Sarcomatoid Transitional Cell Carcinoma of the Renal Pelvis: A Case Report

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Sarcomatoid transitional cell carcinoma (TCC) of the renal pelvis is a rare tumor with sarcoma-like components arising from the malignant transitional epithelium. The prognosis, according to previous reports, is very poor. Due to its rarity, there have been few studies concerning the treatment efficacy. We herein report a case of sarcomatoid TCC arising from the renal pelvis. A 67-year-old female was admitted with the chief complaint of bilateral flank pain. A right renal heterogeneous mass measuring about 5 x 7 x 10 cm was incidentally found on a clinical image. A right radical nephrectomy was performed. Histologically, a TCC in situ and another sarcoma-like area with prominent spindle cells were identified. Immunohistochemistry was positive for cytokeratin. Sarcomatoid TCC of the renal pelvis was confirmed. In spite of no ample evidence of an effective treatment, we recommend a radical, aggressive operation for early localized disease. For the tumor with advanced stage or metastasis, systemic chemotherapy with a gemcitabine-based regimen may be considered for this uncommon renal pelvic tumor due to its urothelial origin in nature. (JTUA 19:48-51, 2008) Key words: renal cancer, sarcomatoid, spindle cell carcinoma, carcinosarcoma.

CASE REPORT

This 67-year-old female patient with a past history of right ureteral stone 20 years ago presented to our clinic with bilateral flank pain for several years. Moreover, intermittent gross hematuria was also noted. On physical examination, bilateral mild knocking tenderness was noted, but there was no palpable abdominal mass. Urine analysis showed predominant occult blood and 6–8 red blood cells (RBCs) per high-power field. Renal sonography revealed bilateral stones and moderate bilateral hydronephrosis. The kidney, ureter, and bladder (KUB) X-ray study revealed multiple radiopaque stones in the bilateral kidney. Under the impression of bilateral renal stones, she was admitted to our ward.

On the day of admission, bilateral percutaneous nephrostomy drainage was performed to relieve the hydronephrosis. Antegrade pyelography showed bilateral caliectasis and multiple renal stones. A ureteroscopic examination incidentally found a right renal pelvic tumor with stone encrustation. A biopsy was taken, and spindle cell sarcoma was our initial diagnosis.

In the following days, a metastatic survey including chest x-ray, an abdominal-to-pelvic computed tomographic (CT) scan, and whole-body bone scan were performed. The CT scan showed a heterogeneous mass measuring about 5 x 7 x 10 cm occupying the right renal pelvis and several enlarged para-aortic lymph nodes were also noted (Fig. 1). A chest x-ray and bone scan showed no evidence of lung or bone metastasis.

Fig. 1. Computed tomographic scan demonstrating a right renal heterogeneous mass measuring about 5 x 7 x 10 cm (white arrows) with enlargement of several para-aortic lymph nodes. Bilateral hydronephrosis and a right renal stone were also noted.

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One week after the initial operation, a radical nephrectomy was performed. A large whitish, intracapsular solid right renal tumor with invasion to the renal sinus and calyceal system was noted during the operation. Several lymphadenopathies over the aortovenous area were also identified. Gross examination revealed a huge solid mass measuring \(8.0 \times 7.0 \times 5.0\) cm occupying the renal pelvis which was whitish, gelatinous, and focally necrotic on cutting (Fig. 2). Microscopic examination revealed 2 main components of the tumor: 1 component with TCC in situ and another sarcoma-like area with prominent spindle cells. Some multinucleated giant cells were also seen. Immunohistochemistry was positive for cytokeratin, indicating an epithelial origin (Fig. 3A-D). In addition, there was involvement of 2 lymph nodes in 4 para-aortic lymph nodes. Sarcomatoid TCC of the renal pelvis with para-aortic lymph node involvement was

**Fig. 2.** Gross appearance of the tumor showing a solid mass measuring \(8.0 \times 7.0 \times 5.0\) cm occupying the renal pelvis with whitish, gelatinous, and focal necrosis on the cut surface.

**Fig. 3.** Sarcomatoid transitional cell carcinoma (TCC) of the renal pelvis. (A) TCC in situ, (B, C) the sarcomatoid component with prominent spindle cells and some multinucleated tumor cells, and (D) a sarcomatoid area showing immunoreactivity for cytokeratin.
the ultimate diagnosis (pT3N2M0). One course of adjuvant chemotherapy with gemcitabine was given. Unfortunately, the patient's condition gradually deteriorated, and a follow-up CT scan 3 months after the radical operation showed recurrent bulky tumor with liver invasion and multiple lymph node involvement. Direct tumor expansion from the previous percutaneous nephrostomy tract was also noted (Fig. 4A,B).

