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CASE OF THE WEEK 9

CLINICAL HISTORY:

A 23 year unmarried woman of Asian origin, not a known hypertensive or diabetic, presented with feeling of generalized weakness and occasional joint pains since 6 months. She also complained of oral ulcers and reduced urinary output since last one week. There was no history of preceding febrile illness or gross hematuria.

EXAMINATION:

BP 150/90 mmHg, Systemic examination- unremarkable

INVESTIGATIONS:

Urine – Albumin trace, RBC- nil, WBC 10-12/hpf.
Haematological profile -Haemoglobin 8.2 g% (82 g/L), Total leukocyte count 16,400/ mm3, ESR 90 mm/ 1st hour
Urea: 80 mg% (28.46 mmol/L), Creatinine 1.9 mg% (167.96 µmol/L)
ANA- positive ,anti dsDNA- positive
pANCA, cANCA, anti GBM antibodies, ASO titres- negative.
Serum C3 & C4 levels: WNL, HBsAg/HIV/HCV- negative
USG abdomen: Bilateral enlarged kidneys (13.9 &14 cms), bilateral hydronephrosis. No calculi or obstructive lesions noted.

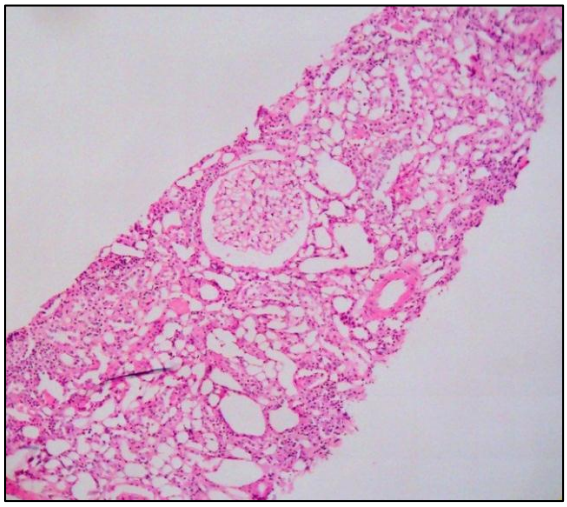
Clinical impression: Systemic Lupus Erythematosus (SLE) with? nephritis

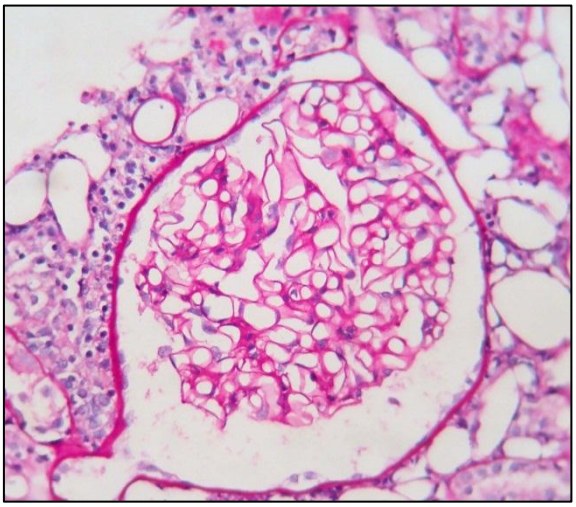
In view of clinical & serological evidence of SLE, and renal dysfunction with enlarged kidney size, a renal biopsy was performed

DIF studies included 6 glomeruli showing segmental mesangial staining for IgM, and were negative for IgA, IgG,C3,C1q and kappa & lambda light chains.

