Case Report

Renal Replacement Lipomatosis Associated with Renal Calculus-Incidentally Detected at Autopsy: A Case Report of Two Cases

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Renal replacement lipomatosis (RRL) is a rare benign condition of the kidney in which there is proliferation of fat within renal sinus, hilum and peri-renalspace, thus replacing the renal parenchyma. The pathogenesis of RRL is unknown, although it is generally associated with aging, renal atrophy, long-standing chronic urinary infections. We report two cases of RRL, one of right kidney and other left, both of them associated with kidney stones. The patients were 45 and 65 years old respectively andwere admitted with fever and pain in abdomen. Autopsy performed showed right RRL associated with staghorn calculus and pulmonary tuberculosis in the first patient and left RRL with renal calculus in the pelvis and left pyelonephritis with perinephric abscess and right sided pyelonephritis in the second patient.

Keywords: Kidney, Pseudotumor, Renal Replacement Lipomatosis

INTRODUCTION

Renal replacement lipomatosis (RRL) is a rare, benign condition in which the renal sinus, hilum, and perirenal spaces are replaced with mature adipose tissue.[1] It can be idiopathic or associated with calculus and pyelonephritis.[2] Patients sometimes present with renal mass and on radiological imaging it can mimic lesions like angiomyolipoma or renal abscess or other tumor with adipose tissue.[3]

CASE REPORTS

Case 1

A 45-year-old female was admitted to our hospital with complaints of fever, breathlessness and pain in abdomen. On examination, she was febrile with low blood pressure and tenderness in abdomen. There was no significant past history. Her general condition was not stable and she expired within 2 hours of admission. Routine blood investigations including complete blood count and blood urea nitrogen were within normal limits.

A complete autopsy performed showed tuberculous bronchopneumonia. The right kidney weighed 600 g and measured 9 cm × 9 cm × 6 cm. The shape of the kidney was maintained. On the external surface, capsule was intact. Cut surface the entire renal parenchyma was replaced by adipose tissue (Figure 1).

The pelviccalycial system was dilated, distorted and harbored a staghorn calculus measuring 1 cm × 2 cm × 2 cm. Lining of pelvis was ulcerated. No normal renal parenchyma was identified. Multiple sections studied shows extensive fatty infiltration of the renal parenchyma. Few atrophic tubules and occasional sclerosed glumeruli were seen admixed with fat. Blood vessels were markedly thickened, and there was moderate inflammatory infiltrate composed of lymphocytes, plasma cells and few macrophages (Figures 2 and 3).

No evidence of pylelonephritis was seen. The pelvicalacial lining was ulcerated and showed inflammation beneath.

The opposite kidney was unremarkable. Other organs were unremarkable except for lungs that showed evidence of bilateral tuberculous bronchopneumonia. Cause of death was ascertained as tuberculous bronchopneumonia. The diagnosis of right RRL associated with renal lithiasis was also made.

Case 2

A 65-year-old male was admitted to our hospital with complaints of fever, abdominal pain constipation and vomiting. On examination, he was febrile and there was tenderness in left renal angle. Clinical impression given was left perinephric abscess and pigtaling of perinephric abscess was done. The condition deteriorated after admission and patient expired within 24 hours of admission. Routine

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blood investigation revealed anemia (hemoglobin 7.2 g/dl) with peripheral neutrophilia (total white blood cell count 41 × 10^3/µl with polymorphs 86%) and elevated blood urea nitrogen of 56 mg/dl. Ultrasonography of left kidney showed grossly altered echotecture, increased echogenicity with perinephric collection. Right kidney showed evidence of Grade I medical renal disease.

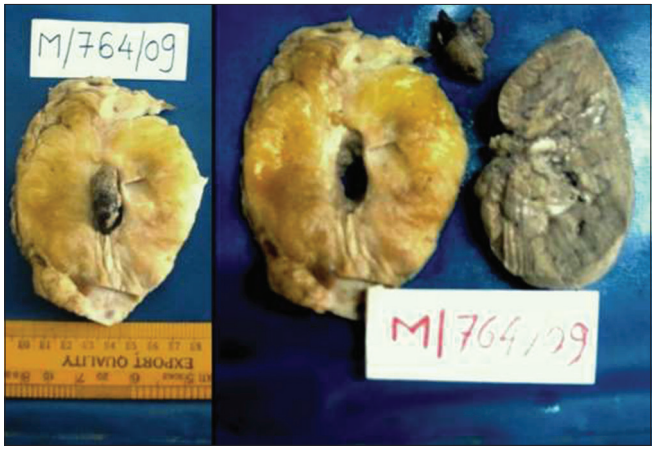

Figure 1: Gross photograph of cut surface of kidneys showing right renal replacement lipomatosis with a calculus and unremarkable left kidney

