Upper Urinary Tract Tumor in a Duplicated Collecting System: Report of Three Cases and Review of the Literature

Kuo-Su Chen, MD; Cheng-Keng Chuang¹, MD; Ching-Herng Wu, MD; Chuang-Chi Liaw², MD; Ning Lee³, MD

Despite the common anomaly of a duplicated collecting system in the urinary tract, urothelial cancer in a duplicated collecting system is a rare occurrence. Herein, we present 2 cases of renal pelvis tumor and 1 case of a ureter tumor which coexisted with a duplicated collecting system. One of the renal pelvis tumors developed bilateral transitional cell carcinoma within the bilateral duplicated pelvis. This has not been reported previously. The tumor of the ureter in the latter case was located at the junction site of the bifurcation. This finding is consistent with the postulation that urine reflux chronically irritates the distal segment of a duplicated ureter, rendering this segment susceptible to malignant change. Recurrence of the tumor is frequently observed, which necessitates an early diagnosis and radical treatment. (Chang Gung Med J 2003;26:377-82)

Key words: duplicated collecting system, transitional cell carcinoma, upper urinary tract tumor.
Case 1
A 65-year-old woman was diagnosed as having incomplete duplication of the left collecting system and hypoplasia of the right kidney 2 years previous. She was a housekeeper who lived in Chiayi City.

Case 2
A 58-year-old man living in Taoyuan County was admitted with a 6-month history of intermittent painless hematuria. Upon admission, physical examination revealed no abnormality. Occupation and drug history were non-contributory. Laboratory examination disclosed an elevated level of blood urea nitrogen (30 mg/dl) and creatinine (1.9 mg/dl), as well as SGOT of 15 U/l, SGPT of 21 U/l, and hemoglobin of 10 g/dl. An initial study with IVU indicated a duplication of the bilateral collecting system (complete duplication on the right side and incomplete duplication on the left). In addition, irregular filling defects were apparent within the calyx of the bilateral upper moieties (Fig. 2). Notably, retrograde pyelography failed to identify any tumor mass, because only the orifice of the right lower moiety ureter could be identified. However, CT revealed a mass on the renal pelvis of the bilateral upper moieties and the left lower moiety. Urine cytology study was positive for TCC.

Next, a bilateral nephroureterectomy and resection of the bladder cuff were performed. Pathologic examination revealed the coexistence of chronic pyelonephritis and transitional cell carcinoma in the bilateral renal pelvis. The patient began maintenance hemodialysis thereafter and regularly received intravesical chemotherapy. Unfortunately, a bladder tumor developed at the end of the first year after the nephroureterectomy. He died of sepsis 2 years after the nephroureterectomy.

Case 3
A 65-year-old woman was diagnosed as having incomplete duplication of the left collecting system and hypoplasia of the right kidney 2 years previous. She was a housekeeper who lived in Chiayi City.
She developed painless intermittent gross hematuria 2 months prior to this admission; however, she did not seek medical assistance. One night, she felt nauseous, and experienced vomiting and severe dyspnea. She was then sent to our emergency department. Laboratory data at that time showed plasma levels of blood urea nitrogen of 137 mg/dl and creatinine of 16.6 mg/dl. A hemogram revealed a white blood cell count of 8.5 × 10^9/l, a platelet count of 163 × 10^9/l, and a hemoglobin level of 8.5 g/dl. Emergent hemodialysis was immediately undertaken.

After admission, ultrasonographic examination of the kidney revealed left hydronephrosis and hydroureter. CT of the abdomen revealed similar findings. Antegrade (Fig. 3) and retrograde pyelography indicated a filling defect over the distal end of the left lower moiety ureter. The patient thus underwent a left total nephroureterectomy. A mass measuring 3 × 3 × 2 cm in size was identified at the distal end of the lower moiety ureter (immediately above the junction site). Histology of the resected mass revealed a TCC. She was put on maintenance hemodialysis after the left nephroureterectomy because her right kidney was non-functioning. She remained in good condition during the initial 6 months after the operation. Unfortunately, follow-up cystoscope 6 month later revealed recurrence of the TCC on the bladder. She is now (2 years after the operation) still undergoing maintenance hemodialysis and receiving regular intravesical chemotherapy.

**DISCUSSION**

Primary tumors of the renal pelvis or collecting system are relatively uncommon in comparison to tumors of the renal parenchyma, representing about 5%-10% of all renal neoplasms and 1% of all genitourinary tumors; bladder, renal pelvis, and ureter tumors occur in a ratio of 51:3:1, respectively. Duplication of the renal pelvis and ureter, although the most common anomaly of the urinary tract, was found only in 1 of 150 autopsies. Thus, an upper urinary tract tumor encountered within this anomaly is very rare. The current case reports represent the 13th to 15th cases reported in the literature.

