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Case Report

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Primary Renal Lymphoma Masquerading as Upper Tract Urothelial Carcinoma

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Abstract

Primary renal lymphoma without widespread nodal disease is a rare entity. Its diagnosis is challenging as it can mimic other more common renal malignancies such as renal cell carcinoma (RCC) or upper tract urothelial carcinoma (UTUC). We report a 65-year-old male who presented a month history of bilateral flank pain. Initial computed tomography urography showed a soft tissue enhancing lesion with irregular margin in the left renal pelvis, extending distally to the left proximal ureter, suspicious of UTUC. He underwent laparoscopic left radical nephrectomy and bladder cuff excision. However, histopathology came back as a left renal in situ follicular B-cell neoplasm (ISFN). He was subsequently referred to haematology service for further management. This case report aims to raise awareness of ISFN and to consider renal lymphoma in a patient presenting with a renal mass. A high index of suspicion and judicious use of tissue biopsy can avoid unnecessary and potentially morbid surgical interventions.

Keywords: Kidney neoplasms; Lymphoma; Nephrectomy; Diagnostic imaging; Biopsy

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INTRODUCTION

Primary renal lymphoma is uncommon with incidence of less than 1%.[1] It is usually seen in middle to advanced age groups. Common presentations include flank pain, palpable abdominal mass, haematuria, constitutional symptoms, fever, night sweats, or deranged renal function.[1] Some of these symptoms overlap with more common renal malignancies such as renal cell carcinoma (RCC) or upper tract urothelial carcino-

ma (UTUC). Features on imaging are non-specific in most of the cases. Accurate diagnosis requires tissue biopsy and immunohistochemical staining. This is crucial because renal lymphoma is managed with systemic chemotherapy while RCC or UTUC requires surgery. Misdiagnosis can lead to unnecessary surgery and potentially affect patient clinical outcomes.

CASE REPORT

A 65-year-old male with underlying diabetes, hypertension, and gout presented with bilateral flank pain for one month. Otherwise, he did not have haematuria, fever, night sweats or constitutional symptoms. He did not have history of renal stone disease nor any family history of malignancy, including renal malignancy.

Clinically, there was no palpable abdominal mass and he did not have inguinal, cervical and axillary lymphadenopathy. His laboratory results showed normal renal function. Screening urinalysis was unremarkable. His contrast-enhanced computed tomography urography (CTU) revealed an enhancing soft tissue lesion with irregular margin in the left renal pelvis extending distally to the left proximal ureter with luminal narrowing and focal caliectasis interpole and lower pole left kidney. (**Figures 1a-f**). The lesion measured about 2.0 x 3.1 cm. There were multiple subcentimetre enhancing lymph nodes found at the aortocaval, paracaval and aortocaval region. No subcutaneous lymph nodes were detected elsewhere on index CTU.

Based on these findings, a presumptive diagnosis of UTUC was made. The patient underwent laparoscopic left radical nephroureterectomy and bladder cuff excision. Intraoperatively, left kidney was removed en bloc with ureter and bladder cuff. No obvious enlarged periaortic or pericaval lymph nodes noted. The patient recovered well from the surgery.

Cut section of the left kidney revealed a firm irregular whitish lesion in the renal pelvis and seen infiltrating into the renal sinus fat (**Figure 2**).

Unexpectedly, his final histopathological analysis showed lymphoid hyperplasia in the renal pelvis (**Figure 3 a-c**). A few lymphoid follicles were involved by in situ follicular B cell neoplasia. The germinal centres of these neoplastic follicles were positive for BCL2

