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RESEARCH ARTICLE

AN ADULT FEMALE PRESENTING WITH ON/OFF RIGHT FLANK PAIN FOR 3 YEARS, INCIDENTALLY,DIAGNOSED WITH PRIMARY RENAL LYMPHOMA: A CASE REPORT

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Abstract

Primary renal lymphoma (PRL) is a rare subtype of non- Hodgkin's lymphoma which represents less than 1% of all renal masses. clinical presentation of PRL is usually non-specific which makes it of a clinical significance. Accurate and early identification of PRL and treatment are imperative to achieve improved outcomes. In this paper, we present a case of a primary renal lymphoma and discusspathophysiology, clinical presentation, imaging characteristics, management, and prognosis.

Introduction:-

Primary renal lymphoma (PRL) is defined as a non- Hodgkin's lymphoma with isolated renal parenchymal involvementand absence of extensive nodal disease[i]. PRL is a rare subtype of both NHL and all renal masses, with incidence representing less than 0.7% and 1% respectively[ii]. The exact pathophysiology of PRL remains unclear, given that the renal parenchyma lacks the lymphatic tissue, however several hypotheses being suggested to explain the etiology of PRL including direct invasion or hematogenous spread[ii]. The clinical presentation of PRL is usually non-specific and can either be genitourinary symptoms or constitutional symptoms of Lymphoma[iii]. The goal standard management of PRL typically includes radical nephrectomy and chemotherapy[i]. PRL is known to be an aggressive tumor with a poor prognosis[ii]. Our case is unique in form that patient had been complaining of symptomatic poor functioning right kidney refusing surgical management of nephrectomy for 3 years with no metastasis and a good health condition.

Case presentation:

Our patient is a45-Year-oldlady, medically free, admitted to our urology department through emergency with on and offright flank pain, radiating to lower abdomen associated with gross hematuria for 7 days for further investigations.

She had been first evaluated in another healthcare facility, 3 years prior to her presentation to our hospital for symptomatic poor functioning right kidney,as evident on renogram, and she was offered right nephrectomy. However, patient refused the option of nephrectomy, and instead was managed conservatively.

At the time of admission, patient was also complaining of lower urinary tract symptoms in form of frequency, urgency and urge incontinence, associated with dysuria. No other associated systematic symptoms.

Her Physical examination was unremarkable. Her laboratory investigations were as follows: hemoglobin 127 g/L, White blood count 6.6 ×109/L,platelets x109/L, serum creatinine 69. No abnormality in coagulation profiles, serum

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electrolytes levels or liver function test, and lactate dehydrogenase. Urine examination and analysis revealed blood (moderate), red blood cells (>30/hpf), bacteria +1, Urine culture showed bacterial growth of Escherichia Coli.

An enhanced Computed tomography (CT) Urogram scan showing multiple mild enhancing infiltrative mass lesions in the right kidney. These lesions are involving the cortex as well the pelvicalyceal system of the right kidney and are associated with faint and delayed contrast excretion from the right kidney (Figure1), metastatic work up was unremarkable for metastasis.

So Patient was managed with Right radical Nephroureterectomy with bladder cuff excision and retrocaval LN Excision.Post-operativehistopathologyindicateddiffuse large B-cell lymphoma germinal center subtype in kidney and ureter, reactive lymph nodes negative for lymphomatous infiltration (Figure 2).

Case was discussed in tumor board meeting and patient were referred to other hospital for adjuvant chemotherapy.

Discussion:-

Primary renal lymphoma (PRL) is a rare subtype of non-Hodgkin's lymphoma with Diffuse large B-cell lymphoma (DLBCL) being the most common histological subtype of PRL[iv].PRLis defined as a non- Hodgkin's lymphoma with isolated renal parenchymal involvementand absence of extensive nodal disease[i]. PRL represent about 0.7% of NHL, and 1% of all renal masses identified[ii], with less than 100 cases being reported worldwide to our knowledge[iii] with male predominance of 2:1 ratio[iv].

