Case Report

Sarcomatoid variant of urothelial carcinoma of renal pelvis with direct invasion of the pancreas and descending colon

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Abbreviations & Acronyms CT = computed tomography HPF = high-power field SVUC = sarcomatoid variant of urothelial carcinoma

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Received 14 February 2022; accepted 13 May 2022. Online publication 4 July 2022 **Introduction:** Sarcomatoid variant of urothelial carcinoma infiltrates the perimeter and occurs occasionally. However, there are only few case reports.

Case presentation: A left renal tumor was incidentally detected in a 75-year-old woman and protruded outside the kidney, infiltrating the pancreatic tail and spleen. Tumor invasion was observed in the adjacent organs; therefore, the left kidney, pancreatic tail, spleen, and, descending colon were resected. Histopathological examination revealed a sarcomatoid variant of invasive urothelial carcinoma. She received two cycles of gemcitabine and carboplatin combination chemotherapy but succumbed to the disease after 5 months.

Conclusion: Sarcomatoid variant of urothelial carcinoma is rare, with aggressive malignancy. The diagnosis was difficult and required surgery. This is the first case of a sarcomatoid variant of urothelial carcinoma with direct invasion into the pancreas and descending colon.

Key words: carcinoma, kidney, pancreas tumor, urothelium.

Keynote message

We detected a left kidney tumor infiltrating the pancreas on computed tomography. Histopathological examination revealed a sarcomatoid variant of invasive urothelial carcinoma. To the best of our knowledge, this is the first case of a sarcomatoid variant of urothelial carcinoma with direct invasion of the pancreas and descending colon.

Introduction

SVUC is a rare tumor (0.1-0.3% of all other cases of urothelial carcinomas,¹) and highly malignant.² SVUC tends to infiltrate the perimeter compared with pure urothelial carcinoma, and such cases are occasionally encountered in clinical practice. However, there are only few case reports and no established standard treatment regimens. To the best of our knowledge, this is the first case of SVUC of the renal pelvis with direct invasion into the pancreas and descending colon.

Case presentation

A 75-year-old woman presented with a high total leukocyte count during preoperative examination for hip osteoarthritis. CT revealed a left kidney tumor for which she visited Hiroshima University Hospital for treatment. She had no other comorbid conditions and had not been diagnosed with cancer. She was 152.6 cm tall, weighed 46.2 kg, and had lost approximately 4 kg in 2 months. Physical examination findings were unremarkable. There was no history of hematuria. Laboratory investigations revealed a hemoglobin level of 11.4 g/dL, total leukocyte count, 8.39×10^9 /L, and serum creatinine level of 1.10 mg/dL (mild renal dysfunction). Urinalysis revealed microhematuria (5–9 red blood cells/HPF) and leukocyturia (50–99 white blood cells/ HPF). All other laboratory parameters showed unremarkable findings. Contrast-enhanced CT



Fig. 1 Computed tomography findings: A 61×45 -mm tumor involving the upper pole of the left kidney. The tumor protrudes outside the kidney and infiltrates the pancreatic tail (a) and spleen (b). The tumor shows an inhomogeneous enhancement and appears to originate from the renal parenchyma. Metastases are observed in the para-aortic lymph nodes (c) and lung fields (d).

revealed a 61×45 -mm deficient tumor involving the upper pole of the left kidney (Fig. 1a,b). The tumor showed inhomogeneous enhancement, protruded outside the kidney, infiltrated into the pancreatic tail, spleen, and descending colon, and originated from the renal parenchyma. Metastases were found in the para-aortic lymph nodes (Fig. 1c), left adrenal gland, and both lung fields (Fig. 1d).

The provisional diagnosis was left renal cell carcinoma with pancreatic and splenic infiltration and left adrenal and pulmonary metastases with tumor stage T4N2M1. As renal cell carcinoma was suspected, cytoreductive nephrectomy was performed for histological confirmation. The tumor invaded the adjacent organs on the ventral side of the left kidney and was indistinguishable from the peritoneum, pancreas, spleen, and descending colon. We detached the perineal kidney from the inferior pole of the kidney and secured the renal arteriovenous. The area around the tumor was carefully exfoliated. The pancreatic tail, which had adhered to the tumor, was dissected using a stapler. The colon was also resected by approximately 20 cm, and an end-to-end anastomosis was performed. Surgical resection was performed within anatomical constraints. The left kidney, pancreatic tail, spleen, and part of the descending colon were removed (Fig. 2). The left ureter was ligated and transected with the upper ureter. A pancreatic fistula developed 2 days after surgery and was managed conservatively (indwelling drainage tube improved the general condition). The patient was discharged 1 month postoperatively. Liver function and pancreatic enzymes improved to normal levels, but renal function deteriorated compared to that before surgery (serum creatinine, 1.43 mg/dL).