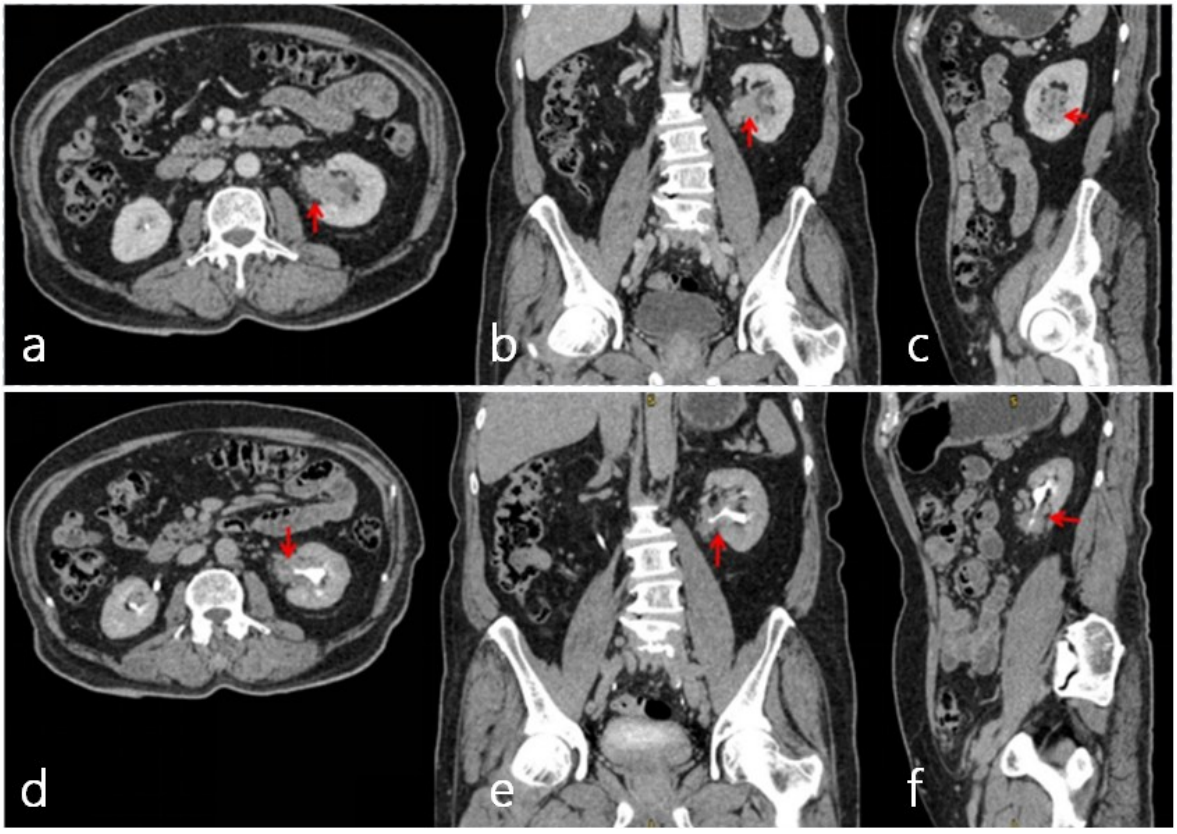

Fig. 1: (a - Axial, b - Coronal, c - Sagittal) : Plain CTU demonstrates enhancing soft tissue lesion with irregular margin in the left renal pelvis, extending distally to the left proximal ureter (red arrow), and (d - Axial, e - Coronal, f - Sagittal) : Delayed/Excretory phase CTU shows enhancing soft tissue lesion causing luminal narrowing and focal caliectasis interpole and lower pole left kidney (red arrow).

CD10 (**Figure 3 d-f**). The abnormal B cells with strong BCL2 and CD10 were limited to germinal centers and did not extend outside of the follicles. Ki67 proliferation index in the neoplastic germinal centers were low (<5%) and the ureter was free of neoplastic cells.

A staging contrast-enhanced CT of the neck, thorax, abdomen and pelvis showed subcentimetre mediastinal, axillary, para-aortic, aorto-caval, paracaval, bilateral common iliac, bilateral external iliac and bilateral inguinal lymph nodes. No splenic or hepatic enlargement. PET CT was done six months post-operative and showed multiple active lymphoma in the thorax, abdomen, pelvis. Extensive discussion regarding treatment options (targeted/ chemotherapy versus active surveillance) was done by haematology service with the patient and the patient opted for active surveillance in view renal toxicity with single functioning kidney.

