# Renal Replacement Lipomatosis with Coexistent Papillary Renal Cell Carcinoma, Renal Tubulopapillary Adenomatosis, and Xanthogranulomatous Pyelonephritis

## An Extremely Rare Association and Possible Pathogenetic Correlation

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## INTRODUCTION

Renal replacement lipomatosis (RRL) is a rare condition, which is characterized by diffuse replacement of renal parenchyma, sinus, and hilum with adipose tissue.(1) It is thought to be associated with physiological as well as several pathological conditions, like aging, chronic inflammation, renal stones, and tuberculosis.(2,3) In the present case, we found coexistence of RRL with papillary renal cell carcinoma (PRCC). Furthermore, renal tubulopapillary adenomatosis, xanthogranulomatous pyelonephritis (XGP), and multiple renal calculi were also found. A brief review of literature along with probable pathogenesis of such a rare coexistence is presented.

## CASE REPORT

A 55-year-old man presented with left flank pain for 6 months and hematuria for the last 2 years. Physical examination was unremarkable. Urinalysis showed 30 to 35 red blood cells/hpf and 20 to 25 pus cells/hpf. An abdominal ultrasonography revealed multiple calculi in the right kidney along with a hypoechoic mass near the upper pole with alteration of vascularity, suggestive of RCC or adrenal mass. Contrast-enhanced computed tomography revealed small contracted right kidney. The perinephric fat was proliferating and compressing parenchyma, suggestive of RRL. A well-defined, 5.3 × 5.3 cm heterogeneously enhanced soft tissue mass was seen at the superior pole suggestive of RCC or oncocytoma. Left kidney was normal in morphology and echotexture.

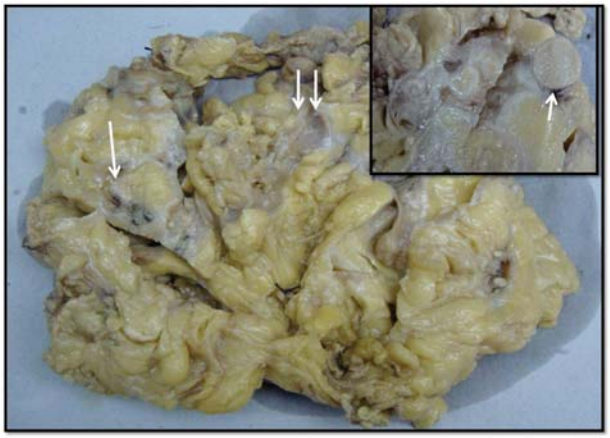

**Figure 1.** Gross photograph of the kidney showing fatty replacement of renal tissue, renal calculi (single arrow), and part of atrophic cystic kidney (double arrows); Inset: Solid foci of papillary RCC (arrow).

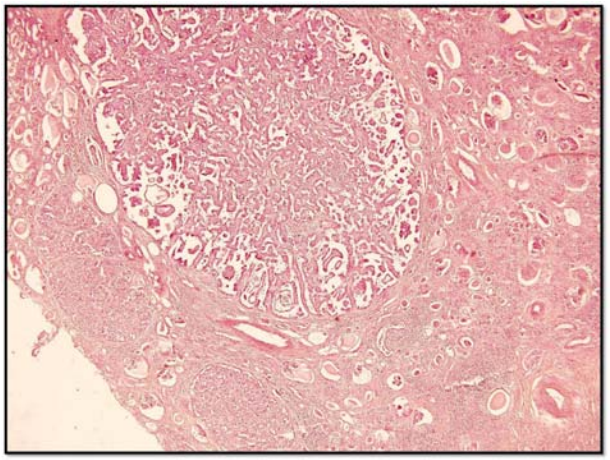

**Figure 2.** Microphotograph of the kidney showing multiple foci of tubulopapillary adenomatosis along with changes of chronic pyelonephritis (Hematoxylin and Eosin stain, ×4).

