

Renal Replacement Lipomatosis: A Rare and Often Ignored Entity

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A 54-year-old elderly hypertensive female presented to urosurgery outpatient department with complaint of increased frequency and on and off pain in the left flank for five years, occurring every 10 to 15 days with moderate to severe intensity and relieved spontaneously with medications. The clinical examination was unremarkable. Her hypertension was under control with medication. Urine routine microscopy, renal function tests and serum electrolytes were within the normal range. A Computed Tomography (CT) of the abdomen suggested a small sized left kidney with a staghorn calculus of size 39×38×15 mm and near total replacement of renal parenchyma by fat density (Fig. 1). A Dimercaptosuccinic acid scan (DMSA) suggested a non-functioning left kidney. Left simple nephrectomy was performed and the specimen was sent to the department of pathology. The left kidney had grossly atrophied and measured 6×4×4 cm, weighed 113 g along with ureter measuring 8.5 cm in length. On serial slicing, the kidney was almost entirely replaced by fibroadipose tissue with loss of corticomedullary distinction along with a large stag-

horn calculus visible in the calyx at the upper pole measuring 3.7×1.3×1 cm (Fig. 2A). Microscopy showed a markedly thinned out renal cortex with interstitial fibrosis and extensive replacement with fibroadipose tissue (Fig. 2B). Marked thyroidization of tubules along with glomerulosclerosis and dense interstitial inflammation was noted comprised of lymphocytes, plasma cells and few neutrophils (Fig. 2C, D). The blood vessels showed moderate fibrointimal thickening (Fig. 2C). No xanthogranulomatous inflammation was noted. A final impression of renal replacement lipomatosis (RRL) with chronic calculous pyelonephritis was rendered. The patient is now on follow up and is doing well.

RRL is a rare benign condition occurring as a manifestation of renal atrophy or chronic damage to renal parenchyma most commonly due to long standing calculus which further leads to inflammation, infection and atrophy.¹ Renal lipomatosis was originally assayed by Kutzmann,² however, little was known regarding the pathogenesis. RRL is an end stage disease and is a severe form of renal sinus lipomatosis (RSL). It is a benign adipocytic proliferation which occupies renal sinus, hilum and almost the whole of the renal parenchyma rendering it non-functioning. Exact pathogenesis is still obscure but it has its associations with aging, obesity, calculi most commonly, renal tuberculosis and post renal transplant leading to atrophy.^{1,3,4} Clinical features are varied and range from fever, recurrent flank pain, urinary tract infection or even as a mass in the abdomen suggesting benign conditions like malakoplakia, xanthogranulomatous inflammation or tumors like angiomyolipoma, lipoma, liposarcoma creating a diagnostic challenge at times on imaging modalities like ultrasonography (USG), CT, or magnetic resonance imaging (MRI).⁵ However, it is CT and histopathological examination which by far facilitate the diagnosis the most.⁴ In our case, the clinical examination was unremarkable and the CT suggested renal lipomatosis which was further confirmed on histological examination. However, either renal replacement lipomatosis is actually a rare entity or is undiagnosed and ignored since, the majority of renal masses

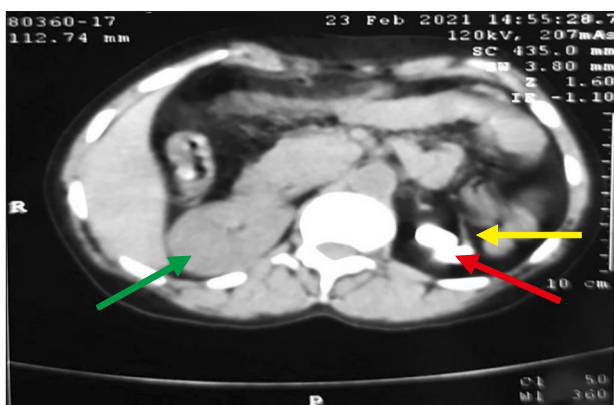


FIG. 1. CT Abdomen axial section at the level of kidney shows a large hyperdense staghorn calculus (red arrow) in the pelvis of left kidney with near total replacement of renal parenchyma by fat density (yellow arrow). Normal kidney is seen on right side (green arrow).

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Article History:

Received February 15, 2022

Revised March 3, 2022

Accepted March 20, 2022

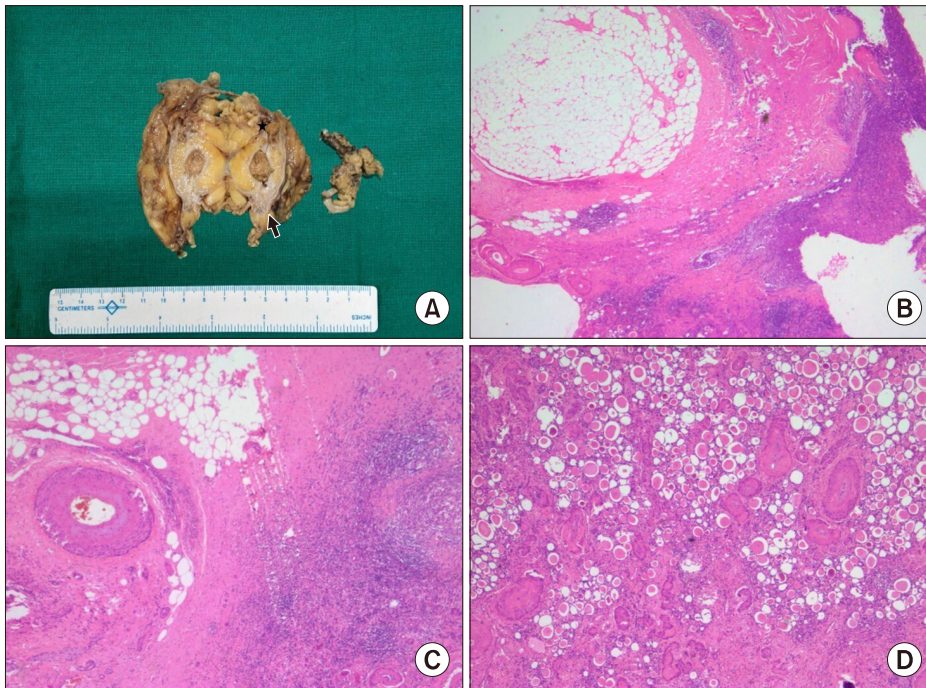


FIG. 2. (A) Gross Image: Shows near total replacement of renal parenchyma with only partly preserved cortical area is seen in lower pole (marked with black arrow). Yellowish staghorn calculus is seen impacted in one calyx at the upper pole (marked with black asterisk). The separately lying ureter is seen. (B, C) Microscopy shows interstitial fibrosis, dense lymphoplasmacytic inflammatory infiltrate and extensive replacement with fibroadipose tissue. In addition, (C) shows blood vessels showed moderate fibrointimal thickening. (D) Marked thyroidization of tubules along with glomerulosclerosis and dense interstitial inflammation was noted comprising of lymphocytes, plasma cells and few neutrophils.

like lesions are malignant. Also, imaging should be done with more scrutiny and the gross specimen should be thoroughly sectioned with adequate representation of the lesion. Thus, the benign nature of RRL, non-specific clinical features and investigations pose a diagnostic dilemma to clinicians. Thus, in light of nonspecific features, renal replacement lipomatosis should be kept in differentials and clinicians, radiologists and pathologists should be aware of it.

CONFLICT OF INTEREST STATEMENT

None declared.

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