Urology Case Reports 59 (2025) 102975

Contents lists available at ScienceDirect

Urology Case Reports

journal homepage: www.elsevier.com/locate/eucr

Oncology

Sarcomatoid renal pelvis carcinoma: A case report

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ARTICLE INFO

Keywords:
Sarcomatoid renal pelvis carcinoma
SRPC
Carcinoma of renal pelvis
Urothelium carcinoma

ABSTRACT

Sarcomatoid carcinoma is a malignant tumor of epithelial origin, some of the cells of which are differentiated in a sarcomatoid direction and are highly invasive. Sarcomatoid renal pelvis carcinoma (SRPC) is very rare. This article reports on an elderly woman with SRPC and discusses the clinical and pathological features and prognosis of SRPC in the hope of providing a reference for the diagnosis and treatment of this disease.

1. Introduction

Sarcomatoid renal pelvis carcinoma (SRPC) is a rare malignant tumor with sarcomatoid changes in tumor morphology, which is more common in middle-aged and elderly people, and is more common in men than in women.[1]. The clinical manifestations of SRPC are nonspecific, with gross hematuria being more common, and others being low back pain and abdominal pain.[2]. Definitive diagnosis of the disease requires pathologic examination, especially immunohistochemical staining. Due to the extremely low prevalence of SRPC and the fact that most of the literature data are case reports, there are no standardized clinical treatment criteria. Here, we report a case of SRPC in a 67-year-old patient with final pathologic staging of pT2N0.

2. Case presentation

In July 2024, a 67-year-old woman was admitted to the hospital with "right-sided low back pain with gross hematuria for 1 month." The patient had recurrent right-sided low back pain that persisted unrelieved for a month prior to admission, accompanied by gross hematuria throughout, with clots visible in the urine. Physical examination reveals that this patient has percussion tenderness over right kidney region, but no tenderness in the right ureteral area. The patient was hemodynamically stable and had no fever. Routine blood and urine tests showed a decrease in hemoglobin to 93 g/l, elevated urine white blood cells to 2926/μl, and elevated urine red blood cells to 328/l. Other laboratory tests did not show any significant difference: white blood cell count 5.72*10^9/l, urine culture (−), creatinine 78 μmol/L. Abdominal unenhanced and contrast-enhanced CT suggests a possible neoplastic lesion of the right renal pelvis, accompanied by mild hydronephrosis of the right renal calyx (Fig. 1A and B). Magnetic resonance imaging of the kidney showed an irregular mass in the right renal pelvis with mild enhancement on enhancement scan, poorly defined lesion, involvement of the renal sinus and lateral renal parenchyma, and mild dilation of the right renal pelvis with hydronephrosis (Fig. 1C and D). The imaging diagnosis was malignant tumor of the right renal pelvis with hydronephrosis. In addition, three urine exfoliative cytology tests were negative.

On July 30, 2024, she underwent a cystoscopy with no evidence of bladder lesions, discoloration, or masses. He then underwent a right laparoscopic assisted radical nephroureterectomy(RUN) after a sleeve resection of the right ureteral opening. The specimen of the mass is hard and friable, and the cut surface is gray and fish-like. Pathology report: consistent with high-grade uroepithelial carcinoma, sarcomatoid type, with acute and chronic inflammation, the tumor invaded to the entire renal pelvis, no renal parenchyma or ureteral involvement was seen, and the pathological stage was T2N0. Immunohistochemical results: Tumor cells PCK(+), Vim(+), CK7(+), CK20(−), GATA-3(partially weak +), CKH(+), P40(−), P53(+ about 80 % mutant), SMA(partially+), Ki-67 (+30–40 %), PAX-8(−)(Fig. 2).

