

RENAL REPLACEMENT LIPOMATOSIS- A CASE REPORT

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PRESENTATION OF CASE

A non-obese 45 yrs. female patient presented with complaints of right loin swelling and pain on & off for 5 yrs. Physical examination revealed tenderness in right lumbar region and no palpable mass. External Genitalia was normal. Investigations like Haemogram, urea, creatinine, urine examination were within normal limits. Ultrasound-Abdomen showed multiple renal calculi, right kidney poor window, left kidney was normal. CECT Abdomen suggestive of Right Renal Replacement lipomatosis with multiple renal calculi present, left kidney was normal. GeneXpert MTB/RIF PCR was not detected in the urine. Radioisotope Renogram with GFR (Figure 1) showed poorly functioning right kidney & normal left kidney, then patient underwent right open nephrectomy.

Renal replacement lipomatosis is a rare benign condition that is characterized by marked proliferation of renal sinus/hilar and perirenal fatty tissue with marked atrophy of renal parenchyma. It is associated with long standing inflammation and calculi. Herein we report a case of 45 yrs. female presented with right flank mass and pain on and off. Investigations showed non-functioning right kidney, subsequently underwent open nephrectomy.

Renal lipomatosis was first described by Kurtzmann in 1931 as reported by Peacock and Balle.¹ Renal sinus lipomatosis, replacement lipomatosis, and fibro-lipomatosis of the kidney are the terms used for this condition and represent a spectrum of changes.² This spectrum is different from renal lipomas that are neoplastic, whereas this condition is thought to be a degenerative condition. Renal Replacement lipomatosis is a rare benign condition that is characterized by marked proliferation of renal sinus / hilar and perirenal fatty tissue with marked atrophy of renal parenchyma.³

It is associated with long standing inflammation and calculi it is usually unilateral. Replacement lipomatosis of the kidney is the end result of severe atrophy of renal

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parenchyma. The clinical importance of RRL is that it simulates fat-containing tumours in the kidney or its vicinity. Clinical presentation, radiological features and pathological findings aid in confirming the diagnosis.

PATHOLOGICAL DISCUSSION

Gross Examination

Received Right nephrectomy specimen measuring 6 x 4 x 2 cms with irregular shrunken kidney and increased peri renal fat. (Figure 2) Cut section was grey yellow to grey brown with No corticomedullary differentiation.

Microscopic Examination

H & E stained multiple s section studied shows renal parenchyma with few atrophic tubules and thyroidisation, hyalinised & thickened blood vessels and replaced by lobules of mature adipose tissue (Figure 3 & 4). Peri nephric fat also shows numerous congested blood vessels – Renal Replacement Lipomatosis.

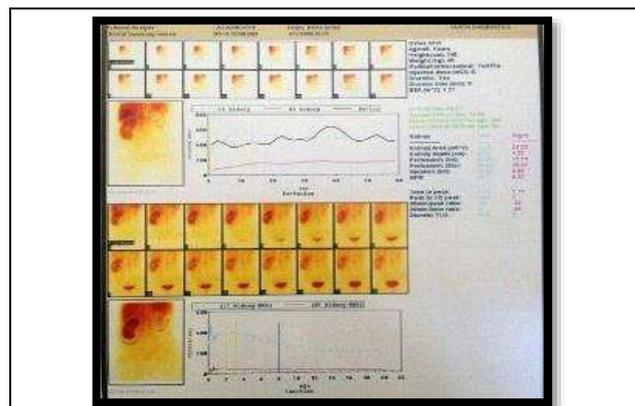


Figure 1. Radio Isotope Renogram with GFR Showing Poor Functioning Right Kidney



Figure 2. Gross Specimen- Shrunken Kidney with No Cortico-Medullary Differentiation & Increased Perirenal Fat

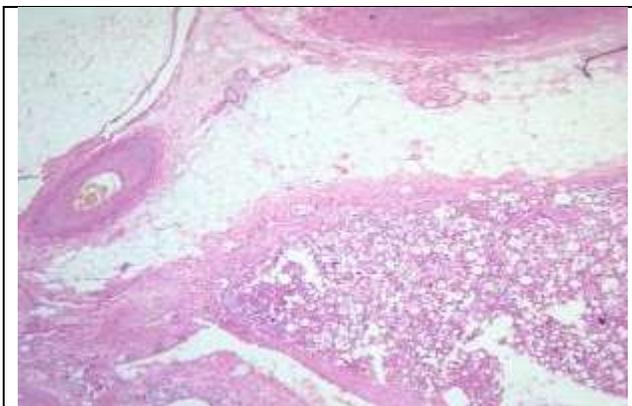


Figure 3. H & E Staining Reveals Renal Parenchyma With Few Atrophic Tubules, Thyroidisation & Replacement With Adipose Tissue (4x)