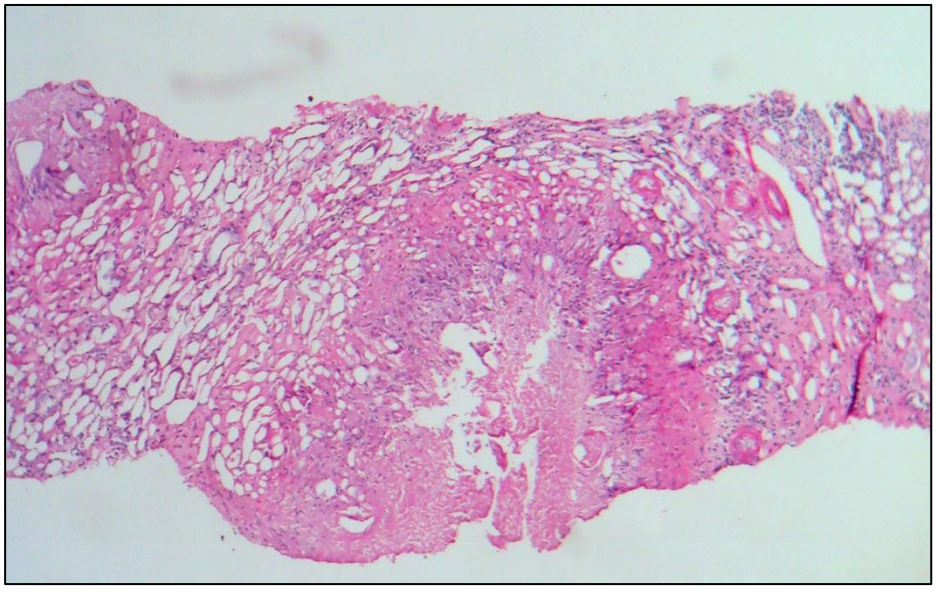
MICROSCOPY

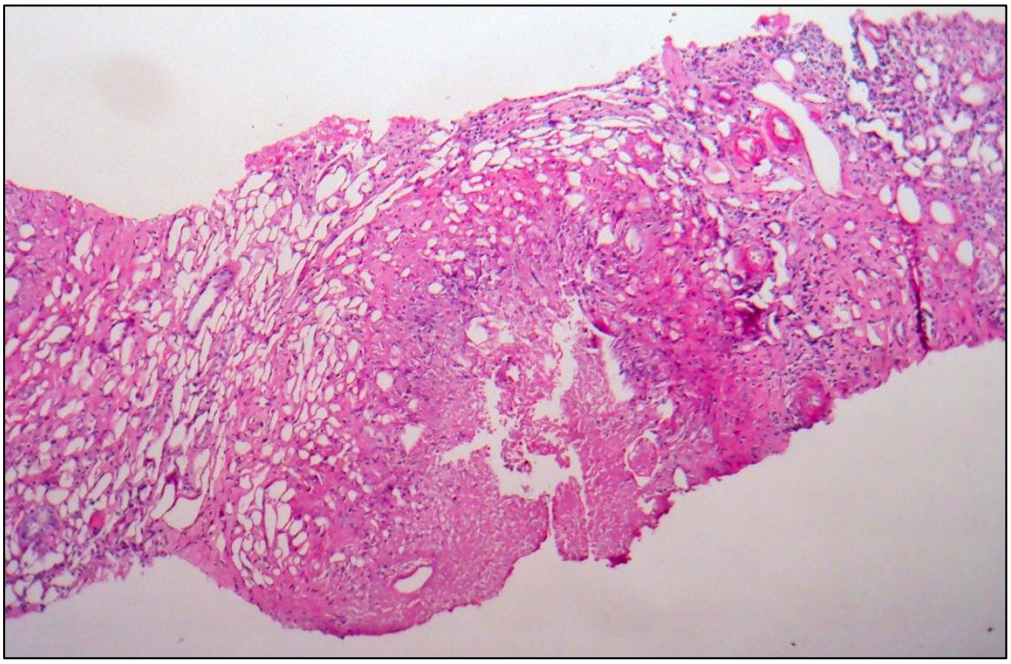
An adequate renal biopsy including both renal medulla and cortical parenchymal tissue was received. Upto 13 glomeruli were seen, none globally sclerosed. The glomeruli appeared relatively unremarkable, without evidence of proliferative activity, subendothelial deposits, crescent formation, capillary wall thickening or tuft necrosis.