While TCC is a multifocal disease, only 1% to 3% of upper urinary tract tumors are bilateral at initial presentation. In fact, there has been no previous report regarding the development of bilateral TCC within a bilaterally duplicated collecting system. Our case no. 2 simultaneously developed tumors at the pelvis of the bilateral upper moieties and the left lower moiety. This is the first reported instance in the literature.

We summarize the clinical and histopathologic characteristics of the 3 cases in the current report and the 12 cases reported in the literature in Table 1. The age of these 15 reported cases ranged from 40 to 81 (mean, 64.3) years, and 9 (60%) were male. The tumor was located at the ureter in 11 cases (73%) and at the renal pelvis in the other 4 (27%). The collecting systems were unilaterally duplicated in 13 cases (93%), bilaterally duplicated in 1 case (7%), and information on the remaining case is unavailable. Among the 13 cases with unilateral duplication, 10 (77%) collecting systems were incompletely duplicated, and 7 (54%) were duplicated on the right side. Of interest, in 10 of the 11 cases of ureter tumors,
including our case no. 3, the tumor was located at the distal part of the duplicated ureter (information regarding the 1 remaining case is unavailable). Those tumors were situated either in close proximity to the junction site of the incompletely duplicated ureter (situated at the junction site in nos. 4, 5, 6, and 7, and near the junction site in nos. 3, 9, 10, and 13) or near the ureter orifice in cases of complete duplication (nos. 11 and 15). It appears that tumors preferentially developed at the lower end of the duplicated ureter. This phenomenon was also reported by Kumon et al.\textsuperscript{1} Given that urine reflux, either vesi-coureteric or ureteroureteral, is frequently encountered within a duplicated ureter, the bifurcation site and distal segment of the duplicated ureter naturally sustain greater pressure due to reflux flow. We hypothesize that reflux flow may chronically irritate the distal part of a duplicated ureter and render this segment of the uroepithelium susceptible to neoplastic change.\textsuperscript{17}

Previously, southern Taiwan was an endemic area for blackfoot disease due to the high arsenic content in well water. A high prevalence of bladder tumors has also been reported in this area. However, none of the currently reported cases had lived in a blackfoot endemic area. Also, none of them had special drug or occupation histories.

As duplication is frequently associated with various anatomical variations, diagnosing a tumor within a duplicated collecting system is sometimes diffi-

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**Table 1.** Summary of Reported Cases in the Literature

<table>
<thead>
<tr>
<th>Case no.</th>
<th>Age (yr)</th>
<th>Gender</th>
<th>Path</th>
<th>Tumor location (moiety)</th>
<th>Site and type of duplication</th>
<th>Symptoms</th>
<th>Diagnostic Tool</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>66</td>
<td>M</td>
<td>TCC</td>
<td>RT pelvis (lower)</td>
<td>RT inc</td>
<td>hematuria</td>
<td>Expl</td>
<td>tot neph</td>
</tr>
<tr>
<td>2</td>
<td>58</td>
<td>M</td>
<td>TCC</td>
<td>Bil pelvis (both)</td>
<td>RT inc</td>
<td>hematuria</td>
<td>IVU, CT</td>
<td>tot neph</td>
</tr>
<tr>
<td>3</td>
<td>65</td>
<td>F</td>
<td>TCC</td>
<td>LT ureter (lower)</td>
<td>LT inc</td>
<td>hematuria</td>
<td>CT, RP</td>
<td>tot neph</td>
</tr>
<tr>
<td>4</td>
<td>52</td>
<td>M</td>
<td>NA</td>
<td>RT ureter</td>
<td>RT inc</td>
<td>NA</td>
<td>NA</td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>67</td>
<td>M</td>
<td>NA</td>
<td>LT ureter</td>
<td>LT inc</td>
<td>NA</td>
<td>NA</td>
<td></td>
</tr>
<tr>
<td>6</td>
<td>70</td>
<td>F</td>
<td>NA</td>
<td>RT ureter</td>
<td>RT inc</td>
<td>NA</td>
<td>NA</td>
<td></td>
</tr>
<tr>
<td>7</td>
<td>67</td>
<td>F</td>
<td>NA</td>
<td>LT ureter</td>
<td>LT inc</td>
<td>NA</td>
<td>NA</td>
<td></td>
</tr>
<tr>
<td>8</td>
<td>62</td>
<td>F</td>
<td>NA</td>
<td>RT ureter</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td></td>
</tr>
<tr>
<td>9</td>
<td>62</td>
<td>F</td>
<td>TCC</td>
<td>RT ureter (lower)</td>
<td>RT inc</td>
<td>hematuria, flank pain</td>
<td>RP, CT</td>
<td>tot neph + cuff</td>
</tr>
<tr>
<td>10</td>
<td>78</td>
<td>M</td>
<td>TCC</td>
<td>RT ureter (lower)</td>
<td>RT inc</td>
<td>hematuria</td>
<td>RP</td>
<td>tot neph</td>
</tr>
<tr>
<td>11</td>
<td>56</td>
<td>F</td>
<td>TCC</td>
<td>RT ureter (lower)</td>
<td>RT inc</td>
<td>hematuria</td>
<td>loc excis</td>
<td></td>
</tr>
<tr>
<td>12</td>
<td>40</td>
<td>M</td>
<td>TCC</td>
<td>RT pelvis (upper)</td>
<td>RT inc</td>
<td>hematuria</td>
<td>IVU, sono</td>
<td>hemineph</td>
</tr>
<tr>
<td>13</td>
<td>74</td>
<td>M</td>
<td>TCC</td>
<td>LT ureter (upper)</td>
<td>LT inc</td>
<td>hematuria</td>
<td>Expl</td>
<td>hemineph</td>
</tr>
<tr>
<td>14</td>
<td>66</td>
<td>M</td>
<td>TCC</td>
<td>LT pelvis (lower)</td>
<td>LT inc</td>
<td>hematuria, weakness</td>
<td>US, RP</td>
<td>tot neph</td>
</tr>
<tr>
<td>15</td>
<td>81</td>
<td>M</td>
<td>TCC</td>
<td>LT ureter (upper)</td>
<td>LT com</td>
<td>hematuria, flank pain</td>
<td>CT</td>
<td>tot neph</td>
</tr>
</tbody>
</table>

