

## Oncology

# Primary non-Hodgkin lymphoma of kidney and ureter mimicking an upper tract urothelial carcinoma: A case report

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## ARTICLE INFO

## Keywords:

Non Hodgkin's lymphoma  
Primary renal lymphoma  
Upper tract urothelial carcinoma

## ABSTRACT

Primary renal lymphoma (PRL) is a rare event, while ureteric involvement by the lymphoma mimicking an UTUC is even rarer. A 30-year-old woman diagnosed for a 6-month history of right flank pain with presumed the UTUC of the right kidney. She underwent radical nephroureterectomy with bladder cuff excision, which reveals the infiltrating papillary mass of the ureter from the proximal region towards the ureterovesical junction. Kidney mass was found mostly solid in the parenchymal region, renal hilus was found adherent towards the wall of the inferior vena cava in which it reveals a similar pathology result of Non-Hodgkin's lymphoma.

## Introduction

Primary renal lymphoma (PRL) is a non-Hodgkin's lymphoma (NHL) emerging from the kidney without any sign of extrarenal lymphatic disease and not an invasion from adjacent lymphomatous mass. PRL is a rare entity, due to the nature of the kidney that devoid of lymphatic tissues. In most cases, PRL often treated as primary renal cancer, due to the similarity of the imaging presentation. Initial clinical presentation often insignificant to highlight the suspicion of PRL and it appears to be similar to renal cell carcinoma (RCC), renal abscess or other kidney tumor. Patients with PRL may or may not present with gross hematuria, acute/chronic kidney failure, and flank pain or weight loss. It has been suggested that flank pain is one of the most common symptoms of PRL.<sup>1</sup>

In the rare case of PRL, ureteric involvement by the lymphoma mimicking upper tract urothelial carcinoma (UTUC) is even rarer. It is difficult to distinguish uretral lymphoma from urothelial carcinoma despite using well-established imaging modalities, such as positron emission tomography/computed tomography (PET/CT), antegrade CT pyelography (ACTP) and CT urography (CTU).<sup>2</sup> In this case, we are to report a case of primary NHL of the kidney with ureter and vena cava involvement mimicking an UTUC.

## Case presentation

A 30-year old woman with chief complaint of right flank and right upper quadrant (RUQ) abdominal pain since 6 months before admission. There was no history of gross or microscopic hematuria. Physical examination revealed hipogastric, right flank and RUQ abdominal tenderness with a palpable mass. There was no peripheral lymphadenopathy. The laboratory results were within normal limits without any sign of leukemic blood picture nor evidence of myelosuppression. Urine cytology showed no sign of malignant cells.

Chest X-ray was normal. Abdominal ultrasound revealed a solid mass in the right kidney and RUQ abdomen suspected right renal mass. Positron Emission Tomographic (PET) examination with MSCT scan contrast axial, coronal and sagittal section with fusion image showed right hypermetabolic solid mass involving the pelvocalyceal system and renal parenchymal tissue with the necrotic area extending from right proximal ureter towards the distal region. The mass appears to infiltrate the right perirenal fat and part of the vena cava inferior as well as to the right retroperitoneum (Fig. 1).

We diagnose this patient with UTUC and planned to underwent radical nephroureterectomy with bladder cuff excision. The surgery was done with flank approach to remove the kidney, part of vena cava and Gibson incision to completely remove the distal ureter. At the 10-days follow-up, the patient remained well without any surgical complications

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<https://doi.org/10.1016/j.eucr.2019.101095>

