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A rare primary renal lymphoma with liver infiltration: A case report and literature review

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ABSTRACT

Primary extra-nodal of Non-Hodgkin Lymphomas (NHL) are rare, and one of the very rare primary loci is renal, namely Primary renal lymphoma (PRL), which is about 0,1–0,7%, and the mortality rate reaches as high as 75%. Here, the author shares a 58-year-old man with an imaging test indicating a mass from the upper pole of the right renal with a sixth segment of the liver infiltration. The patient underwent multidisciplinary embolization and nephrectomy. Histopathologic show the anaplastic lymphoid cells in the kidney and the hepar and lead to chemotherapy with a CHOP regimen for six cycles.

1. Introduction

Primary extranodal lymphomas account for 25–35% of all non-Hodgkin lymphomas (NHL). Notably, one of the exceedingly rare primary sites for NHL is the renal region, constituting only 0.1–0.7% of cases. Typically, this neoplasm is detected post-nephrectomy without a perioperative biopsy, and it is frequently misdiagnosed initially as renal cell carcinoma (RCC). Primary renal lymphoma (PRL) carries a daunting first-year mortality rate, which can soar to as high as 75%.[1]

However, proper management can significantly improve this prognosis, drawing from previous similar cases, particularly those involving metastasis. In this context, the author presents a highly unusual case of PRL featuring liver metastasis, underscoring the need for a multidisciplinary surgical approach. Furthermore, this case is compared with existing literature and is supported by evidence from Pubmed, Google Scholar, and PMC Europe case reports.

2. Case presentation

A 58-year-old man experienced intermittent colic pain in the upper right region of his abdomen for the past year, and this pain had worsened progressively over the last three weeks. A physical examination revealed the presence of a solid mass beneath the liver area. Tests for carcinoembryonic antigen (CEA) and alpha-fetoprotein (AFP) returned within the normal range. An ultrasonography (USG) scan showed a heterogeneous hypoechoic lesion on the right lobe of the liver with an irregular border near the upper part of the right kidney. A contrast-enhanced abdominal computed tomography scan (CT-Scan) revealed a mass measuring 9.3 × 10.9 × 9.4 cm arising from the upper pole of the right kidney, with infiltration into the sixth segment of the liver measuring 5.8 × 5.8 × 4.9 cm, along with hepatomegaly. This presentation raised suspicion of renal cell carcinoma (see Fig. 1).

In managing the patient, renal artery embolization was initially performed using an 8 mm diameter Amplatzer Vascular Plug (AVP) before proceeding with an open radical nephrectomy. A multidisciplinary team carried out the Cattle-Braasch maneuver to remove a complex mass that included the right kidney, Gerota's fascia, a portion of the proximal right ureter, and a fragile tumor mass located at the upper pole (see Fig. 2). A hepatectomy of the sixth liver segment was also performed, and several para-aortic lymph nodes were collected.

The patient was closely monitored in the intensive care unit (ICU) for three days. Microscopic pathological examination revealed anaplastic lymphoid cells in both the kidney and liver, displaying pleomorphic characteristics, round nuclei, thin cytoplasm, and diffuse infiltrative growth, similar to the findings in the para-aortic lymph nodes, and suggesting NHL (see Fig. 2). Thoracic X-ray and bone scintigraphy did not indicate any metastasis.

