Urothelial Carcinoma of the Renal Pelvis and Ureter Associated with Hydronephrosis in a Patient with Horseshoe Kidney: a Case Report

HIDEYUKI ISOBE*1), SHINJI SHIOZAWA*1), SHIGEO HORIE*2)

*1) Department of Urology, Juntendo Tokyo Koto Geriatric Medical Center, Tokyo, Japan, *2) Department of Urology, Juntendo University Graduate School of Medicine, Tokyo, Japan

A 73-year-old woman visited our clinic for right abdominal fullness and dull pain. She had been treated rheumatoid arthritis and CT scan had shown hydronephrosis with a renal stone in the right kidney of horseshoe kidney 2 and a half years ago, although no treatment had been sought because of the lack of symptoms. Although retrograde pyelography showed no filling defect in the upper urinary tract, urine cytology was positive. Subsequent ureteroscopy of the right renal pelvis showed multiple papillary tumors. Right nephroureterectomy and isthmusectomy were performed. The pathological diagnoses were urothelial carcinoma in the renal pelvis and squamous cell carcinoma, accompanied by urothelial carcinoma in the ureter. The present report describes the 35th case of renal pelvic carcinoma occurred in a horseshoe kidney in Japan.

Key words: horseshoe kidney, renal pelvic urothelial carcinoma, hydronephrosis

Introduction

Horseshoe kidney is a renal fusion anomaly that is found in approximately 0.25% of the population. Horseshoe kidney often develops hydronephrosis, urolithiasis and urinary tract infection1 2). Urothelial carcinoma of renal pelvis in a horseshoe kidney is relatively rare. This report describes a case of hydronephrosis, urothelial carcinoma of the right renal pelvis and ureter in a patient with horseshoe kidney, followed by a review of the 34 cases of renal pelvic tumor in a horseshoe kidney previously reported in Japan.

Case report

A 73-year-old woman visited our urology clinic for right abdominal fullness and dull pain. She had been treated rheumatoid arthritis and CT scan had shown hydronephrosis with a renal stone in the right kidney of horseshoe kidney 2 and a half years ago, although no treatment had been sought because of the lack of symptoms. CT scan revealed right hydronephrosis and right renal stone, but no tumors were identified in the urinary tract (Figure–1).

She had no history of pyelonephritis. No hematuria was seen, and urine cytology was negative. A ureteral catheterization drained 1,500 cc of dark red urine. Retrograde pyelography showed the stenosis of right uretero–pelvic junction and no filling defect in the right upper urinary tract. However the urine cytology was positive. Subsequent contrast-enhanced CT after the disappearance of hydronephrosis identified horseshoe kidney (Figure–2).

Corresponding author: Hideyuki Isobe
Juntendo Tokyo Koto Geriatric Medical Center
3-3-20 Shinsuna, Koto-ku, Tokyo 136-0075, Japan
TEL: +81-3-5632-3111 (ext. 3730) FAX: +81-3-5632-3728 E-mail: isobe@juntendo.gmc.ac.jp

[Received Sep. 4, 2015] [Accepted Oct. 21, 2015]
Ureteroscopy of the right renal pelvis was performed in November, 2010 and showed multiple papillary tumors in the right renal pelvis (Figure-3).

Right nephroureterectomy and isthmusectomy were performed in January, 2011. A midline abdominal incision was made, and the posterior peritoneum was incised from the hepatic flexure to the ileocecal junction. The right kidney was markedly adherent to perirenal tissues and was difficult to separate from the duodenum, ascending colon and liver. The right ureter was identified with vessel tape, and the right renal hilus was exposed. One right renal artery and three renal veins were ligated and dissected. The fusion area was approximately 1 cm in width. The feeding artery to the isthmus from the right common iliac artery was ligated and dissected. The isthmus was changed in color to pale, and the kidney was split at the isthmus without left renal hilar clamping for the preservation of left renal function, and the stump was closed with 2-0 chromic z sutures. The patient’s postoperative course was uneventful.

