CROSS-SECTIONAL IMAGING EVALUATION OF RENAL REPLACEMENT LIPOMATOSIS (RRL)

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ABSTRACT

BACKGROUND

Renal replacement lipomatosis is an uncommon benign entity where abundance of fibrofatty tissue proliferation occurs in renal sinus with further extension of proliferated fatty tissues into renal hilum, perinephric and periureteric spaces. It is usually associated with renal atrophy and marked renal parenchymal destruction. Aim of our study was cross-sectional imaging evaluation of Renal Replacement Lipomatosis (RRL).

METHODS

A hospital based cross-sectional retrospective study was conducted. The study group comprised of 16 patients presenting to the Departments of Radio-diagnosis, Surgery and Urology in a tertiary care hospital from May 2014 to April 2016. All patients were initially evaluated clinically and ultrasonographically followed by cross-sectional imaging modality like Computed Tomography (CT), Magnetic Resonance Imaging (MRI) or both.

RESULTS

Out of 16 patients of renal replacement lipomatosis, 15 patients (93.8%) had associated renal pelvic or ureteric calculus while 1 patient (6.2%) had left para-aortic mass without associated calculus. Out of fifteen patients of calculus related RRL, 8 patients (53.3%) had calculus size more than 40 mm, followed by 4 patients (26.7%) who had size from 20 to 40 mm and 3 patients (20%) had size less than 20 mm. The mean CT HU value of calculus was 1334±84.5 in our study. Three patients (18.8%) had only renal hilar fatty excessive deposition, 1 patient (6.2%) had renal hilar and perinephric space fat depositions, 3 patients (18.8%) had renal hilar, perinephric and periureteric spaces depositions and 9 patients (56.2%) had renal hilar and periureteric excessive fatty depositions. Delayed renal functioning was noted in 9 patients (56%), followed by non-functioning in 5 patients (31.2%) and 2 patients (12.5%) had normally functioning kidneys.

CONCLUSION

Cross-sectional imaging like CT and MRI scan helps in diagnosing RRL and proper delineation of extensions of excessive fatty tissue proliferation.

KEYWORDS

Renal Replacement Lipomatosis (RRL), Staghorn, Xanthogranulomatous Pyelonephritis (XGP).

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INTRODUCTION: Renal replacement lipomatosis is an uncommon benign pathological entity, where abundance of fibrofatty tissue proliferations occurs in renal sinus, renal hilum, further extends into perinephric and periureteric spaces and it is usually associated with marked renal atrophy and renal parenchymal destruction. RRL usually associated with longstanding calculus disease.[1] Renal replacement lipomatosis (RRL) is although misdiagnosis - mainly with xanthogranulomatous pyelonephritis (XGP) as these two are very difficult to differentiate radiologically. CT and MRI scan, including the use of fat suppressed sequences played a key role in diagnosing RRL preoperatively. Aim of our study was cross-sectional imaging evaluation of renal replacement lipomatosis.

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MATERIAL AND METHODS: After approval from the Institutional Ethics Review Committee, a hospital based cross-sectional retrospective study was conducted. The study group comprised of 16 patients presenting to the departments of Radio-diagnosis, Surgery and Urology in a tertiary care hospital from May 2014 to April 2016.

Patient Selection: We included both outpatients and inpatients of both sexes presenting with pain abdomen, lump in abdomen or urinary problems. We have included only those patients in whom at least one cross-sectional imaging modality was performed. Patients in whom cross-sectional imaging study was not done were excluded from the study. Informed consent was obtained from patient before undergoing CT or MRI scan. CT scan was done in 15 patients and MRI was done in 3 patients, where 2 patients underwent both CT and MRI scans.