Chemotherapy treatment consisted of six cycles of

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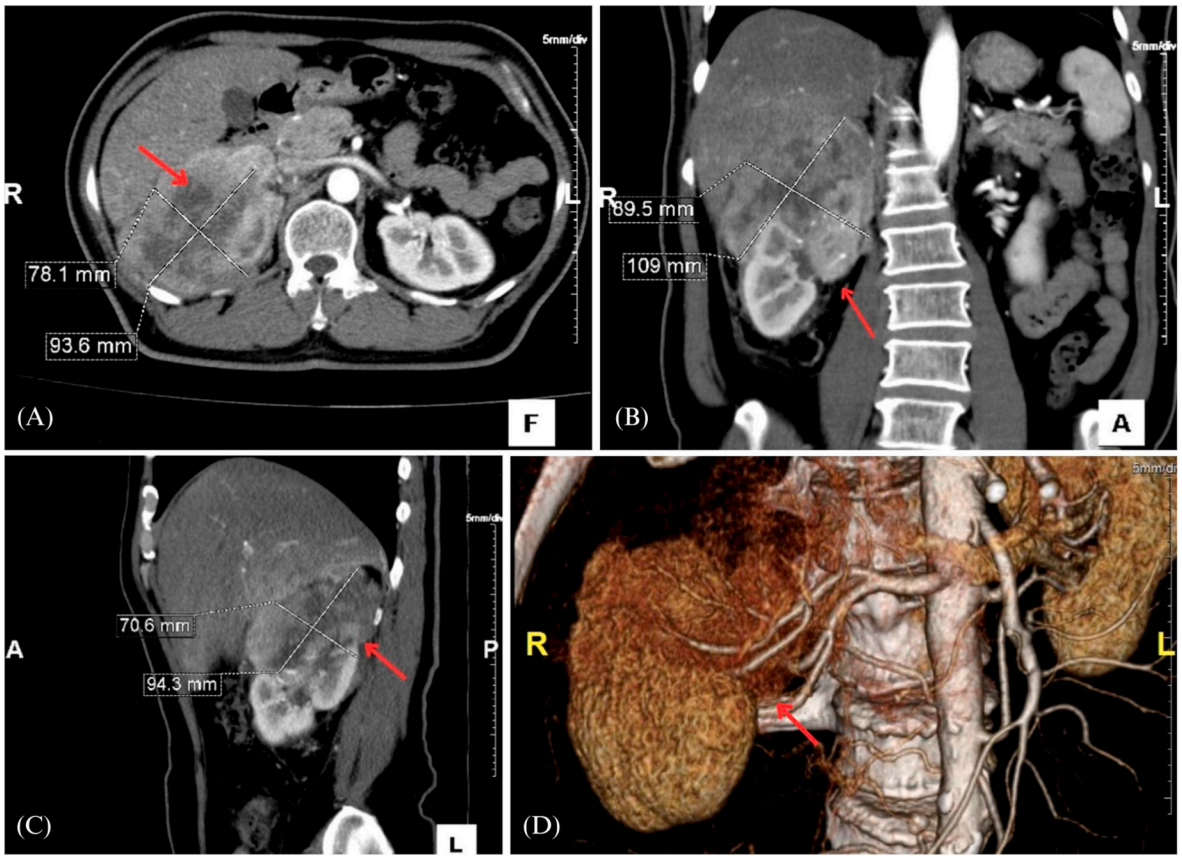

Fig. 1. Imaging test of PRL. A right renal mass (red arrow) infiltrated the six segments of the liver. (a) Axial section of CT-Scan. (c) Coronal section of CT-Scan. (d) Sagital section of CT-Scan. (e) Renal angiography. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

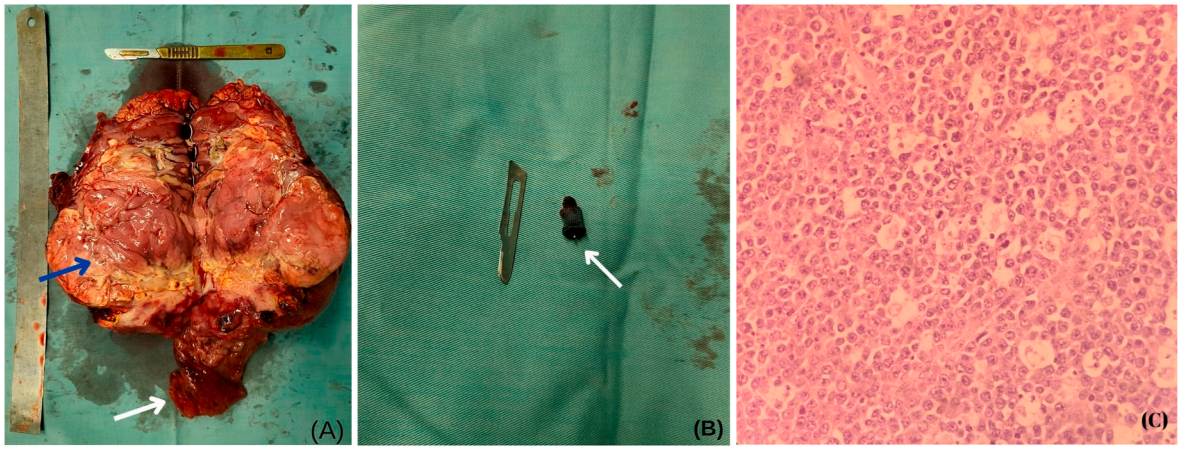

Fig. 2. A renal tumour of PRL. (a) Renal tumour growth on the whole renal with a slice of infiltrating mass in the liver. (b) A renal embolization plug. (c) Histopathology of the tumour.

cyclophosphamide, vincristine, doxorubicin, and prednisone (CHOP) under the supervision of an oncologist. A contrasted CT scan a month after completion of chemotherapy showed no remaining lesion in the right retroperitoneal space. The patient reported no abdominal pain or anemia after the treatment. Follow-up assessments are scheduled annually for up to five years to monitor overall health and any potential recurrence, and this monitoring is ongoing at the time of this report. One year after chemotherapy, the patient had no signs of recurrence based on regular imaging examinations.

