RENAL LIPOMATOSIS

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ABSTRACT

Replacement lipomatosis is thought to be a spectrum of degenerative process. The mildest form is called renal sinus lipomatosis and is usually associated with obesity, atherosclerosis or use of exogenous steroids and it has no clinical significance, while renal replacement lipomatosis is associated with long-standing inflammation and calculi as in this case. The current case report highlights the benefit of imaging techniques for suggesting this condition since early diagnosis and relieving calyceal obstruction can improve renal function and hopefully prevent progressive cortical atrophy while deficient techniques and reports may end with impaired renal function and unfortunately subsequent nephrectomy.

Keywords: *Lipomatosis, Kidney, Lipoma.*

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INTRODUCTION

Renal replacement lipomatosis (RRL) is a rare and benign condition characterized by proliferation of renal sinus/hilar and perirenal fatty tissue with marked atrophy of the renal parenchyma. It is seen with calculus disease in 70% of cases and associated with chronic inflammation and hydronephrosis [(1)].

CASE REPORT

A 49 year old female was admitted to the Teaching Hospital complaining of flank pain which was continuous and dull aching. There were no other significant complaints. She had a previous history of lithotomy. On physical examination, there was a non-tender left lumbar region and no palpable mass. Investigations revealed normal blood urea and serum creatinine, IVU revealed a delay in contrast excretion with mild hydronephrosis, cayceal distortion and significant parenchymal wasting. Abdominal ultrasound revealed two small stones in the left kidney;the largest was of 4 mm in diameter. The right kidney was normal.

Left nephrectomy was performed. Specimen received was measuring 11 x 7 x 4 cm with evident increase in peri-renal fat and intact Gerota's fascia (Figure 1)

On cut surface, the entire renal sinuses were replaced with adipose tissue with atrophied renal parenchyma and moderately dilated pelvicalyceal system (Figure 2). Cortical thickness was less than 2 cm in most areas.

Microscopic examination showed extensive fatty replacement in the renal sinuses, atrophied and compressed renal cortical parenchyma with some sclerosed glomeruli, focal thyroidisation of renal tubules, interstitial fibrosis (Figure 3,4), and thickened blood vessels. There was a clear demarcation between the adipose tissue and renal parenchyma (Figure 4,5). Inflammation was moderate and comprising of lymphocytes, plasma cells and few macrophages. No xanthomatous change or any neoplasm was noted.

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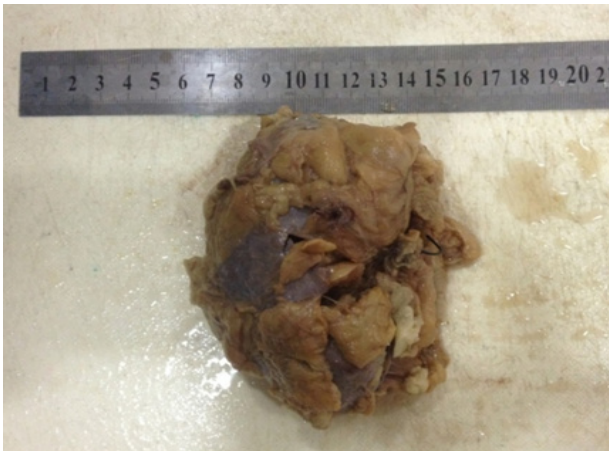

Figure 1. External appearance of the kidney showing increase in perirenal fat.

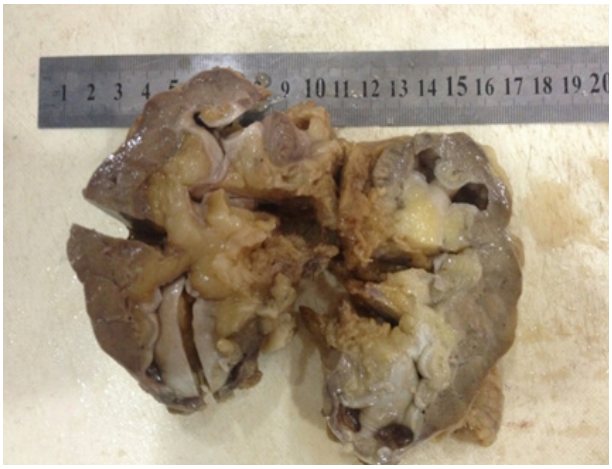

Figure 2. Saggital section shows fat in renal sinus and hilar region with hydronephrosis and thinning of the cortex