**DISCUSSION**

Sarcomatoid carcinoma is a high-grade epithelial neoplasm characterized by a biphasic appearance with 1 part of an epithelial component and another area with a sarcoma-like appearance, which is largely composed of non-specific spindle and/or pleomorphic tumor giant-cells. The actual histogenesis of sarcomatoid carcinoma is still unclear. In a study, because they found that the sarcomatous component exhibited diffuse and intense p53 oncoprotein expression, whereas the carcinosarcomatous component showed a weaker and focal positivity, they assumed that this alteration of the p53 gene might be related to the sarcomatous transformation of the tumor. In another study, it was thought that the sarcomatoid component of sarcomatoid carcinomas may result from either anaplastic changes or de-differentiation related to the process of losing cell adhesion molecules.

In addition to the kidney and bladder, sarcomatoid carcinoma can also occur in the breast, larynx, pharynx, oral cavity, esophagus, and genital tract. Carcinosarcoma is a term frequently confused with sarcomatoid carcinoma. It has a malignant epithelial component as does a sarcomatoid carcinoma. In addition, it has specific features of mesenchymal differentiation, such as chondrosarcomas, osteosarcomas, rhabdomyosarcomas, and liposarcomas, which differ from a sarcomatoid carcinoma. However, these 2 tumors have frequently presented diagnostic difficulties because their morphologies are similar in microscopic appearance with hematoxylin and eosin staining. As a result, an immunohistochemical study is usually necessary for a diagnosis. Immunoreactivity to cytokeratins and CD68 is specific for cells of epithelial origin and diagnostic for a sarcomatoid carcinoma. Carcinosarcomas do not stain for epithelial markers.

Most renal pelvis malignancies consist of TCC. However, a sarcomatoid carcinoma of transitional cell origin in the renal pelvis is very rare. In a clinicopathologic study of 108 cases of high-grade urothelial carcinomas of the renal pelvis, only 8 cases were shown to be sarcomatoid carcinomas. Lopez-Beltran et al. reported 13 cases of sarcomatoid TCC of the renal pelvis since 1961. Metastatic disease or advanced tumor involvement of the parenchyma was noted with the diagnosis in all 13 patients. The prognosis was very poor with 8 patients dying within 2 years of diagnosis and a mean survival time of 11.2 months. Moreover, adjuvant chemotherapy appeared to offer no response or survival advantage.

The Mayo Clinic reviewed 26 cases of sarcomatoid carcinomas of transitional cell origin in the bladder dur-
ing a 59-year period. Sarcomatoid carcinoma of the bladder was more common in men than in women (with a 4:1 ratio), and the mean survival was 9.8 months. The pathologic stage was the main predictor of patient survival. Chemotherapy offered little response and no survival advantage.\textsuperscript{4}

Since sarcomatoid carcinomas of transitional cell origin are a very rare entity, they consistently pose a significant therapeutic challenge to clinicians. Although experience with the treatment of patients with sarcomatoid TCC is limited, Hisataki et al. suggested a radical, extensive operation to treat this disease. However, it remains uncertain whether such an operation contributes to improved survival.\textsuperscript{1} Thiel et al. presented the case of a 61-year-old man with a renal pelvis-confined sarcomatoid carcinoma of transitional cell origin. A laparoscopic nephroureterectomy was performed, and no further adjuvant therapy was given. The patient was without recurrence for more than 1 year.\textsuperscript{8}

Another 2 studies suggested that it seems appropriate to treat these neoplasms in the same manner as conventional high-grade TCC with similar degrees of invasion.\textsuperscript{2,3} However, to our knowledge, no previous report is available for a sarcomatoid TCC of the renal pelvis treated with systemic chemotherapy. Froehner et al.\textsuperscript{3} reported an 80-year-old patient with a sarcomatoid carcinoma of the bladder and lung metastasis. They demonstrated a remarkable response of the residual tumor in the lung and complete remission during 2 years of follow-up after treatment with 4 courses of gemcitabine and cisplatin.

In conclusion, sarcomatoid TCC of the renal pelvis is a very rare high-grade neoplasm with a very poor prognosis. For early localized disease, a radical, aggressive operation should be considered to improve the outcomes; for a tumor with advanced stage or metastasis, the prognosis is extremely poor. Despite no effective treatments having previously been reported, systemic chemotherapy with a gemcitabine-based regimen seems to be reasonable for this uncommon renal pelvic tumor due to its urothelial origin in nature.

REFERENCES