3. Discussion

In the literature review, we identified 28 case reports of PRL with the first reported case dating back to 1953. PRL is often found as a secondary involvement in kidney tumors, even during autopsy (see Table 1). The most common symptoms include flank or abdominal pain, with hematuria being frequent, especially in adults.[1] In children, malaise is the predominant symptom. PRL can manifest in a single kidney or affect both kidneys.[2] Immunohistochemical analyses commonly reveal the presence of CD20, Bcl6, and Bcl3, which indicate B-cell proliferation in

Table 1
Profile of PRL in previous case reports.

(Ref.)	Gender	Age (year)	Mass Location	Main Symptomp	Tumour's Dimension	Metastasis	Treatment	Chemotherapy Regimen	Outcome	Histopathology
Nasrollahi et al. (2022)	Male	50	Right	Flank pain	9 × 8 cm	–	Nephrectomy & Chemotherapy	R–CHOP	Complete remission	DLBCL
Bruce et al. (2020)	Male	12	Bilateral	Malaise	12,6 cm (R) & 11,9 (L)	–	Chemotherapy	R–CHOP	Complete remission	DLBCL
Hiwale et al. (2020)	Male	65	Left	Abdominal pain	9,2 × 9,1 × 6,5 cm	Para-aortic lymphadenopathy	Nephrectomy	No	Unnoted	DLBCL
Bokhari et al. (2020)	Male	21	Bilateral	Shoulder pain	13,7 × 6,7 cm (R) & 14,1 × 6,1 cm (L)	–	Chemotherapy	R–CHOP	Decreasing BUN/ Creatinine	DLBCL
Cheung et al. (2019)	Male	51	Left	Flank pain	12.5 × 12.0 × 12.7 cm	–	Chemotherapy	R–CHOP	Complete remission	DLBCL
Zhao et al. (2019)	Male	68	Right	Flank pain	8.5 × 6.8 cm	–	Nephrectomy & Chemotherapy	R–CHOP	Unnoted	DLBCL
Saddadi et al. (2017)	Male	38	Bilateral	Itching skin lesions	–	–	Chemotherapy	Unnoted	Decreasing BUN/ Creatinine	DLBCL
Erdogmus et al. (2015)	Male	19	Bilateral	Hematuria	16 × 8 cm (R) & 15,5 × 8 cm (L)	–	Chemotherapy	R–CHOP	Normal Renal Function	DLBCL
Haar et al. (2016)	Male	4	Bilateral	Abdominal pain	14 cm (R) & 13,5 cm (L)	Pancreas & spine	Chemotherapy	Inter-B-NHL rituxan 2010 protocol	Unnoted	Burkitt's lymphoma, DLBCL
Chen et al. (2016)	Female	70	Right	Incidental USG finding	3,6 cm	–	Nephrectomy & Chemotherapy	CHOP	Complete remission	DLBCL
Wang et al. (2015)	Male	84	Left	Abdominal mass	–	Intracranial	Chemotherapy	R–CHOP	Complete remission	DLBCL
Hughes et al. (2013)	Male	28	Bilateral	Ophthalmic shingles	–	Lymphoma nodes	Chemotherapy	R–CHOP	Decreasing BUN/ Creatinine	DLBCL
Hart et al. (2012)	Female	82	Right	Incidental USG finding	14 cm (R) & 13,5 cm (L)	Pulmonary nodules, thyroid nodules, liver, aorta, & inferior vena cava duodenum	Chemotherapy	R–CHOP	Unnoted	DLBCL
Valli et al. (2010)	Male	46	Left	Flank pain	12 cm	–	Chemotherapy	R–CHOP	Unnoted	DLBCL
Reuter et al. (2009)	Female	52	Bilateral	Back pain	15,8 × 8,6 cm (R) & 16 × 8,9 cm (L)	Supra- & infra-diaphragmatic lymph nodes	Chemotherapy	R–CHOP	Unnoted	DLBCL
Omer et al. (2007)	Female	21	Bilateral	Abdominal pain	13,7 × 8,5 (R) & 13,6 × 6 cm (L)	–	Chemotherapy	VACOP-B	Complete remission	DLBCL
Becker et al. (2007)	Male	5	Right	Abdominal pain	17,4 cm	Bone marrow	Chemotherapy	CCG-1961 protocol	Died	DLBCL
Diskin et al. (2007)	Male	77	Right	Anorexia	2 cm (mass)	–	Chemotherapy	R–CHOP	Decreasing BUN/ Creatinine	DLBCL
Bozas et al. (2006)	Male	70	Right	Low back pain	9 × 9x9 cm (Weight 240 gr)	–	Nephrectomy & Chemotherapy	R–CHOP	Complete remission	DLBCL
Olusanya et al. (2006)	Male	79	Left	Body aches	10,0 × 9 × 6,5cm	–	Nephrectomy	No, CKD	Decreasing BUN/ Creatinine	DLBCL
Tuzel et al. (2003)	Male	43	Right	Flank pain	9 × 5 cm	–	Nephrectomy	No	Complete remission	MALT, DLBCL
Gellrich et al. (2002) (1)	Male	62	Bilateral	Hematuria		–	Chemotherapy	CHOP	Died	DLBCL
Gellrich et al.	Male	45	Right	Perirenal hematoma	20 cm	Caecum & ascending colon	Nephrectomy & Chemotherapy	B-ALL protocol	Complete remission	Burkitt's lymphoma, DLBCL

