An unusual presentation of primary renal lymphoma: A case report

Darshil Shah, Neelakandan R, Chandru T, Natarajan K

From Department of Urology and Renal Transplantation, Sri Ramachandra Medical College, Sri Ramachandra Nagar, Porur, Chennai, Tamil Nadu, India.

Correspondence to: Dr. Darshil Shah, Department of Urology and Renal Transplantation, Sri Ramachandra Medical College, Sri Ramachandra Nagar, Porur, Chennai-600016, Tamil Nadu, India. E-mail: darshil_27_3@yahoo.co.in

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ABSTRACT

Lymphomatous involvement of the kidney is often seen as a part of the disseminated disease but primary renal lymphoma is very rare. It is essential to differentiate between renal cell carcinoma and renal lymphoma in patients presenting with renal masses. The prognosis is usually poor with a median survival less than a year. We present the case of a 81-year-old male who presented with type B symptoms and was diagnosed to have primary lymphoma of the kidney and discuss briefly about the primary renal lymphoma.

Keywords: *Lymphoma, Renal mass, Non-Hodgkin's lymphoma.*

Primary renal lymphoma (PRL) is a Non-Hodgkin's lymphoma (NHL) involving the kidney in the absence of primarily extra-renal lymphatic disease. As the kidney is an extranodal organ and does not contain lymphatic tissue, PRL is rare. PRL has been shown to account for 0.7% of all extranodal lymphomas in North America and 0.1% of all malignant lymphomas in Japan [1]. No more than 70 cases of PRL have been reported in the literature and the majority are of diffuse large B cell lymphoma.

The precise cause of PRL remains unknown. It has been suggested that PRL originates from the renal capsule and infiltrates the renal parenchyma [2]. Another explanation is that chronic inflammatory conditions of the kidney attract the infiltration of lymphoid cells and eventually evolve into lymphoma. Clinicoradiologically, it may mimic renal cell carcinoma. The prognosis is very poor with a median survival of less than a year. PRL is often considered a systemic disease manifesting initially in the kidneys. Imaging plays an important role in PRL. The most common feature of imaging is that of multiple nodular masses. A preoperative biopsy is worthwhile in patients with atypical radiological features, since it may avoid nephrectomy. The treatment of non-Hodgkin's Lymphoma (NHL) has been revolutionized with the addition of Rituximab to the standard chemotherapy.

Here, we present the case of a present an 81-year-old male who presented with type B symptoms and was diagnosed to have primary lymphoma of the kidney. We report this case as the presentation of the disease is very late with non-specific complaints and radiological features that are suggestive of RCC. Hence, an accurate diagnosis of the entity is very important.

CASE REPORT

An 81-years-old male presented to the Urology OPD with complaints of low-grade fever for 1 month which was associated with generalized weakness. No comorbidities or no surgical history was present. In the family, patient's brother had a history of treatment taken for lymphoma in the past. On examination, no abnormality detected and the vitals were stable (pulse – 76/min, blood pressure- 134/78 mmHg and respiratory rate 14/min).

Complete blood counts (hemoglobin 11.2 gm/dl, total leukocyte counts – 6700/mm^3), renal (serum creatinine 0.8 mg/dl) and liver function tests (total bilirubin - 0.8 mg/dl, serum glutamic oxaloacetic transaminase [SGOT] - 24 IU/dl, serum glutamic pyruvic transaminase [SGPT] - 29 IU/dl) were within normal limits. Serum Lactate Dehydrogenase(LDH) level was 411 U/L, which was significantly higher.

Contrast-enhanced computed tomography (CECT) abdomen showed a soft tissue lobulated mass superior to the right kidney measuring 9x7 cm, abutting the right renal vein. The left kidney was diffusely bulky and lobulated measuring 10x9x14 cm with heterogeneously enhancing soft tissue density replacing the left renal parenchyma with the largest component measuring 6x8x9cm in the posterior aspect of mid-pole (Fig. 1).