CT Protocols: Siemens Somatom Spirit Dual Slice CT Scanner (Siemens Healthcare, Erlangen, Germany) was used in this study. Patients were scanned in supine position. Initial non-contrast CT scan of abdomen was obtained followed by contrast CT scan 40–60 seconds after IV bolus injection of 100 mL of iodinated contrast agents. Delayed scans after 10–15 minutes were obtained. Scanning parameters used were spiral mode with slice thickness of 6 mm and collimation 6 x 2.5 mm, pitch: 1.4; kVp: 130; mAs: 80. Multiplanar Reformatted images were obtained. Recon parameters included slice thickness of 3 mm and recon increment of 1.5 mm.

MRI Protocols: All patients were subjected to MRI scan using Siemens Avanto 1.5 Tesla B15 machine (Siemens Medical Systems, Erlangen, Germany). MR imaging of abdomen was performed in sagittal, coronal and axial planes using a combination of pulse sequences. MRI was performed with patient in supine position. Axial T1WI, T2WI and fat suppressed T2WI weighted fast spin echo images performed followed by sagittal T1WI, T2WI, coronal T1WI, T2WI, fat suppressed T2WI spin echo images using 3-6 mm slice thickness. Heavily T2-weighted MR urography was performed with thin-slice (HASTE) acquisition. Post gadolinium images were obtained in different planes.

Evaluation: Sixteen patients of renal replacement lipomatosis were examined to study the presence or absence of renal or ureteric calculus. The following findings were given close attention: location of calculus, size of calculus, Hounsfield Unit (HU) value of calculus, location of excessive fatty depositions like renal hilar, perinephric space and periureteric location, size of kidney, presence of hydronephrosis and renal function.

STATISTICAL ANALYSIS: Data were presented in terms of percentage and mean. Calculations were done using Microsoft Excel.

RESULTS: In our study, 16 patients were diagnosed with renal replacement lipomatosis. Age of presentation varied from 27 to 72 years with mean age of 50.2 years. Male: Female sex ratio was 1:1.3. Equal 50% of each side of kidney noted. Fifteen patients (93.8%) had associated renal pelvic or ureteric calculus [Figure 1]. One patient (6.2%) had left para-aortic mass, had renal replacement lipomatosis without associated calculus.

Fourteen patients (87.5%) of RRL had renal pelvic calculus where 2 patients (12.5%) had ureteric calculus [Table 1]. Out of fifteen patients of calculus related RRL, 8 patients (53.3%) had calculus size more than 40 mm in their larger dimension followed by 4 patients (26.7) with calculus size from 20 to 40 mm and 3 patients (20%) had calculus size less than 20 mm. The mean CT HU value of calculus was 1334±84.5.

The abundant fatty deposition or proliferations were noted as 3 patients (18.8%) had only renal hilar fatty deposition, 1 patient (6.2%) had renal hilar and perinephric fats [Figure 1,2 &3], 3 patients (18.8%) had renal hilar, perinephric and periureteric fats [Figure 4&5] and 9 patients (56.2%) had renal hilar and periureteric excessive fats [Table 2]. The affected kidney sizes were variable in RRL, where 8 patients (50%) had larger kidneys, 6 patients (37.5%) had smaller kidneys and 2 patients (12.5%) had normal-sized kidney. Ten patients (62.5%) of RRL associated with variable hydronephrosis. Delayed functioning of affected kidney were noted in 9 patients (56%) followed by non-functioning in 5 patients (31.2%) and 2 patients (12.5%) had normally functioning kidneys [Table 3].

Presence or absence of calculus	Frequency	Percent
No cal	1	6.2
Renal pelvic	13	81.2
Ureteric	1	6.2
Both renal pelvic and ureteric	1	6.2

Table 1: Showed the Location of Calculus in 16 Patients of Renal Replacement Lipomatosis (RRL)

Locations of excessive fatty depositions	Frequency	Percent
Renal hilar	3	18.8
Renal hilar and perinephric	1	6.2
Renal hilar, perinephric and periureteric	3	18.8
Renal hilar and periureteric	9	56.2

Table 2: Showed the Locations of Excessive Fatty Depositions in 16 RRL Patients

Renal Function	Frequency	Percent
Normally functioning	2	12.5
Delayed functioning	9	56.2
Non-functioning	5	31.2

