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PRIMARY RENAL NON-HODGKIN LYMPHOMA: A RARE CASE REPORT

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Abstract: Primary renal Non-hodgkin lymphoma is rare. Most of the cases are unilateral though bilateral cases have been reported. Extrarenal involvement is to be excluded by imaging/staging laprotomy for definitive diagnosis. We are presenting a case of 40-year-old Indian female who was attended with history of right flank pain for 3 years, found to have right renal mass on CT scan, underwent right radical nephrectomy and six courses of chemotherapy with CHOP (Cyclophosphamide, Doxorubicin, Vincristine, Prednisolone) regimen subsequent to histo-pathological confirmation. Patient is asymptomatic on follow up of 18 months.

INTRODUCTION

Malignant lymphoma, especially non-hodgkin lymphoma (NHL), may arise in tissues with no infiltration of the lymphnodes, spleen, Waldeyer's ring, or thymus; such cases are referred to as primary extranodal NHL. This accounts for one third of all NHL cases.¹ Diffuse large B-cell lymphoma (DLBCL) is the predominant type of NHL, accounting for approximately 40% of all lymphomas.^{2,3} It has also been reported that 30-50% of NHL cases have extranodal manifestations in various organs, such as the gastric tract, skin, bone, central nervous system, breast, heart, liver or adrenal glands.¹ Although renal involvement is a

common finding in the patients with advanced stages of NHL (ranging from 37 to 47%), primary renal lymphoma (PRL) is quite rare (less than 1 percent of renal lesions) and it is often mistaken for renal cell carcinoma (RCC).⁴ PRL is thought to originate in the lymph node/s of the renal sinus or in the lymphatic network of the renal capsule and to manifest as focal mass/es (solitary or multiple), large infiltrative lesions engulfing the kidney, or diffuse bilateral enlarged kidneys.^{5,6} Most patients additionally have adjacent retroperitoneal adenopathy. Currently, more than 50 cases have been reported in the literature.

CASE PRESENTATION

This 40-years-old female from Haryana (India) presented with history of pain in abdomen for 3 years which was episodic, vague, relieved by painkillers. She noticed weight loss of approximately 5 kg over past 2 months. On physical examination right flank mass approximately 7cm x 8 cm was palpable.

Blood counts, liver and kidney function tests were within normal limits, whereas urinary analysis showed microhaematuria and proteinuria. Ultrasonography (USG) of the abdomen and pelvis showed an irregular outlined hypoechoic mass lesion of size 8cm x 10cm at

the upper pole of right kidney. The contralateral kidney appeared to be normal in size, shape and echotexture.

CECT abdomen and pelvis (Figure 1 A-C) revealed heterogeneously enhancing space occupying lesion on the antero-superior pole of right kidney, extending into suprarenal fossa and abutting inferior hepatic margin. Margin of lesion were poorly circumscribed exophytic. Adrenal gland was not separately visualized from mass. Soft tissue plane with hepatic margin were indistinct and lesion was reaching upto the hilum compressing IVC, encasing renal vascular pedicle, both renal artery and vein.



Figure 1 A - Coronal section



Figure 1 B - Axial section



Figure 1 C - Axial section

IVP showed bilateral normal functioning kidney. FNAC of renal mass was suggestive of malignant round cell tumor.

Nephrectomy was done for diagnosis of renal mass. Biopsy was suggestive of NHL, diffuse large B-cell type. Immunohistochemistry was done and showed LCA +, CD20+ and CK, Vimentin & CD5 negative (Figure 2A-D).

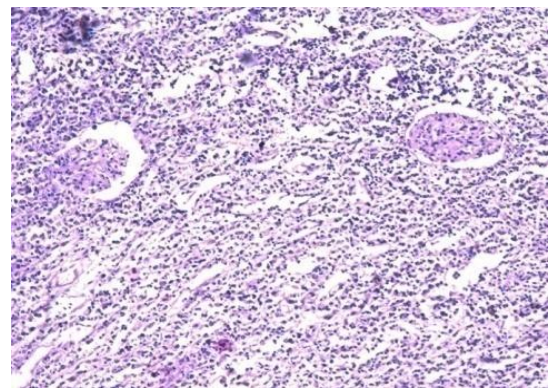


Figure 2 A. Photomicrograph showing atypical lymphoid cells diffusely infiltrating renal parenchyma (H & E, 10X)

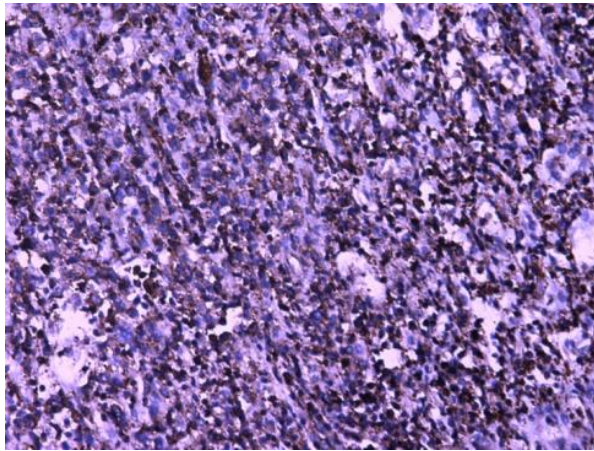


Figure 2 B. Lymphoid cells positive for LCA (IHC, 20X)

