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Case Presentation

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Carcinosarcoma of the Kidney: A Case Report and Literature Review

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Abstract

Carcinosarcomas are biphasic tumors with both epithelial and mesenchymal components. The age of onset is mostly 50-70 years old, and it lack special clinical manifestations. Similar to renal cell carcinoma, most patients present with hematuria, low back pain, and/or abdominal mass as the first symptom. It also lacks special laboratory and imaging examinations, making preoperative diagnosis difficult, and the diagnosis mainly depends on postoperative pathological examination. The malignant degree of renal carcinosarcoma is high, and most of them are already advanced when diagnosed. Radical nephrectomy as soon as possible after diagnosis is the preferred treatment. It is insensitive to radiotherapy and chemotherapy, and has poor prognosis and extremely low 5-year survival rate. Most patients die within 1 year after surgery. The rarity of this disease is the main obstacle to conducting comprehensive clinical trials. It is of great importance to publish the identified renal carcinosarcoma cases.

Keywords: Carcinosarcoma; Malignant tumors; Kidney; Urology

Introduction

Carcinosarcoma occurring in the urinary system is rare, accounting for only about 3% and less than 1% of all renal malignant tumors [1]. Carcinosarcoma of the kidney was first proposed by Farrow in 1946 [2]. The most common occurrence sites of carcinosarcoma are the upper respiratory tract and upper digestive tract [3]. The presence of the sarcomatoid component indicates an aggressive tumor nature [4]. Carcinosarcoma of the kidney is a biphasic tumor and its biphasic nature must be confirmed through immunohistochemical methods when the

pathological diagnosis was established [4].

Case Presentation

A 64-year-old male patient with lumbago and distension for 20 days was admitted to our hospital. The patient was in good health with no basic or special medical history. Physical examination showed no obvious positive signs. Laboratory values were within normal limits. Urinalysis was negative. Ultrasound of urinary system showed a mixed echo mass of about 120×95×101 mm in size in the right kidney, with clear boundaries and irregular shape, convex outward to the envelope, multiple thick strong light spots and non-echoic liquid dark areas can be seen inside, considering the possibility of renal cancer. Magnetic Resonance Imaging (MRI) showed a type of circular mass with a size of about 117×88 mm in the right subrenal pole, with slightly mixed low signal on T1WI and slightly mixed high signal on T2WI, uneven enhancement on enhanced scan, no enhanced necrotic area was visible inside the lesion, high signal on DWI and low signal on ADC, considering the possibility of renal cancer **(Figure 1 and 2)**. Computed Tomography (CT) examination of the lung showed microscopic nodules in the posterior segment of the upper lobe tip of the left lung, LU-RADS 2, and regular review is recommended. Admission diagnosis was space occupying lesion of right kidney. Relevant preoperative examination was completed and surgical conjunctures were excluded. On May 8, 2024, laparoscopic radical resection of right renal carcinoma was performed under general anesthesia. Intraoperative exploration revealed a solid mass in the middle and lower poles of the right kidney, protruded on the surface of the kidney, about 12*11 cm in size, with smooth surface, complete envelope and clear boundary, and the right kidney was completely resected and sent for biopsy.

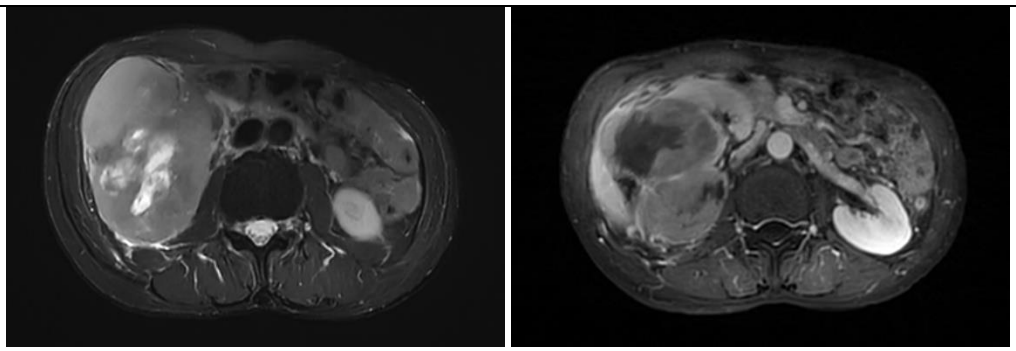

Figure 1 and 2: MRI showed a space occupying in the right kidney.

