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Rare and Controversial Entity of Malignant Genitourinary Tract Tumor: Unilateral Primary Renal Lymphoma of Left Kidney

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Conflicts of Interest: Nil.

Abstract

Extra nodal lymphoma involves various organs. Primary renal lymphoma (PRL) is a very rare entity. Most of the PRL are diagnosed as diffuse large B-cell lymphoma. Pathogenesis of PRL remains controversial. It is essential to diagnose PRL, by exclusion of systemic involvement. We report a case of 60 years old lady who presented with flank pain and mass without hematuria. Histology confirmed the diagnosis of non-Hodgkin's lymphoma, diffuse large B-cell type. Though the prognosis of PRL is very poor but early diagnosis, surgical resection with chemotherapy does improve the survival of patient.

Key words: Renal tumor; primary renal lymphoma; chemotherapy.

Introduction

Primary renal lymphoma (PRL) is a rare and controversial entity of the genitourinary tract tumors, localized to the kidney only. PRL is commonly non-Hodgkin's lymphoma (NHL) and majority of the patients have intermediate or high grade lymphomas of B-cell origin. PRL comprises only 0.7 % of all extra nodal lymphomas and represents less than 1% of renal tumors (1). We report case of a 60 years old lady with flank mass and pain diagnosed as diffuse large B-cell lymphoma of left kidney (LK).

Case Report

A 60 years old lady presented with left side flank mass with dull aching intermittent pain since 1 month. She had undergone hysterectomy for dysfunctional uterine bleeding 6 years back. She did not have hematuria, fever, vomiting, diabetes or hypertension. Lab investigations revealed hemoglobin of 13.4 gm/dL, total leucocyte count of 9.94 x 10^3/µL, with differentials having76% neutrophils, 20% lymphocytes, and 2% each of monocytes and eosinophils. Urine routine examination revealed 30-35 pus cells/ high power field, although the culture was sterile. Serum (S.) creatinine (Cr) was 0.68 mg/dL, blood urea, 26 mg/dL, S.Na^+ 141 mEq/L, S.K^+ 4.58 mEq/L, S. lactate dehydrogenase 438 U/L(normal range: 100 to 190 U/L),blood sugar: 93 mg/dL and she was non-reactive to HBsAg/HIV/HCV by ELISA.

X-ray chest was unremarkable. Ultrasonography revealed a large mass in LK (left kidney) and unremarkable contralateral kidney. Contrast enhanced (CE) CT scan showed a large homogenously hypodense lobulated soft tissue mass measuring 11.3 x 10 x 9.1 cm in LK involving upper pole and infiltrating to renal vein (RV) (Fig 1).

Patient was subjected to open left radical nephrectomy and specimen was sent for histopathological examination. Gross examination revealed LK weighing 800 grams, measuring 16 x 10 x 8 cm in size. Cut section revealed a tumor involving the upper and mid pole of the kidney, measuring 11 x 10 cm in size, homogenous, grey-white in color with focal areas of hemorrhage (Fig 2). Uninvolved renal parenchyma at the lower pole measured 5 x 4 cm in size. The mass appeared to be invading the RV.

Microscopis examination revealed a tumor comprised of monomorphic round cells with hyperchromatic coarse nuclei and prominent nucleoli, having scanty cytoplasm, arranged in sheets (Fig 3a, b). Atypical mitotic figures were also noted. Focal areas of hemorrhage and necrosis were also seen. The RV showed tumor infiltration. On immunohistochemistry CD20, CD38, LCA, Bcl2, MUM-1, and PAX-5, kappa & lambda were positive (Figure-4a,b,c). CD10, Bcl6, CD2, CD3, Vimentin and S-100 were negative. Ki 67 was positive with an Index >80%. Thus diagnosis of diffuse large B cell lymphoma was established.

The patient was offered CHOP regimen (cyclophosphamide, adriamycin, vincristrine and prednisone). After 6 months of follow-up, she is doing well without any evidence of recurrence.

Discussion

Common sites for extra nodal lymphoma include gastro intestinal tract, bone marrow, liver, kidney, breast, skin, thyroid and adrenal gland. Occurrence of PRL is rare because of absence of lymphoid tissue in renal parenchyma. PRL constitutes 0.7 % of all extra-nodal lymphomas (2). Secondary renal involvement is observed in 30-60 % of autopsy studies (3,4). Pathogenesis of PRL is not known but the lymphatics in renal capsule may be the cause for this lesion. Other possibility could be chronic inflammatory diseases like chronic pyelonephritis, Sjogren's syndrome, systemic erythematous lupus, or viral infections which lead to lymphoplasmacytic

