Renal Replacement Lipomatosis “Too Much Fat at Wrong Place”

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INTRODUCTION:
Replacement lipomatosis of the kidney is a rare disorder in which a massive fatty tissue proliferation occurs within the renal sinus, hilum and perirenal region. Replacement lipomatosis is the result of significant atrophy of the renal parenchyma and fatty proliferation, which is usually accompanied by renal stone disease and inflammatory changes in about 70% of cases. We report such case with its CT features.

CASE REPORT:
A 55 year old man presented with a sub-acute history of abdominal distension, yellowish discolouration of urine & sclera, burning micturition, haematuria and decrease appetite. His physical examination revealed pallor, hepatomegaly, splenomegaly, icterus and abdominal distension. Laboratory investigation results showed:

Haematology profile - Hb- 8.6, Tc- 8300
Urine Examination - Colour brown, Albumin- 2+, Pus cell s 2-3,rc
Renal function test - Urea- 19, serum creatinine- 0.9
Liver function test - Total bilirubin- 8.9, SGPT- 37, SGOT- 95, ALP
Ultrasoundography - Hepatomegaly, liver parenchymal disease, splenomegaly, minimal ascites, left renal mass

Computed Tomography - Left kidney shows stag horn calculus with excessive fat proliferation seen in renal sinus, renal pelvis and perinephric region with cirrhosis of liver and portal hypertension. Right kidney shows small calculi without hydrenephrosis or hydroureter.

DIAGNOSIS – Renal pelvis lipomatosis

DISCUSSION
Kutzman was amongst the first ones to describe the unusual entity in 1931 as reported by peacock and balle. Renal sinüs lipomatosis, Replacement lipomatosis and fibrolipomatosis of kidney are the terms used for this rare condition and represent a spectrum of changes. It is different from renal lipomas which are neoplastic, whereas this condition is thought to be degenerative process. Renal sinus lipomatosis, the mildest form is usually seen in the sixth and seventh decade associated with obesity, atherosclerosis or use of exogenous steroids. It has no clinical significance. Increased proliferation in the renal sinus exerts a mass effect on the intrarenal collecting structures, infrequently producing symptoms of obstruction. At the other end of the spectrum is Renal Replacement Lipomatosis where the entire renal parenchyma is replaced with adipose tissue, usually secondary to renal calculus and renal tuberculosis in 70% of the cases and occasionally after renal infarction. Usually it is unilateral in occurrence and rarely idiopathic. 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. In this case the patient presented in the fifth decade, which is the recorded age group, though it has also been reported in a young patient in second decade with posterior mediastinal lipomatosis. The usual complaint in literature is flank pain, dull aching, and non-radiating which was seen in the present case. They can also present with complaints of urinary tract infections due to urinary stones or mass in the abdominal cavity, as seen in our patient. Blood urea nitrogen and serum creatinine is usually within the normal limits. They are normal if the disease is unilateral with other well-functioning kidney. CT scan can confirm the fatty infiltration in the kidney and its distribution. In this case, the fatty infiltration on CT scan had led to the radiological diagnosis of lipomatous tumor. Xanthogranulomatous pyelonephritis and fat-containing neoplasms including lipoma, liposarcoma, and angiomyolipoma are the usual differential diagnosis reported in the literature with imaging techniques. The latter group can be excluded by absence of parenchymal atrophy. Ultrasoundography may suggest the diagnosis by demonstrating parenchymal atrophy or a hyperchoic renal sinus mass with a stone, however it is not diagnostic. 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.

CONCLUSION
The important aspect of this case is that renal replacement lipomatosis is a relatively uncommon entity and may be confused with renal neoplasms on radiology. It is associated with poorly or non-functioning kidney and histologically there is gradual replacement of renal parenchyma with mature adipose tissue leading to end stage renal disease. Renal replacement lipomatosis may be missed if not considered due to lack of experience, so we hope more physicians, urologists, radiologists, and pathologist become aware of this entity and recognize
REFERENCES