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# Rare case of renal replacement lipomatosis with xanthogranulomatous pyelonephritis

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**Abstract**
We report a relatively rare case of renal replacement lipomatosis (RRL) with coexistent xanthogranulomatous pyelonephritis (XP), depicting most of the radiological findings RRL and XP has several similarities in terms of clinical background as triggering mechanism is same i.e. chronic calculus disease and inflammation in most of them. Being rare entity diagnosis is usually missed due to lack of experience.

**Keywords:** renal replacement lipomatosis, xanthogranulomatous pyelonephritis

## Introduction

Renal replacement lipomatosis is a relatively rare, benign condition in which renal parenchyma especially sinus, hilum and perirenal space is replaced by mature adipose tissue. [1, 2] Xanthogranulomatous pyelonephritis is a granulomatous inflammation characterized by destruction of renal parenchyma and replacement by lipid laden macrophages. This is a case of RRL and XP in same kidney, a rare coexistence.

**Case Report:** A 70 year old male admitted to our hospital, with complaints of right lumbar pain and low grade fever with previous history of right renal calculi. His physical examination revealed a vague mass in right lumbar region and fullness in flank region. Urinalysis showed RBCs and WBCs. A plain radiograph revealed two radio-opaque shadows (calculi) in right renal region, one of them was staghorn calculus with enlargement of renal outline (Figure 1). Ultrasonography revealed hyperechoic (fat) mass replacing the right kidney with thinned parenchyma having two calculi, one was central staghorn calculus with hydronephrosis (Figure 2A and 2B). Left kidney was normal.

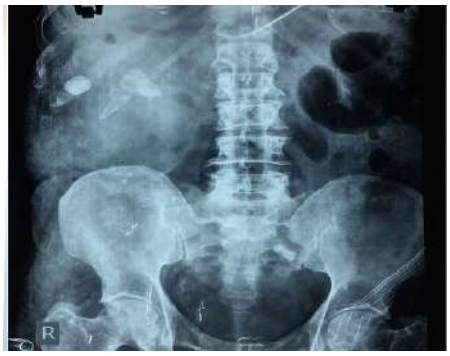


**Fig 1:** X-Ray KUB showing right renal calculi

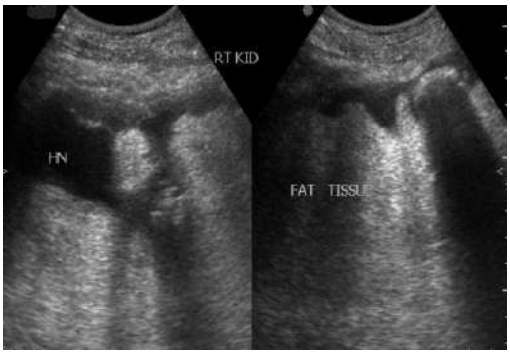


**Fig 2A:** USG image showing fatty tissue in right kidney

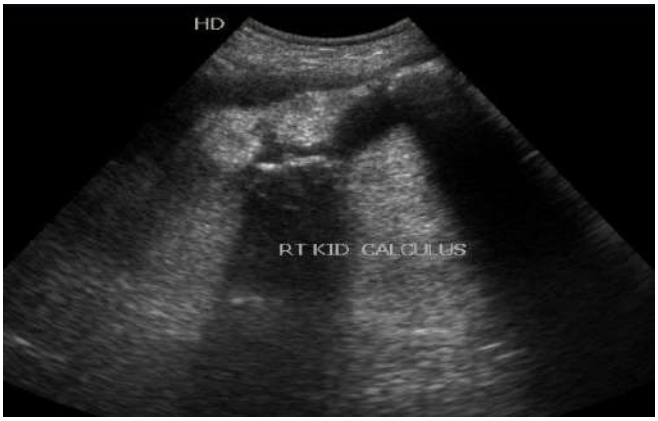


**Fig 2B:** USG Image Right Renal Calculi

Contrast enhanced Axial CT showed predominantly low density and roundish mass with extreme parenchymal atrophy with high density staghorn calculus with one more calculus and abundant low density fat and fat stranding in right kidney (Figure 3A and 3B).