The pathological diagnoses were urothelial carcinoma in the renal pelvis, G2>G1, squamous cell carcinoma, G2 > G3, pT3b, INFβ, v1, ly1, and urothelial carcinoma in the ureter, G2, pTis (Figures-4, 5). Urine cytology was recognized as class III following three successive postoperative follow up. The patient was subsequently performed for bladder mucosal random biopsy to examine if she had carcinoma in situ in the bladder. The pathological diagnosis was urothelial carcinoma G1, pTis. She subsequently developed a left ureteral tumor at 17 months after surgery, which caused hydronephrosis. Her renal function gradually declined, and hemodialysis was initiated. She refused further urological treatments. She ultimately died of metastases to the lymph nodes and liver at 20 months after surgery.
**Table 1** Urothelial carcinoma of renal pelvis and ureter in a horseshoe kidney in Japan

<table>
<thead>
<tr>
<th>Author</th>
<th>Year</th>
<th>Age</th>
<th>Sex</th>
<th>Complaint</th>
<th>Location</th>
<th>Complication</th>
<th>Cytology</th>
<th>Pathology</th>
<th>Prognosis</th>
<th>Adjuvant Therapy</th>
</tr>
</thead>
<tbody>
<tr>
<td>Matsushima</td>
<td>1975</td>
<td>27</td>
<td>M</td>
<td>Hematuria</td>
<td>Lt P</td>
<td>Nfk</td>
<td>Unknown</td>
<td>UC, G2, pT?</td>
<td>13MoNED</td>
<td>Unknown</td>
</tr>
<tr>
<td>Ouhashi</td>
<td>1985</td>
<td>60</td>
<td>F</td>
<td>Tumor palpable</td>
<td>Lt P</td>
<td>Nfk</td>
<td>4</td>
<td>UC, G3, pT3</td>
<td>NED</td>
<td>Unknown</td>
</tr>
<tr>
<td>Araki</td>
<td>1988</td>
<td>57</td>
<td>M</td>
<td>Hematuria</td>
<td>Lt P</td>
<td>None</td>
<td>Unknown</td>
<td>Unknown</td>
<td>11MoNED</td>
<td>Chemotherapy</td>
</tr>
<tr>
<td>Murai</td>
<td>1989</td>
<td>60</td>
<td>F</td>
<td>Hematuria</td>
<td>Lt P</td>
<td>None</td>
<td>4</td>
<td>Poorly diff. UC, pT3</td>
<td>60MoNED</td>
<td>Chemotherapy</td>
</tr>
<tr>
<td>Shimada</td>
<td>1990</td>
<td>64</td>
<td>M</td>
<td>Lt. flank pain</td>
<td>Lt P</td>
<td>Nfk</td>
<td>Unknown</td>
<td>UC, G3 &gt; SCC, AC, pT?</td>
<td>11MoNED</td>
<td>Chemotherapy</td>
</tr>
<tr>
<td>Ounishi</td>
<td>1991</td>
<td>59</td>
<td>M</td>
<td>Hematuria</td>
<td>Lt P</td>
<td>Hydronephrosis</td>
<td>5</td>
<td>UC, G2, pT?</td>
<td>Unknown</td>
<td>Unknown</td>
</tr>
<tr>
<td>Ozawa</td>
<td>1991</td>
<td>73</td>
<td>M</td>
<td>Unknown</td>
<td>P</td>
<td>Hydronephrosis</td>
<td>5</td>
<td>UC, G3, pT1</td>
<td>2MoDeath</td>
<td>Unknown</td>
</tr>
<tr>
<td>Tsuji</td>
<td>1991</td>
<td>49</td>
<td>M</td>
<td>Unknown</td>
<td>P</td>
<td>Unknown</td>
<td>2</td>
<td>UC, G1, pT?</td>