Postoperative pathological examination results reported: (right kidney) renal tissue 15.5*9,5*12.5 cm, attached ureter 0,8cm in length, 0.4 cm in diameter, section of a 12.5*9*11.2 cm mass, malignant tumor, combined with immunohistochemical results consistent with carcinosarcoma, cancer mainly manifested as chromophobe cell carcinoma: There was more necrosis, cancer tissue invaded the renal capsule and perirenal fat, and no cancer invasion was found at the margin of renal vein and ureter incision **(Figure 3 and 4)**. Cancer immunophenotype: EMA(+), E-cad(+), PAX-8(+), CX-7(+), CD10(-), Vim(-), RCC(-), CD68(-), SOX-10(-), SMA(-). Sarcoma immunophenotype: EMA(-), E-cad(-), PAX-8(-), CX-7(-), CD10(-), Vim(-), RCC(-), CD68(+), SOX-10(-), SMA(+), CAIX(+), Cyclin D1(+), DES(-), S100(-), CD34(+). Solid tumor 1021 gene detection method: second generation gene sequencing (NGS) test results: a. Somatic mutation: 7 mutation sites were detected, 3 of which

were related to targeted drugs: TP53p.S215R, PDGFRA amplification, KIT amplification; b. Germ line variation: pathogenic/possibly pathogenic variation is not detected; c. Tumor Mutation Load (TMB) : 3.84 Muts/Mb, TMB-L; d. Microsatellite Stabilization (MSS) type. On May 31, 2024, the patient underwent whole-body PET-CT: 1. Postoperation resection of the right kidney, no signs of recurrence were found in the operative area. Please refer to the clinic. 2. Small nodular lymph nodes with increased glucose metabolism were scattered in the outer and lower margin of liver (near peritoneum), upper mesenteric region (close to peritoneum), and pelvic region (around mesenteric region and rectum). Lymph node metastasis may be large, except peritoneal metastasis. 3. Plvic fluid; 4. The mesenteric space in the middle and lower abdomen is slightly blurred, and flaky shadows of increased glucose metabolism can be seen. Consider the possibility of inflammatory changes, please combine clinical; 5. Prostate hyperplasia and calcification focus; 6. No obvious abnormal glucose metabolism and placeholder shadow were found in other areas. CT reexamination on June 11, 2024: Changes after resection of right renal carcinoma; Mass shadow in the operative area, slight thickening of the peritoneum, multiple nodules in the peritoneum and abdominal pelvic cavity, abdominal pelvic effusion, tumor recurrence and peritoneal metastasis? Lymph node metastasis is not excluded, please combine clinical. Prostate hyperplasia and calcification. Left iliac crest density, undetermined, bone metastases? Bone Island? Other? Please combine clinical and bone scan. In combination with imaging and clinical findings, the patient was considered to have tumor recurrence and metastasis one month after surgery. The patient received immunotherapy and targeted therapy with terriplizumab and antirotinib on June 17, 2024, and was followed up until August 31, 2024. The patient had received immunotherapy and targeted therapy for four times, during which no further tumor development was observed. Follow-up will continue.