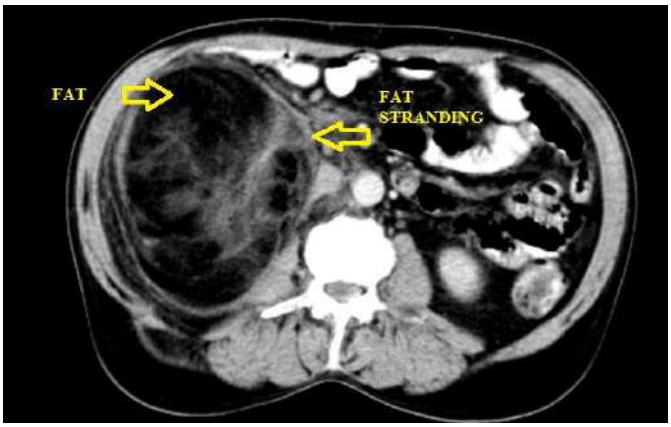


**Fig 3a:** CT image showing fatty replacement of right kidney

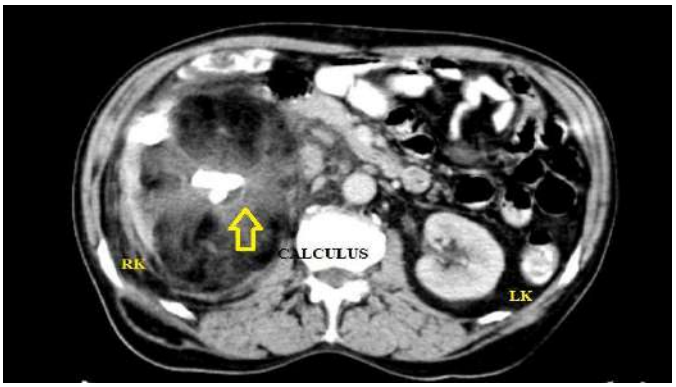


**Fig 3b:** CT image showing right renal calculi and fat in right kidney

Magnetic resonance imaging on COR T1, T1FS, T2 showed fatty replacement of right kidney (Figure 4A, 4B and 4C). post right nephrectomy diagnosis was renal replacement lipomatosis with xanthogranulomatous pyelonephritis.