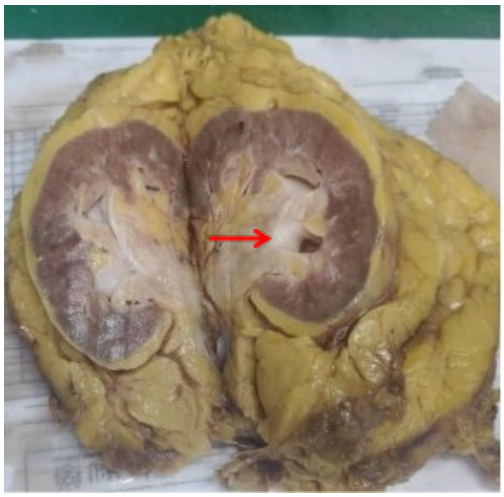

Fig. 2: Cut section of the left kidney revealed an irregular whitish lesion in the renal pelvis and seen infiltrating into the renal sinus fat (red arrow).

DISCUSSION

Renal lymphoma is an uncommon malignancy, typically arising as part of a systemic lymphoproliferative disorder rather than as a primary renal neoplasm. Secondary renal involvement occurs in up to 30–60% of patients with non-Hodgkin lymphoma (NHL), whereas primary renal lymphoma remains a rare entity, with an incidence of only 0.7%.[1] In contrast, UTUC is a relatively more common malignancy of the renal pelvis and ureter, often presenting with haematuria, flank pain, or obstructive symptoms. The diagnosis of primary renal lymphoma remains challenging and requires a high index of clinical suspicion due to the absence of pathognomonic features. Renal tissue biopsy for histopathological examination and immunohistochemical staining are the most accurate diagnostic tools for confirming the diagnosis of renal lymphoma.

