Uratcatal Adenocarcinoma with a C'oncomitant Uratcatal Remnant
- A Case Report -

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Primary adenocarcinoma of urinary bladder is a rare tumor. Most of these tumors are nonurachal adenocarcinoma, and the rest are urachal adenocarcinoma. Especially, urachal adenocarcinoma has been estimated to account for 20 to 39 percent of primary bladder adenocarcinomas. The differential diagnosis between urachal adenocarcinoma and nonurachal adenocarcinoma is important because of different treatment modalities. Urachal adenocarcinoma is similar to nonurachal adenocarcinoma in location and morphologic features, and so the differential diagnosis is sometimes difficult. Since urachal carcinoma originates from an urachal remnant, demonstrating the urachal remnant or associated lesion like urachal cyst or sinus will provide the pathologist with a valuable aid for diagnosis. Here we report a typical case of urachal adenocarcinoma with a well-preserved urachal remnant at the dome of the urinary bladder in a 65-year-old woman.

CASE REPORT

A 65-year-old woman was referred to our hospital to evaluate a painless gross hematuria. This gross hematuria developed 17 months ago, and there was no improvement with the medical treatment she received at a local clinic. She had a past history of taking anti-tuberculosis medication when she was 20 years old. Otherwise, she had neither specific symptoms nor a palpable abdominal mass. Pelvic computerized tomographic (CT) scanning shows an intraluminal protruding mass at the anterior wall of the bladder. The mass was well enhanced and infiltrated to the perivesical fat (Fig. 1). Cystoscopy revealed the anterior wall mass and dome tubercle. Considering the radiological and cystoscopic findings, the possibility of urachal carcinoma was raised. En bloc partial cystectomy was subsequently done, and intraoperative frozen biopsy demonstrated adenocarcinoma with a predominant signet ring cell component.

The resected specimen was a partial cystectomy specimen with perivesical fat and the median umbilical ligament. The urinary bladder measured 5.3 × 5 cm. There was an ulcerofungating tumor in the center of the resected bladder, measuring 3 × 2.5 × 1 cm. The central portion of the mass showed extensive mucosal ulceration, and the cut section of the tumor revealed extensive...
mucinous materials. The cut surface revealed that the mucinous mass was transmurally infiltrative with focal involvement of perivesical fat. Serial sections from the bladder to the umbilical ligament showed a mucin-contained dilated small cyst in the perivesical fat below the mass (Fig. 2). Microscopically, this tumor mass was centered in the muscularis propria rather than in the mucosa, and it was sharply demarcated from the normal surface epithelium. There was no cystitis cystica or glandularis in the adjacent mucosa. The tumor showed abundant extracellular mucin and floating signet ring cell clusters, compatible with mucinous type of adenocarcinoma (Fig. 3). Near the tumor, cystic tubular

Fig. 1. Contrast enhanced CT demonstrates a 2.7 cm-sized, well-enhanced mass (asterisk) in the anterior bladder.

Fig. 2. Partial cystectomy specimen with median umbilical ligament shows an ulcerofungating tumor (arrow) in the center of the resected bladder, measuring $3 \times 2.5 \times 1$ cm. Inset: The cut section reveals mucinous tumor centered in the muscularis propria. A tubular structure with partly cystic dilatation is noted, microscopically confirmed to urachal remnant (asterisk).

Fig. 3. The tumor is composed of nests and sheets of signet ring cells floating in extracellular mucin with nonneoplastic urothelial covering.

Fig. 4. Urachal remnant is composed of pseudostratified columnar epithelia and concentric bundles of smooth muscle, and occasionally filled with inspissated mucinous material.
structures were found. The cystic structures were lined by pseudostratified columnar epithelia with mucous cells, surrounded by smooth muscle, and occasionally filled with inspissated mucinous material (Fig. 4). These histologic findings were reminiscent of an urachal remnant. We failed to find any preneoplastic change of the urachal remnant such as epithelial dysplasia or any direct connection between the tumor and urachal remnant. On immunohistochemical staining, the carcinoma cells were positive for CEA, CK20, MUC-2 and p53, and negative for CK7, MUC-5AC, MUC-6 and chromogranin A. In contrast, the urachal remnant showed strong positivity for CK7 and chromogranin A. Prostate specific antigen (PSA) immunostainings were all negative (Table 1) (Fig. 5).

**Table 1.** Summary of immunohistochemical expressions in urachal adenocarcinoma and urachal remnant

<table>
<thead>
<tr>
<th></th>
<th>Urachal adenocarcinoma</th>
<th>Urachal remnant</th>
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<tbody>
<tr>
<td>Cytokeratin 7</td>
<td>-</td>
<td>+++</td>
</tr>
<tr>
<td>Cytokeratin 20</td>
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<td>-</td>
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<tr>
<td>CEA</td>
<td>+</td>
<td>-</td>
</tr>
<tr>
<td>MUC 2</td>
<td>+++</td>
<td>-</td>
</tr>
<tr>
<td>MUC 5AC</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>MUC 6</td>
<td>-</td>
<td>-</td>
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<tr>
<td>Prostate specific antigen</td>
<td>-</td>
<td>-</td>
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<tr>
<td>Chromogranin A</td>
<td>-</td>
<td>+, Scattered</td>
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**DISCUSSION**

Primary adenocarcinoma accounts for less than 1% of malignant bladder tumors. Adenocarcinoma of urinary bladder is categorized into two groups, urachal or nonurachal origin, according to the strict clinical and pathologic criteria. Previous studies have suggested several criteria for making a diagnosis of urachal carcinoma: first, its location in bladder dome; second, an epicenter in the bladder wall (not the mucosa); third, the absence of cystitis glandularis or intestinal metaplasia in the urothelium; forth, other primary sites are excluded. Our case is a typical urachal carcinoma that met with all these diagnostic criteria.

The urachus connects the apex of the bladder to the umbilicus and contains the allantois. After birth, this tubular structure undergoes progressive atrophy and becomes a cord of fibrous tissue.
attached to bladder wall and umbilicus. 32% of normal adults have tubular urachal remnants, and variable kinds of lesions including urachal sinus, fistula, cyst, and neoplasm can be developed in the urachal remnant. Some authors have addressed the identification of urachal remnant as one of the major criteria for urachal adenocarcinoma. Actually, there are a few cases that the urachal remnant was detected in urachal adenocarcinoma. In the present case, we observed the concomitant urachal remnant that had the specific histology seen in urachal adenocarcinoma. It was a great help to us for confirming the diagnosis.

Urachal adenocarcinomas have been histologically classified into colonic, colloid (mucinous), and signet ring cell types. The mucinous type of urachal adenocarcinoma is relatively common, and our case was also the mucinous type. The immunohistochemical expressions of adenocarcinoma of the urinary bladder including urachal adenocarcinoma have been reported to be similar with those of colonic adenocarcinoma. Based on the results of immunohistochemical staining, the urachal adenocarcinoma components showed characteristics similar to the colonic adenocarcinoma; positive for CEA, CK20 and MUC-2, and negative for CK7, MUC-5A and MUC-6. Interestingly, the urachal remnant in our case revealed a somewhat different immunoeexpression: it showed a strong positive reaction for CK7 and chromogranin A. Until now, the immunohistochemical study for the urachal remnant has been rarely mentioned yet. Immunohistochemical staining for PSA and argyrophilic histochemistry have been introduced as a specific marker. We performed PSA immunostaining, which showed as negative, while chromogranin A staining as a marker of neuroendocrine differentiation was positive. Herein we report a typical urachal adenocarcinoma associated with presence of an urachal remnant that showed unique immunohistochemical findings.

REFERENCES