Table 3: Renal Functions in 16 RRL Patients

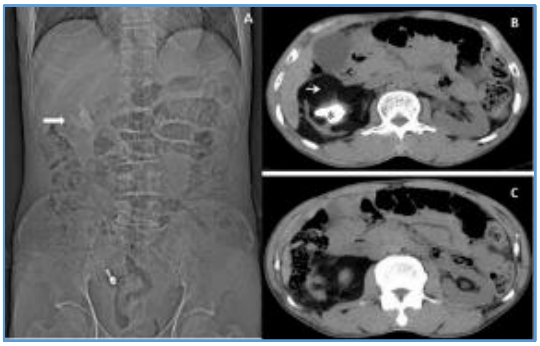

Figure 1

Figure 1: A 55-year-old male patient, presented with right flank pain and on & off fever. CT topogram (A) showed an irregular radio-opaque density in right renal fossa (block arrow). Axial non-contrast CT images (B& C) of abdomen showed a large fat density mass replacing the right kidney mainly filling renal hilum extending into perinephric space associated with markedly atrophic residual renal parenchyma (arrow). A large right staghorn calculus was noted within the atrophied renal tissue (*).

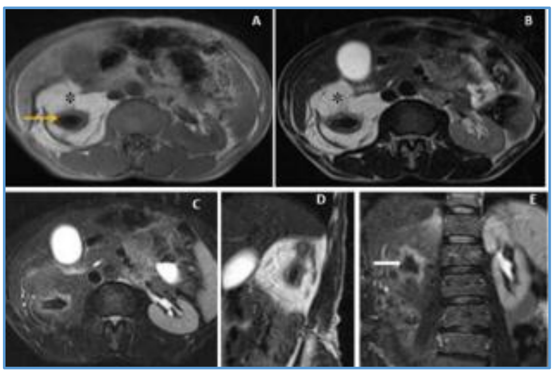

Figure 2

Figure 2: MR Images of same patient in Figure 1. Axial T1WI and T2WI MR images (A&B) and fat saturated T2WI (C) images showed a large irregular lobulated T1 and T2 hyperintense (*) lesion replacing the right kidney which got suppressed on fat saturation images. It surrounds the irregular outlined atrophied right renal tissue which bearing an irregular marginated T1 and T2 hypointense staghorn calculus (yellow arrow). Sagittal T1WI image (D) showed a large T2 hyperintense fatty lesion surround the T1WI hypointense calculus. Fat saturated coronal T2WI image showed large irregular T2 hypointense staghorn calculus (Block arrow).

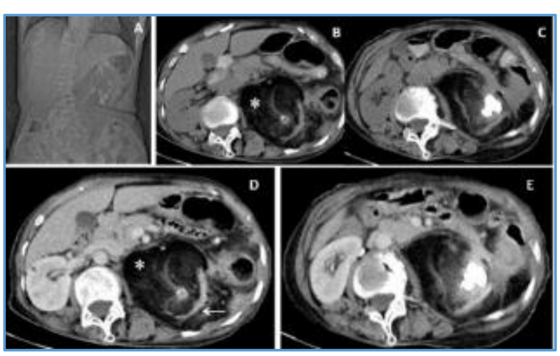

Figure 3

Figure 3: A 63-year-old female patient presented with history of back pain and lump in left side of abdomen. CT topogram (A) showed left-sided staghorn calculus. Axial non-contrast (B &C) and (D &E) contrast enhanced CT images showed an irregular larger staghorn calculus in left renal pelvis with atrophied left renal tissue. Extensive fatty tissue replacement of left kidney (*) was noted. Extension of fatty tissue was into left renal perinephric space. Peripherally located minimally enhancing atrophic residual left renal tissue was noted (Arrow).

