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CLINICAL CASE REPORT

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A Renal Inflammatory Myofibroblastic Tumor Similar to Cystic Renal Cell Carcinoma

One Case Report

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Abstract: We describe and analyze the clinical course and imaging findings of a case of a renal inflammatory myofibroblastic tumor (IMT) that is similar to cystic renal cell carcinoma.

''Solitary cysts'' on the left kidney were found during a health examination of a 60-year-old female. The patient also had hypertension. She had undergone surgeries twice for limb trauma fracture and had no definite record of hepatitis. There was no tenderness with percussion of the kidney area or edema in the lower extremity. The renal function results, including serum creatinine, blood urea nitrogen, and blood urea, were within the normal range. No gross hematuria or microscopic hematuria was found. An 8.7 cm × 9.2 cm mixed echogenic mass at the upper pole of the left kidney was observed with ultrasound, the majority of which was an anechoic mass that was slightly protruding from the renal capsule and had well-circumscribed borders. After a bolus injection of an ultrasound contrast agent, the mass had rapid enhancement with fast fading. An approximately 9.4 cm × 10.1 cm round-like cyst lesion at the upper pole of the left kidney was revealed by computed tomography (CT) examination of the abdomen; it had edge finishing with well-circumscribed borders. The upper inner wall of the lesion was thick with crescentic soft tissue. The solid content had gradual enhancement on enhanced CT scans. A kidney tumor was considered based on the CT findings.

Based on the preoperative examination, the left renal cystic masses were resected. Intraoperative frozen sections were used to further clarify the nature of the lesion, and no significant malignant cells were observed; therefore, the kidney was not removed. The pathological diagnosis was renal IMT. After surgery, the patient recovered and did not have recurrence or metastasis over the course of long-term follow-up.

CT images of our patient with renal cystic disease are categorized as Fuhrman grade IV and typically indicate the presence of malignant lesions. However, gradual enhancement of the solid content in our case is different from typical cystic renal cell carcinoma. The nature of the lesion was further identified using intraoperative frozen sections, which helped avoid unnecessary nephrectomy.

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Abbreviations: ALK = anaplastic lymphoma kinase, CEUS = contrast-enhanced ultrasound, CT = computed tomography, IMT = inflammatory myofibroblastic tumor, MRI = magnetic resonance imaging, PET/CT = positron emission tomography/computed tomography, US = ultrasonography.

INTRODUCTION

Inflammatory myofibroblastic tumor (IMT) is a rare, benign lesion that can affect nearly all parts of the body.[1] IMT is common in the lungs, which is followed by the abdomen, retroperitoneum, and pelvis, but it rarely occurs in the kidney.[1–3] Fisch and Brodey[4] reported the first case of IMT in the kidney, which was followed by other case reports. IMT in the kidney has low clinical specificity because of its diverse imaging findings; therefore, the lesions must be surgically removed to determine the diagnosis. Finally, the prognosis of IMT in the kidney is good.[2,5] Therefore, clinicians need to be familiar with the imaging findings so that they can develop an appropriate treatment strategy before surgery. We report 1 case of renal IMT that was found during a health examination, and its imaging findings were similar to cystic renal cell carcinoma. However, observation of intraoperative frozen sections allowed us to rule out kidney cancer, preventing unnecessary nephrectomy. The patient signed informed consent forms, allowing for publication of the relevant clinical and imaging data from her case.

CONSENT

In our case, the patient signed related informed consent for the publication of clinical data and images.

CASE REPORT

Solitary cysts were found in the left kidney of a 60-year-old female farmer during a health examination 2 weeks ago. She did not have a backache, soreness in the waist, urinary urgency, dysuria, chills, fever, or other discomfort when she visited our clinic. The kidneys, ureter, and bladder (KUB), and intravenous pyelogram (IVP) examination showed left renal pelvis pressure signs. The patient was hospitalized with a left renal cystic lesion. She was in good mental health and had a normal appetite with no significant changes in body weight. The patient was diagnosed with hypertension and prescribed oral Captopril. She underwent right-hand trauma fracture surgery 6 years before and left leg fracture surgery 5 years before. Her postoperative recovery was good. The patient had no history of hepatitis, diabetes, tuberculosis, or blood transfusion. She was not in the habit of smoking or drinking heavily. She had no history of drug or food allergies.

