Renal Replacement Lipomatosis With Coexisting Xanthogranulomatous Pyelonephritis in a Pregnant Woman

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Renal replacement lipomatosis of the kidney is a rare entity characterized by renal sinus and perirenal fat proliferation often caused by renal calculi. Most of the renal parenchyma is replaced by fat and the kidney is usually small, atrophic and nonfunctioning. We report magnetic resonance imaging and ultrasonography findings of a pregnant woman with xanthogranulomatous pyelonephritis and renal replacement lipomatosis coexisting in the same kidney.

INTRODUCTION

Renal replacement lipomatosis (RRL) is a rare disorder where extensive proliferation of fatty tissue occurs within the renal sinus, hilum, and perirenal region. It is often associated with renal calculi, longstanding chronic inflammation, and urinary infection.1-3 Renal replacement lipomatosis and xanthogranulomatous pyelonephritis (XP) have several overlapping clinical features and imaging findings. It is extremely rare that both conditions coexist in the same kidney. To our knowledge, only few such cases have been reported in the literature.1-5 We report the magnetic resonance (MR) and ultrasonography findings of our patient with XP and RRL, additionally associated with nephrocutaneous fistula.

CASE REPORT

A 30-year-old pregnant woman presented in her 12th week of pregnancy with a discharging sinus in the left loin for the past 2 weeks. She gave a history of drainage of the left loin abscess 4 months earlier. History of episodic left flank pain and fever for the past 2 years was present. No significant finding was noted on physical examination apart from the discharging sinus in the left loin. Urine examination showed numerous pus cells. Culture of urine and discharge from the sinus grew Staphylococcus aureus.

Ultrasonography of the left renal fossa showed hyperechoic mass with thin hypoechoic rim of residual renal parenchyma (Figure 1). The reniform shape of the kidney was preserved. Dense echogenic foci with distal acoustic shadowing was embedded in the renal sinus fat suggesting the presence of calculi.

Intravenous urography and abdominal computed tomography was not performed considering the pregnant status of the patient. Magnetic resonance imaging revealed atrophy of the left kidney, which was almost completely replaced with adipose

Figure 1. Ultrasonography shows multiple echogenic foci (C) with distal acoustic shadowing in the left kidney.
tissue. Marked proliferation of the fatty tissue was also seen in the perinephric space (Figures 2 to 4). Multiple small rounded signal voids was seen in the central renal sinus (Figure 2), suggesting calculi. The fistulous tract was noted extending from the left kidney to the left loin, being hyperintense on T2 weighted images (Figure 3). Renal replacement lipomatosis with coexistent XP was strongly suggested on the basis of MR findings. After 2 weeks of conservative treatment, she underwent nephrectomy, and the final pathological diagnosis was XP associated with massive RRL. Both the patient and the fetus did well postoperatively.

DISCUSSION
In RRL, renal parenchyma is gradually replaced by fibrous and fatty tissue. It is usually secondary to chronic infection and renal calculi in more than 70% of cases. Clinically, patients present with recurrent urinary tract infections, fever, and flank pain. Ultrasonography features suggestive of RRL include an echogenic mass representing the lipomatous tissue with central high-density echoes with distal acoustic shadowing caused by the calculus. Computed tomography demonstrates a lipomatous mass, with negative attenuation values replacing renal parenchyma along with calculi. Magnetic resonance imaging has high sensitivity and specificity to identify fat as hyperintense signal on T1 and T2 weighted images. Although it is not as good as computed tomography in the demonstration of calculus, it is superior for the demonstration of the extent of the disease and its complications such as fistula, especially if surgery is indicated. In a pregnant patient, MR imaging provides sufficient information without the need for the contrast material and risk of ionized radiation. Important differential diagnoses that should be considered include XP, angiomyolipoma, and liposarcoma. Associated findings like renal atrophy, calculus, and hydronephrosis can exclude the possibility of lipomatous tumors.
Xanthogranulomatous pyelonephritis can be extremely difficult to differentiate from RRL as both entities show overlapping clinic-etiologic features. However, chronic obstruction in XP causes destruction of renal parenchyma, and histological examination reveals lipid-laden macrophages (foamy histiocytes) diffusely infiltrating the renal parenchyma, in contrast to RRL where fat cells remain outside of the atrophied renal parenchyma. Computed tomography typically shows an enlarged kidney with multiple hypodense egg-shaped areas of fluid collection along with renal calculi. Xanthogranulomatous pyelonephritis is graded into 3 stages: stage I (nephric) is a localized disease confined to the renal parenchyma; stage II (perinephric) lesions involve perinephric fat; and stage III (paranephric) lesions extend beyond the Gerota’s fascia into the retroperitoneum.

Although coexistence of XP and RRL is extremely rare in the same kidney, one should become aware of this entity, so that it is not mistaken for a fatty neoplasm. Magnetic resonance imaging is important in pre-operative diagnosis and distinguishing it from other conditions, as it accurately demonstrates the morphologic changes, extent of the disease, and its complications.

CONFLICT OF INTEREST
None declared.