The patient was perfused with 5 % dextrose 30 ml with pirarubicin 30 mg in the bladder once at 3 weeks postoperatively, and was advised regular intravesical instillation. Because the patient's urinary pain was obvious after intravesical instillation and difficult to relieve after treatment, the patient stopped instillation on his own. A follow-up CT at the community hospital in three months after surgery did not show any

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https://doi.org/10.1016/j.eucr.2025.102975
Received 10 December 2024; Received in revised form 29 January 2025; Accepted 1 February 2025
Available online 3 February 2025

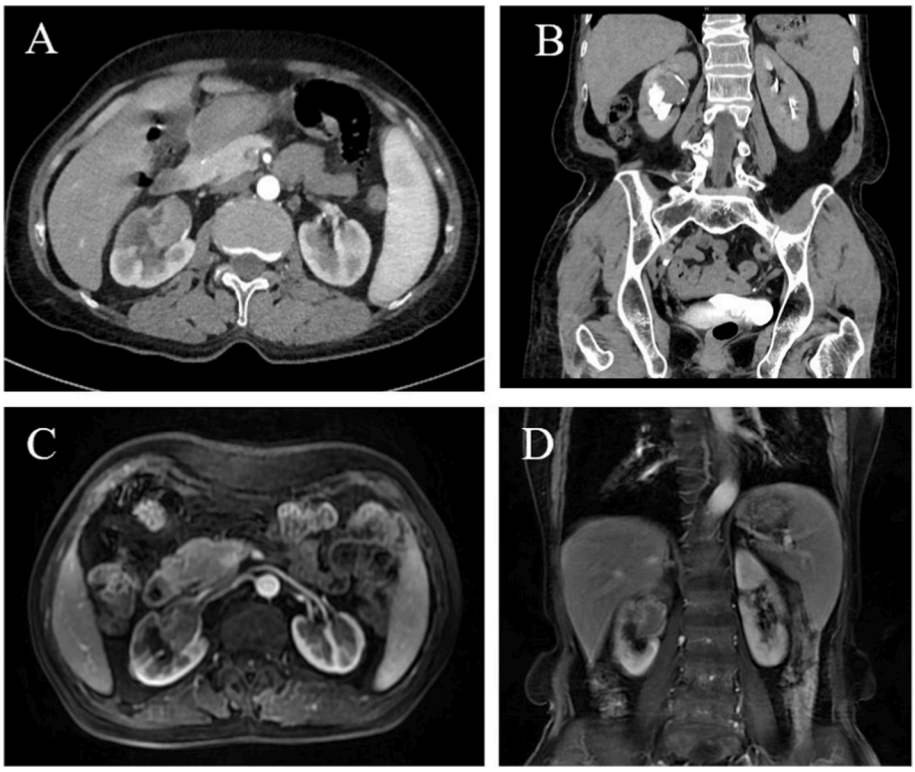

Fig. 1. Imaging manifestations of Sarcomatoid renal pelvis carcinoma. A-B) CT suggests a low-density soft tissue mass in the right renal pelvis with clear borders and uniform moderate enhancement. It was accompanied by mild hydronephrosis of the right renal calyx. C-D) MRI of the kidney showed an irregular TI isosignal, T2 isosignal mass, about 2.4*3.2cm in size, with obvious high signal in DWI diffusion restriction, obvious low signal in ADC, mild enhancement in enhancement scan, unclear boundary of the lesion, involving the renal sinusoids and lateral renal parenchyma.

