Abstract

Rationale: Primary carcinosarcoma of the upper urinary tract is rare. Ureteral duplication is one of the most common urinary tract malformations. Additionally, the association between ureteral duplication and malignancy is unknown. To the best of our knowledge, no cases of malignant tumors diagnosed as carcinosarcoma with ureteral duplication have been reported. We herein report the case of a patient with carcinosarcoma of the ureteropelvic junction associated with incomplete ureteral duplication.

Patient concerns: A 60-year-old Japanese woman presented with painless gross hematuria. She had a history of total hysterectomy and chemotherapy for endometrioid carcinoma 5 years before. She had no history of occupational chemical exposure.

Diagnoses: Radiographic imaging revealed right incomplete ureteral duplication, hydronephrosis, and a polypoid tumor in the ureteropelvic junction of the lower moiety of the right kidney. Urine cytology showed a small amount of degenerated atypical epithelial and nonepithelial cells. The transureteral biopsy specimen showed dysplastic urothelial cells and atypical myoid spindle cells. These findings were indefinite for malignancy.

Interventions: The patient underwent right nephroureterectomy. Pathological examination of the resected tumor showed a biphasic neoplasm composed of carcinomatous and sarcomatous components. The sarcomatous component was immunohistochemically positive for vimentin, desmin, h-caldesmon, and α-SMA and negative for pancytokeratin (AE1/AE3), low molecular weight cytokeratin (CAM 5.2), EMA, E-cadherin, GATA3, uroplakin 2, and p63. Based on these findings, we diagnosed the tumor as carcinosarcoma.

Outcomes: The postoperative course was uneventful. No additional therapy was administered. The patient has remained alive without recurrence for 21 months since surgery.

Lessons: Carcinosarcoma can arise from ureteral duplication. Although the majority of carcinosarcomas of the upper urinary tract are diagnosed at an advanced stage and have a poor prognosis, some can have a less aggressive course. Further studies are needed to determine the association between ureteral duplication and malignancy.

Abbreviations: CT = computed tomography, UPJ = ureteropelvic junction.

Keywords: carcinoma, carcinosarcoma, cytology, leiomyosarcoma., pelvis, ureter, ureteral duplication

1. Introduction

Carcinosarcoma is a biphasic neoplasm with mixed malignant epithelial and mesenchymal components whose definition and histogenesis have long been discussed.[1–3] This neoplasm is highly aggressive in general and is often associated with poor prognosis. Carcinosarcoma is rare but can occur in a variety of organs or systems, such as the uterus, ovary, lung, breast, esophagus, and urinary tract. For the urinary tract, the majority of reported cases of carcinosarcomas are cases of the lower urinary tract, predominantly located in the urinary bladder. Several clinicopathological studies with a series of carcinosarcoma cases of the urinary bladder have been published.[8–11] In contrast, there have been only approximately 20 cases of carcinosarcoma of the upper urinary tract reported to date. Due to its rarity, the clinicopathological features of carcinosarcoma of the upper urinary tract remain incompletely elucidated.

Ureteral duplication is one of the most common urinary tract malformations found in approximately 0.8% of autopsy series.[12] There are 2 subtypes of duplicated ureter: complete
ureteral duplication refers to 2 separate ureters with 2 separate orifices in the bladder, while incomplete ureteral duplication refers to 2 ureters joining together to form a single ureter before entering the bladder. Embryologically, complete ureteral duplication occurs when 2 separate ureteric buds arise from the mesonephric duct, while incomplete ureteral duplication is caused by premature bifurcation of a single ureteric bud prior to fusion with metanephros. Complications of ureteral duplication include urinary reflux, ureterocele, and ureteral obstruction. However, the association between ureteral duplication and malignancy remains unclear.

Herein, we report a rare case of carcinosarcoma arising from the ureteropelvic junction (UPJ) of incomplete ureteral duplication. Informed consent for publication of this case report has been obtained from the patient. We also discuss the significance of distinguishing carcinosarcoma from another related entity and review the relevant literature.

