A CASE REPORT ON PRIMARY RENAL LYMPHOMA - A DIAGNOSTIC DILEMMA

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Abstract

Primary Renal Lymphoma (PRL) is diagnosed when lymphoma affects the kidneys and there is no evidence of lymphatic manifestation outside the kidneys. PRL is very rare, with an incidence of less than 1%. This case report highlights Computed Tomography (CT) features of PRL in a 68-year-old female with histology-confirmed renal lymphoma. She presented with worsening left flank pain for two months, associated with constitutional symptoms. Abdominal examination reveals a tender mass in the left hypochondriac region, extending to the left lumbar region. Ultrasound revealed an irregular, solid hypoechoic lesion with cystic spaces in the left kidney. Multiphasic CT renal revealed a large ill-defined mass with poor enhancement in both corticomedullary and nephrographic phases, with no significant washout in the delayed phase. It has a low attenuation value of +30 to +50HU on non-enhanced CT and contains no calcification. Meanwhile, in other renal malignancies, particularly Renal Cell Carcinoma (RCC), a renal mass will exhibit significant enhancement with contrast washout in the delayed phase. RCC often demonstrates early infiltration into the Inferior Vena Cava (IVC). The patient is treated with chemotherapy, and a follow-up Positron Emission Tomography-Computed Tomography (PET-CT) presents a smaller left renal mass. In conclusion, a hypovascular renal mass on CT should raise the suspicion of a PRL, and early tissue diagnosis is important to avoid unnecessary nephrectomy.

Keywords: Primary Renal Lymphoma, Multiphase Computed Tomography, Ultrasound abdomen, Diffuse Large B Cell Lymphoma (DLBCL)

Introduction

Renal lymphoma is a rare cancer originating in the lymphatic system and affecting the kidneys (1). There are two major types of lymphoma: Hodgkin and non-Hodgkin lymphoma, with Diffuse Large B Cell Lymphoma (DLBCL) being the most common subtype of non-Hodgkin lymphoma (2). Hodgkin lymphoma with kidney involvement and Primary Renal Lymphoma (PRL) of the DLBCL subtype are very rare, with an incidence of less than 1%. Renal lymphoma can be categorised into two main types: primary and secondary. PRL is when lymphoma affects the kidneys exclusively without evidence of lymphatic manifestation outside the kidneys (3). Secondary renal lymphoma is when the kidneys are involved in the presence of widespread nodal or extranodal lymphoma. Patients with an immunosuppressed status, such as those who have undergone organ transplantation, or those with an immunocompromised status, such as Human Immunodeficiency Virus (HIV) infection, are more vulnerable to developing renal lymphoma (3).

Case presentation

This is a case report of a 68-year-old female with comorbid hypertension, congestive cardiac failure, hyperlipidemia, and bronchial asthma. She had presented with worsening left flank pain for two months. It was associated with constitutional symptoms such as loss of appetite and significant loss of weight. Abdominal examination revealed a hard, immobile tender mass at the left hypochondriac region extending to the left lumbar region, measuring 8 x 6 cm; able to get below the mass yet unable to get above the mass. Her blood investigations, including renal profile, were unremarkable. However, her urinalysis was positive for infection (leukocyte 2+) and had traces of Red Blood Cells (RBC) (1+).

During her initial visit to a private clinic for a similar complaint, an ultrasound was performed, which revealed an irregular solid hypoechoic lesion with cystic spaces in the left kidney. Given the infective picture of the urinalysis, she was treated for Urinary Tract Infection (UTI). However, due to increasing left flank pain, she was later referred to

our centre for further management. A plain radiograph was then conducted at our centre, which revealed a soft tissue opacity on the left side of the abdomen, displacing bowel loops (Figure 1). Following this, a multiphasic renal Computed Tomography (CT) was performed, which presented a large ill-defined heterogeneously enhancing mass (Figure 2a) in the left kidney, infiltrating the left renal vein with a filling defect observed within (Figure 2c) and encasing the left renal artery (Figure 2b).