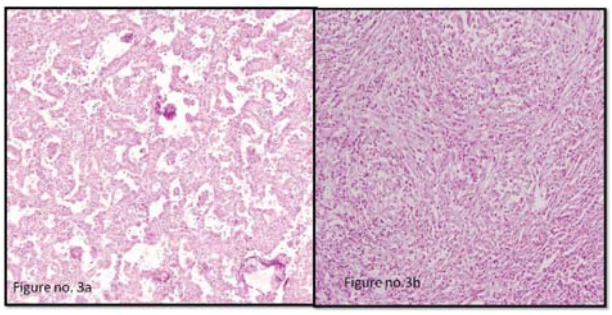


**Figure 3.** (A) Microphotograph showing papillary RCC with psammomatous calcification (Hematoxylin and Eosin stain, 10×); (B) Kidney showing xanthogranulomatous pyelonephritis comprising foamy cells along with inflammatory cells (Hematoxylin and Eosin stain, ×20)

Based on clinical and imaging findings, right nephrectomy was performed. Nephrectomy specimen measured 15 × 12 × 10 cm. On cut surface, parenchyma was largely replaced by adipose tissue with small foci of residual atrophied renal parenchyma (Figure 1). There was dilatation of pelvicalyceal system with presence of multiple stones in the calyces. A solid, focally papillary area measuring 2 × 2 cm was identified at the upper pole (Figure 1; Inset) along with multiple small yellow-colored nodules, largest one of which measured 1.5 × 1 × 1 cm.

Microscopic examination from yellow nodules showed foci of tubulopapillary adenomatosis (Figure 2). Sections from papillary area showed PRCC (Figure 3A). Adjacent areas revealed thyroidization of tubules along with diffuse interstitial inflammation comprising lymphocytes, macrophages, and xanthoma cells, suggestive of XGP (Figures 2 and 3B).

## DISCUSSION

Renal replacement lipomatosis is a benign condition where mature fibroadipose tissue proliferates and replaces the renal parenchyma, which in turn becomes atrophic. It can involve the renal sinus, renal hilum, and perirenal space to variable extent.[1] Other conditions which may be associated with fibrofatty proliferation of the renal sinus, renal hilum, and/or perirenal space include obesity, Cushing's syndrome, corticosteroid excess, or be idiopathic. However, the preservation of renal parenchyma in such cases distinguishes these entities from RRL.[4,5] Renal replacement lipomatosis is usually secondary to renal calculus and renal tuberculosis, occasionally occurs after renal infarction or may be idiopathic.[1,6] The present patient was not obese; there was no history suggestive of Cushing syndrome or corticosteroid and anti-tuberculosis drug intake.

Computed tomography scan provides a definite radiological diagnosis in RRL and is considered to be superior to ultrasonography, which is not very sensitive for detecting fatty proliferation.[7] Furthermore, CT scan is considered to be the best imaging modality to differentiate RRL from other fat-rich lesions, such as renal angiomyolipoma, retroperitoneal lipoma, and liposarcoma.[8]

Renal replacement lipomatosis is frequently associated with XGP. Both share several similarities in terms of possible

pathogenesis. Both conditions are associated with inflammation. It has been proposed that inflammation may cause induction of fatty proliferation, which in turn compensates the loss of renal tissue, thus leading to RRL.[9,10]

Although there are only limited data available, it is known that the kidneys harboring RCC could reveal various premalignant lesions ranging from dysplasia to adenoma.[11,12] Such findings suggest an adenoma-carcinoma sequence in PRCC. In a study of 542 nephrectomy specimens, 7% showed papillary adenoma, out of which 47% arose in the setting of PRCC and 15.7% in association with clear cell RCC.[12] Adenomas associated with PRCC are usually multiple.[12] The present subject had PRCC with presence of multiple tubulopapillary adenomatosis, thereby corroborating adenoma-carcinoma sequence. Furthermore, our patient had the unique coexistence of RRL with XGP, probably secondary to renal calculi.

In conclusion, apart from the novelty of being a rare case, possibly first of its kind, this case also suggests the possible predisposing factors and pathogenesis of RRL and PRCC. We hope more urologists, radiologists, and pathologists become aware of this unique entity for its early diagnosis and proper management.

## CONFLICT OF INTEREST

None declared.

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