Clinical Case Report

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Inflammatory myofibroblastic tumor of kidney together with ipsilateral perinephric and periureteric fibrosis

A case report and literature review

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Abstract

Rationale: Both inflammatory myofibroblastic tumor (IMT) and retroperitoneal fibrosis are rare lesions, but kidney involvement is more rare. It is the first study about IMT of the kidney in a patient with perinephric and periureteric fibrosis and we hold that fibroblast proliferation may be an intermediate status in oncogenesis of IMT. But further investigation is necessary in order to better clarify the relationship between fibroblast proliferation and IMT.

Patient concerns: A 54-year-old female presented no positive signs except dull back pain after overwork.

Diagnoses: On the basis of the urinary ultrasonography and computed tomography (CT) scan, we strongly suspected a renal cell carcinoma.

Interventions: Considering the little remaining function of the right kidney and the possibility of malignancy, we performed a laparoscopic right radical nephrectomy.

Outcomes: According to the analysis of the postoperative paraffin section and immunohistochemistry assay, a final diagnosis of IMT and retroperitoneal fibrosis nodules was made.

Lessons: Both IMTs are rare lesions and its etiology and pathogeny are unclear. It is the first study about IMT of the kidney in a patient with perinephric and periureteric fibrosis. This report suggested that fibroblast proliferation may be an intermediate status in oncogenesis of IMT, but further investigation is necessary in order to better clarify the relationship between fibroblast proliferation and IMT. The preoperative diagnosis of renal IMT remains difficult. Preoperative fine-needle aspiration or percutaneous biopsy and intraoperative frozen section were applied to confirm the diagnosis to avoid unnecessary nephrectomy, especially in patients with renal insufficiency, bilateral masses, or a solitary kidney.

Abbreviations: ALK = anaplastic lymphoma kinase, CT = computed tomography, EBV = Epstein–Barr virus, ECT = emission computed tomography, GFR = glomerular filtration rate, IMT = inflammatory myofibroblastic tumors, LCA = lymphocytotoxic antibody, SMA = smooth muscle antigen, UPJO = ureteropelvic junction obstruction.

Keywords: fibrosis, hydronephrosis, inflammatory myofibroblastic tumor, intermediate status, kidney

1. Introduction

Inflammatory myofibroblastic tumor (IMT) is a distinctive myofibroblastic neoplasm with a predilection for the lung.[1] In the urological tract, these tumors can occur in the bladder and prostate, but kidney involvement is rare.[2] The preoperative diagnosis of renal IMT remains difficult due to its nonspecific manifestations and the mimicry of malignancy by imaging, such as renal cell carcinoma, which possibly leads to nephrectomy.[3–6] To our knowledge, only about 30 cases of IMT of kidney have been reported in the English literature thus far. The majority of these cases were misdiagnosed as renal carcinoma before surgery. In the present report, we report 1 case of IMT of kidney together with ipsilateral perinephric and periureteric fibrosis and systematically review the literature of IMT to facilitate study the etiology, pathogeny, and clinical diagnosis of IMT. It is the first study about IMT of the kidney in a patient with perinephric and periureteric fibrosis and we hold that fibroblast proliferation may be an intermediate status in oncogenesis of IMT. But further investigation is necessary in order to better clarify the relationship between fibroblast proliferation and IMT.

2. Case report

A 54-year-old female was referred to the Department of Minimal Invasive Urology of Provincial Hospital affiliated to Shandong University (Jian, China) in July 2016. Right hydronephrosis was detected by ultrasound during a health examination 1 week