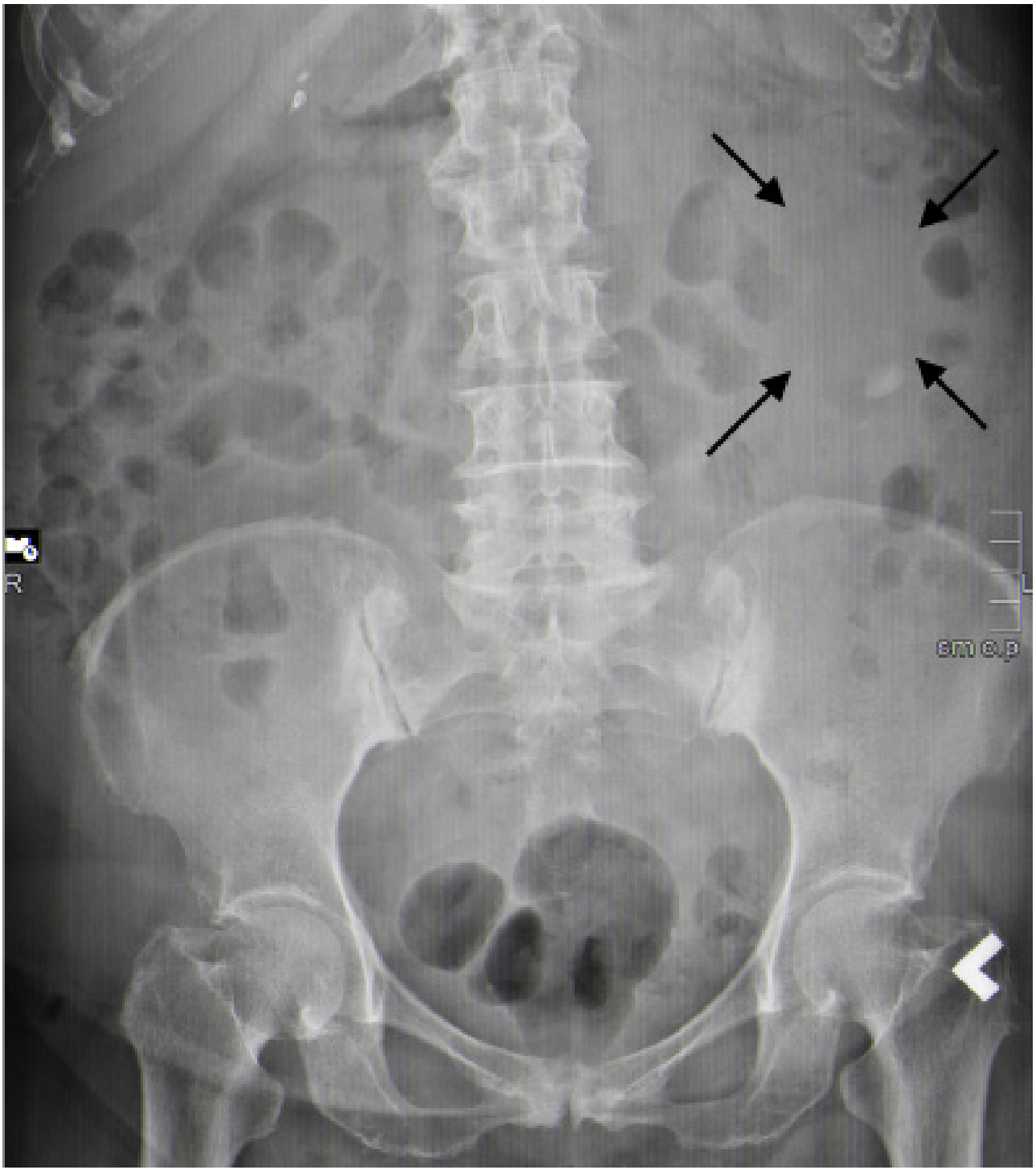

Figure 1: Erect abdominal radiograph. Soft tissue opacity *(black arrows)* is seen at the left renal bed, slightly displacing the bowel loops.