(*continued on next page*)

Table 1 (*continued*)

(Ref.)	Gender	Age (year)	Mass Location	Main Symptomp	Tumour's Dimension	Metastasis	Treatment	Chemotherapy Regimen	Outcome	Histopathology
(2002) (2)										
Gellrich et al. (2002) (3)	Male	70	Right	Flank pain	14 × 11	Paracarotid lymph nodes	Chemotherapy	CHOP	Died	DLBCL L
Mita et al. (2002)	Male	77	Right	Kidney mass	–	–	Nephrectomy	No	Complete remission	MALT, DLBCL L
Basavaraj et al. (2000)	Male	71	Left	Hematuria	–	–	Conservative	No	Complete remission	Lymphoplasmacytoid type
Gelder et al. (1992)	Female	58	Bilateral	Anorexia	17 cm	–	Chemotherapy	CHOP	Complete remission	DLBCL
Sheil et al. (1991)	Female	29	Bilateral	Malaise	13 cm (R) & 14 cm (L)	–	Chemotherapy	MACOP-B Protocol	Complete remission	DLBCL

this context.

Histologically, PRL presents with glomerular or interstitial infiltration of lymphoma cells, accompanied by an expanded interstitium. This appearance can sometimes be misinterpreted as pseudo-proliferative glomerulonephritis.[1] Chen et al. have classified PRL into six subtypes: marginal zone lymphoma, follicular lymphoma, small lymphocytic lymphoma, Burkitt lymphoma, diffuse large B-cell lymphoma (DLBCL), and natural killer (NK)/T-cell lymphoma.[2] DLBCL is the most common subtype and may appear alongside other subtypes, such as Burkitt lymphoma.[1,3,4]

Imaging studies often reveal a hypoechoic mass with a heterogeneous texture in the renal area and effacement of renal sinuses in USG. Contrast-enhanced CT-Scan typically shows kidney enlargement with indeterminate boundaries, which can be valuable in identifying the edge of homogenous perinephric encasing the normal parenchyma.[1,4]

The diagnosis of PRL can be challenging, but Bruce et al. have simplified the criteria to include lymphomatous renal inflammation, enlarged kidneys without obstruction, and no extrarenal lymphoma at diagnosis. However, these criteria may not cover all cases.[5] A multidisciplinary approach is crucial for managing PRL, particularly when metastasis is present. In this case, liver metastasis complicated matters, involving a team of urologists, digestive surgeons, thoracic cardiovascular surgeons, and oncologists. Abdominal regional lymph nodes can also serve as potential sites for the spread of lymphoma cells. Specific organs, such as the liver, thyroid, colon, lungs, pancreas, spine, and intracranial sites, have been reported as metastatic targets.[3]

While palliative nephrectomy for renal malignancies is associated with minimal intraoperative blood loss of only 5.8% of patients preceded by embolization, the role of nephrectomy in PRL remains debatable, although it is generally recommended in cases of unilateral PRL.[1] Robotic-assisted partial nephrectomy, although potentially offering higher excision accuracy than open laparotomy, was not chosen due to its higher cost. Although relatively rare, metastasis can occur in other organs, such as the meninges, central nervous system, bones, lymph nodes, adrenal glands, and liver.[1] In this case, liver involvement led to adhesions in the diaphragm, necessitating a multidisciplinary approach to management.

The standard treatment regimen for PRL, especially the DLBCL subtype, typically includes Rituximab, Cyclophosphamide, Doxorubicin Hydrochloride, Vincristine Sulfate, and Prednisone (R–CHOP), often in conjunction with nephrectomy. R–CHOP has shown better outcomes in terms of survival probability compared to CHOP alone.[4] However, the median survival for PRL is still relatively low, ranging from 1 to 2 years, underscoring the need for advanced treatment strategies.[1]

4. Conclusion

The mortality rate for PRL is associated with a lower median survival, necessitating early diagnosis and intervention. Using a multidisciplinary approach for comprehensive management can potentially lead to improved outcomes. Treatment choices for PRL are often a subject of debate due to its rarity. Nevertheless, combining chemotherapy and palliative nephrectomy has shown promise in enhancing survival rates.

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