Received 10 October 2019; Accepted 10 December 2019

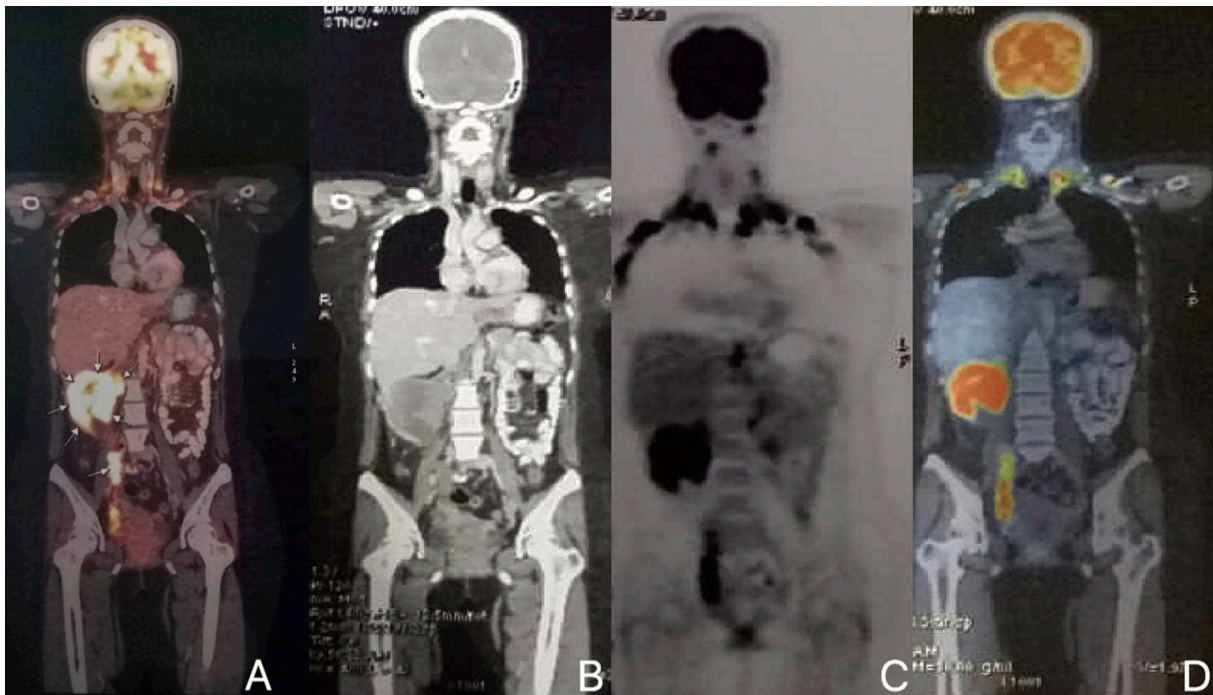
Available online 13 December 2019

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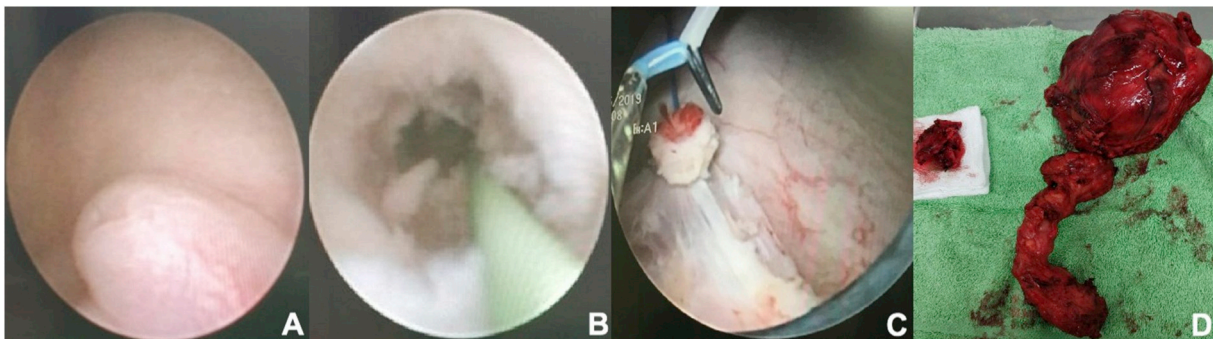
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**Fig. 1.** A, Fused PET/CT image shows lesion (arrow) that is marked FDG avid. B, PET/CT scan contrast shows right kidney mass. C, PET/CT scan contrast shows right kidney mass extending to the right proximal ureter to distal. D, FDG PET/CT scan demonstrates very intense uptake at all sites of a right renal solid mass extending the right proximal ureter to distal.



**Fig. 2.** A, Right ureteral opening B, Ureteroscopy revealed proximal ureter mass. C, Cystoscopy resection of bladder cuff procedure. D, The specimen of the right kidney, the ureter and vena cava wall.