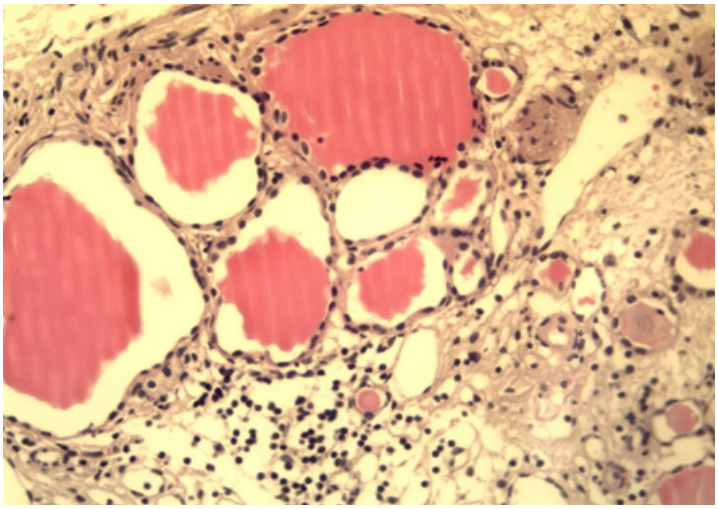

Figure 3. Renal cortex showed scattered areas of thyroidization with mild chronic inflammatory cell infiltration. Absence of xanthoma cells excludes xanthogranulomatous pyelonephritis. H&E x400.

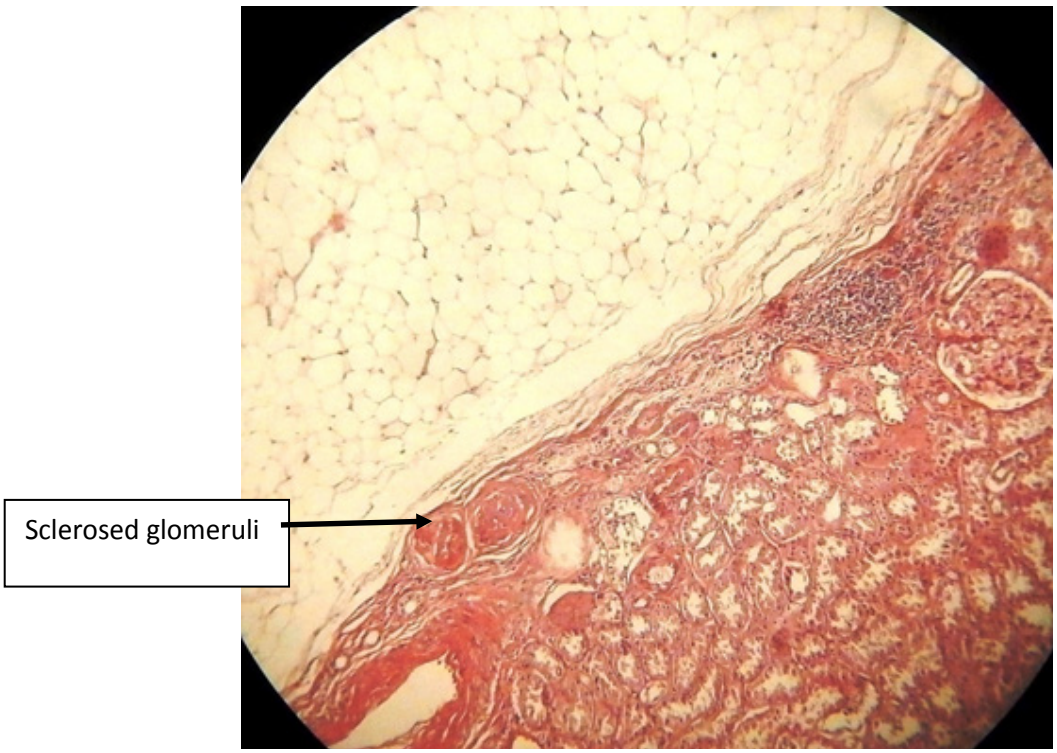

Figure 4. Sharp demarcation of the fatty replacement of the renal parenchymal tissue with compression atrophy of the adjacent renal tissue and sclerosis of the some renal corpuscles (arrows). H&E x100

