Case Report

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Replacement Lipomatosis of Kidney: How to Diagnose and Analyze the Important Differential Diagnosis along with a Case Report

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Introduction

Replacement lipomatosis of the kidney (RLK) is a rare disorder in which a massive fatty tissue proliferation occurs within the renal sinus, hilum and perirenal region. It is also known as replacement fibrolipomatosis [1]. Usuallya varying amount of fat and fibrous tissue is always present within the renal sinus and along the branches of the renal segmental arteries, which becomes more prominent with aging, obesity and use of steroids (renal sinus lipomatosis) [2,7]. RLK represents the extreme form of renal sinus lipomatosis in which recurrent pyelonephritis and calculi are associated with severe renal parenchymal atrophy that usually occurs unilaterally. Renal calculus disease is associated in more than 70% of cases [2,3].

Clinical symptoms, including urinary tract infections, fever and low grade constant flank pain, usually result from the associated infection and calculus disease [4]. Frequently, renal function is poor or the kidney is non-functioning [2].Occasionally RLK can simulate renal malignancies hence it is important to analyze its differential diagnosis carefully [2].

Pathologically the reniform shape of the kidney is maintained. Kidney is enlarged with gross fibrofatty appearance. Renal cortex is extremely atrophied and thinned out with marked proliferation of hyperplastic fat cells in the renal sinus that do not permeate the renal parenchyma but merely replace it as it atrophies [1].

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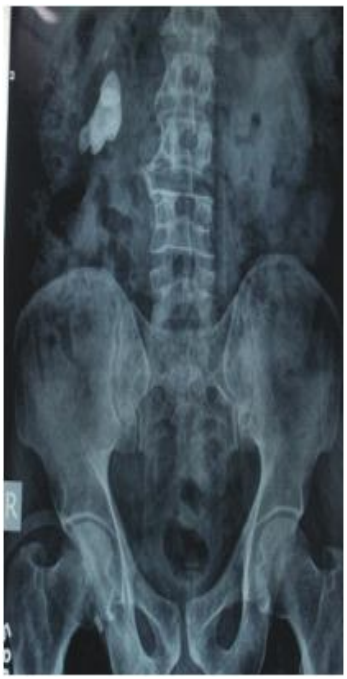
Important imaging methods used to diagnose this disease are intravenous urography (IVU), ultrasonography, CT and MRI. CT is the investigation of choice with staghorn calculus [9], atrophied renal parenchyma and abundant fatty tissue centrally which has characteristic attenuation of fat [1,2,7].

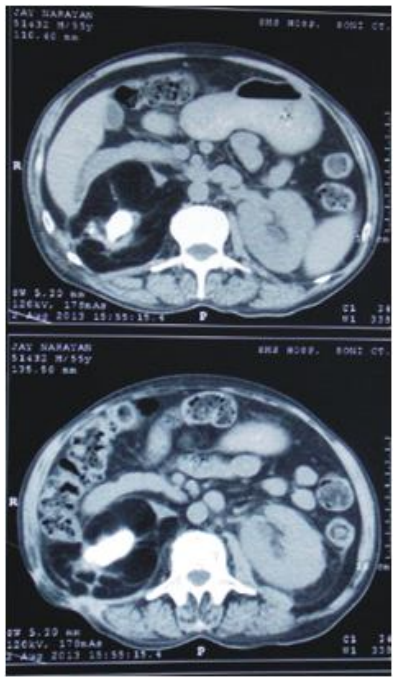
We present NC CT, X-ray KUB and renal scintigraphy findings of a patient with RLK.