Fluorodeoxyglucose Positron emission tomography (FDG PET) showed FDG avid mass lesions involving the right suprarenal region and the left kidney (Fig. 2). On suspecting metastatic renal cell carcinoma (RCC), we performed CT-guided core needle biopsy of the left renal mass which revealed NHL diffused large B cell lymphoma with positivity for CD20, CD 79a, CD 45 and Vimentin with Ki 67 labelling index of 65% (Fig. 3).

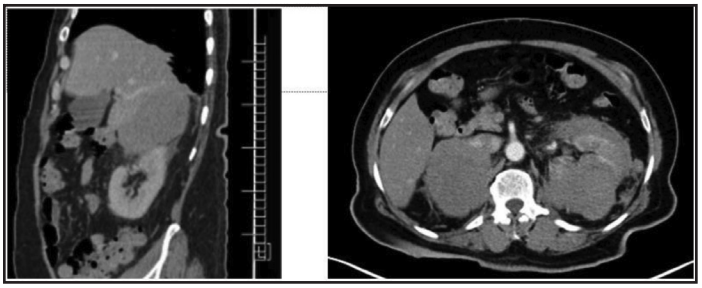

Figure 1: CECT abdomen: soft tissue lobulated mass superior to the right kidney measuring 9x7 cm abutting the right renal vein, left kidney was diffusely bulky and lobulated measuring 10x9x14 cm with heterogeneously enhancing soft tissue density replacing the left renal parenchyma with largest component measuring 6x8x9 cm in the posterior aspect of mid pole.

Hence, it is a case of suspected RCC metastasis to the kidney, which turned out to be Primary renal lymphoma as diagnosed by percutaneous biopsy. The patient was advised chemotherapy with Cyclophosphamide, Doxorubicin, Vincristine, prednisone (CHOP) and Rituximab. Chemotherapy gave very good results without much morbidity. The patient is doing well on follow-up.

DISCUSSION

The kidney is not a lymphoid organ; hence, the very existence of lymphomas of the kidney was questioned by some investigators. The proposed pathogenic mechanisms include seeding via hematogenous route, origin in the sub-capsular lymphatics, an extension from retroperitoneal disease or inflammatory disease with a lymphoplasmacytic infiltrate [3]. Primary renal NHL is defined as an NHL arising in the renal parenchyma and not an invasion from an adjacent lymphomatous mass.

PRL usually affects adults. The clinical presentations include flank pain, hematuria, abdominal mass, fever and weight loss [1,4]. Acute renal failure is also a rare clinical presentation described in the literature [5,6]. The role of imaging is very crucial for diagnosis. The most commonly encountered pattern is that of multiple soft tissue masses or diffused renal mass with minimal enhancement after contrast compared to surrounding renal parenchyma [7].

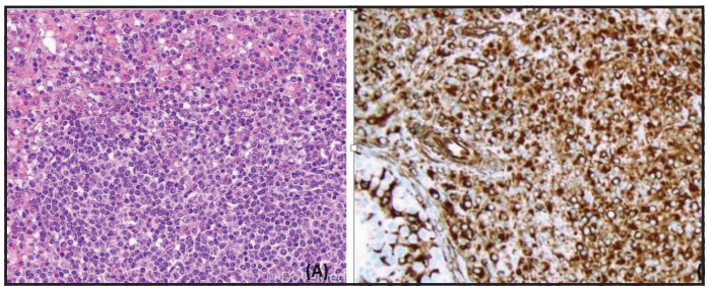

Figure 3: Histopathology revealed NHL diffused large B cell lymphoma with positivity for CD20, CD 79a, CD 45 and Vimentin with Ki 67 labelling index of 65%.