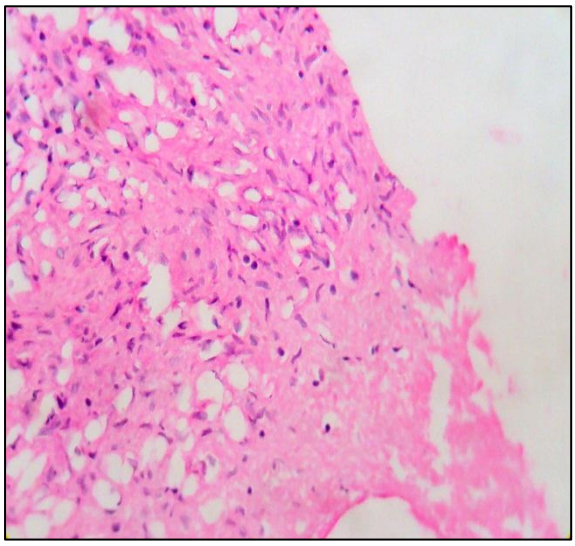


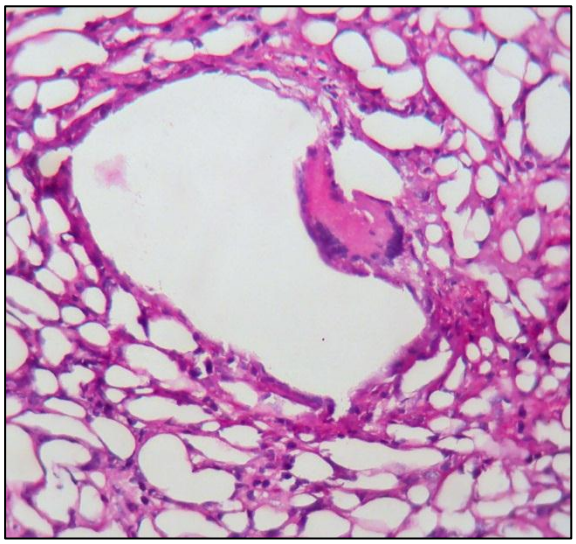


The interstitum showed several foci of chronic (lymphoplasmacytic) inflammation and few discrete areas of necrotizing epithelioid cell granulomas with focal giant cell reaction. The necrotic foci were unrelated to vascular structures in the multiple step sections examined. Stain for AFB was negative