The rare incidence of PRL is believed to be due to the lack of lymphatics in the renal parenchyma[iv], and the pathophysiology remains unclear of how PRL originates in the renal parenchyma. Several hypothesis have been suggested to explain the etiology of PRL, including direct invasion through lymphatic vessels found in the renal capsule to the renal parenchyma, or through hematogenous spread, which may explain the bilaterality of PRL[ii].

Approximately, 50% of patients with PRL are asymptomatic and discovered incidentally as renal mass, others may present with flank pain, hematuria, abnormal renal functions, or constitutional symptoms of lymphoma, such as fever, and weight loss[i]. In our case the patient presented with on and off right flank pain, radiating to lower abdomen associated with gross hematuria.

In PRL, dissemination to extrarenal sites is common and confers a poor prognosis[iii].

Accurate and early identification of PRL is critical, owing to the aggressive nature of the disease and different management options needed for treatment than other types of renal tumors. Stallone proposed three rigid diagnostic criteria for PRL, which remains in use: (i) lymphomatous renal infiltration; (ii) non-obstructive unilateral or bilateral kidney enlargement; and (iii) no extrarenal localization at the time of diagnosis.

Imaging features of PRL include hypovascularity and low enhancing lesion on computed tomography urogram (CTU), single or multifocal nodules, or diffuse renal enlargement[iii].

Management of unilateral PRL typically includes nephrectomy like renal cell carcinoma, however, in case of bilateral PRL, treatment with systemic chemotherapy using an R-CHOP regimen is the treatment of choice[i].

Neoadjuvant chemotherapy for unilateral PRL before nephrectomy is essential and thought to improve renal function within 2–4 weeks of initiating therapy[ii]. Unfortunately, our patient did not receive neoadjuvant chemotherapy, owing to her atypical presentation and localized disease over 3 years duration, despite the aggressive behavior of PRL. She underwent right radical Nephroureterectomy with bladder cuff excision and retrocaval LN Excision.

Although management options for PRL have improved, PRL still carries poor prognosis and the 1-year mortality rates can be as high as 75%. [ii]

Conclusion:-

Although PRL is a rare renal tumor, it must not be overlooked from differential diagnoses of renal tumors, and strict diagnostic criteria should be established for accurate and early diagnosis, management andfollowupof PRL.