Histopathological examination of the resected mass revealed an invasive urothelial carcinoma with sarcomatoid



Fig. 2 Surgical specimen: the left kidney, left adrenal gland, spleen (a), pancreatic tail (b), and part of the descending colon (c) were resected (indicated by the white arrow). A grayish-white tumor infiltrating the renal parenchyma is observed (indicated by the yellow arrow).

differentiation (Fig. 3a,b). Tumor cells infiltrated the left adrenal gland, pancreas (Fig. 3c), and transverse colon (Fig. 3d). No malignant findings were observed in the spleen. Immunohistologically, the tumor cells were positive for p63,



CK7, AE1, AE3, vimentin, and INI-1; partially positive for CD19, EMA, and GATA3 and negative for CK20.

No tumor was detected in the bladder during postoperative cystoscopy.

Postoperative chemotherapy with gemcitabine $(800/m^2)$ and carboplatin (4.0 AUC) was administered. Considering the patient's general condition, the dosage was slightly reduced, and the administration duration was 4 weeks. However, chemotherapy was ineffective, and CT revealed pulmonary, pleural, and hepatic metastases. Chemotherapy was discontinued because of the patient's impaired general condition; immunotherapy was also ruled out. There was no sufficient time for performing genetic testing or individualized treatment. The patient succumbed to the disease 5 months after surgery.

Discussion

Sarcomatoid carcinoma is a rare variant of urothelial carcinoma of the renal pelvis. Reportedly, 85% and 50% of patients with SVUC present with tumor sizes of >2 cm and organ metastases, respectively.² Additionally, SVUC has outward infiltration, SVUC of the renal pelvis often extends into the renal parenchyma.³ Moreover, SVUC is highly malignant with poor prognosis. Approximately 70% of patients with SVUC die within 2 years of the initial diagnosis.⁴ SVUC originating from the renal pelvis has the worst prognosis with a mean survival of <9 months.⁵

SVUC is detected on physical examination and may manifest as gross hematuria and back pain. Early diagnosis is rare because clinical symptoms, such as gross hematuria or back pain develop late during the disease course. Several reports indicate incidental postoperative detection of SVUC.^{3,6} Differential diagnosis of SVUC of the renal pelvis is difficult as imaging findings are similar to those of renal sarcomas and renal cell carcinomas. Furthermore, only a few cases of SVUC of the renal pelvis have been reported.⁷ The diagnosis

Fig. 3 Pathological findings, hematoxylin–eosin (H-E) staining: sarcomatoid carcinoma of the renal pelvis. The tumor contains spindle cell carcinomatous components (a, b). Tumor cells infiltrate the pancreas (c) and descending colon (d). The tumor part is indicated by the arrow.

becomes increasingly complex in cases with local infiltration. Cases in which the preoperative diagnosis was mistaken for renal cell carcinoma have been reported.^{6,8} Apparently, SVUC tends to infiltrate leading to a suspected diagnosis of RCC. Consistent with previous reports,^{2,8,9} preoperative diagnosis was challenging in our case.

No standard treatment regimens have been developed for SVUC. Current guidelines recommend platinum-based combination chemotherapy as the first-line treatment for metastatic upper tract urothelial carcinomas. However, radical surgery is not recommended.¹⁰ Although successful treatment results have been reported with combination chemotherapy (cisplatin and gemcitabine) in SVUC of the urinary bladder,¹¹ no studies have described the effects of chemotherapy in cases with SVUC of the renal pelvis. Moreover, adjuvant chemotherapy has not been associated with an improved prognosis.^{12,13} Recent evidence suggests that SVUC responds favorably to treatment with pembrolizumab,¹⁴ and immunotherapy should therefore be considered.

In our case, the preoperative diagnosis was challenging. In reflection, we would have performed urinary cytopathology, cystoscopy, and retrograde ejaculation preoperatively, considering enough the possibility of urothelial carcinoma. However, after surgery, chemotherapy was administered following conclusive postoperative histopathological diagnosis. However, the prognosis was poor, highlighting the importance of an accurate differential diagnosis of SVUC from other renal cell carcinomas for improved overall survival. Further research is warranted to develop definitive treatment strategies for achieving optimal treatment outcomes of SVUC.

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Author Contributions

Yoshinori Nakano: Writing – original draft. Hiroyuki Kitano: Writing – original draft; writing – review and editing. Keisuke Hieda: Supervision; writing – review and editing. Takashi Babasaki: Investigation; writing – review and editing. Kenshiro Takemoto: Writing – review and editing. Tetsutaro Hayashi: Supervision; writing – review and editing. Aya Kido: Investigation. Daiki Taniyama: Investigation. Kazuhiro Sentani: Investigation; supervision. Nobuyuki Hinata: Supervision.

Conflict of interest

The authors declare no conflict of interest.

Approval of the research protocol by an Institutional Reviewer Board

Not applicable.

Informed consent

Consent to participate and for publication were obtained from the patient.

Registry and the Registration No. of the study/trial

Not applicable.

Data accessibility statement

The authors confirm that the data supporting the findings of this study are available within the article.

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