<td>NED</td>
<td>Unknown</td>
</tr>
<tr>
<td>Sugano</td>
<td>1992</td>
<td>60</td>
<td>M</td>
<td>Hematuria</td>
<td>Lt P</td>
<td>None</td>
<td>4</td>
<td>UC, G2, pT3</td>
<td>10MoNED</td>
<td>None</td>
</tr>
<tr>
<td>Murata</td>
<td>1994</td>
<td>69</td>
<td>M</td>
<td>Hematuria</td>
<td>Rt P</td>
<td>Hydronephrosis</td>
<td>3b</td>
<td>SCC &gt; UC, G2, pT3</td>
<td>Unknown</td>
<td>Chemotherapy</td>
</tr>
<tr>
<td>Imazono</td>
<td>1995</td>
<td>63</td>
<td>M</td>
<td>Pollakisuria</td>
<td>P</td>
<td>Nfk</td>
<td>4</td>
<td>UC, G2, pT4</td>
<td>13MoNED</td>
<td>Unknown</td>
</tr>
<tr>
<td>Tsuru</td>
<td>1997</td>
<td>71</td>
<td>F</td>
<td>Appetite loss</td>
<td>P</td>
<td>Nfk</td>
<td>1</td>
<td>UC, G3 &gt; SCC, pT4</td>
<td>Cancer death.</td>
<td>None</td>
</tr>
<tr>
<td>Higashida</td>
<td>1997</td>
<td>44</td>
<td>M</td>
<td>Lt. flank pain</td>
<td>P</td>
<td>Nfk</td>
<td>4</td>
<td>UC, G2, pT1</td>
<td>8MoNED</td>
<td>None</td>
</tr>
<tr>
<td>Kitani</td>
<td>1998</td>
<td>91</td>
<td>M</td>
<td>Unknown</td>
<td>P</td>
<td>Unknown</td>
<td>3b</td>
<td>UC, G3 &gt; SCC, pT2</td>
<td>6MoNED</td>
<td>Unknown</td>
</tr>
<tr>
<td>Sugawara</td>
<td>1999</td>
<td>68</td>
<td>M</td>
<td>Hematuria</td>
<td>Lt P</td>
<td>None</td>
<td>Unknown</td>
<td>UC, G3 = SCC, pT4</td>
<td>3MoNED</td>
<td>Chemotherapy</td>
</tr>
<tr>
<td>Kagawa</td>
<td>1999</td>
<td>75</td>
<td>M</td>
<td>Unknown</td>
<td>P</td>
<td>None</td>
<td>5</td>
<td>UC, G3 &gt; G2, pT4</td>
<td>Unknown</td>
<td>Unknown</td>
</tr>
<tr>
<td>Matsumoto</td>
<td>2000</td>
<td>58</td>
<td>M</td>
<td>Atypical cells</td>
<td>Rt P</td>
<td>None</td>
<td>4</td>
<td>UC, G2, pT1</td>
<td>8MoNED</td>
<td>None</td>
</tr>
<tr>
<td>Yamamoto</td>
<td>2001</td>
<td>69</td>
<td>M</td>
<td>Back pain</td>
<td>Lt P</td>
<td>None</td>
<td>Unknown</td>
<td>UC, G3 &gt; G2, pT3, pL1, PV1 *</td>
<td>Cancer death.</td>
<td>None</td>
</tr>
<tr>
<td>Ishida</td>
<td>2001</td>
<td>59</td>
<td>M</td>
<td>Hematuria</td>
<td>P</td>
<td>None</td>
<td>5</td>
<td>UC, G3 &gt; SCC, pT2</td>
<td>Unknown</td>
<td>Unknown</td>
</tr>
<tr>
<td>Ueda</td>
<td>2002</td>
<td>41</td>
<td>M</td>
<td>Abdominal fullness</td>
<td>Lt P</td>
<td>Hydronephrosis</td>
<td>3b</td>
<td>Mucinous AC</td>
<td>Cancer death.</td>
<td>Chemotherapy</td>
</tr>
<tr>
<td>Kosugi</td>
<td>2002</td>
<td>74</td>
<td>M</td>
<td>Unknown</td>
<td>P</td>
<td>Hydronephrosis</td>
<td>4</td>
<td>UC, G3 &gt; G2, pT3</td>
<td>21MoNED</td>
<td>Unknown</td>
</tr>
<tr>
<td>Narita</td>
<td>2003</td>
<td>57</td>
<td>F</td>
<td>Lt. flank pain</td>
<td>P</td>
<td>Hydronephrosis</td>
