

Inflammation and Infection

Massive Renal Replacement Lipomatosis With Foci of Xanthogranulomatous Pyelonephritis in a Horseshoe Kidney



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ABSTRACT

Renal replacement lipomatosis (RRL) is a rare condition that occurs at the end of the spectrum of renal tissue replacement by fat. Xanthogranulomatous pyelonephritis (XGP) is a granulomatous inflammation characterized by destruction of renal parenchyma and replacement by lipid-laden macrophages. We present the case of a 75-year-old man who complained of severe anemia 34 years after right nephrolithotomy. Computed tomography revealed a huge low-density mass with renal parenchyma atrophy on the right side of horseshoe kidney. Right nephrectomy was performed. Pathological diagnosis was RRL with XGP. This is the first report of RRL with XGP in a horseshoe kidney.

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Introduction

Renal replacement lipomatosis (RRL) is a rare condition and is defined as growth of fat within the renal sinus, hilum and perirenal space with the atrophic renal parenchyma. RRL is related to a chronic inflammatory disease state, and is reportedly associated with renal calculi in 76% to 79% of previously reported cases.¹ The clinical importance of RRL is that it simulates fat-containing tumors in the kidney or its vicinity. Xanthogranulomatous pyelonephritis (XGP) is a granulomatous inflammation characterized by destruction of renal parenchyma and replacement by lipid-laden macrophages. A few reports showed a coexistence of XGP with RRL.² We here report a rare case of RRL concomitant with XGP in a horseshoe kidney.

Case presentation

A 75-year old man was admitted with general fatigue and breathlessness. He also claimed a severe weight loss (−20 kg) during 2 years. The patient had undergone nephrolithotomy once on the right side of horseshoe kidney 34 years before. The laboratory examination showed severe anemia (hemoglobin = 7.2 g/dl), elevated serum CRP level (6.3 mg/dl) and low serum albumin level (1.5 g/dl). A white blood cell count was within normal limit. Urine examination showed increased white blood cell count. A contrast-enhanced computed tomography (CT) revealed a low-density

mass (17.1 × 13.9 × 19.4 cm) with renal parenchyma atrophy and hydronephrosis on the right side of horseshoe kidney. Residual nephrostomy tube was identified in the inner side of the right kidney. The mass expanded at the renal sinus, hilum and perirenal space, and deviated surrounding organs (Fig. 1). No renal calculi were seen. Well-differentiated liposarcoma was strongly suspected from radiological findings. The patient underwent the resection of the right kidney, surrounding abnormal fatty mass. During the operation, there were severe adhesions around the right kidney. A specimen was removed. Cut section of the specimen grossly showed atrophic renal parenchyma surrounded by tumorous fatty tissues continuous to renal sinus and perirenal fat (Fig. 2). The microscopic findings showed a proliferation of large mature adipose cells with inflammatory cell infiltration (Fig. 3A). No malignant cells were identified. Sclerosed glomeruli, thyroidization of tubules, interstitial fibrosis and foci of aggregates of lipid-laden macrophages were found in the atrophic kidney (Fig. 3B). The pathological diagnosis was RRL with XGP. He unfortunately died of aspiration pneumonia on 37th postoperative day.

Discussion

RRL is a rare condition characterized by the proliferation of the renal sinus, renal hilum, and perirenal fatty tissue with the atrophic renal parenchyma. In its most extensive RRL, a total atrophy of renal parenchyma was found with complete fibrofatty replacement.³ It is usually unilateral and occurs as a result of severe renal atrophy or destruction usually due to chronic calculus disease, chronic pyelonephritis and renal tuberculosis. The coexistence of RRL with XGP

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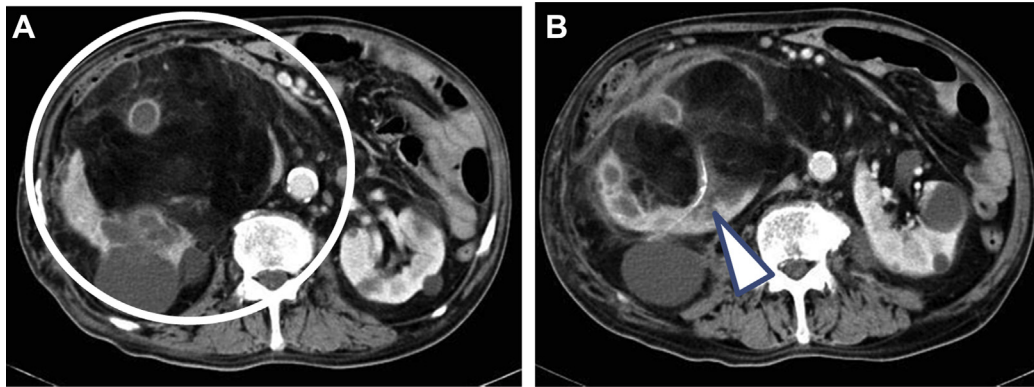


Figure 1. (A) CT scan showed the 17.1 × 13.9 × 19.4 cm fat-density mass (circle) with renal parenchyma atrophy, hydronephrosis on the right side of horseshoe kidney (B) residual nephrostomy tube (arrow head).