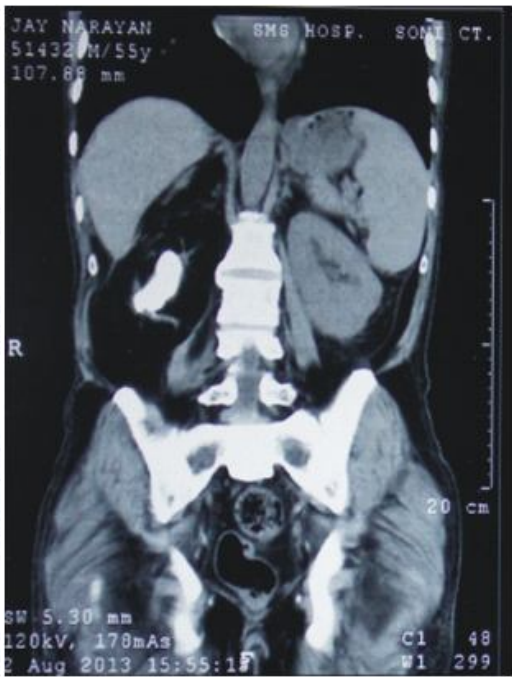
Case report

This 55 year old male patient was admitted with complaints of recurrent right flank pain and occasional dysuria. No significant past history. X-ray KUB revealed a staghorn calculus in right kidney. USG showed enlarged hypo-echoic right kidney with a central hyperechoic calculus and atrophied renal parenchyma. Serum creatinine was 2.0 on repeated examination. NCCT shows a staghorn calculus 42x28mm size in right kidney with complete loss of renal parenchyma and replacement by fatty tissues. Left kidney appears normal in size and shape. DTPA scan shows non visualized right kidney with border-line function of left kidney. A differential diagnosis of RLK and xanthogranulomatous pyelonephritis (XGP) was made although the CT attenuation values suggest a more likely possibility of RLK. Patient underwent right simple nephrectomy. On gross examination there was complete replacement of right kidney with fatty tissues with parenchymal atrophy and a large staghorn calculus. Histopathological examination revealed lobules of fatty tissues interspersed with fibrous bands that replace the marked atrophic renal parenchyma.