2. Clinical summary

A 60-year-old Japanese woman presented with a several-day history of gross hematuria. She had no fever or pain. She had a history of total hysterectomy and postoperative adjuvant chemotherapy (paclitaxel and carboplatin) for endometrioid carcinoma (G1, pT1bN0) 5 years before. A preoperative CT scan revealed right unilateral incomplete ureteral duplication. Two months after the surgery, right hydronephrosis, which was not preoperatively present, was detected on CT scan, probably because of the ureteral stricture caused by postoperative scarring. A double-J ureteral stent was inserted and remained in place for 11 months. She developed pyelonephritis several times during the placement of the stent. One year and 7 months after the surgery, wall thickening at the UPJ of the lower moiety of the right kidney was observed on CT scan. Two years and 7 months after the surgery, a 6mm polypoid nodule was detected at the right lower UPJ, whose size increased to 9mm a year later. There were no signs of recurrence of endometrioid carcinoma. She also had histories of Wolff-Parkinson-White syndrome, splenectomy for thrombocytopenic purpura, glaucoma, hypercholesterolemia, and carpal tunnel syndrome and was a hepatitis B virus carrier. She had a 7-year history of smoking 10 cigarettes per day. She denied any occupational chemical exposure. She had no family history of urinary tract malignancy.

Retrograde pyelography showed right incomplete ureteral duplication, hydroureteronephrosis, and a filling defect at the right lower UPJ (Fig. 1A). A contrast-enhanced CT scan revealed a round, uniformly enhanced polypoid tumor 12 mm in diameter (Fig. 1B), which was 3 mm larger than that observed a year before. Cystoscopy showed no tumor in the bladder. Ureteroscopy revealed a nodular tumor at the right lower UPJ. Urine cytology showed a small amount of degenerated atypical epithelial and nonepithelial cells, which were indefinite for malignancy (Fig. 2).

Following cytology, the transureteral biopsy specimen showed dysplastic urothelial cells and atypical spindle cells. The atypical spindle cells were immunohistochemically positive for desmin and h-caldesmon, suggesting that there was a neoplasm with smooth muscle differentiation. With high suspicion of clinical malignancy, right nephroureterectomy was performed. The postoperative course was uneventful. The patient was followed up without additional therapy and remained alive without evidence of local recurrence or metastasis 21 months after the surgery.

3. Pathological findings

The tumor fell off of the UPJ during the surgical procedure and was submitted separately from the nephroureterectomy specimen for pathology (Fig. 3). Macroscopically, the tumor was 14 × 10 mm in size and had a spherical shape with a smooth surface. It had no stalk, which made it difficult to identify the attachment site to the ureteral wall. The cut surface showed a solid, yellowish-white appearance with hemorrhage.

Figure 1. Radiographic imaging of the tumor. (A) Right retrograde pyelography showed incomplete ureteral duplication, hydroureteronephrosis, and a filling defect at the ureteropelvic junction of the lower moiety (arrow). (B) Enhanced CT (coronal view) shows a round, uniformly enhanced polypoid tumor protruding into the lumen (arrow). Mild ureteral wall thickening was also observed immediately distal to the tumor.
Microscopically, the tumor consisted of two components: carcinomatous and sarcomatous components (Fig. 4). The tumor predominantly consisted of the sarcomatous component, which exhibited proliferation of atypical spindle cells with fibrous cytoplasm and enlarged, hyperchromatic, irregularly shaped nuclei (Fig. 4I). The surface of the tumor was covered with atypical epithelial cells with enlarged, hyperchromatic, irregularly shaped nuclei, which formed irregular glandular structures, suggestive of adenocarcinoma (Fig. 4B and C). A few nests of carcinomatous components were also observed inside the tumor (Fig. 4D and E). Some of the carcinomatous components had intercellular bridges, suggesting squamoid differentiation. There was no obvious “transition” between the 2 components.