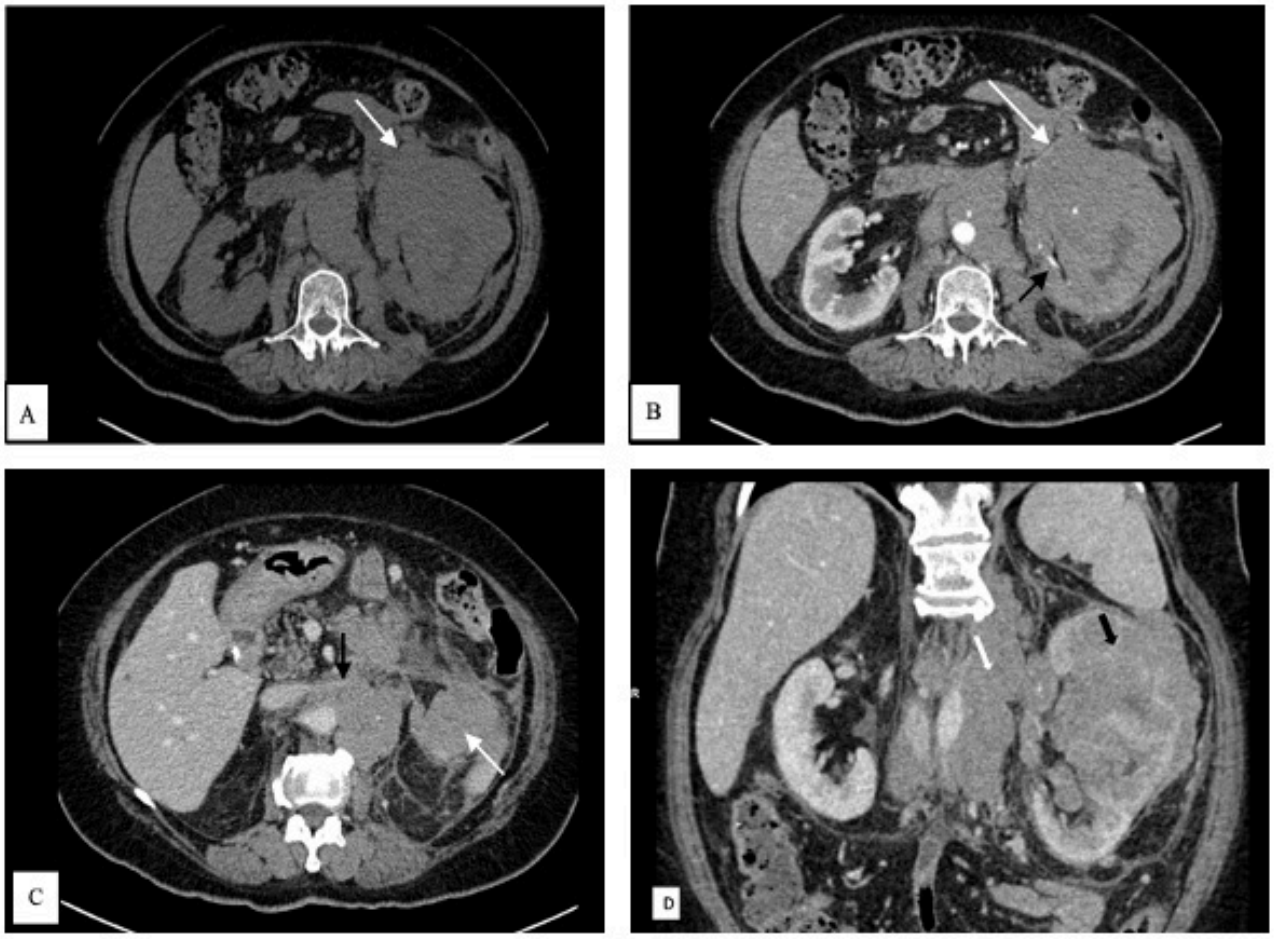

Figure 2: Multiphasic CT renal images in axial and coronal views. **(A)** Non-enhanced phase (at the level of midpole of left kidney): The left kidney is enlarged with a large irregular isodense mass occupying the entire left kidney (*white arrow*). **(B)** Corticomedullary phase (at the level of midpole of left kidney): Large irregular hypodense mass (*white arrow*) seen in the left kidney encasing the left renal artery (*black arrow*). **(C)** Nephrographic phase (at the level of the upper pole of the left kidney): Ill-defined heterogeneously enhancing left renal mass (white arrow) with thrombosis of the left renal vein (black arrow). **(D)** Nephrographic phase (coronal view): Ill-defined heterogeneously enhancing left renal mass (black arrow) with matted abdominal lymph nodes (white arrow).

