Renal replacement lipomatosis is a rare disease characterized by renal parenchymal atrophy and proliferation of inflammatory and fatty cells within the renal sinus, hilum, and perirenal region. The exact pathogenesis is unknown, however more than 75% of cases are associated with renal stones. Here, we provide images from a case with 10 years of follow-up, outlining the radiographic progression of the disease through time. Although only histopathology can confirm the diagnosis, high clinical suspicion in combination with the radiographic findings can differentiate the disease from other fat-containing entities.

Ten years ago, an asymptomatic 60-year-old man was investigated for persistent microscopic hematuria. At that point, on clinical examination, there was a palpable mass in his left flank. A computed tomography (CT) showed a 1.7 cm calculus in the left renal pelvis that was surrounded by an exuberant, inflammatory process (Fig. 1). A dimercaptosuccinic acid (DSMA) scan revealed a 34% differential function in the left kidney with normal serum creatinine (Fig. 2). After a course of antibiotics, he had a left ureteroscopy and laser lithotripsy with complete stone fragmentation. For the next 5 years, he remained stone free and was discharged from the clinic. Recently, after admission to hospital with pyrexia, he had an update CT scan, which showed a mild prominence of the left collecting system with characteristics of renal replacement lipomatosis (RRL) (Fig. 3).

RRL, also known as replacement fibrolipomatosis, is a rare entity that usually occurs in the 6th and 7th decade. It is mostly related with nephrolithiasis or xanthogranulomatous pyelonephritis. CT is the most used imaging modality, as it recognizes the fatty nature of the lesion and detects stones simultaneously. RRL has to be differentiated from other fat-containing tumors such as renal lipoma, angiomyolipoma, and liposarcoma.

Figure 1. Axial, coronal, and sagittal (left to right) noncontrast CT showing a staghorn calculus in the left renal pelvis surrounded by an exuberant inflammatory stranding in the perirenal fat. This is in contrast to the normal appearing contralateral right kidney.
Figure 2. DMSA scan showing lower uptake of radioactivity in the left kidney corresponding to the staghorn calculus and renal scarring in comparison to the normal right kidney. The differential uptake is 66% within the right kidney and 34% in the left kidney.
Figure 3. (A) Noncontrast (axial, coronal, and sagittal; left to right) and corresponding; (B) excretory phases of a split-bolus CT urogram shows proliferation of the renal sinus and perirenal fat of the left kidney. Also note the stretched, dilated, and displaced renal pelvis and thinning of the renal parenchyma. This is in contrast to the normal appearing contralateral right kidney.

References