Solitary lesion makes it a difficult diagnosis as it resembles the more common RCC [8]. The differentiating features include an absence of calcification, post-contrast homogenous attenuation, absence of renal vein thrombus and absence of a mass effect on renal vessels and pelvicalyceal system in PRL. However, this situation still demands a biopsy to rule out RCC. Other less commonly seen patterns include enlarged non-enhancing kidneys, direct invasion of renal sinus and hilum by bulky retroperitoneal mass or a diffuse perirenal infiltration encasing the kidney. Most of the patients also have adjacent retroperitoneal adenopathy [9]. MRI is currently becoming the imaging modality of choice for evaluation of renal lesions. Lower signal intensity on unenhanced T1-weighted images than normal renal cortex and less enhancement on early gadolinium-enhanced images differentiates Renal Lymphoma from RCC.

Diffuse Large B Cell Lymphoma (DLBCL) is the most common histology though encountering a follicular lymphoma, small lymphocytic lymphoma or MALToma is not unusual [10]. The prognosis is reported poor universally. Median survival is less than a year. PRL is considered as a systemic disease, presenting with renal manifestation. However, nephrectomy can be avoided if a preoperative diagnosis is made. Patients with atypical features of RCC, therefore, should undergo a preoperative percutaneous renal biopsy. The sensitivity and specificity of renal biopsy are 70% to 92% and 100%, respectively, with accuracy close to 90%. The treatment of renal lymphoma depends on the primary histological subtype. The addition of Rituximab to the standard CHOP chemotherapy may improve the dismal outcome reported so far.

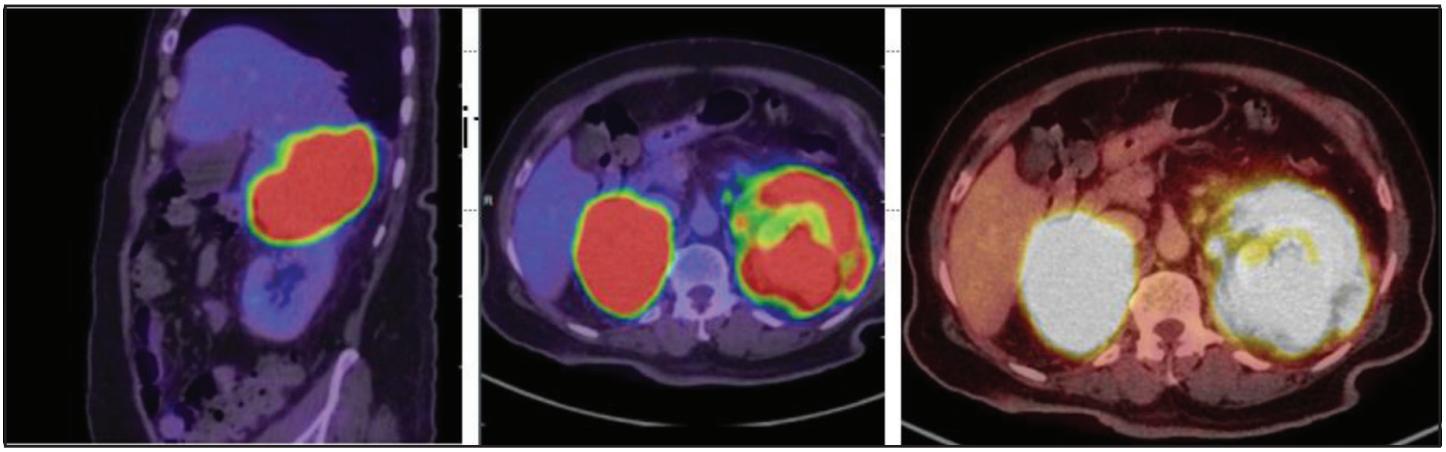

Figure 2: FDG PET: FDG avid mass lesions involving the right suprarenal region and the left kidney.

CONCLUSION

Primary renal lymphoma can radiologically and clinically mimic renal cell carcinoma. Making a preoperative diagnosis with biopsy can avoid unnecessary nephrectomies and initiate chemotherapy at the earliest.

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