This mass demonstrates poor enhancement in both corticomedullary and nephrographic phases, with no significant washout in the delayed phase. It causes distortion of the left renal upper pole calyces and left renal pelvis with delayed contrast excretion in the delayed phase. However, no left hydronephrosis was noted. There was also the involvement of abdominal lymph nodes, lung, spleen, and bone, suggesting part of lymphomatous involvement. A biopsy of the left renal mass was performed under ultrasound guidance, and the sample was submitted for Histopathological Examination (HPE).

The HPE concluded that the tissue samples are consistent with High-grade B cell lymphoma; the morphological features were in keeping with DLBCL.

In conclusion, she was treated for left renal lymphoma with extensive metastasis (Stage 4 renal DLBCL) and referred to a haematologist. Thereon, she was started on systemic chemotherapy. After completion of the fourth cycle of R-CHOP (rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisolone), four months after diagnosis, Positron Emission Tomography-Computed Tomography (PET-CT) was performed (Figure 3) to reassess. It demonstrated a significant metabolic reduction of the left kidney mass and abdominal nodes.

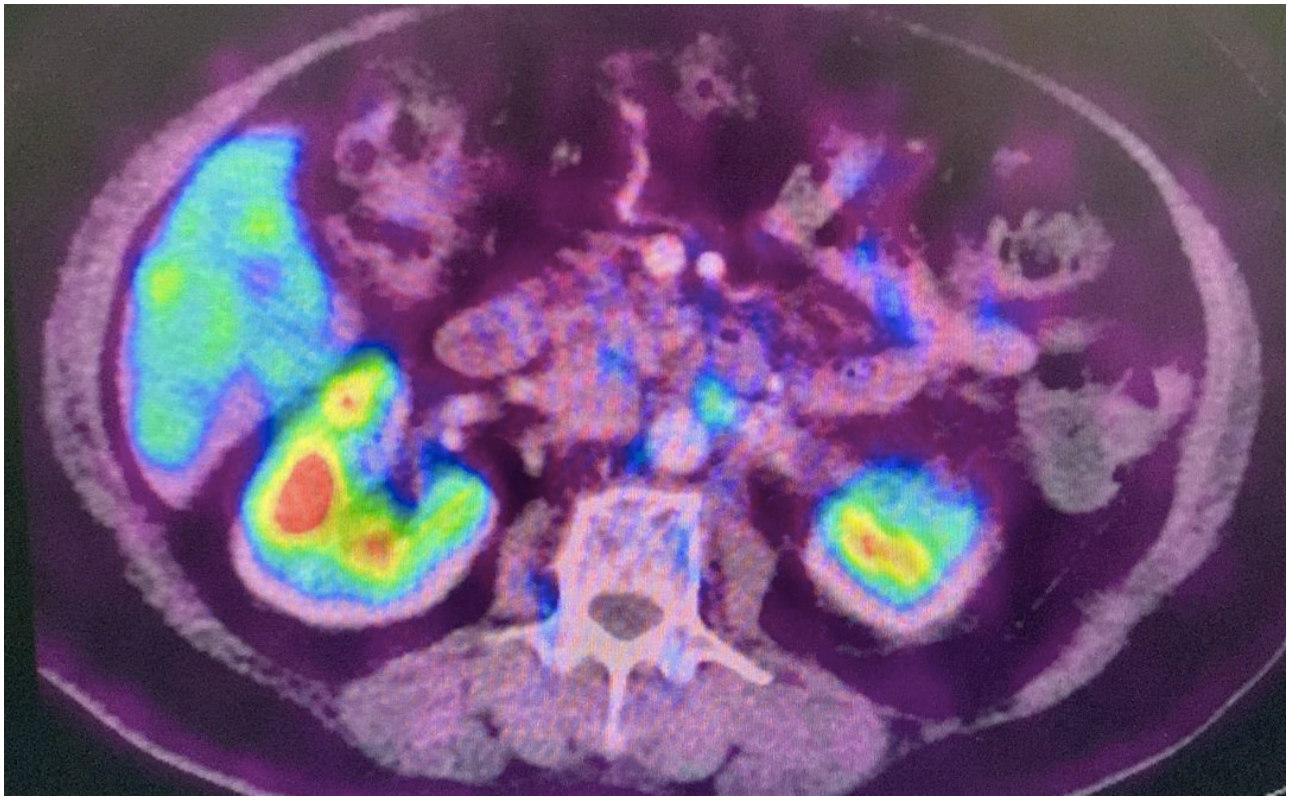

Figure 3: PET-CT image at the level of the upper pole of the left kidney shows resolved renal mass and matted abdominal lymphadenopathy following chemotherapy.

Discussion

PRL is a rare type of lymphoid cancer, with a prevalence of less than 1% of extranodal lymphomas (4, 5). Diagnosing PRL can be controversial and difficult, as a normal kidney does not contain lymphoid tissues. However, some hypotheses suggest that PRL may originate from its renal capsule, which is rich in lymphatic tissue and penetrates the renal parenchyma (5). Additionally, chronic inflammatory conditions of the kidney may attract lymphoid cells that can eventually develop into lymphoma by penetrating through the renal parenchyma (6).

It is crucial to distinguish between PRL and Renal Cell Carcinoma (RCC) as managing both these diseases varies, and timely diagnosis is crucial. The treatment for PRL involves chemotherapy, while RCC requires nephrectomy for better outcomes. Although ultrasound may be the first imaging method used to detect a gross mass, it adds little value to the diagnosis as the ultrasound features of PRL can be non-specific and mistaken for pyelonephritis or abscess (7-12). However, multiphase CT is more effective in differentiating between PRL and conventional RCC.