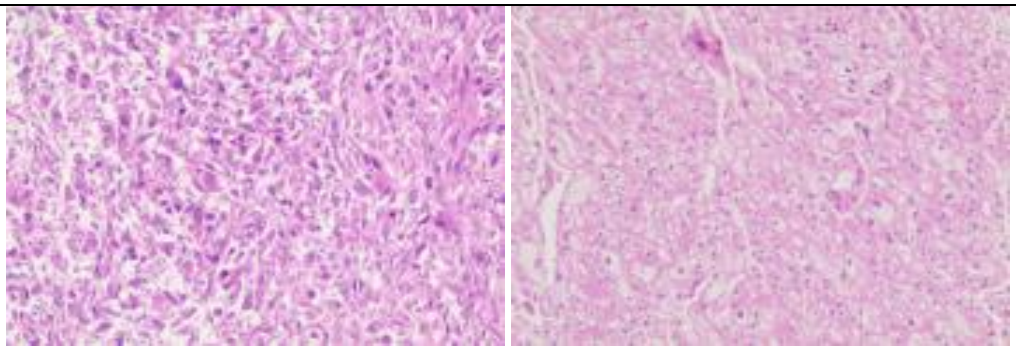

Figure 3 and 4: Immunohistochemical revealed carcinosarcoma.

Discussion and Literature Review

Renal carcinosarcoma is a rare and highly malignant tumor of the urinary system that shares similar clinical features with renal cell carcinoma. It typically presents with hematuria, flank pain, and an abdominal mass. This tumor is characterized by the presence of both malignant epithelial and mesenchymal components **[4]**. Notably, these components coexist without exhibiting an invasive relationship, meaning there is no distinct transitional zone between them on histological examination. In contrast, sarcomatoid carcinoma, a distinct cancer type, has a histological appearance resembling soft tissue sarcoma, but it remains a malignancy of epithelial origin. The dual phenotype of epithelial and mesenchymal differentiation in sarcomatoid carcinoma results in a hypertransitional zone between the two components, which serves as an important pathological marker for

distinguishing it from carcinosarcoma [5]. In this case, the diagnosis of renal carcinosarcoma was confirmed through postoperative pathology and immunohistochemistry. The limited number of reported renal carcinosarcoma cases may be attributed to several factors. Firstly, its diagnosis is challenging due to the overlapping clinical manifestations with other renal tumors, such as renal cell carcinoma, renal sarcomatoid carcinoma, and various renal space-occupying lesions. Additionally, when symptoms subside, the condition may be overlooked, leading to delayed diagnosis and treatment. Secondly, there are no specific hematological or biochemical markers for renal carcinosarcoma, and urine cytology, particularly urine exfoliative cytology, offers limited diagnostic value, with the possibility of multiple negative results. Imaging studies often reveal space-occupying lesions in the kidney, but it is difficult to differentiate between specific lesion types. Consequently, the definitive diagnosis typically relies on pathological biopsy. Furthermore, the sarcomatoid component of the tumor, which arises from mesenchymal tissue, lacks natural barriers, contributing to its high malignancy and rapid growth. This tumor is prone to invade surrounding renal tissues, resulting in local recurrence and distant metastasis, which complicates treatment and leads to poor prognosis [1]. The 5-year survival rate is low, and mortality remains high, with most patients reported to have died within one year post-surgery due to metastatic spread. The role of adjuvant chemoradiotherapy remains to be determined through further research. However, recent studies suggest that PD-L1 inhibitors may represent a promising new treatment option for renal carcinosarcoma [6].

Conclusion

Renal carcinosarcoma is a rare and aggressive malignancy with poor prognosis due to its high degree of malignancy and tendency for early metastasis. Diagnosis is challenging, often requiring pathological confirmation due to its overlapping features with renal cell carcinoma and renal sarcomatoid carcinoma on imaging. Radical nephrectomy remains the cornerstone of treatment, followed by careful postoperative monitoring. Despite aggressive treatment, the prognosis remains poor, emphasizing the importance of early diagnosis and individualized therapeutic strategies to improve survival and quality of life.

Ethical Approval

The study received approval from the Ethics Committee of Second Xiangya Hospital and obtained written informed consent from the patient.

Conflicts of Interest

The authors confirm that they do not have any conflicts of interest. All authors have read and approved this manuscript. All authors consent to the publication of this case.

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