\textsuperscript{a} Cases no. 4-8 are cited from a review of the Japanese literature reported by Kumon et al.\textsuperscript{11}  
\textbf{Abbreviations:} com: complete; CT: computed tomography; expl: exploratory laparotomy; hemineph: heminephroureterectomy; inc, incomplete; IVU: intravenous urography; loc excis: local excision with ureteroureterostomy; LT: left; NA: not available; path: pathologic diagnosis; RP: retrograde pyelography; RT: right; US: ultrasonography; tot neph: total nephroureterectomy; tot neph + cuff: total nephroureterectomy and resection of the bladder cuff.
cult. Actually, tumors were found only during an exploratory laparotomy in 2 (nos. 1 and 13) of these 15 cases.\(^{(1-7)}\)

A total nephroureterectomy with excision of the bladder cuff has been the standard treatment for upper urinary tract tumors. Seven of these cases underwent a total nephroureterectomy. A heminephroureterectomy was performed in 2 patients: 1 case with a right upper pelvis tumor (no. 12) and another case with a ureter tumor on the left upper moiety (no. 13). Local excision with a ureteroureterostomy was performed in a case with a ureter tumor on the right lower moiety (no. 11). Long-term follow-up was unavailable in these cases. Thus, it is not clear whether the heminephroureterectomy or local excision was appropriate for these patients. Considering the fact that tumor recurrence is high in the ureteric stump after local resection, a total nephroureterectomy seems to be a better treatment choice. A heminephroureterectomy may be considered only in cases of complete duplication, or when the opposite-side kidney is nonfunctioning. Both cases no. 2 and 3 in the present study who received a total nephroureterectomy with excision of the bladder cuff suffered tumor recurrence within 1 year of the operation (no. 1 was finally lost to follow-up). This may necessitate an early diagnosis and radical treatment in such patients.

REFERENCES

雙套腎合併上泌尿道腫瘤：病例報告及文獻回顧

陳國書 莊正鈺 吳景恆 廖宗琦 李 宇

雙套腎是泌尿道常見的先天結異常，但是合併雙套腎的泌尿道惡性腫瘤則很少見。在本文中，我們報告了兩例雙套腎及一例輸尿管腫瘤出現在雙套腎病人身上。在兩例腎盂腫瘤中，有一例是雙側同時出現移形細胞腫瘤；在腫瘤細胞的病例中，這是首例。另外一例輸
尿管腫瘤出現在雙套輸尿管交叉處的上端，文獻也有類似的報告，我們懷疑雙套輸尿管可能
造成尿逆流的刺激，進而造成輸尿管腫瘤容易出現在雙叉處上端尿逆流段。在本文中，我們
也對過去的文獻做回顧。(臺灣醫誌 2003;26:377-82)

關鍵字：雙套腎，移形細胞腫瘤，上泌尿道腫瘤。