On admission, she had a body temperature of 36.6°C, heart rate of 70 beats/min, respiratory rate of 20 breaths/min, and

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blood pressure of 117/97 mm Hg. The patient had a ruddy complexion with no eyelid edema or swollen lymph nodes in the neck. The patient did not have tenderness on percussion of the kidney area. The abdomen was soft and flat with no tenderness or rebound tenderness. The liver and spleen were not enlarged. No abdominal masses were felt on palpation. There was no lower extremity edema. Laboratory tests revealed renal function results as follows: serum creatinine of 59 μmol/L (reference range, 44–133 μmol/L), blood urea nitrogen of 3.16 mmol/L (reference range, 2.86–8.20 mmol/L), and blood urea of 274 μmol/L (reference range, 90–420 μmol/L). Liver function tests showed a white globulin ratio of 1.4 (reference range, 1.5–2.5), alanine aminotransferase of 69 U/L (reference range, 3–50 U/L), aspartate aminotransferase of 62 U/L (reference range, 3–40 U/L), and total calcium of 2.02 mmol/L (reference range, 2.08–2.60 mmol/L). The remaining biochemical indicators were in the normal range. Urine occult blood, urine protein, and urine bilirubin were normal with ureteroscopic white blood cell of 0–3/hp. The quantitative examination of hepatitis B showed hepatitis B surface antigen-negative 0.6 S/N (S/N $\geq$ 2.0 positive), hepatitis B e antigen-negative 0.2 S/CO (S/CO $\geq$ 1.0 positive), hepatitis B core antibody inhibition rate-positive 96.7% (inhibition rate $\geq$50% positive), hepatitis B e antibody inhibition rate-positive 98.6% (inhibition rate $\geq$60% positive), hepatitis B surface antibody-positive 24.2 IU/L (concentration $\geq$10.0 IU/L positive), and hepatitis B core antibody IgM-negative 0.2 S/CO (S/CO $\geq$ 1.0 positive). In addition, the patient was negative for hepatitis B surface antigen, negative for hepatitis C antibody, negative for human immunodeficiency virus antibody, and negative for Treponema pallidum antibody. There were no abnormalities in routine blood and stool examinations or in the blood coagulate functions. Chest x-ray showed no abnormalities.

Ultrasonography (US) showed an 8.7 cm × 9.2 cm mixed echogenic mass at the upper pole of the left kidney; the majority of the mass was echoless, and it was slightly protruding from the renal capsule with well-circumscribed borders. After a bolus injection of US contrast agent, the mass was rapidly enhanced, while there was a ''developing defect'' within the tumor, and the contrast agent in the mass quickly washed out. Contrast-enhanced ultrasound (CEUS) imaging indicated a left renal cell carcinoma with necrosis. Kidney CT examination revealed an approximately 9.4 cm × 10.1 cm round-like cyst lesion at the upper pole of the left kidney with edge finishing and well-circumscribed borders. The upper inner wall of the lesion was thick with crescentic soft tissue density. With enhanced CT scanning, the solid content had mild enhancement in the early stage and obvious enhancement in the later stage. The left renal calices were under slight partial pressure without hydronephrosis of the renal pelvis. Based on CT imaging, a kidney tumor was considered (Figure 1).

Based on the preoperative examination, the left renal cystic masses were resected. During surgery, local thickness was found at the cystic wall, which was approximately 2 cm × 5 cm cm and had a hard texture. To further evaluate the lesion, it was sent for frozen sections. The frozen analysis suggested that there were no obvious malignant cells. As a result, the affected kidney was not resected. Finally, the pathological diagnosis was left renal IMT (Figure 2). The following were the immunohistochemistry findings: smooth muscle actin (SMA) (++), CD68 (+), anaplastic lymphoma kinase (ALK) (−), CD34 (−), CD10 (+), and Ki67 interspersed +. The patient recovered and was discharged 9 days after the surgery. One month after operation, the patient was reexamined by US in our institution, which showed that the left kidney had no evidence of recurrence, and no obvious abnormal occurred in right kidney. The subsequent ultrasound or CT examinations were accomplished in local hospital. With a follow-up of 5 years and 3 months, the patient remained asymptomatic.