<td>Unknown</td>
<td>UC, G7, pT?</td>
<td>Unknown</td>
<td>Unknown</td>
</tr>
<tr>
<td>Kobori</td>
<td>2003</td>
<td>60</td>
<td>M</td>
<td>Unknown</td>
<td>P</td>
<td>Nfk</td>
<td>Unknown</td>
<td>UC, G2, pT1</td>
<td>18MoNED</td>
<td>Unknown</td>
</tr>
<tr>
<td>Minagawa</td>
<td>2004</td>
<td>40</td>
<td>M</td>
<td>Hematuria</td>
<td>Lt P</td>
<td>Hydronephrosis</td>
<td>4</td>
<td>UC, G1, pT4</td>
<td>4MoNED</td>
<td>None</td>
</tr>
<tr>
<td>Mizusawa</td>
<td>2004</td>
<td>80</td>
<td>F</td>
<td>Occult blood in urine</td>
<td>Lt P</td>
<td>None</td>
<td>5</td>
<td>SCC, pTa with thrombus to renal vein</td>
<td>Cancer death.</td>
<td>None</td>
</tr>
<tr>
<td>Urunishira</td>
<td>2005</td>
<td>71</td>
<td>F</td>
<td>Hematuria</td>
<td>Rt P</td>
<td>Nfk</td>
<td>1</td>
<td>UC, G1, pT1</td>
<td>28MoNED</td>
<td>None</td>
</tr>
<tr>
<td>Tamura</td>
<td>2007</td>
<td>67</td>
<td>F</td>
<td>Unknown</td>
<td>P</td>
<td>Hydronephrosis</td>
<td>1</td>
<td>UC, G2, pT1</td>
<td>NED</td>
<td>Unknown</td>
</tr>
<tr>
<td>Murakami</td>
<td>2007</td>
<td>59</td>
<td>F</td>
<td>Pollakisuria</td>
<td>Lt U</td>
<td>Hydronephrosis</td>
<td>4</td>
<td>UC, G2 &gt; G3, pT3</td>
<td>Unknown</td>
<td>Unknown</td>
</tr>
<tr>
<td>Otsuka</td>
<td>2008</td>
<td>36</td>
<td>M</td>
<td>Hematuria</td>
<td>Lt P</td>
<td>None</td>
<td>Unknown</td>
<td>UC, G2, pT2</td>
<td>5MoNED</td>
<td>Chemotherapy</td>
</tr>
<tr>
<td>Mochizuki</td>
<td>2009</td>
<td>73</td>
<td>M</td>
<td>Hematuria</td>
<td>Rt P</td>
<td>None</td>
<td>Unknown</td>
<td>UC, G2, pT2</td>
<td>Unknown</td>
<td>Unknown</td>
</tr>
<tr>
<td>Mochizuki</td>
<td>2009</td>
<td>60</td>
<td>M</td>
<td>Hematuria</td>
<td>Lt P</td>
<td>None</td>
<td>5</td>
<td>Unknown</td>
<td>Unknown</td>
<td>Unknown</td>
</tr>
<tr>
<td>Ryuge</td>
<td>2009</td>
<td>63</td>
<td>M</td>
<td>Hematuria</td>
<td>Rt U</td>
<td>Hydronephrosis</td>
<td>4</td>
<td>UC, G2, pT3</td>
<td>10MoNED</td>
<td>Chemotherapy</td>
</tr>
<tr>
<td>Matsumoto</td>
<td>2010</td>
<td>74</td>
<td>M</td>
<td>Hematuria</td>
<td>Lt P</td>
<td>None</td>
<td>Unknown</td>
<td>UC, G3, pT1</td>
<td>11MoNED</td>
<td>Unknown</td>
</tr>
<tr>
<td>Matsumoto</td>
<td>2011</td>
<td>64</td>
<td>M</td>
<td>Hematuria</td>
<td>Lt P</td>
<td>None</td>
<td>5</td>
<td>UC, G2, pTa</td>
<td>Unknown</td>
<td>Unknown</td>
</tr>
<tr>
<td>Present case</td>
<td>2011</td>
<td>73</td>
<td>F</td>
<td>Abdominal fullness</td>
<td>Rt P, U</td>
<td>Nfk</td>
<td>3b</td>
<td>UC, G2 &gt; G1, SCC, G2 &gt; G3, pT3, UC, G2, pT4</td>
<td>Cancer death.</td>
<td>None</td>
</tr>
</tbody>
</table>