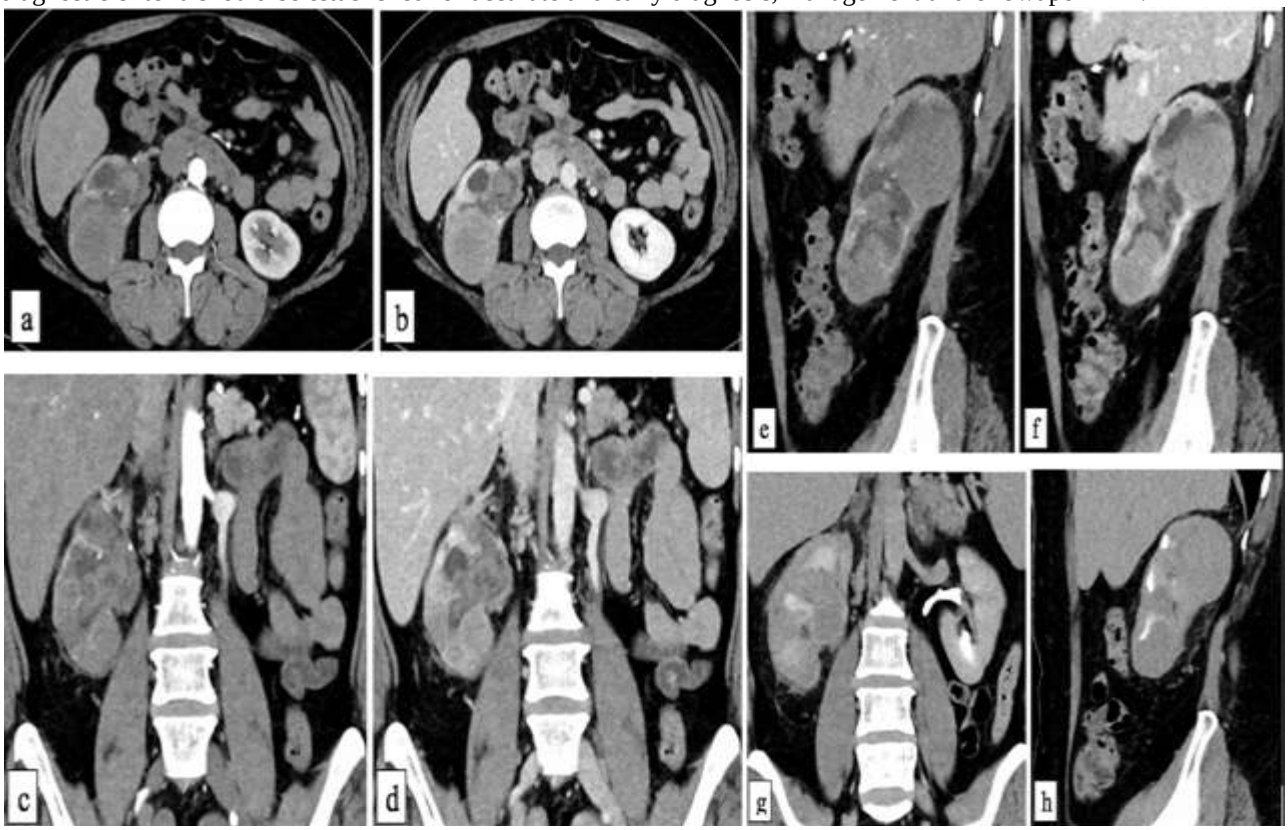

Figure 1:- Axial (a=arterial phase, b=venous phase), coronal (c=arterial phase, d=venous phase), sagittal (e=arterial phase, f=venous phase), coronal (g=delayed urography phase) and sagittal (h= delayed urography phase) images from the contrast enhanced CT abdomen showing multiple mildly enhancing infiltrative mass lesions in the right kidney. These lesions are involving the cortex as well the pelvicalyceal system of the right kidney and are associated with faint and delayed contrast excretion from the right kidney. No significant retroperitoneal lymphadenopathy is seen. Right renal vein and inferior vena cava (not shown) are patent.

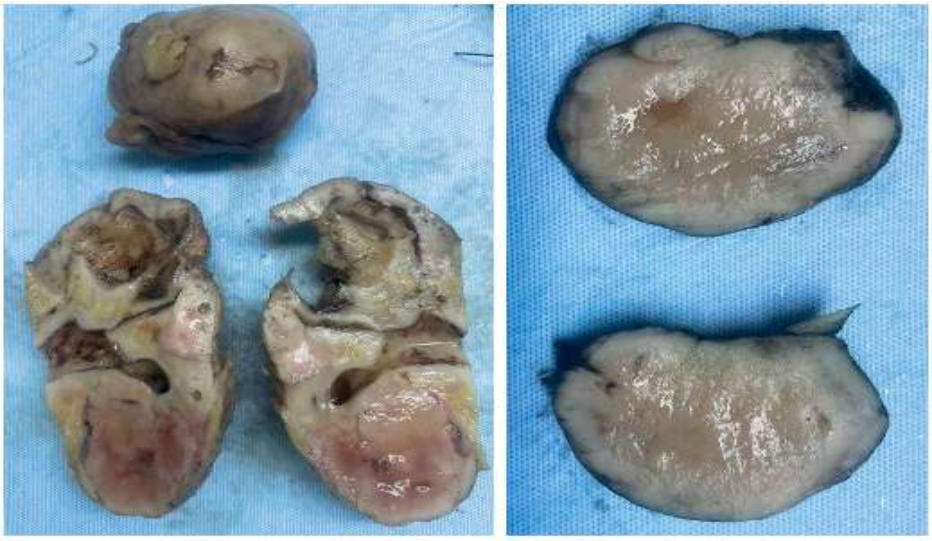

Figure 2:- Gross section of the kidney showing fleshy ill defined gray nodules.