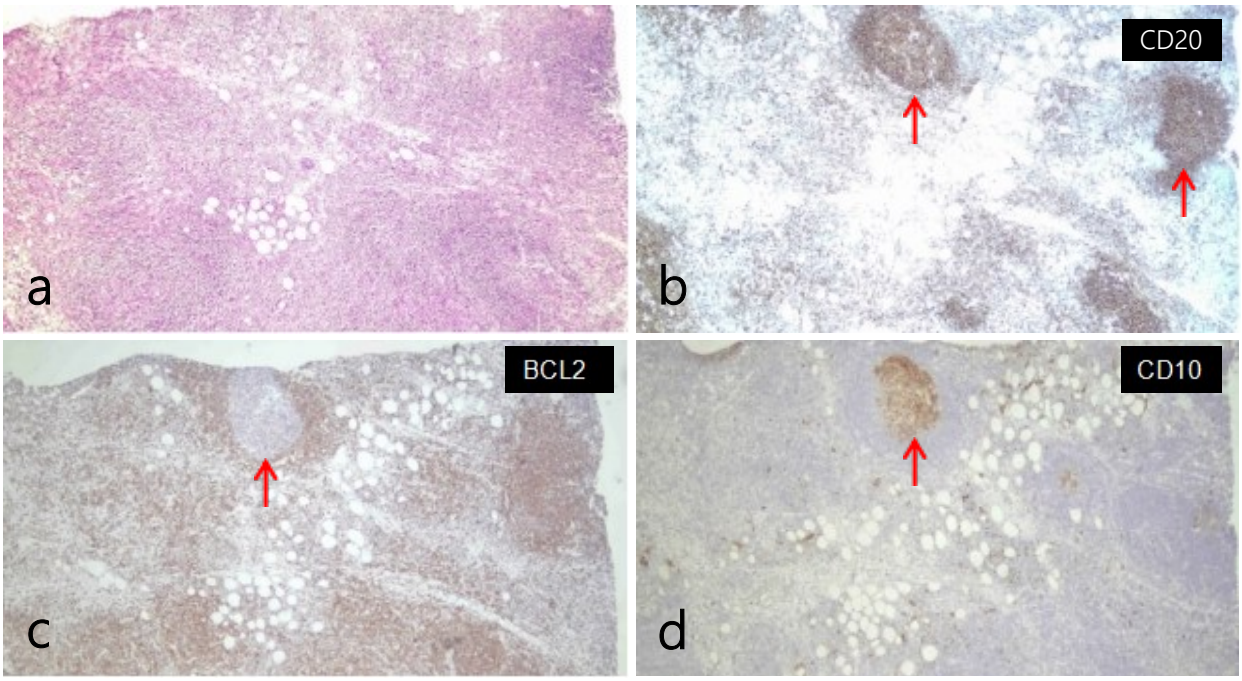

Fig. 3: a) Hematoxylin and eosin (H&E) staining showed lymphoid hyperplasia composed of reactive lymphoid follicles with prominent germinal centres; b) These reactive lymphoid follicles are highlighted by CD 20 (red arrow): c and d) Germinal centres of the neoplastic follicles are positive for BCL2 and CD10 on immunohistochemical staining (red arrow) (x40 magnification).

CTU has a sensitivity of 92% and specificity of 95% in detecting UTUC.[2] Typical features of UTUC on unenhanced phase are soft tissue masses with lower density than renal calculi. In contrasted CT, it shows early enhancement while in the excretory phase, they appear as filling defects or luminal narrowing in the urinary tract.[3] However, its ability to differentiate UTUC from renal lymphoma is limited. In this case, imaging via CTU showed an enhancing soft tissue lesion in the renal pelvis and proximal ureter—features that are often suggestive of UTUC. However, lymphoma can also present as an infiltrative soft tissue lesion, or even mimic collecting system tumours.[4-5] There are some features that may favour lymphoma over UTUC such as lack of haematuria and widespread lymphadenopathy without a clear primary tumour elsewhere. The presence of extensive retroperitoneal lymphadenopathy may suggest lymphoma, but can also be seen in metastatic UTUC, making differentiation difficult without tissue diagnosis.[5]

A percutaneous image-guided core needle biopsy is safe and can provide definitive diagnosis in the majority of renal masses, particularly when lymphoma is a differential consideration.[6] Misdiagnosis can lead to unnecessary surgical intervention which can potentially affect patient clinical outcomes. Based on the workup done, our patient did not have strong clinical (no B symptoms), biochemical (normal LDH and urinalysis) or imaging features (no extensive lymphadenopathies) that would suggest a diagnosis of primary renal lymphoma. On a retrospective note, a diagnostic tissue biopsy of the lesion might provide information regarding nature of the lesion and probably change the surgical management of this patient.

Histologically, our patient's lesion was characterised by reactive lymphoid follicles, some of which were involved by in situ follicular B-cell neoplasia (ISFN)—a rare and often incidental finding. ISFN is recognised as a precursor lesion to follicular lymphoma, characterised by BCL2- and CD10-positive germinal centres that remain confined within the reactive follicles. The low Ki-67 proliferation index and confinement of BCL2+ cells to the follicles distinguish ISFN from overt follicular lymphoma. Though ISFN may not always require immediate treatment, its presence warrants further haematologic evaluation for systemic involvement.[7]

Primary renal lymphoma is mainly treated with chemotherapy. Chemotherapy alone could preserve organ function and might improve renal function.[1] However, studies showed that combination of chemotherapy and surgery produce longer survival times than therapy alone.[8] Early detection and effective treatment is required to improve the prognosis.

CONCLUSIONS

This case report highlights the importance of considering lymphoma in the differential diagnosis of renal masses, particularly when imaging findings are inconclusive. A preoperative renal biopsy should be considered in such cases to avoid unnecessary surgical intervention and delayed appropriate treatment.

Take Home Message

- Primary renal lymphoma is a rare entity with no specific pathognomonic features.
- Accurate diagnosis requires tissue biopsy and immunohistochemical staining.
- Renal lymphoma can be managed with systemic chemotherapy, whereas RCC or UTUC usually requires radical nephrectomy.
- Awareness is important as accurate diagnosis will avoid unnecessary surgery.

Abbreviations

RCC	Renal cell carcinoma
UTUC	Upper tract urothelial carcinoma
CTU	Computed tomography urography
NHL	Non Hodgkin lymphoma
IFSN	In situ follicular B-cell neoplasia

Declarations

All the authors declared no competing interests.

Ethical Consideration

Written consent was obtained from all patients for publications of the clinical details and accompanying images.

Acknowledgement

None

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