered the urology department referred from a nephrology service where she was being studied for impaired renal function. She was referred due to an incident finding of a left renal mass. Contrast enhanced abdominal MRI showed a poorly defined left renal mass in the hilum with parenchyma extension, associated to a small lymph node conglomerate and pyelocaliceal ectasia. The unusual diffuse perirenal involvement led us to think of a diagnosis different to renal cell carcinoma. Ureteroscopy, pelvic urothelial biopsy and urinary cytology were performed ruling out upper tract urothelial carcinoma. At this point, we suspected a PRL. A percutaneous biopsy of the renal lesion was performed, which resulted in nonspecific lymph node tissue. A laparoscopic renal biopsy was ordered and the results showed a low-grade marginal renal B-cell lymphoma with kappa free light chain restriction and plasmacytoid differentiation, this confirmed the diagnosis of PRL and the need to start chemotherapy.

**Conclusions:** In the presence of an atypical renal mass, the spectrum of differential diagnoses should be broadened and an adequate diagnostic approach should be made from the clinical and radiological point of view. The use of percutaneous or laparoscopic renal biopsies will help increase the diagnosis of these pathologies and reduce unnecessary radical surgical procedures.

#### Introduction

Primary urinary tract lymphoma is a rare condition with a reported incidence of 1 case per 1,000,000 inhabitants per year [1]. The existence of the Primary Renal Lymphoma (PRL) has been debated due to the extranodal nature of the kidney and the absence of lymphocytes at this level [2-4]. Until

now, the cause of PRL is not known [4]. Two theories have been proposed to explain its pathophysiology: an origin at the renal capsule with subsequent parenchymal infiltration, [4] and a chronic inflammation as a predisposing condition for the infiltration of lymphocytes in the kidney [5].

The PRL is defined as the presence of a Non-Hodgkin's Lymphoma (NHL) in the kidney without a primary lymphatic extrarenal involvement [4]. It represents 0.68% of all extranodal lymphomas in North America [6]. In Colombia we do not have any epidemiological data about this neoplasm. Less than 120 cases of LRP have been reported worldwide, with large B-cell NHL as the most frequent subtype [1,4].

We present a case of PRL in our country, in a 64-year-old female patient with impaired renal function as the reason for consultation.

### Case Presentation

A 67-year-old female patient was admitted to the urology service in the outpatient clinic referred from nephrology, where she was being studied for an impaired renal function (Cr: 1.83 mg/dL). Her only symptom was intermittent left flank pain of variable intensity without any impact in lifestyle quality. On admission, no alterations were found in the physical examination.

The patient was referred due to an incident finding of a left renal mass in ultrasound. Contrast enhanced abdominal MRI showed a poorly defined mass with a diameter of 44 x 45 mm, isointense in the T1 sequences and slightly hyperintense in the T2 sequences, compromising the entire left renal hilum and extending and invading the renal parenchyma. Although the mass respected the renal vein and moved it anteriorly, it extended to the retroperitoneum. Additionally, it was associated with a lymph node conglomerate of 24 x 28 mm, with a slight and heterogeneous uptake of contrast in the enhanced phase (Figure 1 and 2).

Due to its atypical radiologic pattern (diffuse perirenal involvement, parenchymal extension and presence of an associated lymph node conglomerate) different diagnosis from a renal cell carcinoma were considered.

DMSA renal scintigraphy showed a functional contribution of 42% and 58% by the left and right kidney respectively.



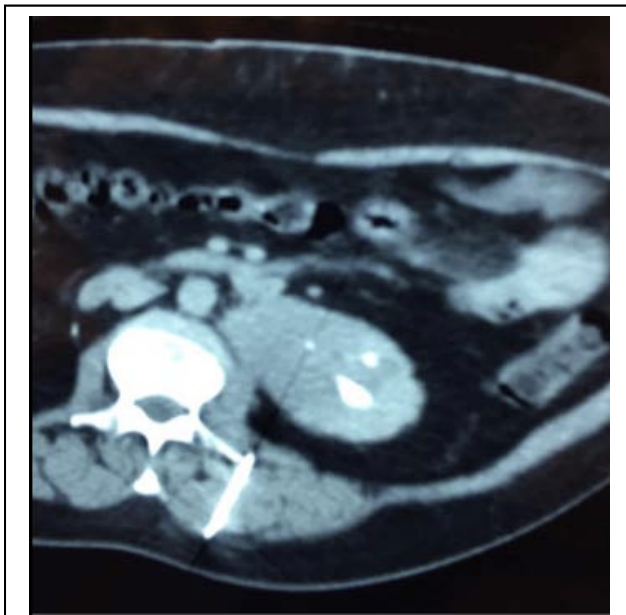
**Figure 1:** Coronal view of the contrast enhanced abdominal MRI where a poorly defined mass is evident, compromising the entire left renal hilum, and extending and invading the parenchyma.