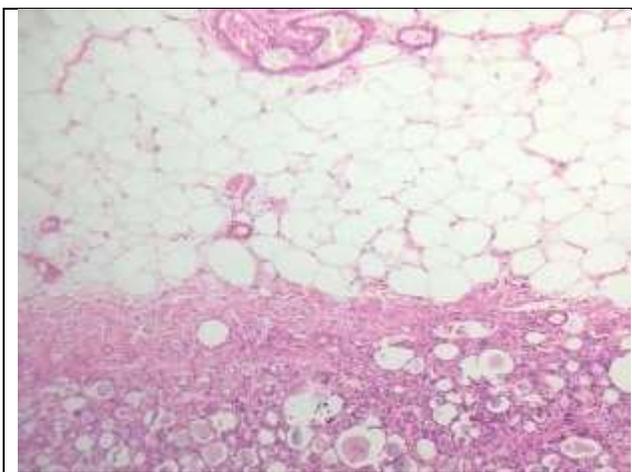


Figure 4. Low Power (10X)

DIFFERENTIAL DIAGNOSIS

- Xantho granulomatous pyelonephritis.
- Angiomyolipoma.
- Renal lipoma.
- Well differentiated Liposarcoma.

DISCUSSION OF MANAGEMENT

Renal sinus lipomatosis /replacement lipomatosis/fibrolipomatosis are the terms used for this condition & represent a spectrum of changes. Renal sinus lipomatosis – mild form, seen in 6th-7th decade, associated with obesity, atherosclerosis & steroids usage. Pathogenesis of Renal replacement lipomatosis involves two mechanisms, Compensatory mechanisms in which adipose tissue occupies the space that is produced by the atrophied/ destroyed kidney and there is an inflammatory induction of fatty proliferation to compensate for renal tissue loss. There is no specific clinical feature to diagnose this condition. Patients usually present with complaints of recurrent flank pain, fever, weight loss and mass per abdomen. Usually its unilateral and Hydroureteronephrosis with chronic infection leading to parenchymal atrophy. Renal calculi is associated with 70% cases. Renal calculi obstruct collecting system. This condition most commonly follows calculus disease. However, associations with conditions such as aging, renal

tuberculosis and post renal transplantation have also been reported.^{4,5}

The presence of atrophic renal parenchyma distinguishes this condition from other causes of fibro-fatty proliferation in and around the kidney, as in obesity, Cushing's disease or excessive corticosteroid therapy and idiopathic.⁶ It has been reported to occur in transplanted kidneys also. USG shows parenchymal atrophy or a hyper echogenic renal sinus mass with calculi. CT is the imaging method of choice. Histopathology is the method of choice for the definite diagnosis and to exclude the other possibilities.

Thorough sampling is recommended to exclude smooth muscle element & tortuous prominent vessels seen in angiomyolipoma. Absence of xanthoma cells excludes xanthogranulomatous pyelonephritis. xanthogranulomatous pyelonephritis CT scan shows pus filled, dilated calyces and xanthogranulomatous tissue (-15 to +15 HU {Hounsfield units}) reflecting the presence of intracellular fat droplets as against the pure fatty tissue seen in renal replacement lipomatosis.⁷ Absence of lipoblasts excludes well differentiated liposarcoma. Lack of well circumscribed intra parenchymal benign lipomatous mass exclude lipoma.

Pelvic lipomatosis is characterised by overgrowth of none capsulated, non-malignant, but infiltrative adipose tissue. It is usually symmetric and limited to the pelvis rarely to retro peritoneum.⁸ Pelvic lipomatosis and renal lipomatosis are of different entity, but both can present with renal failure

FINAL DIAGNOSIS

Renal replacement lipomatosis is a rare condition associated with chronic calculus disease, chronic pyelonephritis, renal tuberculosis. The diagnosis of RRL can be elusive with an erroneous initial impression of an xanthogranulomatous pyelonephritis, angiomyolipoma or liposarcoma. CT scan is the investigation of choice to differentiate it from renal neoplasms.

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