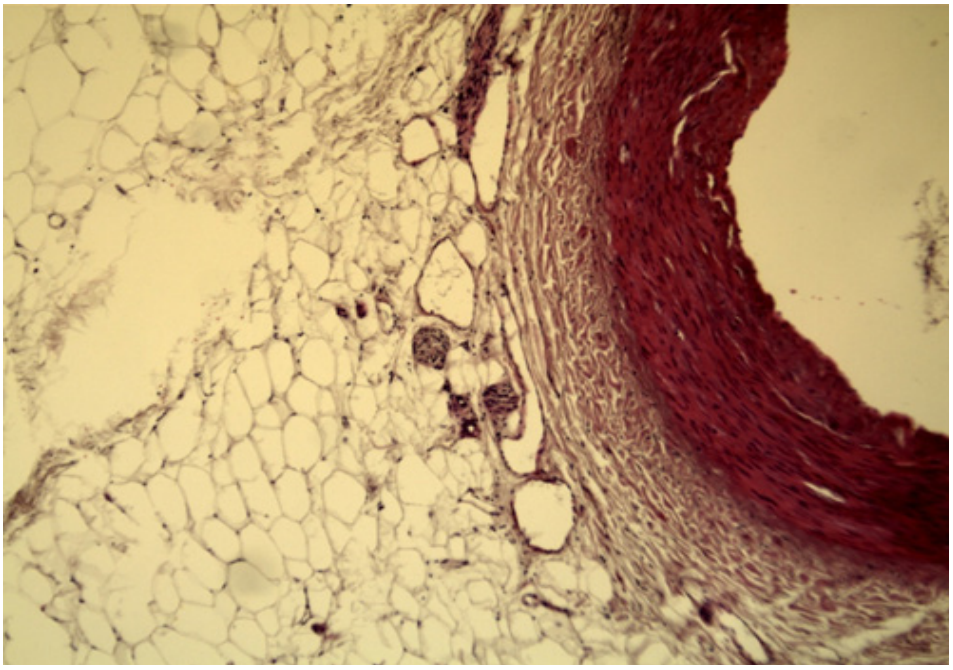

Figure 5. Hilar region shows renal vein (right side) with adjacent fatty replacement of renal sinus. H&E x100

DISCUSSION

Renal sinus lipomatosis, replacement lipomatosis, and fibrolipomatosis of kidney are the terms used for this rare condition and represent a spectrum of changes [2]. It is different from renal lipomas which are neoplastic, whereas this condition is thought to be a degenerative process [3, 4]. Renal sinus lipomatosis, the mildest form is usually seen in the sixth and seventh decade associated with obesity, atherosclerosis or use of exogenous steroids, and it has no clinical significance [3]. At the other end of the spectrum is RRL where the entire renal parenchyma is replaced with adipose tissue and is associated with long-standing inflammation and calculi in 76%-79% of cases [5]. Usually it is unilateral in occurrence and rarely idiopathic [6]. We think that our case lies within this spectrum. Two theories have been proposed for its pathogenesis. Some suggest a compensatory mechanism in which the adipose tissue occupies the space that is produced by the atrophied or destroyed kidney or there is an inflammatory induction of fatty proliferation to compensate for renal tissue loss [6]. Early diagnosis and relieving calyceal obstruction can improve renal function and hopefully prevent progressive cortical atrophy [7].

In literatures the peak age is in the fifth decade as in our case, although it is also reported in the second decade [2, 4]. An idiopathic case, in 30 years age female was reported [8]. The main clinical presentation is usually dull flank pain as in this case with repeated attacks of urinary tract infection associated with renal stones. Blood urea and serum creatinine are usually within the normal limits if the disease is unilateral with other well functioning kidney as in our case [2, 4].

Ultrasonography may suggest the diagnosis by demonstrating parenchymal atrophy or a hyperechoic renal sinus mass with a stone; however, it is not diagnostic [6] . In this case two small stones were found in the left kidney with cortical atrophy, no further findings and measurement . Usually IVU demonstrates a non-functioning or poorly functioning kidney [9] as in this case where IVU demonstrates a delay in contrast excretion with mild hydronephrosis, calyceal distortion and significant parenchymal wasting so this case came with late presentation. CT scan and magnetic resonance imaging scan appear to be an accurate method for demonstrating the distribution of adipose mass within the renal sinus and perirenal space [6], CT appears to be the imaging method of choice [10], but it was not performed for this case. By imaging techniques differential diagnoses of lipomatous neoplasm like lipoma, liposarcoma or angiomyolipoma or xanthogranulomatous pyelonephritis was made [6].

Histopathology is the method of choice for the definite diagnosis and to exclude the other possibilities. Thorough sampling is recommended to exclude smooth muscle element and tortuous prominent vessels seen in angiomyolipoma. The bright-yellow fat tissue in the renal sinus that is similar to the perirenal fat in renal lipomatosis while a pale-yellow fatty tissue in xanthogranulomatous pyelonephritis is a helpful feature to differentiate between both [11]. The absence of xanthoma cells excludes xanthogranulomatous pyelonephritis although they are known to co-exist [11]. Absence of lipoblasts excludes well differentiated liposarcoma, and the lack of well circumscribed intraparenchymal benign lipomatous mass exclude lipoma.

In conclusion, imaging techniques are of great benefit in suggesting the differential diagnosis in the early stage of this condition otherwise histopathological examination gives the definitive diagnosis but obviously in nephrectomy specimens.

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