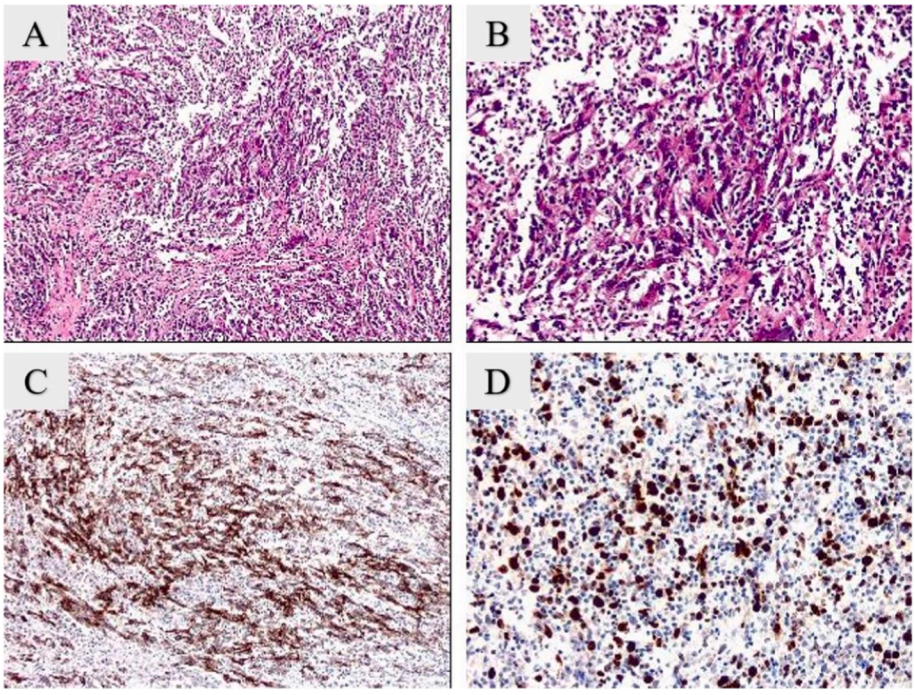

Fig. 2. Pathological examination of Sarcomatoid renal pelvis carcinoma. A-B)Hematoxylin and eosin-stained tumor. C) Immunohistochemical CK (+). D) Immunohistochemical P53+ about 80 % (mutant).

tumor recurrence. Now the patient is regularly followed up in our department.

3. Discussion

Sarcomatoid carcinoma is a highly malignant tumor that can occur in various organs including breast, larynx, pharynx, oral cavity, skin, esophagus, kidney, bladder, and female genital tract,[3], and is rare in the renal pelvis. SRPC tends to develop at the age of 60 years or older, with a male-to-female incidence ratio of 3:1[1]. Similar to uroepithelial carcinoma, the clinical manifestations of SRPC are nonspecific, with more frequent gross hematuria, and other symptoms such as lower back pain and abdominal pain.[2]. In our case, the patient had several previous cases of painless hematuria which could be relieved by himself, which may be related to this disease.

In SRPC, imaging features are helpful in the diagnosis but are not specific. The diagnosis of SRPC is mainly dependent on pathology. Sarcomatoid carcinoma mainly originates from malignant spindle cells of epithelial tissues, and has the ability to differentiate to mesenchymal tissues and epithelial tissues in both directions.[4]. The main component of sarcomatoid carcinoma is still epithelial cell carcinoma, but some of the carcinoma cells differentiate into sarcoma form, and the diagnosis can be made when the sarcoma component is more than 50 %.[5]. Current guidelines recommend RNU as standard treatment for high-risk non--metastatic UTUC and platinum-containing combination chemotherapy as first-line treatment for metastatic UTUC.[6]. In addition, Anraku[7] et al. had a case report of a patient with SRPC who had multiple systemic metastases at the time of presentation to the hospital, and almost all of the lesions, including the left kidney tumor, shrank after immunotherapy with nivolumab and ipilimumab. This suggests that immunotherapy can be a promising treatment for sarcomatoid uroepithelial carcinoma.

SRPC has a high degree of malignancy, a low degree of differentiation, rapid development, infiltrative growth, easy to involve adjacent renal parenchyma, peripheral fat, muscle, blood vessels, easy to develop peripheral lymph nodes and systemic metastasis, and a very poor prognosis.[8]. In this case, after RNU, 5 % dextrose 30 ml with pirarubicin 30 mg was performed for bladder instillation once at 3 weeks postoperatively, and the instillation was stopped because the patient could not tolerate it. No tumor recurrence was seen on follow-up CT three months after surgery, and the patient recovered well on follow-up.

4. Conclusion

SRPC is a rare and highly malignant tumor, the clinical manifestations and imaging manifestations are similar to those of uroepithelial carcinoma without specificity, and the diagnosis mainly relies on pathological diagnosis. In patients without metastases, surgical treatment is helpful. In patients who develop metastases, chemotherapy can reduce the rate of tumor recurrence. In recent years, the use of PD-1 inhibitors has been reported to achieve some efficacy in the treatment of this type of tumor recurrence.The prognosis of SRPC type is poor, so increasing the clinician's knowledge of RPSC can help in early diagnosis and treatment, and improve the prognosis.

CRediT authorship contribution statement

Xuechao Zhang: Writing – original draft. **Yanan Jin:** Writing – original draft. **Weiran Zhang:** Writing – original draft. **Yawei Zhang:** Writing – review & editing. **Haifeng Wang:** Writing – review & editing.

Conflicts of interest

The authors declare no conflict of interest.

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