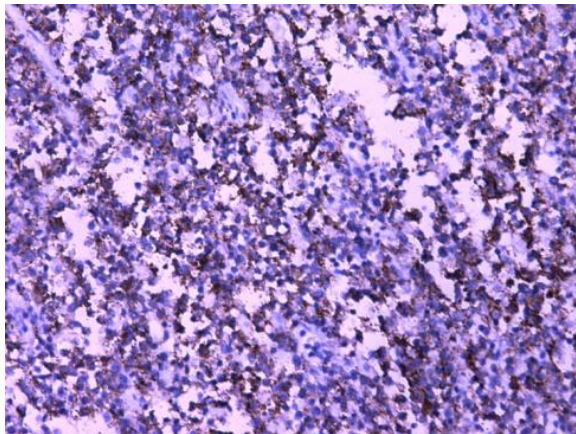


Figure 2 C. Lymphoid cells positive for CD20 (IHC, 20X)

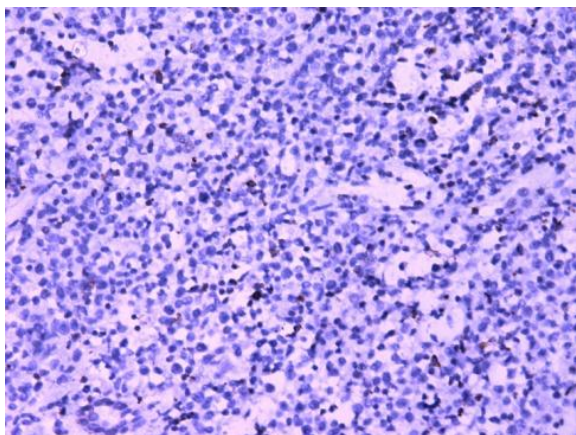


Figure 2 D. Lymphoid cells negative for CD5 (IHC, 20X)

Physical examination and imaging showed no disease elsewhere in the body. After radical nephrectomy patient was given six cycle of chemotherapy (CHOP) cycle being three weekly (Inj. Cyclophosphamide

750mg/m² D1, Inj. Doxorubicin 50mg/m² D1, Inj. Vincristine 1.4mg m² D1 and Tablet Prednisolone 100 mg PO D1-D4). Rituximab could not be offered due to economical constraints.

Post-chemotherapy ¹⁸F-FDG whole body PET-CT showed no definitive evidence of metabolically active disease anywhere in body (Figure 3). Patient remains symptom-free as of now (18 months after chemotherapy).

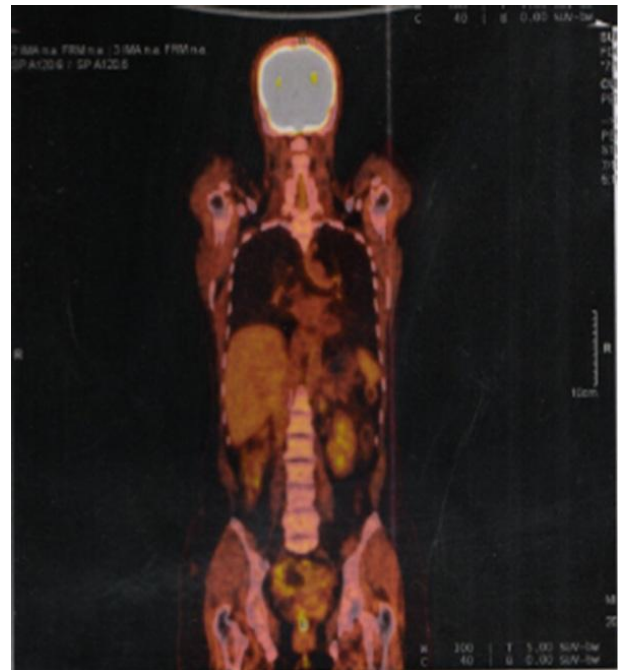


Figure 3 PET-CT showing no definite evidence of metabolically active disease

DISCUSSION

The term 'Primary renal lymphoma' (PRL) is attributed when the disease is localized to the kidney without any sign of other organ involvement or in which renal involvement is the presenting manifestation.⁷ PRL is a rare and uncertain entity because renal parenchyma lacks lymphatic tissue. Therefore, the status of PRL as a primary disease or the first manifestation of a rapidly progressive systemic disease is controversial.

PRL is often mistaken for a RCC and the diagnosis is performed after a radical nephrectomy. However, imaging studies may

provide evidence for suspected cases of PRL. Ultrasonographic images typically depict an unspecific homogeneous hypoechoic mass. Therefore a CT scan is preferred to differentiate PRL from RCC. PRL usually appears as hypervascularized mass with minimal and characteristic homogeneous contrast enhancement. Other indirect signs of PRL include- renal size enlargement, sinus or hilum direct infiltration by a bulky mass, or diffuse retroperitoneal infiltration. In contrast, indirect signs that may indicate RCC include- presence of calcifications, venous thrombosis, or an obstructive mass effect over the renal vessels or urinary tract. However, a percutaneous biopsy can confirm the diagnosis. Bone gammagraphy and a bone marrow biopsy should be performed to exclude extrarenal dissemination.

Systemic chemotherapy is currently the first treatment option for PRL. Though most authors believe that CHOP protocol should be an elective option (as it is in non-Hodgkin's BCL), there is no agreed-upon standard

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