The immunohistochemical findings of the tumor are summarized in Table 1. The carcinomatous component was positive for pancytokeratin (AE1/AE3; Leica Biosystems; Tokyo, Japan), low molecular weight cytokeratin (CAM 5.2; Becton Dickinson; San Jose, CA, USA), EMA (E29; Roche Diagnostics; Tokyo, Japan), E-cadherin (NCH-38; Nichirei Biosciences; Tokyo, Japan), GATA3 (L50-823; Biocare Medical; Pacheco, CA, USA), uroplakin 2 (BC21; Biocare Medical) (focal) and p63 (4A4; Nichirei Biosciences; Tokyo, Japan) (focal) and negative for vimentin (V9; Roche Diagnostics; Tokyo, Japan), desmin (D2-R-11; Leica Biosystems; Tokyo, Japan), h-caldesmon (h-CD; Agilent, Santa Clara, CA, USA), and α-SMA (1A4; Dako; Santa Clara, CA, USA) (Fig. 4F–H). In contrast, the sarcomatous component was positive for vimentin, desmin, h-caldesmon, and α-SMA and negative for all epithelial markers listed above.
Anaplastic lymphoma kinase 1 (ALK-1; Dako; Santa Clara, CA, USA) was negative through the tumor. Based on these findings, we diagnosed the tumor as carcinosarcoma.

The nephroureterectomy specimen showed incomplete ureteral duplication, hydronephrosis, and ureteral dilation (Fig. 3A). The tumor was likely to have originally located on the UPJ of the more severely dilated ureter. Microscopically, the ureteral mucosa of the more dilated mucosa showed severe inflammation with lymphoid follicle formation, while the other did not. However, we could not find any evidence of sarcoma, invasive carcinoma, carcinoma in situ or dysplastic epithelium in the background ureteral tissue.
Table 1
Immunohistochemical findings of the carcinosarcoma in the ureteropelvic junction.

| Antibodies to | Carcinomatous component | Sarcomatous component |
|--------------|-------------------------|-----------------------|
| pancytokeratin (AE1/AE3) | + | – |
| low molecular weight cytokeratin (CAM5.2) | + | – |
| EMA | + | – |
| E-cadherin | + | – |
| uroplakin 2 | + (focal) | – |
| p63 | + (focal) | – |
| GATA3 | – | + |
| vimatin | – | + |
| h-caldesmon | – | + |
| α-SMA | – | + |
| chromograninA | – | – |
| synaptophysin | – | – |
| CD56 | – | – |
| myoD1 | – | – |
| myogenin | – | – |
| p53 positive rate | 20% | 3% |
| Ki-67 labeling index | 55% | 15% |

4. Discussion
Carcinosarcoma is a biphasic malignant tumor with carcinomatous and sarcomatous components. Sarcomatoid carcinoma is another term that is occasionally improperly used to refer to tumors exhibiting such features. Distinction between carcinosarcoma and sarcomatoid carcinoma has been a matter of controversy. The widely accepted criterion for distinguishing carcinosarcoma and sarcomatoid carcinoma is the presence or absence of epithelial characteristics in its sarcomatous component; carcinosarcoma lacks any immunohistochemical or ultrastructural evidence of epithelial differentiation in its sarcomatous component, whereas sarcomatoid carcinoma retains epithelial markers.[6,7,15,16] Based on this definition, the present case corresponds to carcinosarcoma. However, according to the latest World Health Organization classification of tumors of the urinary system, all urothelial carcinomas with sarcomatous components are collectively classified as sarcomatoid variants of urothelial carcinomas.[17]

The histogenesis of carcinosarcoma is hypothesized as follows: (1) collision tumor, which is developed through independent occurrences of carcinoma and sarcoma in close proximity that merge into one lesion; (2) composition tumor, which is developed through malignant transformation of both epithelial and mesenchymal components of the same tissue; (3) combination tumor, which is developed through divergent differentiation and malignant transformation of a single pluripotent stem cell; and (4) conversion tumor, which is developed through epithelial-to-mesenchymal transition of carcinoma.[18,19] In terms of clonality, the former 2 mechanisms are referred to as monoclonal theory, while the latter can be called monoclonal theory. The molecular evidence accumulated to date supports the monoclonal theory.[16,20-22] Additionally, many studies have revealed that carcinomatous and sarcomatous components showed significant overlap of genetic aberrations, suggesting the late occurrence of divergent differentiation during tumor development.[16,20,21] In other words, the sarcomatous component of carcinosarcoma is likely derived from the carcinomatous component in most cases. For the present case, however, the histogenesis is uncertain because we did not perform genetic analysis.