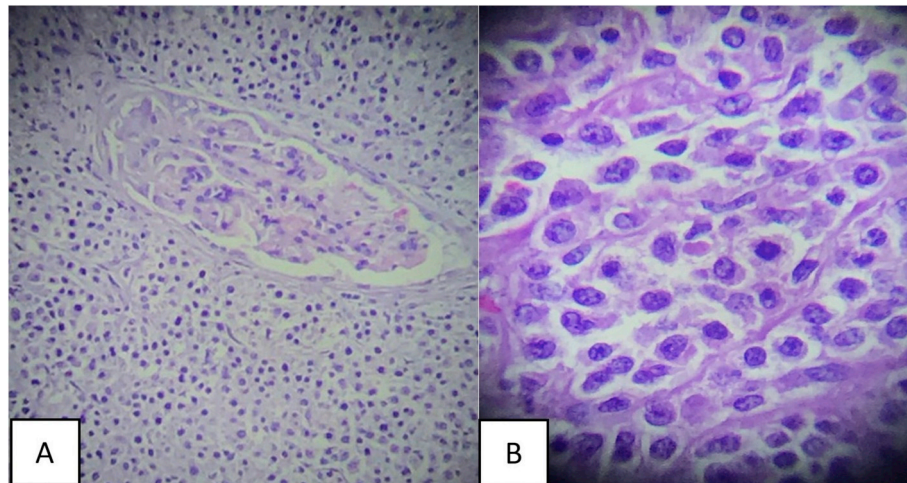
(Fig. 2). Histopathologic examination revealed tumor cells spread diffuse among the connective septa tissue with extensive necrotic areas. It consists of the distribution of oval-shaped lymphoid cells, generally large to medium size, vesicular and hyperchromatic nuclei. The core membrane is irregular with 1–2 core immature cells. Atypical mitosis is easy to find. The conclusion of the histopathologic examination is Non-hodgkin malignant lymphoma. The histopathologic findings were consistent from the right kidney, ureter and vena cava wall contained the same type of tumor (Fig. 3).

## Discussion

Primary renal lymphoma is a rare entity, comprising 0.1%–0.7% of the extranodal lymphoma. Puente Duanay once hypothesized that inflammatory processes recruit lymphoid cells into renal parenchyma and "untimely oncogenic event takes place".<sup>3</sup> Another study has revealed that because the renal capsule is rich in lymphatics, PRL originates from the renal capsule and penetrates the renal parenchyma. Another hypothesis suggests that the lymphomatous process in the perirenal

adipose tissue with secondary involvement of the kidney causes PRL. While PRL is a rare case, involvement of the upper urinary tract by malignant lymphoma usually occurs within the context of disseminated disease. Primary renal pelvis and ureter lymphomas are extremely rare, and the available literature is limited to isolated reports.<sup>4</sup>

Image study of choice for evaluation of renal masses including renal lymphoma is the helical CT scan. In the helical CT scan, renal lymphoma has five common patterns: multiple renal masses, solitary renal mass, renal invasion from contiguous retroperitoneal disease, perirenal disease, and diffuse renal infiltration.<sup>3</sup> In this case, the patient was diagnosed with upper tract urothelial carcinoma, initially shown on PET/CT with MSCT scan contrast axial, coronal and sagittal section with fusion image. Imaging modalities showed mass from the right kidney extending to the proximal and distal ureter, perfectly resemble the UTUC. Moreover, from the PET scan there were no other malignant activities outside the right kidney and ureter. Thus, we naturally assume the clinical diagnosis as UTUC. For an upper tract urothelial carcinoma, radical nephroureterectomy with bladder cuff excision is the treatment of choice. After radical nephroureterectomy with bladder cuff incision, a diagnosis



**Fig. 3.** Histopathological examination of kidney, ureter and vena cava specimen revealed diffuse infiltration of the interstitium by monomorphous atypical lymphocyte. A. H&E 100x magnification, B. H&E 400x magnification.

of Non-Hodgkin's lymphoma was confirmed.

In most cases of PRL and primary lymphoma of the upper urinary tract, the diagnosis of lymphoma was made after surgery, as well as in this case, due to the similarity of the clinical presentation with other renal tumor and in this case, similarity with UTUC. Biopsy, either core biopsy or open biopsy rarely chosen before surgery, especially if the imaging modalities is very suspicious for malignancy. In a case report by Hamada et al. with renal pelvis and ureter lymphoma, they performed open biopsy of the lymph node and periureteral tissue rather than performing a radical surgery. In their case, they were able to avoid radical nephroureterectomy after cycles of chemotherapy.<sup>5</sup> In their case, multiple LN swelling and other findings suggested that the malignant lymphoma originated from the renal pelvis and ureter rather than urothelial cancer, so they decide to performed biopsy. In this case, no clinical finding suspicious to renal and upper tract lymphoma were found, so biopsy was not the option. However, if with initial biopsy and chemotherapy we could avoid nephroureterectomy in cases of primary renal and upper tract lymphoma, the decision of biopsy before surgery must be seriously considered.

### Conclusion

PRL is an extremely rare disease, it should be considered in the differential diagnosis of an upper tract urothelial carcinoma. Early diagnosis

and management can improve the outcome in these patients.

### Source of support

None.

### Appendix A. Supplementary data

Supplementary data to this article can be found online at <https://doi.org/10.1016/j.eucr.2019.101095>.

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