**Figure 2:** Axial view of the contrast enhanced abdominal MRI, in the contrast enhanced phase.

We ruled out an upper-tract urothelial carcinoma with ureteroscopy, pelvic urothelial biopsy and selective urinary cytology. For this reason, a percutaneous biopsy of the mass was performed by the interventional radiology service (Figure 3). However, the histopathological reading reported lymphatic tissue corresponding to a benign lymph node. At this point, we considered that the findings could be related to a PRL. Given the operative characteristics of the trucut needle biopsy and the limitations to obtain a representative sample, we considered necessary a new laparoscopic renal biopsy in order to obtain a greater tissue sample and thus perform specialized studies.

The procedure was performed and the sample was sent to pathology. Histopathological results were compatible with a PRL. Given the low prevalence of this pathology, the sample was sent to an external laboratory in order to obtain a second opinion and to complement studies with immunohistochemistry. A primary low-grade marginal renal B-cell lymphoma with kappa free light chain restriction and plasmacytoid differentiation negative for the presence of mutations in exon 5 of the MYD88 gene was reported.



**Figure 3:** Percutaneous biopsy guided by CT in charge of the interventional radiology service.

The patient was referred to the hemato-oncology service, who discarded differential diagnoses and performed a bone marrow biopsy –reported within normal limits. They considered a diagnosis of and extranodal PRL and started a 4-dose course of chemotherapy with Rituximab.

**Discussion**

The PRL does not have a clinical presentation that differentiates it from other renal masses such as clear cell renal carcinoma, renal metastasis or renal abscesses [4], which makes its diagnosis difficult. The most frequent symptom is pain at the involved flank [7], and it can also be associated with macroscopic hematuria, acute or chronic renal failure, or weight loss [4].

Through the different case reports, several criteria necessary for the diagnosis of PRL have been described. However, there is no global consensus, which makes its diagnosis even more difficult. PRL diagnosis is histopathological [8], by biopsy or after a nephrectomy, once a possible extrarenal lymphomatous origin has been ruled out.

A CT scan is the preferred diagnostic imaging for the study of patients with suspected renal masses, including the LRP [9]. Nonetheless, MRI is a valid diagnostic test, mainly in cases in which there is a negative CT with high clinical suspicion or contraindications for the use of intravenous contrast (allergic or deterioration in renal function) [10], as in the case of our patient.

The most characteristic radiologic pattern of lymphomatous lesions in the kidney is the presence of multiple homogenous masses that show lower contrast uptake when compared with the renal parenchyma [11] (Table 1). By convention, this type of tumors generates little mass effect and therefore does not compress the collecting system in most of the patients [11]. In some cases these lesions growth enough to compress the calyces, but without altering the reniform shape of the kidney [11].

**Table 1:** Characteristic radiological patterns of lymphomatous tissue in the kidney.

CT imaging patterns suggestive of renal lymphomatous tissue.	
Multiple renal masses	60%
Adjacent renal invasion from retroperitoneal compromise	25-30%
Perirenal or diffuse renal infiltration	25-30%

Systemic chemotherapy with or without radiotherapy is the treatment of choice for PRL [12]. However, the option of surgical management in cases of unilateral PRL has also been described [12]. In a case review carried out by Chen et al, a shorter survival time was found in patients with PRL treated with chemotherapy as monotherapy (15.8 months), compared to patients managed with surgery and chemotherapy (49.4 months) [3].

We present a case of LRP in a high-complexity institution in Colombia, in a female patient of 64 years of age with impaired renal function and finding of incidental left renal mass as the reason for consultation at our urology service in the outpatient clinic.

We consider that this case is an atypical presentation, given the finding of lymph node conglomerates and secondary dilation of the urinary tract. Different procedures were required in order to rule out differential diagnoses, and initial inconclusive histopathological findings were obtained which could have led to radical surgical behavior. However, they were confirmed by a new laparoscopic biopsy and histopathology review.

### Conclusion

In the presence of an atypical renal mass, the spectrum of differential diagnoses should be broadened and an adequate diagnostic approach should be made from the clinical and radiological point of view. The use of percutaneous or laparoscopic renal biopsies will help increase the diagnosis of these pathologies and reduce unnecessary radical surgical procedures.

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