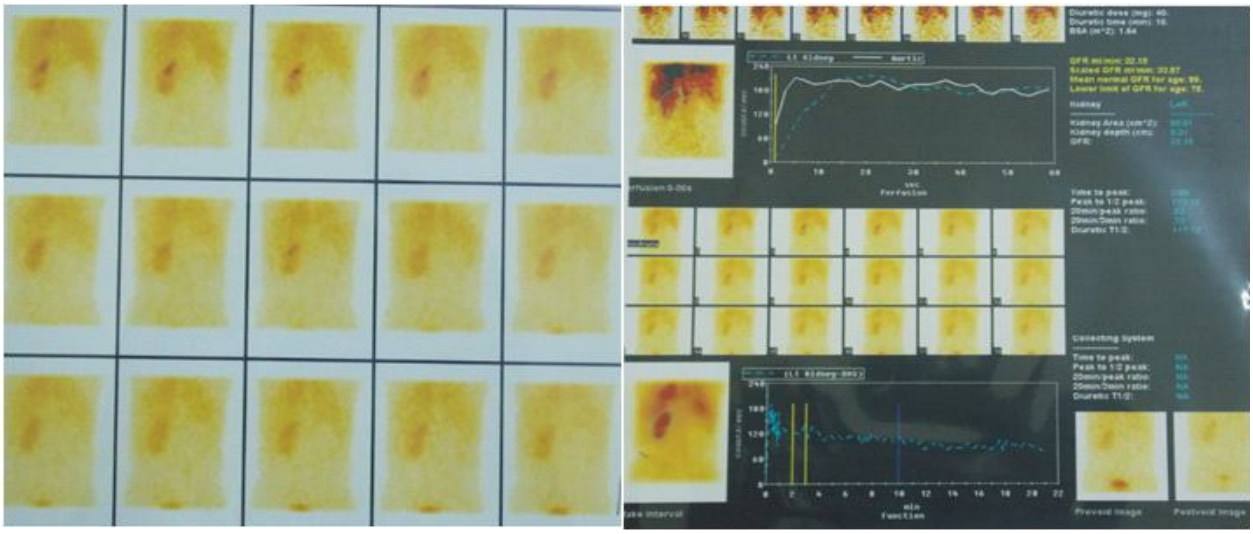


X-ray KUB

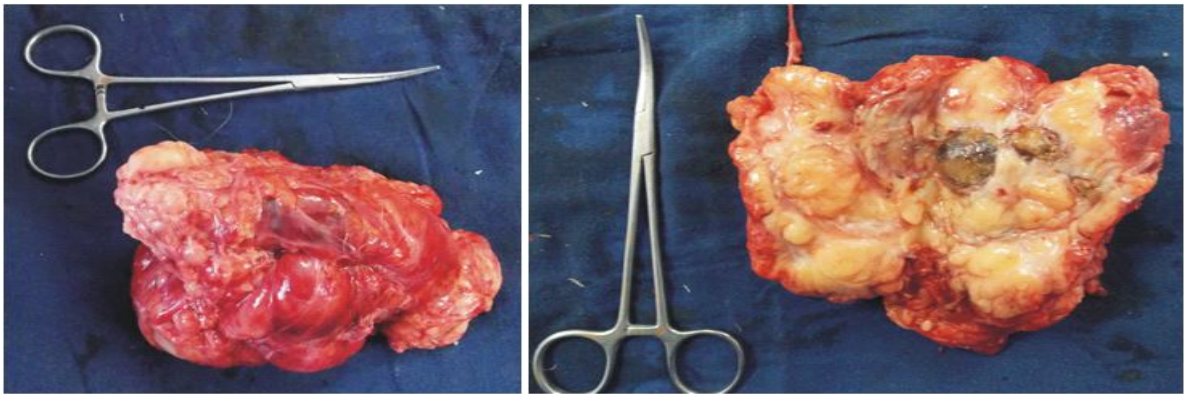




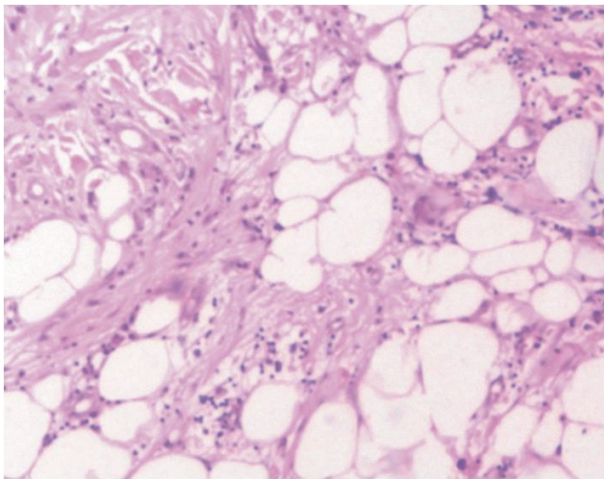
NCCT



DTPA SCAN: Right non functioning kidney.



Gross Appearance Eand Cut Section: Central staghorn calculus with fatty replacement of the kidney.



MICROSCOPIC APPEARANCE: large fat cells replacing the renal parenchyma

Discussion

There is a varying amount of fat within the renal sinus that covers major branches of the renal artery. Although the fatty tissue in the renal sinus and perinephric region normally increases with both age and obesity but the volume of renal parenchymal tissue remains the same [2,5]. Renal sinus lipomatosis infrequently produces symptoms because of the absence of caliceal obstruction [7]. RLK is an aggressive variety of renal sinus lipomatosis that develops secondary to chronic obstruction, renal stone disease with recurrent inflammation and renal parenchymal atrophy [8]. These continuum fatty changes can be confused with renal lipomas and neoplasms leading to diagnostic challenges on imaging [2]. Two theories have been proposed in the pathogenesis of RLK. First, a compensatory mechanism in which proliferated fatty tissue replaces the atrophied or destroyed. Secondly, the inflammatory induction of fatty proliferation compensates for renal tissue loss [2].

Useful imaging methods are IVU, USG, CT and MRI [7,10]. X-ray KUB may reveal calculi whereas IVP demonstrate a poorly or non-functioning kidney. Ultrasonography may suggest the diagnosis by demonstrating parenchymal atrophy, expansion of hyper-echoic renal sinus mass with a stone; however, it cannot depict confidently the perirenal changes. Although USG may be highly suggestive, CT appears to be an accurate method for demonstrating the different features of RLK, i.e. parenchymal atrophy, calculi and the characteristic distribution of a dipose mass within the renal sinus and perirenal space, with negative attenuation values similar to those of a dipose tissue between –77 (HU) to –102 HU [2]. MRI can suppress the fathomogeneously and provides useful information to aid in the differential diagnosis between fatty tumors of the kidneys [1,2,10].

The important differential diagnoses are- Xanthogranulomatous pyelonephritis and fat-containing renal neoplasms including lipoma, liposarcoma and angiomyolipoma [2,8].

Xanthogranulomatous pyelonephritis is a chronic renal inflammation condition associated with stone obstruction, in which lipid-laden macrophages infiltrate the renal parenchyma; however, in RLK, fat cells remain outside the renal parenchyma. In CT examination the accompanying pyonephrosis and abscesses has attenuation values between -15 HU and +15 HU [2]. Both Xanthogranulomatous pyelonephritis and RLK can also coexist [2,6,8].

The key features of angiomyolipoma are areas of fat attenuation within the tumor, contrast-enhancing soft tissue and saccular aneurysms [2]. Lipoma is found within the renal parenchyma. Renal liposarcoma is usually arises from the renal capsule or wall of renal vein and therefore is peripheral in location. Additionally, in these tumors the absence of renal parenchymal atrophy, more or less preserved function of the kidney and absence of stone help to make a differential diagnosis [2].

Treatment option is nephrectomy[2] although there has been a case report of resolution of RLK following stone surgery.

Therefore in conclusion, the important aspects of this disease are: (1) RLK is a relatively rare disorder (2) RLK is usually associated with a poorly or non-functioning kidney (3) RLK may be confused with fat containing renal neoplasms.

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