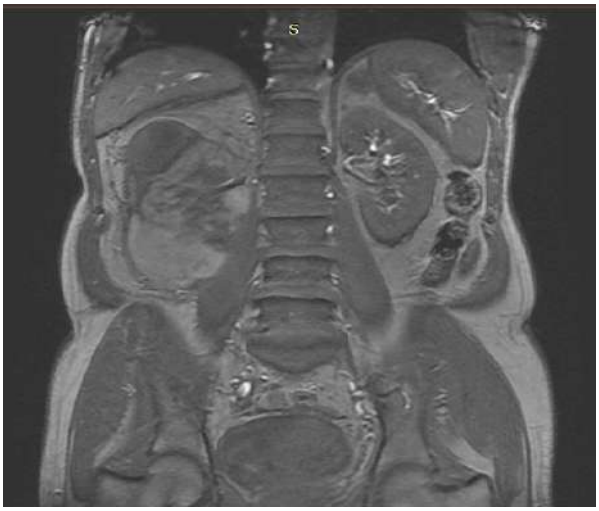


**Fig 4a:** COR T1 image showing TY replacement of right kidney

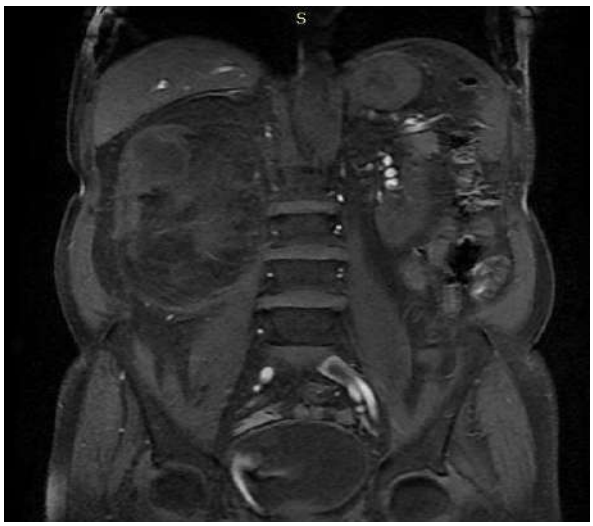


**Fig 4b:** COR T1 FS image showing suppression of fat of kidney

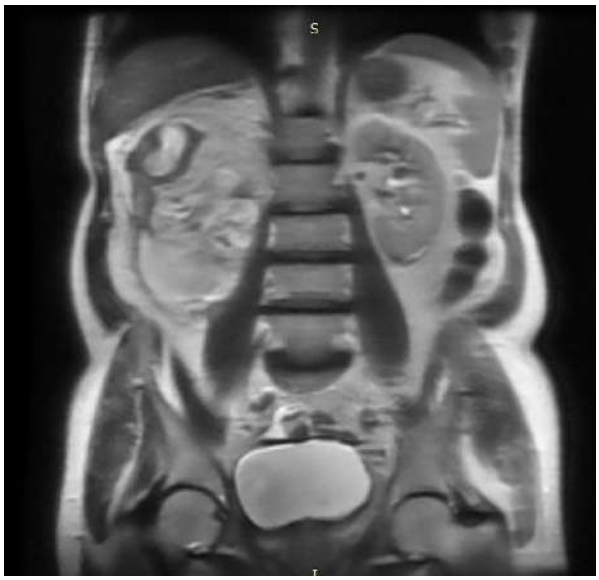


**Fig 4c:** COR T2 image showing fatty replacement of right kidney

## Discussion

Renal Replacement Lipomatosis (RRL) also known as replacement fibrolipomatosis is an uncommon benign condition occurring as end result of renal atrophy due to mature fibroadipose tissue proliferation and replacement of renal parenchyma involving sinus, hilum and perirenal space to variable extent. It is advanced form of sinus lipomatosis. In 70% cases it is associated with calculi, renal tuberculosis and renal infarction. [2-4] Patient has no specific symptoms but generally present as flank pain, mass, weight loss and fever. XP is a chronic renal inflammatory disease associated with indolent bacterial infection. Organisms most commonly involved are P. mirabilis, E.coli, Enterobacter species, P.aeruginosa and S.aureus. Inflammatory process begins in renal pelvis and later extends into medulla and cortex, which are gradually destroyed and replaced by lipid laden macrophages (xanthoma cells). [5] Pathogenesis is not clearly understood however few theories agree with primary factors being interrelated as bacterial infection, obstruction and calculus disease. Other factors are venous occlusion and hemorrhage, abnormal lipid metabolism, lymphatic blockage, failure of antimicrobial therapy, altered immunologic competence, and renal ischemia. [6] XP patients are typically middle aged women. Disease is usually diffuse but may be focal (tumefactive) also. Patient may be asymptomatic or may present as flank pain, fever weight loss, palpable mass, UTI, hematuria.

CT is optimum modality in radiology for diagnosing and differentiating XP and RRL. Abdominal radiograph shows staghorn calculus or large calcification overlying the region of kidney with a large mass filling the space. Intravenous urography demonstrates calculi and a poorly functioning or non-functioning kidney. On USG in XP, kidney appears enlarged with loss of identifiable landmarks with central echogenic focus having acoustic shadow representing large calculus or staghorn calculus. It may also show multiple calculi of varying sizes. Renal parenchyma demonstrates diffuse hypoechoic pattern corresponding to areas of inflammatory reaction or abscess. In RRL, there is hyperechoic mass representing lipomatous tissue with calculi usually staghorn or other calculus. It is difficult to demonstrate residual parenchyma which will present as hypoechoic rim. [7] On CT XP, findings are pathognomonic in most cases with diffuse reniform enlargement with ill-defined central low attenuation, apparent cortical thinning, staghorn calculus and unilateral decrease or (more commonly) absence of renal excretion of contrast material. [8, 9] Multiple fluid density, rounded areas almost replace parenchyma with radial distribution which represent dilated calyces and abscess described as “bunch of grapes” or “bear claw”. Focal (tumefactive) form shows poorly enhancing mass adjacent to calyx or in one pole with associated calculi, in most cases it is misdiagnosed as neoplasm. In RRL on CT, findings are suggestive of marked parenchymal atrophy with characteristic distribution of adipose tissue within renal sinus and perirenal space with areas of negative attenuation values similar to those of adipose tissue. Calculi are also seen. MRI is helpful in patients having contrast allergy or in pregnancy. It is not good as CT in demonstrating calculus but superior in demonstration of extent of disease. Findings are similar to those of CT. HASTE sequence is an ultrashort sequence so not affected by respiratory motion artifact. It is a heavily T2 weighted sequence hence depicting hydronephrosis, renal cysts and perirenal edema. TRUFI [True (FISP) fast imaging with steady precession] is a fast sequence providing great anatomic detail. [10] VIBE sequence depicts excretory function of kidney without motion related artifacts.

Pathologically RRL shows increased lipid content outside renal parenchyma whereas XP shows lipid laden macrophages(foam cells) infiltrating parenchyma.[11] Differentials of RRL are XP, other fat containing lesions as lipoma, liposarcoma and angiolipoma. Renal liposarcoma produces no defect in renal parenchyma and is located peripherally (between kidney and renal capsule) and in cases of angiomyolipoma and lipoma atrophy of renal parenchyma, calculi and absence of renal contrast concentration and excretion are not observed.[3] Although coexistence of RRL and XP is very rare in same kidney, one should become aware and not to confuse with neoplasm.

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