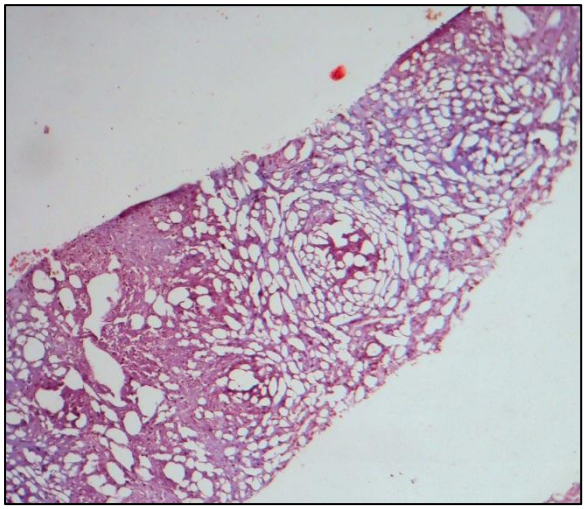


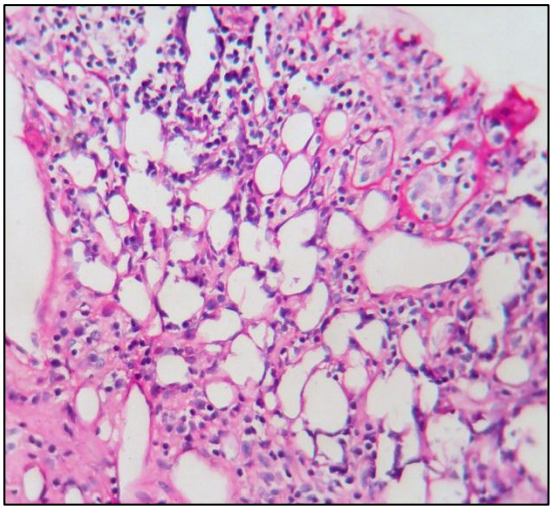






Additionally, the entire interstitium showed a “foamy” appearance on low magnification images. On closer inspection it was evident that many of these “ foamy” spaces showed peripherally located nuclei (adipocytes). Thus, a near diffuse infiltration of renal interstitium by adipocytes, widely replacing the nomal interstitial parenchyma (renal replacement lipomatosis) was seen. The adipocytes were also seen infiltrating in the foci of necrotizing granulomatous inflammation. Blood vessels were unremarkable.





FINAL DIAGNOSIS:

1. Necrotizing granulomatous tubulointerstitial nephritis

2. Renal replacement lipomatosis

3. No evidence of renal involvement in SLE

FOLLOW UP:

- CT scan confirmed the replacement of renal parenchyma by fat , which was also noted in the renal sinus and perinephric areas in a charactersitic distribution.
- Systemic evaluation showed small cervical and axillary lymph nodes. FNAC was perfromed which showed necrotizing granulomatous lymphadenitis. AFB was positive in FNAC from axillary lymph nodes.

DISCUSSION:

Renal replacement lipomatosis (RRL) , renal sinus lipomatosis and fibrolipomatosis are terms used interchangeably to denote conditions of varying severity where normal renal sinus and perirenal fat increases in amount and leads to variable replacement of renal parenchyma [1]. *Peacock & Balle* [2] were among the first one to describe this entity and according to many authors, these conditions represent a spectrum of changes with renal sinus lipomatosis being the mildest (and most common) form and RRL the other extreme where a significant proportion of renal parenchyma is replaced by fat .

Renal infiltration by fat (usually sinus lipomatosis) has been seen in the sixth or the seventh decade accompanied by atrophy of normal renal parenchyma (senile atrophy). This probably has no clinical significance. Similar phenomenon has also been observed in association with obesity and atherosclerosis. Majority of cases of RRL are associated with renal stones, infections including tuberculosis and rarely renal infarction [3-5]. Few cases have no apparent identifiable cause and are labelled idiopathic (idiopathic RRL). Cases co existing with xanthogranulomatous pyelonephritis and occurring in the renal allograft have also been described [6-8].

Radiological features of RRL are characteristic. Ultrasonography features suggestive of RRL include an echogenic mass representing the lipomatous tissue with central high-density echoes with distal acoustic shadowing if a calculus is present. Renal size is usually enlarged but may be small in advanced cases with abundant renal fibrosis. Computed tomography (CT) demonstrates a lipomatous mass, with negative attenuation values replacing renal parenchyma. Magnetic resonance imaging (MRI) has high sensitivity and specificity to identify fat as hyperintense signal on T1 and T2 weighted images. Although it is not as good as computed tomography in the demonstration of calculus, it is superior for the demonstration of the extent of the disease. [9-13]

Present case is a unique clinical presentation in a patient with SLE, clinically suspicious for lupus nephritis but detected with necrotizing granulomatous tubulointerstitial nephritis and extensive renal replacement lipomatosis on renal biopsy. There was no evidence of renal involvement in SLE, at least in this point of time.

Key to diagnosis is awareness regarding this entity amongst renal pathologists. The diagnosis can be challenging in cases where radiological investigations have not been performed and in unusual clinical situations as the present one. As the patient was diagnosed with tuberculosis during later investigations, the necrotizing granulomatous tubulointerstitial nephritis and RRL noted in the biopsy also likely to have a common etiological link! Emphysematous pyelonephritis can bear a superficial resemblance to RRL, however closer inspection revealing the peripheral nuclei in adipocytes and the clinical scenario (patients with emphysematous pyelonephritis are generally very ill) will lead to correct diagnosis in most cases.

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