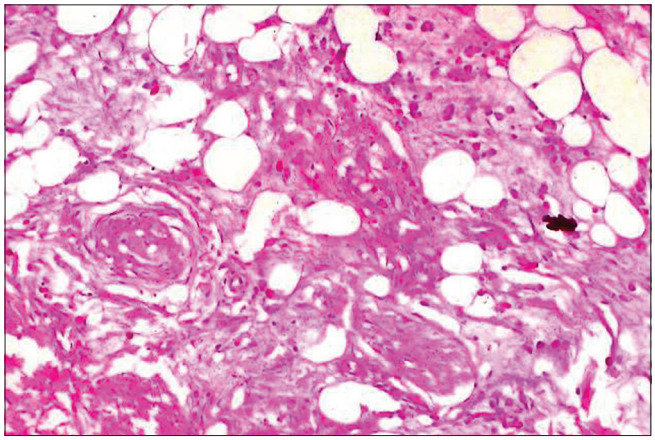

Figure 2: Histology showing scleorsedglomeruli, mature adipose tissue with few inflammatory cells (H and E, ×400)

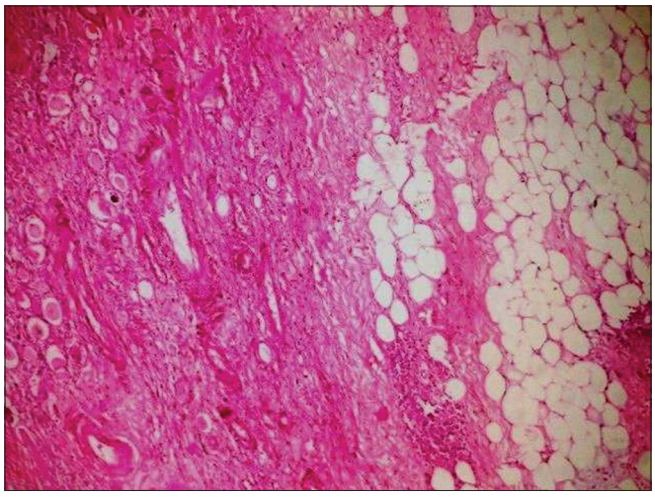

Figure 3: Histology showing fatty infiltration of renal parenchyma with atrophic tubules and thyroidization of tubules (H and E, ×100)

A complete autopsy was performed. The left kidney weighed 200 g and measured 10 cm × 7 cm × 4 cm, the shape of the kidney was maintained. Cut surface showed entire renal parenchyma was replaced by adipose tissue, and there was oozing of pus from the upper pole of the left kidney. The pelvicalycial system was distorted and a calculus measure 1 cm × 1 cm × 1 cm was present in renal pelvis. Normal renal parenchyma was identified (Figure 4).

Multiple sections studied shows extensive fatty infiltration of the renal parenchyma with multiple microabscesses. Occasional atrophic tubule and dense inflammatory infiltrate composed of lymphocytes, neutrophils, and few plasma cells were seen. The right kidney weighed 310 and external surface showed pus flakes and deep scars. Section studied shows few sclerosed glomeruli, patchy atrophy and thyroidization of tubule. Interstitum shows mixed inflammatory infiltrate composed of lymphocytes and polymorphs.

Urinary bladder showed thickening of wall and section showed moderate cystitis. Prostate was enlarged showed features of benign prostatic hypertrophy.

Cause of death was ascertainedsepticemia following acute on chronic pyelonephritis.

Diagnosis of RRL of the left kidney was made.

DISCUSSION

RRL, renal sinus lipomatosis and fibrolipomatosis of kidney are the terms representing different spectrum of same condition.[4]

Figure 4: Left kidney is enlarged, completely replaced by adipose tissue with a calculus at renal pelvis. Right kidney shows pus flakes and deep scars

Renal sinus lipomatosis, which is usually unilateral, occurs in elderly and is associated with obesity, atherosclerosis, and exogenous steroids. There is the proliferation of fat and fibrous tissue within the renal sinus. Whereas, RRL represents an extreme form of renal sinus lipomatosis. This is generally associated with infection (especially Tuberculosis), long-term hydronephrosis, and renal calculi.[1]

There is severe renal parenchymal atrophy. It is usually unilateral. The patients present in the fifth decade and usually complains of dull aching flank pain.[1] Sometimes they also present with urinary tract infection due to urinary stones or as a mass in the abdominal cavity.

Computed tomography (CT) scan, magnetic resonance imaging scan was not done in our case but these investigation appear to be the accurate methods for demonstrating the distribution of adipose mass in the renal sinus and perirenal space.[5]

Macroscopically the kidney is usually enlarged and appears to be a fibro-fatty mass in the lumbar region. Although the shape of the kidney is usually maintained.[1] The entire renal parenchyma is replaced by adipose tissue. Pelvicalycial system is generally dilated and may harbor renal stones obstructing the outflow tract as seen in our both cases. Histologically the renal parenchyma is totally replaced by mature adipose tissue admixed with sclerosed glomeruli and atrophied renal tubules. There is a variable amount of chronic inflammatory infiltrate.

Finally, the RRL can be easily confused with renal lipomatous neoplasms and xanthogranulomatous pylonephritis, especially on imaging that has a different line of treatment.[3]

CONCLUSION

RRL is a rare condition and should be considered in the differential diagnosis of cases in which there is a replacement of the renal parenchyma by adipose tissue of a hypo-functioning or non-functioning kidney leading to end-stage renal disease. Accurate diagnosis is possible with CT scan and magnetic resonance imaging scan and thus helping in the proper management.

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