On a plain CT, a PRL will typically appear as a homogeneous isodense lesion with an irregular margin. However, in the corticomedullary phase, the mass will appear hypodense in comparison to the rest of the renal parenchyma, unlike with RCC, where the mass will demonstrate strong enhancement. In the nephrographic phase, a PRL will display heterogeneous enhancement with an unclear border and will appear hypodense in comparison to the rest of the renal parenchyma. On the other hand, in RCC, the hypervascular mass will be well-demarcated from the homogeneously enhancing renal parenchyma. This is attributable to the fact that a PRL lacks blood supply, while RCC is a highly vascularised tumour.

Apart from that, in PRL, the renal mass has an attenuation value of +30 to +50HU on non-enhanced CT (13). The mass contains no calcification and rarely infiltrates into the Inferior Vena Cava (IVC) to cause thrombosis, unlike RCC (14). In our patient, all these features, the hypodense mass with low attenuation value on plain CT, poor enhancement on nephrogenic phase, and presence of patent IVC, were prompting the radiologist to suspect PRL.

Hence, a hypovascular mass on CT should raise the suspicion of a PRL in the absence of extrarenal lymphomatous manifestation at the time of diagnosis (15).

Conclusion

Differentiating PRL from other renal masses, especially RCC, can be challenging due to the rarity of renal lymphoid malignancy. In view of the aggressive nature of the disease, raising a high index of suspicion in the case of a hypovascular renal mass on CT, with early tissue diagnosis, is essential. This can avoid unnecessary nephrectomy as PRL has a good outcome with systemic chemotherapy.

In our case follow-up, the patient was referred to haematology, and chemotherapy was commenced. Follow-up PET-CT exhibited a significant reduction of the left kidney mass, proving a good outcome.

Acknowledgement

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Ethical Clearance & Informed consent

We declare that ethical approval was not required for our study, and there are no ethical issues. Written informed consent was obtained from the patient for the publication of this case report.

Competing interests

The authors declare that they have no competing interests.

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