Malignant neoplasms accompanied by ureteral duplication are extremely rare; only 23 cases have been registered in English-language publications to date (Table 2).[23-43] Among them, urothelial carcinoma predominated, and only 2 cases were reported as sarcomatoid carcinoma with uncertainty of epithelial marker expression in the sarcomatous component.[15,40] Whether ureteral duplication is a risk factor for malignancy remains unclear. In contrast, horseshoe kidney, which is another common urinary tract malformation, is a risk factor for pelvic tumor.[12,44] The higher incidence of pelvic tumors in horseshoe kidneys is attributed to chronic irritability, such as urinary stasis, urinary tract infection, and stone formation.[12,44] Based on this speculation, it is reasonable to hypothesize that ureteral duplication is also a risk factor for malignancy because this anomaly is occasionally accompanied by urinary reflux and infection. Of note, 1 of the duplicated ureters of the present case, where the tumor is considered to have arisen, showed severe lymphocytic inflammation, which was evidently different from the other duplicate ureter.

The differential diagnosis of carcinosarcoma includes sarcomatoid carcinoma, carcinoma with reactive stromal proliferation, and carcinoma with a benign heterologous component. Sarcomatoid carcinoma can be differentiated by immunohistochemical studies based on the definition of carcinosarcoma/sarcomatoid carcinoma mentioned above. Carcinomas with reactive stromal proliferation and benign heterologous components can be differentiated by careful examination for malignant cytohistological features. If the sarcomatous component shows smooth muscle differentiation, as in the present case, inflammatory myofibroblastic tumor should be distinguished. A recent study showed that ALK-1 protein expression and ALK gene rearrangement were identified in approximately 60% of inflammatory myofibroblastic tumor cases in the urinary bladder, while sarcomatoid urothelial carcinoma was negative for ALK-1 expression and ALK rearrangement, indicating the utility of these markers in differential diagnosis.[45]

The prognosis of carcinosarcoma/sarcomatoid carcinoma of the upper urinary tract has not been systematically studied due to the paucity of cases. In the urinary bladder, to the best of our knowledge, 2 studies comparing the prognosis of carcinosarcoma and sarcomatoid carcinoma have been reported.[10,11] One study of 41 cases showed no significant difference in survival between carcinosarcoma and sarcomatoid carcinoma.[10] Conversely, another study of 301 cases concluded that the survival rate of sarcomatoid carcinoma is better than that of carcinosarcoma.[11] As noted in a recent review, the latter study has a limitation of not performing a central pathology review.[15] However, stratifying by T stage in survival analysis, which was not conducted in the former study, provides superiority of the latter study. Indeed, in the former study, there were more pT4 cases in the sarcomatoid carcinoma group than in the carcinosarcoma group, leaving the possibility that failure to show a significant difference in survival was due to selection bias. Given that carcinosarcoma and sarcomatoid carcinoma of the urinary tract may differ in clinical outcome, we believe that the distinction should be made between these 2 entities for further investigation.

In summary, carcinosarcoma of the upper urinary tract accompanied by ureteral duplication is extremely rare. Further studies are warranted to elucidate the association between
## Table 2

Clinicopathological characteristics of 24 cases of malignant tumor accompanied by ureteral duplication.