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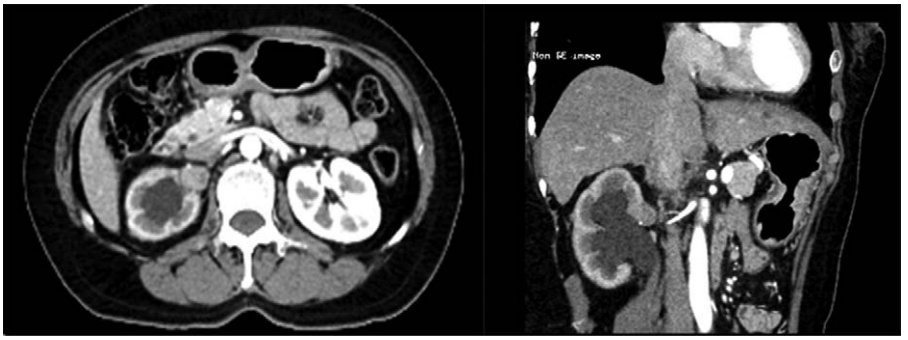

Figure 1. Computed tomography image of inflammatory myofibroblastic tumor of the kidney showed the presence of a slightly enhanced mass.

before her visit. There were no positive signs except dull back pain after overwork. Routine blood examination was normal. Urinalysis revealed an elevated number of red blood cells. The renal function results were as follows: serum creatinine of 87.9 mmol/L (reference range, 40–105 μmol/L) and blood urea nitrogen of 7.8 mmol/L (reference range, 2.8–7.14 mmol/L). Other laboratory values were within normal limits.

Urinary ultrasonography showed expansion of the right renal pelvis, and a thick and stiff upper section of the right ureter. Computed tomography (CT) scan revealed the presence of a slightly enhanced mass, 1.5 x 1.5 cm in size, in the upper pole of the right kidney, close to the renal holus (Fig. 1). The renal emission CT (ECT) showed left kidney glomerular filtration rate (GFR) of 47.39 mL/min, and right kidney GFR: 4.25 mL/min, for a total renal GFR of 51.64 mL/min. This suggested that the right kidney function was severely impaired. Retrograde urography was performed before surgery. Under the cystoscope, no abnormality was observed in the bladder. A ureteral catheter was inserted 15 cm in the right ureter until resistance was met. Retrograde urography showed an obvious stenosis at the upper segment of ureter, with slight dilation of the pelvis and upper segment of ureter (Fig. 2).

On the basis of the radiologic findings, we strongly suspected a renal cell carcinoma. Considering the little remaining function of the right kidney, we performed a laparoscopic right radical nephrectomy. During the surgical procedure, we found the kidney seriously adhered to adjacent structures, especially the inferior vena cava and duodenum. After careful dissection, a laparoscopic right radical nephrectomy was consequently performed. The postsurgical, macroscopical anatomy of the specimen revealed a hard and brown tumor with an obscure boundary located in the kidney near the holus. The renal parenchyma was thick and pale. The upper ureteral wall was thick and stiff, with a small nodule adhered to it.

Analysis of the postoperative paraffin section showed inflammatory cells and fibroblasts that were proliferated such as focal nodular hyperplasia. Immunohistochemistry assay was positive for smooth muscle antigen (SMA), vimentin, caldesmon, desmin, anti-lymphocytotoxic antibody (LCA), CD68, and CD38, but negative for HMB45, S-100, and CD34. On the basis of these results, a final diagnosis of IMT was made (Fig. 3). The fibroblasts exhibited nodular hyperplasia with collagen degeneration and were closely adhered to the ureter and the surrounding tissue, which caused ureteral stenosis. According to the immunohistochemistry assay results (positive for SMA, negative for HMB45, S-100, CD34, and desmin), the per-ureter lesion was diagnosed as retroperitoneal fibrosis nodules.