**Discussion**

A horseshoe kidney is the most common fusion anomaly, with an incidence of 1:400 in the general population and a male–female ratio of 2:1. Complications include hydronephrosis, urinary tract infection and urolithiasis as the result of structural abnormalities. Blackard et al. described 72 cases...
of renal malignant tumors occurring in the background of a horseshoe kidney. Subsequent studies suggest an associated incidence of renal cell carcinoma of 40 – 50%, while associated renal pelvic tumor is less common (7 – 30%) \(^3-^5\). In general, it is suggested that chronic urinary tract infection or untreated urolithiasis contribute to the development of urothelial carcinoma. The incidence of urothelial carcinoma is higher for horseshoe kidneys than for non–fused kidneys (25% vs. 7.7%).

To our knowledge, 34 cases of urothelial carcinoma of the renal pelvis or ureter in a horseshoe kidney have been previously reported in Japan (Table-1). Therefore, the present report describes the 35th case. Among these 35 cases, patient age ranged from 27 to 80 years (mean age, 66 years), and the male – female ratio was 3 : 2. The tumor was located in the renal pelvis in 33 cases and in the ureter in three cases. The most common signs or symptoms were macroscopic hematuria (17 cases; 49%) and urinary tract obstruction (20 cases; 57%). In 25 of the 35 cases in which urine cytology was assessed, urine cytology was positive in 17 (68%), false positive in four (16%) and negative in four (16%). Cytological examinations appeared to be more sensitive for diagnosis of urothelial carcinoma in patients with a horseshoe kidney than in patients with a non–fused kidney. Among the 33 cases in which was pathologically diagnosed, 31 cases (94%) were transitional cell carcinoma, eight cases (24%) were squamous cell carcinoma, and two cases (6%) were adenocarcinoma. A mixture of urothelial carcinoma and squamous cell carcinoma was found in seven cases (21%), and a mixture of urothelial carcinoma, squamous cell carcinoma and adenocarcinoma was found in one case (3%). Among the 29 cases in which pathological grade was described, the pathological grade was G1 in three cases (10%), G2 in 14 cases (48%) and G3 in 12 cases (42%). Among the 27 cases in which pathological stage was described, the pathological stage was pTis in one case, pTa and pT1 in 10 cases, pT2 in three cases and pT3 and over in 14 cases. In the case including squamous cell carcinoma, the tumor grade and histological T stage were higher than those any other case, and outcomes were poor, even though the patients received chemotherapy. This may be because chemotherapy is not so effective for squamous cell carcinoma of the urinary tract. The present case had urothelial carcinoma and squamous cell carcinoma with lesions in the renal pelvis accompanied by carcinoma in situ in the middle ureter.

Anatomy of the isthmus and the major vessels is sometimes complicated in horseshoe kidney. According to Kölln et al., blood supply to the isthmus can be from the 1) renal artery, 2) the aorta, or the 3) common iliac arteries \(^6\). To our knowledge, 12 cases have multiple renal arteries and in these cases, the blood supply to the isthmus are from renal artery in 5 cases, aorta in 5 cases, and common iliac artery in 2 cases. In our case, blood supply to the isthmus was from the right common iliac artery, and the identification and the ligation of blood vessels was relatively simple. Three–dimensional and multi–row detector CT was informative \(^7\).

**Conclusion**

We described a case of urothelial carcinoma of renal pelvis and ureter in a horseshoe kidney and reviewed the literature.

Clinicians should be aware of the possibility of malignancy in the setting of hydronephrosis in a horseshoe kidney.

**References**