| Case | References [number] | Age (years) | Sex | Diagnosis | Tumor location | Duplication type | Symptoms | Treatment | pT stage | Metastasis | Local recurrence | Outcome | Follow-up period |
|------|---------------------|-------------|-----|-----------|---------------|-----------------|-----------|-----------|----------|------------|------------------|----------|-----------------|
| 1    | Leadbetter et al. [24] | 66          | M   | papillary carcinoma | right upper moiety pelvis | complete | painless hematuria | nephroureterectomy | not described | not described | not described | not described | not described | not described |
| 2    | Gutiérrez et al. [25] | 56          | F   | urothelial (transitional cell) carcinoma | right lower moiety ureter (distal) | not described | painless hematuria | nephroureterectomy | pT2         | not described | not described | not described | not described | alive | 2 months |
| 3    | Budd [26]           | 40          | M   | urothelial (transitional cell) carcinoma | right upper moiety pelvis | complete | painless hematuria | nephrectomy | not described | not described | not described | not described | not described | not described |
| 4    | Shenevanszky et al. [27] | 67          | M   | urothelial (transitional cell) carcinoma | right lower moiety ureter | incomplete | painless hematuria | nephroureterectomy | not described | not described | not described | not described | not described | alive | 24 months |
| 5    | Gasser et al. [28]  | 47          | M   | urothelial (transitional cell) carcinoma with osseous metaplasia | left ureter | not described | flank discomfort, abdominal distension, dysuria | nephroureterectomy | not described | not described | not described | not described | not described | not described |
| 6    | Gopi-Atte et al. [29] | 74          | M   | urothelial (transitional cell) carcinoma | left upper moiety ureter (distal) | incomplete | painless hematuria | nephroureterectomy | pT2         | not described | not described | not described | not described | alive | 24 months |
| 7    | Arce et al. [30]    | 66          | M   | urothelial (transitional cell) carcinoma | left lower moiety pelvis | incomplete | painless hematuria | nephroureterectomy | not described | not described | not described | not described | not described | alive | 3 months |
| 8    | Döbbi et al. [31]   | 81          | M   | urothelial (transitional cell) carcinoma | left upper moiety ureter (distal) | complete | dysuria, abdominal pain | nephroureterectomy | pT1         | not described | not described | not described | not described | alive | 7 months |
| 9    | Chung et al. [32]   | 42          | F   | leiomyosarcoma | left upper moiety pelvis | incomplete | none | nephrectomy | pT2a        | not described | not described | not described | not described | not described | alive | 6 months |
| 10   | Tan et al. [33]     | 62          | M   | urothelial (transitional cell) carcinoma | left ureter (middle) | incomplete | painless hematuria, dysuria, abdominal pain, etc. | nephroureterectomy | pT1         | not described | not described | not described | not described | alive | 24 months |
| 11   | Zéras et al. [34]   | 38          | M   | urothelial (transitional cell) carcinoma | left ureter | not described | none | nephroureterectomy | pTis        | not described | not described | not described | not described | not described | alive | 6 months |
| 12   | Kawai et al. [35]   | 67          | F   | urothelial (transitional cell) carcinoma | right ureter | incomplete | hematuria | nephroureterectomy | pT3         | not described | not described | not described | not described | alive | 6 months |
| 13   | Hatatani et al. [36] | 43          | M   | sarcomatoid urothelial (transitional cell) carcinoma | left upper moiety pelvis | not described | flank pain | radical nephrectomy | not described | not described | not described | not described | (+) dead | 14 months |
| 14   | Li et al. [37]      | 74          | M   | urothelial (transitional cell) carcinoma | right upper moiety pelvis | incomplete | painless hematuria | nephroureterectomy | pT1         | not described | not described | not described | not described | (+) dead | 12 months |
| 15   | Chen et al. [38]    | 66          | M   | urothelial (transitional cell) carcinoma | right lower moiety pelvis | incomplete | painless hematuria | nephroureterectomy | not described | not described | not described | not described | (+) dead | 6 months |
| 16   | Chen et al. [39]    | 58          | M   | urothelial (transitional cell) carcinoma | right upper moiety pelvis | incomplete | painless hematuria | nephroureterectomy | not described | not described | not described | not described | (+) dead | 24 months |
| 17   | Chen et al. [40]    | 65          | F   | urothelial (transitional cell) carcinoma | left lower moiety ureter (distal) | incomplete | painless hematuria, myocardial infarction, etc. | nephroureterectomy | pT1 or less | not described | not described | (+) alive | 24 months |
| 18   | Ursi et al. [41]    | 68          | F   | urothelial (transitional cell) carcinoma | right upper moiety ureter (proximal) | complete | painless hematuria | nephroureterectomy | not described | not described | not described | not described | not described | alive | 24 months |
| 19   | Bans et al. [42]    | 51          | M   | urothelial (transitional cell) carcinoma | right upper and lower moiety ureter (distal) | incomplete | painless hematuria | nephroureterectomy | pT2         | not described | not described | not described | not described | not described | alive | 24 months |
| 20   | Chen et al. [43]    | 77          | M   | sarcomatoid urothelial (transitional cell) carcinoma | left lower moiety pelvis | complete | abdominal pain, fever, anorexia | nephroureterectomy | pT3         | not described | not described | not described | not described | (+) alive | not described |
| 21   | Kao et al. [44]     | 87          | F   | urothelial (transitional cell) carcinoma | left lower moiety ureter (proximal) | not described | painless hematuria, dysuria, abdominal fullness | radiotherapy | not described | not described | not described | not described | not described | not described | 6 months |
| 22   | Ogawa et al. [45]   | 71          | F   | squamous cell carcinoma | left upper moiety pelvis | incomplete | hematuria, back pain, radiculopathy | nephroureterectomy | pT4         | not described | not described | (+) alive | 6 months |
| 23   | Zhang et al. [46]   | 65          | M   | urothelial (transitional cell) carcinoma | right upper moiety pelvis | complete | painless hematuria | nephroureterectomy | pT1         | not described | not described | not described | not described | (+) alive | 6 months |
| 24   | Present case        | 60          | F   | carcinosarcoma | right lower moiety ureteropelvic junction | incomplete | painless hematuria | nephroureterectomy | pT1         | not described | not described | not described | not described | alive | 21 months |