3. Discussion

IMT is an uncommon tumor, first reported in the lung in 1937 by Bahadori and Liebow.[1] IMT invasion sites have been reported to be mainly in the lung, but it can affect various organs (from the central nervous system to the gastrointestinal tract).[2] It has been reported to affect individuals of both sexes (females more than males) and has been seen in a wide range of age groups (reported cases in patients between the ages of 3 and 72 years).[7] However, in the urinary tract, reports of IMT of the kidney have been extremely rare.[2]

IMT was initially called as pseudoneoplasm, as the myofibroblast was identified and this cell type was eventually recognized as the principal cell type in the inflammatory pseudotumor, which designated this tumor as IMT.[8] Because the term "pseudoneo-

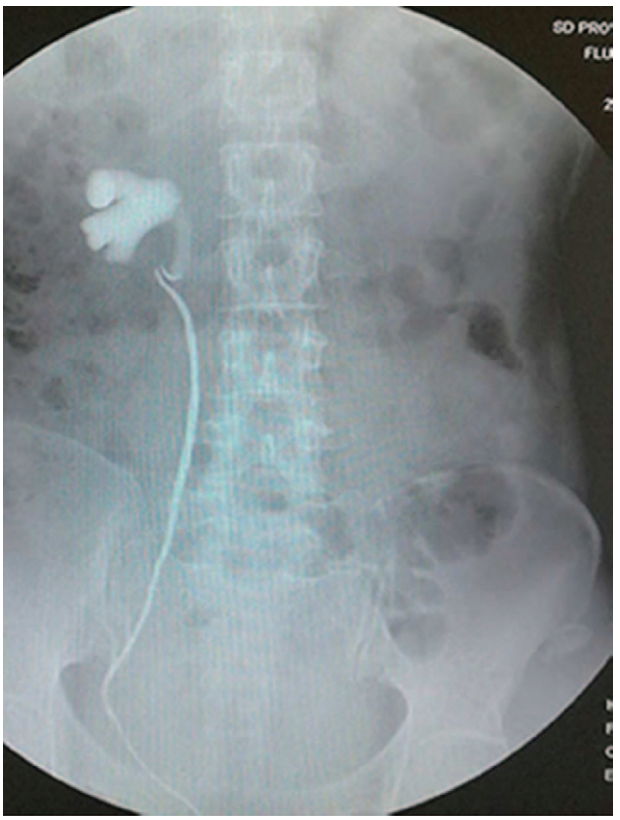

Figure 2. Retrograde urography image showed an obvious stenosis at the upper segment of ureter and hydronephrosis.

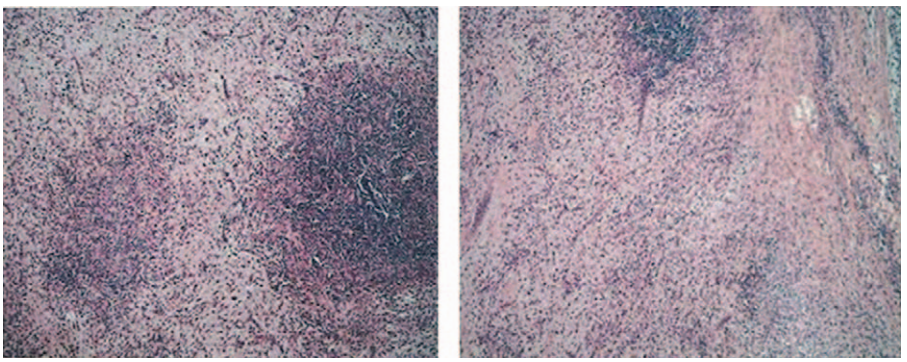

Figure 3. Postoperative paraffin section showed inflammatory cells and fibroblasts and immunohistochemistry assay was positive for smooth muscle antigen.