infiltration eventually leading to lymphoproliferative disorder (1,5). PRL comprise NHL involving both B and T-cells. Most of the patients have intermediate or high grade lymphoma of B-cell origin. Diffuse large B-cell lymphoma (DLBCL) is the largest sub-type of NHL accounting for 40% reported by registry of Danish lymphoma group (6). The peak incidence of DLBCL is in the 6thand 7th decade of life (5,7). However most of the cases of PRL have been observed between 4th to 6th decades with male predominance. Clinically patients present with abdominal mass, pain and hematuria (7, 8, 9). Immunophenotypically DLBCL is most commonly positive for B-cell lineage markers like CD20, Bcl2, follicular center cell markers CD10 in 40% and Bcl6 in 60% cases. Plasma cell associated markers CD38 and Mum1 are also positive. Ki-67 shows high proliferative index. As the histology revealed small round monomorphic population of tumor cells, we performed all the IHC markers along with Ki-67 to find out the propensity for proliferation which would also help in deciding the prognosis.

Yasunaga et al has reported six out of eight cases of DLBCL (7). A case report by Abdulla Ladha & Ghulam Haider et al also reported a case of PRL diagnose as a DLBCL(10). Lymphadenopathy or hepatosplenomegaly were not present at time of diagnosis in most of the cases. Our patient had singular involvement of RV invasion without any lymphadenopathy or metastasis, case similar to one reported by Vazquer Alongo et al (11). Celik et al reported a case of PRL with metastasis to hilar lymph node (LN) surrounding the renal artery along with splenomegaly, sparing RV (4). Adamasco Cupisti et al reported bilateral PRL with uninvolved perinephric fat, LN or distance metastasis (9). It is very essential to diagnose PRL without any systemic involvement like hepatosplenomegaly or LN/distant metastasis both by physical examination as well as radiological examination. PRL has poor prognosis with rapidly invading systemic lymphatic tissue and metastasis to lung and breast. Combined surgical resection and chemotherapy are necessary for better survival and regular follow-up to rule out any recurrence. However 75% mortality rate has been reported after one year of diagnosis (1, 4, 11).

Conclusion

PRL is a rare extra-nodal lymphoma with unknown etiology. It has a rapidly progressive and aggressive course. Early diagnosis and surgical resection with systemic chemotherapy may improve survival rate.

Conflict of interest

None of the authors had any conflict of interest regarding the publication of this case report.

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References

[1]. Vázquez-Alonso F, Puche-Sanz I, Sánchez-Ramos C, Flores-Martín J, Vicente-Prados J, Cózar-Olmo JM. Primary renal lymphoma: long-term results of two patients treated with a chemotherapy + rituximab protocol. Case Rep Oncol Med. 2012; 2012:726424.

[2]. Qiu L, Unger PD, Dillon RW, Strauchen JA. Low-grade mucosa-associated lymphoid tissue lymphoma involving the kidney: report of 3 cases and review of the literature. Arch Pathol Lab Med 2006; 130:86-89.

[3]. Urban BA, Fishman EK. Renal lymphoma: CT patterns with emphasis on helical CT. Radiographics 2000; 20:197-212.

[4]. Celik A, Yildiz Y, Ozekincl S and Mizrak B. Non-Hodgkin's Lymphoma of Kidney, Presenting as a Solid Mass: Case Report. J Clin Exp Pathol 2012, 2:5.

[5]. Moslemi MK, Tahvildar SA, Ashtari AA. Primary Lymphoma of the Kidney in an Adult Male - The First Reported Case from Iran. Case Rep Oncol 2010; 3:72-76.

[6]. Møller MB, Pedersen NT, Christensen BE. Diffuse large B-cell lymphoma: clinical implications of extranodal versus nodal presentation a population-based study of 1575 cases. Br J Haematol 2004; 124:151-159.

[7]. Yasunaga Y, Hoshida Y, Hashimoto M, Miki T, Okuyama A, Aozasa K. Malignant lymphoma of the kidney. J Surg Oncol 1997; 64:207-211.

[8]. Cyriac S, Rejiv R, Shirley S, Sagar GT. Primary renal lymphoma mimicking renal cell carcinoma. Indian J Urol 2010; 26:441-443.

[9]. Cupisti A, Riccioni R, Carulli G, Paoletti S, Tognetti A, Meola M, Francesca F, Barsotti G, Petrini M. Bilateral primary renal lymphoma treated by surgery and chemotherapy. Nephrol Dial Transplant 2004; 19:1629-1633.

[10]. Ladha A, Haider G. Primary renal lymphoma. J Coll Physicians Surg Pak 2008; 18:584-585.

[11]. Vázquez Alonso F, Sánchez Ramos C, Vicente Prados FJ, Pascual Geler M, Ruiz Carazo E, Becerra Massare P, Funes Padilla C, Rodríguez Herrera F, Cózar Olmo JM, Tallada Buñuel M. Primary renal lymphoma: report of three new cases and literature review. Arch Esp Urol 2009; 62:461-465.