DISCUSSION

IMT was previously referred to as ''inflammatory pseudotumor.'' Because of its varied pathological manifestations, different names have been used for IMT, including plasma cell granuloma, inflammatory myofibroblastic proliferation, and xanthomatous pseudo tumor.[6] With an improved understanding of its pathological and immunohistochemical features, the name IMT has become widely accepted.[7] The etiology and pathogenesis of IMT remains unclear. Microbial infections and their immune suppression status may play important roles in IMT occurrences; for example, viral DNA sequences are found in the spindle cells of IMT, and there is a higher incidence in individuals with immunosuppression conditions or who are taking corticosteroid treatment.[8–11] Trauma and chronic hepatitis B infection are also thought to play a role in tumorigenesis.[12] ALK gene rearrangement has been confirmed in some IMTs, suggesting that ALK rearrangement is correlated with tumor progression.[13] Interestingly, the patient in our report had hepatitis B infection without persistent chronic infection; also, she had a history of trauma to 2 limbs without trauma at her left waist. Our case did not have systemic disease or immunosuppression.

The distribution of renal IMT varies for different age groups, and it is rare in children.[3,14] The incidence of IMT is more common in men. Although there are no specific clinical symptoms of IMT, patients commonly report pain and hematuria.[3,14] There are usually no obvious abnormalities in laboratory tests, except for the presence of microscopic hematuria.[3] No clinical symptoms were found in the physical examination in our case, which is different from previous reports. Lesions are generally solitary, though there can occasionally be multiple lesions, and they are usually 1.0 to 10.0 cm in size.[14,15] US is usually nonspecific and hyperechoic or hypoechoic. CT shows ill-defined borders and slight homogeneous enhancement; there is occasionally significant enhancement with a clear edge.[14,15] For some cases, a renal IMT may also have a thick-walled cystic mass and ill-defined borders, which is occasionally accompanied by calcification.[14,15,16] The magnetic resonance imaging (MRI) findings for renal IMT vary, but they commonly include hypointensity on T1-weighted image and T2-weighted image sequences.[14] On enhanced MRI, the tumor has a poor blood supply.[14,15] Unlike previous reports, our case was primarily anechoic due to its cystic content. Ours is the first report with CEUS findings for a renal IMT in which the outflow type of enhancement was similar to the enhancement type for cystic renal cell carcinoma. On CT, unlike in reported thick-walled cystic lesions, such as renal cell carcinoma tumor with necrosis, our case had a clear edge and mural nodules, which is similar to cystic renal cell carcinoma. For cystic renal cell carcinoma, a conservative surgical plan is optional.[17] In our case, the significant solid content and Fuhrman grade IV status are typically indicative of malignant lesions. However, unlike the typical early significant enhancement of a cystic renal cell carcinoma, our case had gradual enhancement on enhanced CT. Therefore, a neoplastic lesion was considered based on the CT findings.

CEUS technology can dynamically indicate blood perfusion state in tumor tissue by ultrasound contrast agent. In our