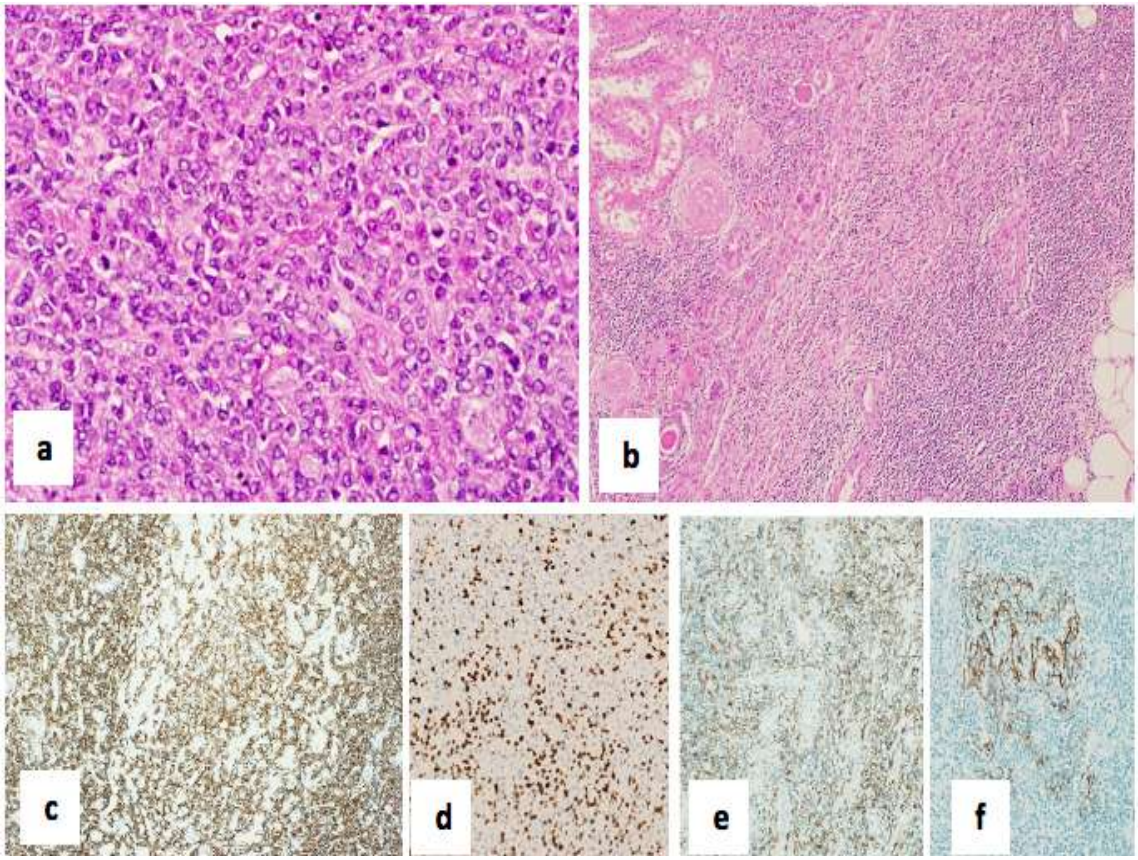

Figure 3:- Large B cell lymphoma of the kidney , developed in top of marginal zone lymphoma.
a: HE x 400: monotonous large lymphomatous cell proliferation. b- HE x 200: small cell proliferation showing centrocyte –like cellsc- the large and small cells express diffusely and strongly CD20.d- Ki67 labelling index in high in the large cell proliferation(70%). e- The small cell proliferation marginal zone type expresses CD43. f- CD21 highlights the infiltration of the residual follicular germ center by the small cell.

Guarantor of submission:
The corresponding author is the guarantor of submission.

Source of Support:
None.

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Consent statement
Written consent was obtained from the patient for publication of this case report.

Conflict of interest:
Authors declare no conflict of interest.

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