Primary Renal Lymphoma: A Rare Cause of Renal Enlargement

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Abstract
Primary renal lymphoma (PRL) is a rare entity seen predominantly adult males. Patients usually have nonspecific manifestations and diagnosis may be delayed until they present with appreciable renal mass with recognizable clinical manifestations. Diagnosis depends upon ultrasound, CT scan (with exclusion of primary lymphoma at other site and lymph node enlargement), histopathology and immunohistochemistry. We present a case of fifty years old female who presented with dull abdominal pain and fever. On ultrasound a huge renal mass replacing the whole of the kidney was found. Ultrasound and CT scan for lymphadenopathy, organomegaly or any fluid was unremarkable. Histopathology of nephrectomy specimen received, showed replacement of renal tissue by sheets of large sized malignant lymphoid cells with vesicular nuclei. Immunohistochemistry confirmed its non epithelial origin and B-cell Non Hodgkin’s lymphoma.

Key words: Renal lymphoma, non-Hodgkin’s lymphoma, primary renal lymphoma, bilateral renal lymphoma, diffuse large B-cell lymphoma

Introduction
Primary renal lymphoma is an uncommon variant of extranodal non-Hodgkin’s lymphoma (NHL). Primary Renal lymphomas are defined as lymphomas arising in the renal parenchyma and not presenting as invasion from an adjacent lymphoid tissue. It is an uncommon site for extranodal non-Hodgkin’s lymphoma (ENL).¹ It mostly affects middle aged and elderly people and has an incidence of 0.7% among all cases of ENL.² Pathogenesis is unclear and diagnosis is often delayed due to nonspecific clinical manifestations. The proposed pathogenetic mechanisms include: origin in the subcapsular lymphatics, seeding via hematogenous route, an extension from retro peritoneal disease or inflammatory disease with a lymphoplasmacytic infiltrate.³ Renal lymphoma has insidious clinical onset and clinical manifestations usually present either as single nodule or multiple nodules. The clinical presentations include flank pain, hematuria, abdominal mass, fever and weight loss. Infrequently they present with acute renal failure. Pathological data are scanty; few reports indicate it has a very poor prognosis.⁴ Early detection therefore depends on combination of computed tomographic scan (CT scan) and examination of adequate biopsy material. MRI is currently becoming the imaging modality of choice for evaluation of renal lesions particularly differentiating renal lymphoma from renal cell carcinoma. The prognosis is very poor with median survival less than a year. The most common feature is that of multiple nodular masses. Early detection by preoperative biopsy is suggested in patients with atypical radiological features. The most common histological subtype encountered is diffuse large B cell lymphoma (DLBCL).⁵

PRL is a rare disease and only a few cases have been reported previously. We report a case of 50 years old female, whose nephrectomy specimen was received at a local laboratory.

Case Report
A large nephrectomy specimen was received at a local laboratory. The patient in whom nephrectomy was performed was 50 years old female from KPK (Khyber Pakhtun Khuwan) and she presented with abdominal pain and fever. Her ultrasound showed huge right renal mass involving whole kidney and impression on ultrasound was renal carcinoma/Renal lymphoma). There was no lymphadenopathy or organomegaly on ultrasound. Her CT abdomen was unremarkable except for this renal mass. The nephrectomy specimen which was received showed 12x11x8 cm mass replacing whole of the kidney. The mass had irregular surface. Cut surface showed a homogenous fish flesh appearance Microscopic examination showed replacement of renal tissue by homogenous population of large sized malignant lymphoid cells with vesicular nuclei. Immunohistochemistry showed that these cells were positive for CD 45 and CD 20 and negative for CD 3 and cytokeratin.

Discussion
Primary renal lymphoma is a rare lesion that represents in less than 1% of the kidney’s lesions. Renal lymphoma may be seen in conjunction with multisystemic, disseminated lymphoma or as tumor recurrence. It may also be seen as primary disease and rarely in immunocompromised
patients. Metastatic renal involvement is seen in 1/3rd of lymphomas and it is more common in NHL than HL. Burkitt’s and AIDS related lymphomas have greatest affinity for kidneys. Most primary tumors present as single or multiple nodules that usually involve the whole kidney. It usually affects adults with male preponderance. However few cases of PRL at a younger age have been reported. Many classes of PRL including small, large, intermediate and mixed cell types have been identified and neoplastic cell types may express both B and T cell markers. In our case morphologically they were diffuse large B cells and immunologically also they were positive for CD20. Rose F. et al reported 4 cases of primary renal lymphoma. Among these two were diffuse B cell type and two were NHL small B cell type. In another case reports of three cases of PRL all three cases were found to be diffuse large B cell type on histopathology and one of these cases had lymphoma in a transplanted kidney secondary to immunosuppression. Primary renal lymphoma of T cell origin are unusual but very few case reports have been reported. In 10 to 20% of cases lymphoma affects both kidneys, and few case reports of bilateral renal lymphoma are available. Rare cases of localized amyloid deposition in renal lymphoma have been reported. PRL is a rare disease hence poses difficulty in diagnosis. Diagnosis is made on the basis of exclusion of primary extra-renal disease, tissue histology and immunohistochemistry. Differential diagnosis of renal lymphoma includes pyelonephritis, inflammatory pseudotumors, transitional cell carcinoma and renal cell carcinoma. Infections can be easily ruled out. Renal neoplasms such as transitional cell carcinoma, renal cell carcinoma, nephroblastomas, neuroblastomas and neuroendocrine tumors though have characteristic morphological features, but in less typical forms may simulate renal lymphomas. Immunohistochemistry would help to differentiate such cases.

Treatment of PRL remains controversial because pathogenetic mechanisms are uncertain. It has been established that most cases of PRL are high-grade lymphomas mainly of B-cell phenotype, they are treated by chemotherapy with or without radiotherapy. However, results are unsatisfactory due to rapid progression of disease and treatment resistance. Recently, rituximab has been added and is expected to improve the outcome of PRL of B-cell origin.

**Conclusion**

Primary renal lymphoma is a rare entity but should be kept in mind in differential diagnosis of isolated renal mass (even in young patients), to ensure early and proper chemotherapy.
References