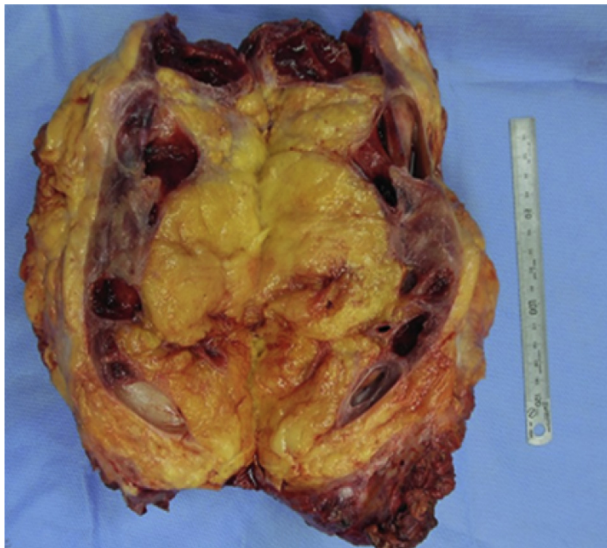


Figure 2. On cut surface, the specimen revealed atrophic renal parenchyma surrounded by overgrown fat tissues at renal sinus and perirenal space.

has been reported in a few cases.² Our report is the first case of RRL with XGP in a horseshoe kidney.

CT is reportedly the best imaging modality to diagnose RRL and differentiate it from other fat containing lesions such as renal angiomyolipoma, retroperitoneal lipoma, and liposarcoma. Angiomyolipoma is usually a well-circumscribed focal mass. In the diffuse form, angiomyolipoma will distinguish themselves from RRL by their parenchymal involvement. Lipoma is well-circumscribed and encapsulated. Liposarcoma is typically located in a retroperitoneal extrarenal location and is different from RRL. And in all three of these entities, renal function is expected to be preserved. RRL is often linked to renal calculus disease and chronic renal infection. But there is sometimes a difficult case to differentiate RRL from liposarcoma.² In our case, renal atrophy was not severe. We firstly diagnosed that tumor as well-differentiated liposarcoma and could not diagnose RRL preoperatively.

The horseshoe kidney is the most common renal fusion anomaly. Impaired drainage of the collecting system and associating ureteropelvic obstruction predispose the patient to urinary tract infection and urolithiasis.⁴ In our case, the patient had undergone nephrolithotomy once on the right side of horseshoe kidney, but no residual renal calculi were found. Hydronephrosis and chronic pyelonephritis caused by impaired drainage of the collecting system, renal atrophy, chronic infection due to the residual nephrostomy tube, or aging may be associated with the occurrence of RRL.

XGP is an atypical form of chronic pyelonephritis. Its exact etiology remains unknown, but it usually occurs in association with

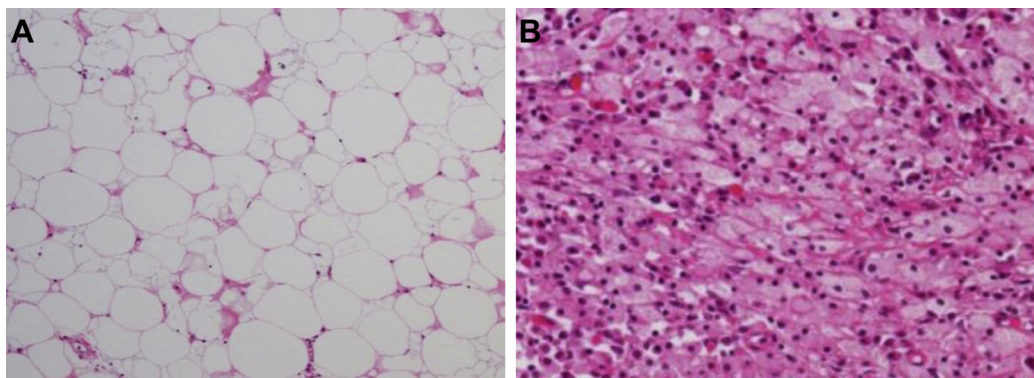


Figure 3. (A) Within the renal sinus and perirenal space, mature adipocytes with interspersed inflammatory cells (lymphocytes and plasma cells) were observed. (HE stain; magnification, ×200). (B) Foci of aggregates of lipid-laden macrophages were identified in the renal parenchyma. (HE stain; magnification, ×200).

nephrolithiasis, urinary tract obstruction, and/or chronic urinary tract infection.⁵ In our case, foci of aggregates of lipid-laden macrophages were identified in the renal parenchyma. XGP may be occurred by chronic pyelonephritis due to impaired drainage of the collecting system by horseshoe kidney and/or by chronic infection due to the residual nephrostomy tube.

Conclusion

We reported the first case of RRL with XGP in a horseshoe kidney. This was a very difficult case to differentiate RRL from liposarcoma preoperatively.

Conflict of interest

None.

Financial disclosure

The authors declared that this study has received no financial support.

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