*We determined the pT factors of these cases from pathological findings described in the articles based on the TNM Classification of Malignant Tumours, 8th edition.

**This patient had synchronous contralateral ureteral urothelial carcinoma without ureteral duplication.

*** Transitional cell carcinoma arose from a urothelial polyp in this case.
ureteral duplication and malignancy as well as the clinicopathological correlation in carcinosarcoma/sarcomatoid carcinoma of the urinary tract.

Author contributions
Conceptualization: Kentaro Tsuji, Hisashi Oshiro.
Data curation: Kentaro Tsuji, Atsushi Ito, Shinsuke Kurokawa, Takeo Nakaya, Hisashi Oshiro.
Formal analysis: Kentaro Tsuji, Atsushi Ito, Shinsuke Kurokawa, Hisashi Oshiro.
Funding acquisition: Hisashi Oshiro.
Investigation: Kentaro Tsuji, Atsushi Ito, Shinsuke Kurokawa, Takeo Nakaya, Hirotoishi Kawata, Hisashi Oshiro.
Methodology: Kentaro Tsuji, Taichiro Yoshimoto, Hirotoishi Kawata, Mio Tamba-Sakaguchi, Hisashi Oshiro.
Project administration: Hisashi Oshiro.
Resources: Noriyoshi Fukushima, Hisashi Oshiro.
Software: Hisashi Oshiro.
Supervision: Takeo Nakaya, Taichiro Yoshimoto, Hirotoishi Kawata, Mio Tamba-Sakaguchi, Noriyoshi Fukushima, Hisashi Oshiro.
Validation: Shinsuke Kurokawa, Taichiro Yoshimoto, Hirotoishi Kawata, Mio Tamba-Sakaguchi, Noriyoshi Fukushima, Hisashi Oshiro.
Visualization: Mio Tamba-Sakaguchi, Noriyoshi Fukushima, Hisashi Oshiro.
Writing – original draft: Kentaro Tsuji, Hisashi Oshiro.
Writing – review & editing: Kentaro Tsuji, Hisashi Oshiro.
Hisashi Oshiro orcid: 0000-0002-5036-9282.