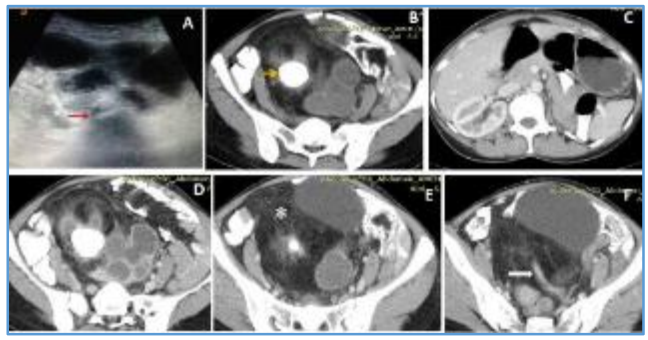

Figure 4

Figure 4: A 40-year-old female presented with right lower abdominal pain. Ultrasonography image (A) showed staghorn calculus (red arrow) with surrounding hyperechoic lesions around the calculus and associated hydronephrosis. Non-contrast CT (B) and contrast CT (C, D, E&F) images showed normal position of right kidney with ectopically located left kidney in right side of lower abdominopelvic cavity. Large staghorn calculus (yellow arrow) was noted with excessive fatty tissue deposition around the calculus, renal hilum, perinephric space, perivesical space and encircle the left ureter (*) causing superior and anterior displacement of urinary bladder. Hydronephrotic changes noted affected ectopic left kidney.

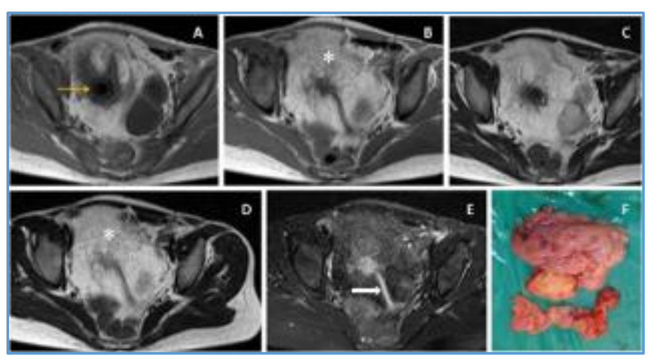

Figure 5

Figure 5: MR Images of same patient in Figure 4, Axial T1WI (A&B) and T2WI(C &D) images showed a large irregular lobulated T1 and T2 hyperintense (*) lesion replacing the ectopically located left kidney in right lower abdominopelvic cavity with a centrally located irregularly marginated T1 and T2 hypointense staghorn calculus (yellow arrow). Fat suppressed T2WI axial image showed suppression of fatty tissue around the left ureter (block

arrow). Postoperative nephrectomy specimen showed abundance of lobulated fatty mass.

DISCUSSION: Renal replacement lipomatosis is usually associated with chronic infection, longterm obstruction and calculi. It has been reported that renal calculous disease is found in more than 70% of cases.[1]

The mechanism of RPL is regarded as proliferation of fat secondary to renal atrophy and stimulation of chronic inflammation. The features on CT scan are extreme parenchymal atrophy and a characteristic distribution of the proliferated fat within the renal sinus, hilum and perirenal space.[2,3]

RRL, renal sinus lipomatosis, and fibrolipomatosis of the kidney represent a spectrum of changes in which normal renal sinus and perirenal fat increase in amount and replace the renal parenchyma. Renal sinus lipomatosis, the mildest form, is associated with obesity, renal atrophy of varying causes (e.g. aging and atherosclerosis), Cushing's syndrome or the use of exogenous steroids. [4] This mild form infrequently produces symptoms because of the absence of calyceal obstruction[5] and is a common finding at autopsy.[4] Invasion of adipose cells from the peripelvic fat into the kidney occurs along the blood vessels in the renal sinus.[6] At the other end of the spectrum is RRL, where the entire renal parenchyma is replaced with adipose tissue, usually secondary to calculous disease and longstanding inflammatory/infectious disease (e.g. renal tuberculosis).[4]