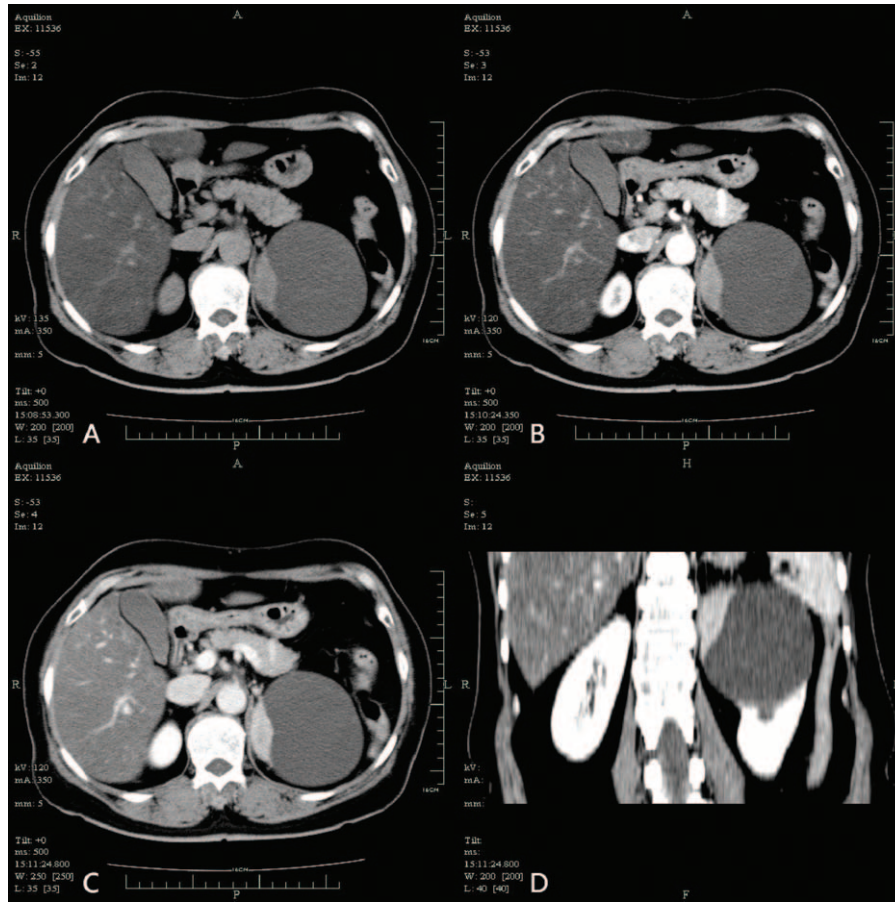

FIGURE 1. Abdominal CT scans of the patient. (A) Abdominal plain CT scans showing a round-like cystic mass with edge finishing, clear boundaries, and upper inner wall with significant soft tissue on the left kidney pole. (B, C) Enhanced CT scans showing solid content with gradual enhancement and cystic content with no enhancement. CT = computed tomography.

case, ultrasound contrast agent rapidly entered into the solid component of renal IMT in arterial phase, which presented on focal hyperenhancement. When the contrast agent quickly washed out in delayed phase, echo intensity within tumor was significantly lower than renal parenchyma, which showed low echo. Although the solid component in our case also had significant enhancement in early phase, but less than CEUS, it might be caused by 2 different contrast methods. Ultrasound contrast is more real time and sensitive for microvascular perfusion state. In our case, the enhanced degree in the late stage of enhanced CT was lower than renal parenchymal strengthen degree. The solid component showed relatively low density, which was similar with that in CEUS. Its enhancement value gradually increased by measuring the value, which showed delayed enhancement. We presume that it might indicate that CT contrast agent was accumulated in nonvascular tissues. In positron emission tomography/computed tomography (PET/CT), renal IMTs have high fluoro-2-deoxy-d-glucose (FDG) uptake, and transitional cell carcinoma is considered because it is different from the typical renal cell carcinoma FDG uptake.[18] Therefore, when kidney tumor imaging reveals atypical kidney cancer, the differential diagnosis needs to include renal IMT.

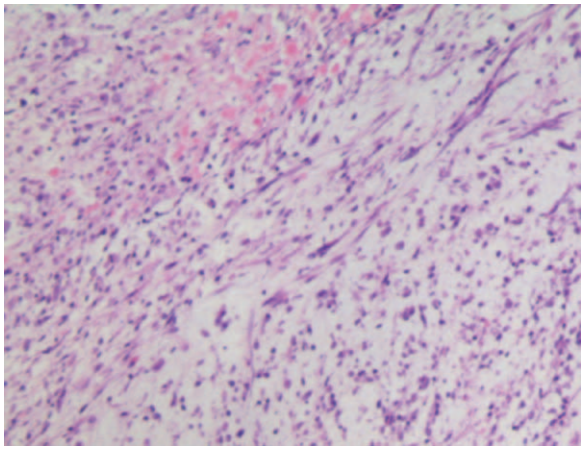

FIGURE 2. Pathology of the patient, including myofibroblastic proliferation, fusiform, rare mitotic count, and a high number of plasma cells.

Some have thought that biopsy does not contribute to preoperative diagnosis of renal IMT.[19] However, in a recent report, core needle biopsy was applied to confirm the diagnosis, and the case underwent spontaneous resolution.[20] Therefore, the value of the kidney biopsy for IMT merits further evaluation. Most renal IMT cases have undergone surgical resection, and nephrectomy is usually performed due to the mimicry of

malignancy on imaging findings, such as renal carcinoma or Wilms tumor.[5,15,21,22] In some cases that were not treated by resection, corticosteroid therapy was somewhat beneficial.[9,10] In our case, because the preoperative imaging was similar to cystic renal cell carcinoma, surgical resection was implemented, and no recurrence was observed during follow-up.

CONCLUSION

Therefore, renal IMT may occasionally present as a cystic lesion with gradual enhancement on enhanced CT, and surgical removal of renal IMT is an optional treatment. The preoperative imaging and biopsy should be evaluated. If necessary, intraoperative frozen section can be performed to help avoid unnecessary nephrectomy in renal IMT patients.

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