References
[1] Cantrell LA, Blank SV, Duska LR. Uterine carcinosarcoma: a review of the literature. Gynecol Oncol 2015;137:581–8.
[2] del Carmen MG, Bierer M, Schorge JO. Carcinosarcoma of the ovary: a review of the literature. Gynecol Oncol 2012;125:271–7.
[3] Koss MN, Hochholzer L, Frommelt RA. Carcinomas of the lung: a clinicopathologic study of 66 patients. Am J Pathol 1999;23:1514–26.
[4] Wargozz ES, Norris HJ. Metaplastic carcinomas of the breast. III. Carcinosarcoma. Cancer 1989;64:1490–9.
[5] Sano A, Sakurai S, Kato H, et al. Clinicopathological and immunohistochemical characteristics of esophageal carcinosarcoma. Anticancer Res 2006;26:1775–80.
[6] Aramendi T, Fernandez-Acenero MJ, Villanueva MC. Carcinosarcoma of the colon: report of a rare tumor. Pathol Res Pract 2003;199:345–8.
[7] Russ DA, Kayser C, Neubauer J, et al. Carcinosarcoma of the pancreas: case report with comprehensive literature review. Pancreas 2017;46:1225–33.
[8] Perret L, Chaubert P, Hesseler D, et al. Primary heterologous carcinosarcoma (metaplastic carcinoma) of the urinary bladder: a clinicopathologic, immunohistochemical, and ultrastructural analysis of eight cases and a review of the literature. Cancer 1998;82:1535–49.
[9] Wang J, Wang FW, Kessinger A. The natural history and outcomes of the patients with carcinosarcoma involving kidney and renal pelvis. Adv Urol 2011;2011:693964.
[10] Lopez-Beltran A, Pacelli A, Rothenberg HJ, et al. Carcinosarcoma and sarcomatoid carcinoma of the bladder: clinicopathological study of 41 cases. J Urol 1998;159:1497–503.
[11] Wright JL, Black PC, Brown GA, et al. Differences in survival among patients with sarcomatoid carcinoma, carcinosarcoma and urothelial carcinoma of the bladder. J Urol 2007;178:2302–6. discussion 7.
[12] Wein AJ, Kavoussi LR, Partin AW, et al. Campbell-Walsh Urology. 11th ed. Philadelphia: Elsevier; 2016.
[13] Diemer RA, Chow JS, Kwatra NS, et al. The duplicated collecting system of the urinary tract: embryology, imaging appearances and clinical considerations. Pediatr Radiol 2017;47:1526–38.
[14] Fernbach SK, Feinstein KA, Spencer K, et al. Ureteral duplication and its complications. Radiographics 1997;17:109–27.
[15] Cheng L, Zhang S, Alexander R, et al. Sarcomatoid carcinoma of the urinary bladder: the final common pathway of urothelial carcinoma dedifferentiation. Am J Surg Pathol 2011;35:e34–46.
[16] Torenbeek R, Hermens MA, Meijer GA, et al. Analysis by comparative genomic hybridization of epithelial and spindle cell components in sarcomatoid carcinoma and carcinosarcoma: histogenetic aspects. J Pathol 1999;189:338–43.
[17] Moch H, Humphrey PA, Ulbright TM, et al. World Health Organization Classification of Tumours of the Urinary System and Male Genital Organs. 4th ed. Lyon: IARC Publications; 2016.
[18] Petersen RO, Sesterhenn I, Davis CJ. Urologic pathology. 2nd ed. Philadelphia: JB Lippincott; 1992.
[19] McCluggage WG. Malignant biphasic uterine tumours: carcinosarcomas or metaplasmic carcinomas? J Clin Pathol 2002;55:321–5.
[20] Fuji H, Yoshida M, Gong ZX, et al. Frequent genetic heterogeneity in the clonal evolution of gynecological carcinosarcoma and its influence on phenotypic diversity. Cancer Res 2000;60:114–20.
[21] Singh MT, Wang M, MacLennan GT, et al. Histogenesis of sarcomatoid urothelial carcinoma of the urinary bladder: evidence for a common clonal origin with divergent differentiation. J Pathol 2007;212:420–30.