Cross-sectional imaging especially MRI can differentiate RRL from a fat containing renal tumour like angiomyolipoma, lipoma or liposarcoma. RRL is usually centred within renal sinus and renal hilum without mass effect upon the renal collecting system, while renal tumoural lesion exerts mass effect over renal collecting system. The three possible mechanisms of RRL are pressure atrophy of renal parenchyma by abundant proliferated renal sinus fat; invasion of renal parenchyma by proliferated fat; and finally fatty replacement of renal parenchyma associated with renal parenchymal destruction and renal atrophy.[7]

RRL and XGP have similar aetiopathogenic, clinical and radiological features. Both are characterised by atrophy and destruction of renal parenchyma, often associated with unilateral chronic renal infection, hydronephrosis or pyonephrosis, and calculous disease. The main difference between them is that in RRL as first reported by Brown in 1861,[6] the atrophic renal parenchyma in RRL is replaced by fatty tissue proliferation.[6,8] While XGP, initially described as staphylomycosis in 1916 by Schlagenhaufer, the xanthoma cells (lipid-laden macrophages) infiltrate and substitute necrotic renal tissue resulting in a lipomatous degeneration.[4,9] In XGP, there is increased lipid content in the inflammatory foam cells, whereas in RLK there is proliferation of large fat cells in the renal sinus, renal hilum, perinephric and periureteric spaces. The features on US and CT in xanthogranulomatous pyelonephritis are different.[1,4,10] XGP on USG, there was expansion of the hyperechoic mass of lipomatous tissue. [1]In XGP, CT scan shows pus filled, dilated calyces and xanthogranulomatous tissue showings CT attenuation varying from (-15 to +15 HU) reflecting the presence of intracellular fat droplets as against the pure fatty tissue seen in RRL.[11]

CT is the most accurate method of demonstrating the distinctive features of renal replacement lipomatosis. The calculi and the atrophied renal parenchyma were depicted easily. The abundant fatty tissue centrally has the characteristic attenuation of fat.[10] Although ultrasonography may suggest the diagnosis, CT demonstrated the distinctive features most accurately replacement lipomatosis. The CT features of RRL clearly distinguish this entity from hydronephrosis, cysts, and nonfat containing tumours based on attenuation values. Fat containing neoplasms arising in the parenchyma, renal sinus, renal capsule, or perinephric space, such as angiomyolipoma, lipoma, and liposarcoma can also be readily differentiated from RRL.

Heavily T2-weighted pulse sequences of MRI is used to obtain static water images of the urinary tract. T2-weighted MR urograms have proved excellent in the visualisation of the markedly obstructed collecting system, even if the renal excretory function is quiescent. T2-weighted MRU is less suitable for the imaging of abnormalities that occur in the nondilated urinary tract. The HASTE MR sequence is a breathing-independent T2–weighted spin-echo ultrafast sequence with great sensitivity for fluid detection. With it, breathing-related ghosting artifact, bowel motion, and magnetic susceptibly difference artifact from air and bowel are largely eliminated. It allows for differentiation of dilated ureter and distal ureter calculus without exogenous contrast. Diffuse and homogenous suppression of perirenal fat tissue with fat suppression sequence is also useful in differentiation from tumours including fat and arising in the parenchyma, renal sinus, renal capsule, or perinephric space, such as angiomyolipoma, lipoma, and liposarcoma.[12,13]

Recently, MRI and MR urography with gadolinium contrast have come up as the most accurate diagnostic tools for replacement lipomatosis showing the presence of perirenal fat intensity signal, obstruction, dilated ureter, level of obstruction, and atrophic kidney.[5,14]

Since majority of affected kidney in RRL is non-functioning, hence, for such patients, nephrectomy offers the adequate treatment.

CONCLUSION: The diagnosis of RRL is difficult to establish with conventional radiographic methods, particularly if the affected kidney is non-functioning. Although USG may show highly suggestive findings of fat proliferation in renal hilum; however, perinephric space and periureteric region fatty depositions are not visualised well on USG, hence cross-sectional imaging like CT and MRI scan helps in diagnosing RRL and proper delineation of extensions of excessive fatty tissue.

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