Figure Legends

Figure 1: Large homogenously hypodense lobulated soft tissue mass in left kidney.

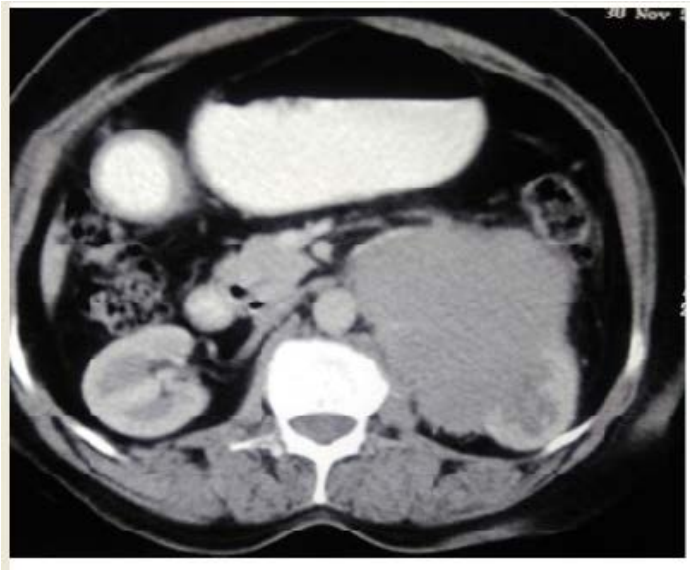

Figure 2: Tumor involving upper and mid pole of left kidney homogenously grey-white in color.

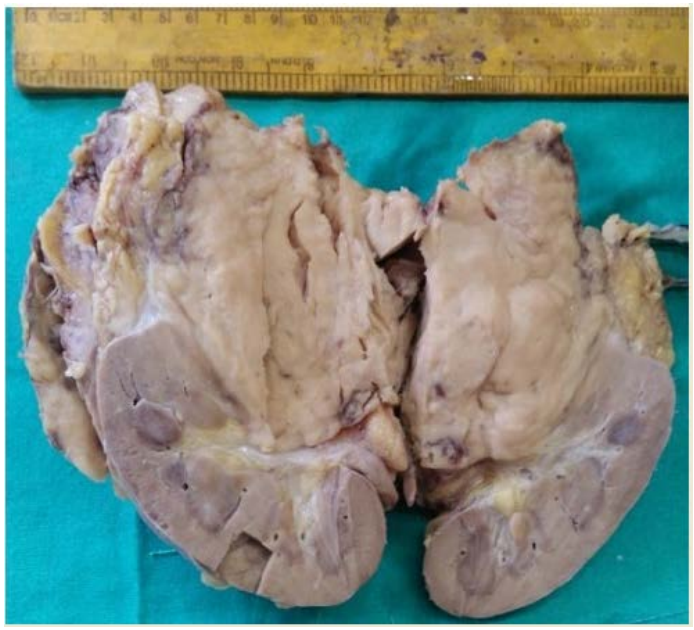

Figure 3-a: Monomorphic round cells, hyperchromatic coarse nuclei and prominent nucleoli, arranged in sheets. Hematoxyline & Eosine, 40x.

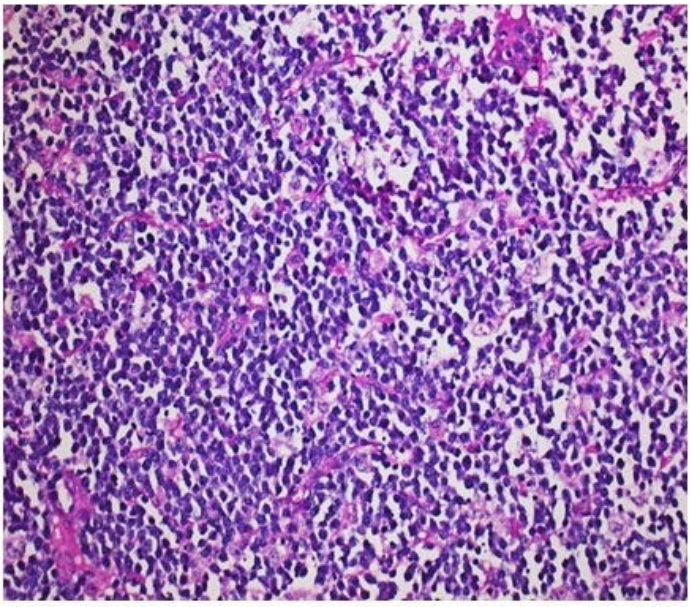

Figure 3-b: Monomorphic round cells, hyperchromatic coarse nuclei and prominent nucleoli, arranged in sheets. Hematoxyline & Eosine, 400x.