[22] Armstrong AB, Wang M, Elebe JN, et al. TP53 mutational analysis supports monoclonal origin of biphasic sarcomatoid urothelial carcinoma (carcinosarcoma) of the urinary bladder. Mod Pathol 2002;15:113–8.
[23] Leadbetter WF, Schulz MD, et al. Papillary carcinoma (Grade II A), involving upper half of bifid pelvis. N Engl J Med 1946;235:599–602.
[24] Tudor RG, Clear JD. Conservative surgery in the management of carcinoma in a duplex ureter. J R Coll Surg Edinb 1986;31:323–4.
[25] Budd JS. Primary transitional cell carcinoma of the renal pelvis in a duplicated collecting system. Br J Clin Pract 1987;41:1063–4.
[26] Sreenevasan G, Por PK, Mukherjee AK, et al. Carcinoma in one limb of an incompletely duplicated ureter. Br J Urol 1987;60:79–80.
[27] Gassner JR, Cullen GM, Walsh JP. Osseous metaplasia of transitional cell carcinoma within a duplicated ureter. AJR Am J Roentgenol 1989;153:1101–2.
[28] Gepi-Atte S, Gingell JC. Ureteric tumour in a duplex system. Br J Urol 1991;68:106.
[29] Assae D, Frank RG, Gerard PS, et al. Transitional cell carcinoma of the renal pelvis in an incompletely duplicated collecting system. N Y State J Med 1992;92:500–1.
[30] Dudak SD, Ament RA. Transitional cell carcinoma in a duplicated ectopic ureter. Urology 1995;46:231–3.
[31] Chung YG, Kang SG, Yoon SM, et al. Leiomyosarcoma arising from the blind end of a bifid renal pelvis. Yonsei Med J 2007;48:557–60.
[32] Tan LB, Tserng BR, Huang WH, et al. Synchronous bilateral carcinoma in one limb of a duplicated collecting system. J Urol 1996;155:3196–9.
[33] Zargar A, Rashidkis G, Nakopolou L, et al. Transitional cell carcinoma arising from a fibroepithelial ureteral polyp in a patient with duplicated upper urinary tract. J Urol 1997;157:2252–3.
[34] Kawamura H, Sasaki N. Transitional cell carcinoma in the blind-ending branch of the bifid ureter. Br J Urol 1990;68:307–8.
[35] Hisataka T, Takahashi A, Taguchi K, et al. Sarcomatoid transitional cell carcinoma originating from a duplicated renal pelvis. Int J Urol 2001;8:704–6.
[36] Li JD, Lin JS, Yao WJ. Synchronous transitional cell carcinoma in both moieties of an incomplete duplex system. Urology 2002;59:944–5.
[37] Chen KS, Chuang CK, Wu CH, et al. Upper urinary tract tumor in a duplicated collecting system: report of three cases and review of the literature. Chang Gung Med J 2003;26:377–82.
[38] Usual A, Cimentepe E, Koc A, et al. A case of primary ureteral carcinoma in association with unilateral complete duplication of the ureter. Int J Urol Nephrol 2003;35:489–90.
[39] Boris RS, McIntyre L, AlAlassi O, et al. An unusual case of ureteral tumor in a duplex system. Int J Urol Nephrol 2006;38:473–4.
[40] Chen GM, Chen SW, Xia D, et al. Sarcomatoid carcinoma of the renal pelvis in duplex kidney. Chin Med J 2011;124:2074–6.
[41] Kao JL, Huang CH, Chang YM, et al. Ureteral cancer in a duplicated ureter. QJM 2013;106:777–8.
[42] Ogawa M, Morikawa T, Toyoshima T, et al. Squamous cell carcinoma in a duplicated renal pelvis. Int J Clin Exp Pathol 2014;7:7957–61.

[43] Zhang Y, Yu Q, Zhang Z, et al. Renal pelvis urothelial carcinoma of the upper moiety in complete right renal duplex: a case report. Int J Clin Exp Pathol 2015;8:15422–5.

[44] Mizusawa H, Komiyama I, Ueno Y, et al. Squamous cell carcinoma in the renal pelvis of a horseshoe kidney. Int J Urol 2004;11:782–4.

[45] Sukov WR, Cheville JC, Carlsson AW, et al. Utility of ALK-1 protein expression and ALK rearrangements in distinguishing inflammatory myofibroblastic tumor from malignant spindle cell lesions of the urinary bladder. Mod Pathol 2007;20:592–603.