plasm" is nonspecific and with the improved understanding of the pathological and immunohistochemical features, IMT has been more widely accepted as the name for this type of tumor.[9,10] The pathogenesis of IMT remains unclear. It is still controversial whether IMT is a kind of reactive hyperplasia lesion or a tumor disease.[8] Oncocytogenetics have determined that IMT is related to chromosomal abnormality, suggesting that it may be a neoplasm rather than a reactive or postinflammatory process.[11] However, we found that the renal IMT coexisted with ipsilateral perinephric and periureteric fibrosis in the present case. Pathology analysis revealed that fibroblasts exhibited nodular hyperplasia with collagen degeneration and closely adhered to the kidney, ureter, and the surrounding tissue. Immunohistochemical staining tended to the diagnosis of retroperitoneal fibrosis nodules. Retroperitoneal fibrosis is a rare fibroinflammatory disease that develops around the abdominal aorta and the iliac arteries, and spreads into the adjacent retroperitoneum, where it frequently causes ureteral obstruction and renal failure.[12] Renal failure due to obstruction of the ureters resulting in hydronephrosis develops in 42% to 95% patients.[13] Bilateral retroperitoneal fibrosis was reported by Tanaka et al,[14] Shiber et al,[15] and Bjorndalen and Hastings.[16] However, in our case, the patient only presented with right retroperitoneal fibrosis nodules. This suggested that fibroblast proliferation may be an intermediate status in oncogenesis of IMT. de Suray et al[17] reported liver IMT and retroperitoneal fibrosis in a single patient; however, no specific relationship between IMT and fibroblast proliferation was observed.

The etiology of IMT also remains unknown. The arrangement of the anaplastic lymphoma kinase (*ALK*) gene may play an important role in IMT occurrences, as Schweckendiek et al[18] reported that approximately half of all IMTs show a rearrangement of the *ALK* gene. In addition, viral hepatitis B, Epstein–Barr virus (EBV), and Moraxella infection are also thought to play a role in tumorigenesis. Trauma, vascular causes, and autoimmune disorders have also been proposed.[18–20] Chronic pyelonephritis was observed in the present case, which might represent an inciting event of IMT.

There are no specific clinical symptoms of IMT. Patients commonly report flank pain, painless gross hematuria, ureteropelvic junction stenosis with hydronephrosis, and asymptomatic.[21] In this present report, the patient complained of dull back pain after overwork, painless microscopic hematuria, and ureteropelvic junction obstruction (UPJO) with hydronephrosis. The imaging findings of IMT of kidney are also nonspecific. On ultrasonography, the tumor can be seen as a heterogeneous echoic mass, appearing either hyperechoic or hypoechoic.[22] By CT, a renal IMT shows ill-defined, hypovascular, homogeneous borders,[2] but there is occasional significant enhancement with a clear edge. Calcification was observed in some cases with necrosis in the center.[23,24] Like previous reports, in our case, the ultrasound showed a solid, medium-low echogenic mass and the right ureter upper-middle section was not completely obstructed due to the hydronephrosis. By CT scan, the tumor showed significant enhancement with a clear boundary, homogeneous density, and flecks like calcification.

Generally, the preoperative diagnosis of IMT remains difficult.[3] Nephrectomy is usually performed in most of the renal IMTs due to the mimicry of malignancy by imaging.[4–6] In previous reports, preoperative fine-needle aspiration or percutaneous biopsy and intraoperative frozen section were applied to confirm the diagnosis.[25,26] These may help avoid unnecessary nephrectomy, especially in patients with renal insufficiency, bilateral masses, or a solitary kidney.

Most of the cases reported previously revealed no association with malignancy. Kapusta et al[21] found no recurrences or associated metastases in a series of 12 cases of renal IMT. However, Gwynn and Clark [27] described a rare case of IMT in a 46-year-old man, which encased a focus of renal cell carcinoma. IMTs usually present a good prognosis. In the present case, the patient has remained asymptomatic without any evidence of recurrence during 12 months of follow-up.

4. Conclusion

IMT is a rare lesion and its etiology and pathogeny are unclear. It is the first study about IMT of the kidney in a patient with perinephric and periureteric fibrosis This report suggested that fibroblast proliferation may be an intermediate status in oncogenesis of IMT, but further investigation is necessary in order to better clarify the relationship between fibroblast proliferation and IMT. The preoperative diagnosis of renal IMT remains difficult. Preoperative fine-needle aspiration or percutaneous biopsy and intraoperative frozen section were applied to confirm the diagnosis to avoid unnecessary nephrectomy, especially in patients with renal insufficiency, bilateral masses, or a solitary kidney.

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