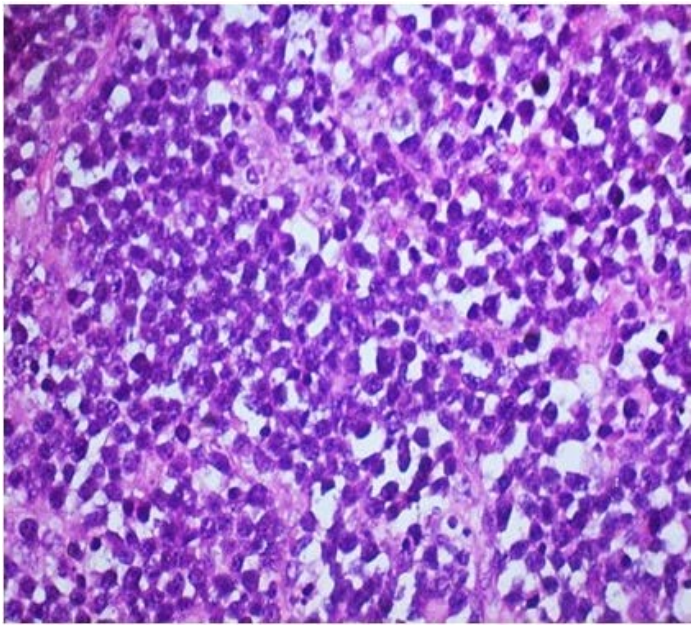

Figure: 4,a: Immunohistochemestry (IHC) Tumour cells are CD20 positive, 200x.

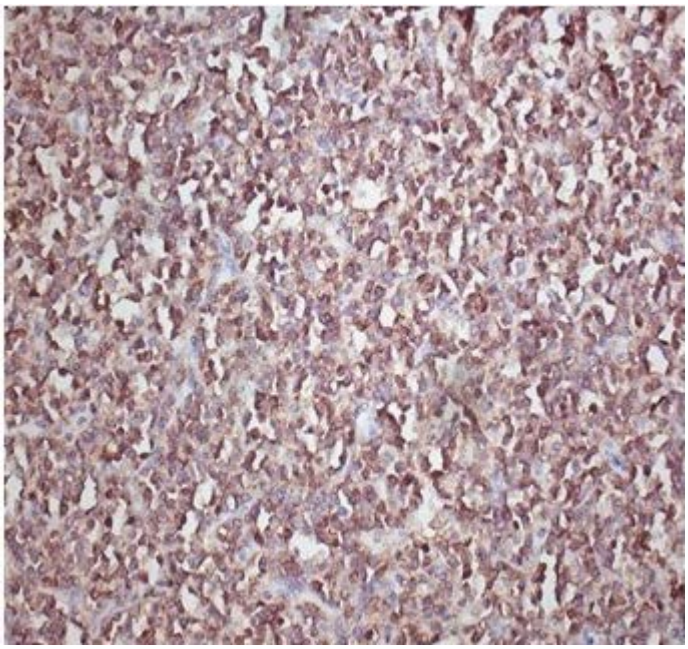

Figure: 4 - b: Tumour cells are CD38 positive, 200x

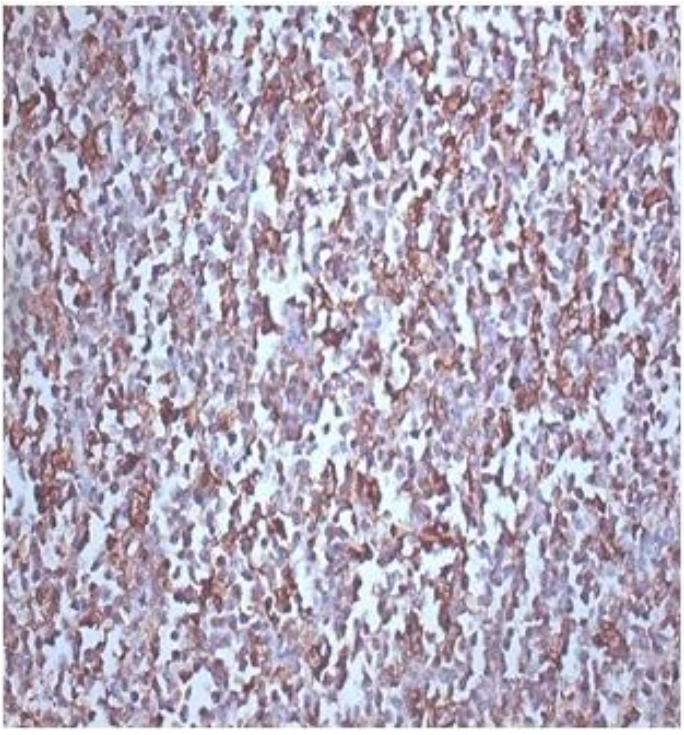

Figure: 4-c : Tumour cells are Ki67 positive, 200x.