# Primary Perinephric Lymphoma.

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#### Citation

P Joshi, J Date, S Shivde, J Rakesh, J Singh, L Raja, G Devashree. *Primary Perinephric Lymphoma*.. The Internet Journal of Surgery. 2009 Volume 24 Number 1.

#### **Abstract**

The differential diagnosis of masses in the perinephric space and the peripelvic space of the renal hilum would include tumor, fluid, inflammation, or various proliferative diseases. More broadly, abnormal tissue at the periphery of the kidney may be subcapsular or perinephric in origin, and these processes are not always distinguishable. Radiologic evaluation of renal and perinephric masses is gaining an increasing importance. We report one such case of a right perinephric mass which posed a diagnostic dilemma.

#### INTRODUCTION

Masses in the perinephric space including the peripelvic space of the renal hilum may be due to tumor, fluid, inflammation or various proliferative diseases. Abnormal tissue at the periphery of the kidney may be subcapsular or perinephric in origin and these processes are not always distinguishable. Radiologic evaluation of renal and perinephric masses is important. More than half the renal malignancies are discovered incidentally during imaging performed for unrelated reasons. <sup>[1]</sup> In cases of perinephric lesions, if precise pre operative diagnosis by imaging is not possible, a biopsy or surgical exploration remains the mainstay in diagnosis and management.

## **CASE**

A 72-year-old non-diabetic male patient came with history of right flank pain and fever for 3 weeks. On clinical examination there was a tender lump in the right hypochondrium. Ultrasonography revealed upper hydroureter and hydronephrosis with a perirenal lesion of mixed ecogenicity suggestive of infective/inflammatory process such as abscess.

A contrast enhanced CT scan of the abdomen was performed which revealed a right perinephric lesion encasing the kidney with inhomogeneous enhancement (figure 1, 2 & 3). It was mildly vascular and showed encasement of upper ureter and renal vessels. There was proximal hydroureter with hydronephrosis. It was thought to be a perirenal hematoma. However, the possibility of an inflammatory lesion could not be ruled out.

Figure 1: CT Scan

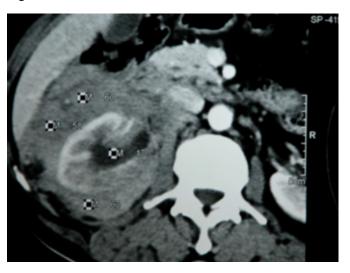
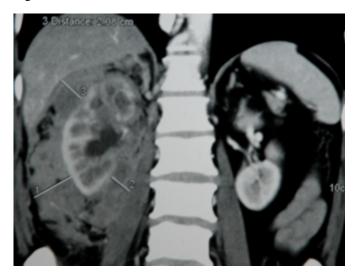
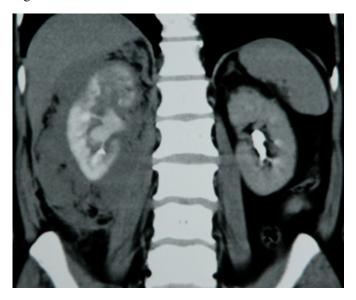


Figure 2: CT Scan

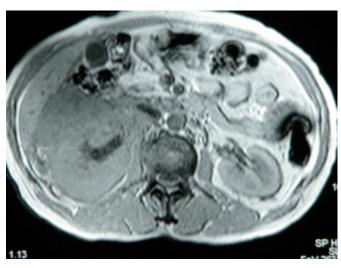


**Figure 3**Figure 3: CT Scan

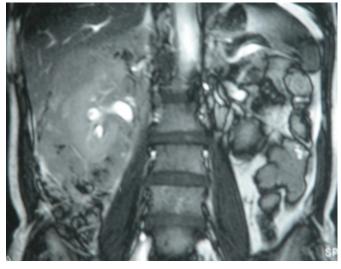


An MRI scan was done using a T1, T2 & True FISP protocol .It was done to differentiate between inflammatory lesion and hematoma (figure 4 & 5). It revealed a mass lesion which was isointense on T1 and hyperintense on T2. There was neither a renal space-occupying lesion nor any abdominal lymphadenopathy. It strongly suggested the mass to be of inflammatory origin.

Figure 4: T1 MRI



**Figure 5**Figure 5: True FISP protocol MRI.



A right ureteral stent was inserted for the upper tract obstruction. As the lesion was uncharacteristic on all the imaging modalities and the patient remained symptomatic with fever and pain inspite of stenting, it was decided to explore the patient. The plan was to characterize the lesion and sample the tissue for microbiology and pathology tests. Intraoperatively, the entire kidney was seen encased by dense tissue up to the upper ureter. Upper ureterolysis was done & biopsy of the lesion was sent for frozen section. Both the frozen and final histopathology reported the lesion as perinephric lymphoma (Non-Hodgkin's variety) (figure 6). The patient was put on a four-drug chemotherapy regime viz CHOP (cyclophosphomide, doxorubicin, vincristine, prednisolone).

#### DISCUSSION

The spaces around the kidney can be divided in subcapsular and perinephric spaces. A variety of conditions like tumor, fluid, inflammation or infilterative disease may involve the above spaces with potentially overlapping radiologic features. Close attention to knowledge of the associated clinical and imaging features can facilitate a confident and specific diagnosis in many cases. However, in cases where radiologic imaging remains inconclusive as in our case, histopathology examination may be the only method to make the final diagnosis and guide further management.

Renal involvement is found in 15–50% of patients with lymphoma at autopsy. The characteristic features of lymphoma can usually be documented only in 3-8% of patients at CT staging<sup>[2,3]</sup>. Perinephric lymphoma is one of the described patterns of renal involvement. It is typically homogeneous, hypovascular, mildly enhancing, and associated with non-obstructive encasement of retroperitoneal vessels.<sup>(4)</sup>

# CONTENTS OF THE PERINEPHRIC SPACE (PRS)

Kidney, adrenal, renal vasculature, upper ureter, lymphatic networks and fibrofatty tissue are the contents of the PRS.

#### WHY PERINEPHRIC?

As per anatomic description, each of the paired perirenal spaces is an inverted cone of tissue that lies lateral to the lumbar spine. It is confined anteriorly by the anterior renal fascia (ARF/Gerota fascia/fascia of Toldt) and posteriorly by the posterior renal fascia (PRF/fascia of Zuckerkandl). Laterally these two fasciae fuse to form the lateroconal fascia. Superiorly, its extent is open to the bare area of the liver<sup>(2)</sup>. Inferiorly, the ARF and PRF gradually converge towards a point about 8 cm inferior to the lower pole of the kidney.

Our patient had lymphoma tissue wrapping the kidney and non-obstructive involvement of renal vessels. The upper ureter was also encased causing hydroureter. The pathology was confined to the defined perinephric space as described by Bechtold et al.<sup>(2)</sup>

## WHY DO WE CALL THIS "PRIMARY"?

About 25-40 % of Non-Hodgkin's lymphoma present with a primary extranodal lymphoma. Numerous papers dealing with extranodal Non-Hodgkin's Lymphoma arising from almost every organ in the body have been published. <sup>(5)</sup>In our case, this patient had pure involvement of perinephric tissue without renal parenchymal or lymphnodal involvement, hence it is called "primary".

So we present a case of a primary extranodal Non-Hodgkin's Lymphoma involving only the perinephric space. Contemporary imaging modalities such as CT scan and MRI failed to characterize this isolated lesion in the absence of typical multiple systemic features of lymphoma.

As per the review of literature, this is the first case to be reported having primary perinephric lymphoma without any systemic or local nodal involvement.

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