Case Report

Extramedullary plasmacytoma of the ureter

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Abbreviations & Acronyms
AMy = amylase
CT = computed tomography
EAU = European Association of Urology
EMP = extramedullary plasmacytoma
Ig = immunoglobulin
SBP = solitary bone plasmacytoma
T-BIL = total bilirubin
UC = urothelial carcinoma
UTUC = upper tract urothelial carcinoma

Introduction: We report a rare case of an extramedullary plasmacytoma of the ureter. Case presentation: A 77-year-old man was referred to our hospital because of left hydronephrosis. Computed tomography showed a tumor in the left ureter, and tumor invasion into the periureteric fat was suspected. Urine cytology showed atypical cells whose nuclei were hyperchromatic (class IIIb). The left ureteral tumor was diagnosed as urothelial carcinoma (cT3N0M0) preoperatively. Subsequently, we performed laparoscopic radical nephroureterectomy with bladder cuff excision. The tumor was pathologically diagnosed as a plasmacytoma. Postoperative positron emission tomography did not reveal abnormal uptake, and bone marrow aspiration findings were normal. Consequently, the left ureter tumor was diagnosed as an extramedullary plasmacytoma of the ureter. Conclusion: Extramedullary plasmacytoma commonly occurs in the upper respiratory tract or upper gastrointestinal tract. Extramedullary plasmacytoma of the ureter is rare. This is only the fourth reported case of extramedullary plasmacytoma of the ureter. Key words: extramedullary plasmacytoma, hydronephrosis, tumor, ureter, urothelial carcinoma.

Keynote message
The ureter is a rare location for an EMP. Therefore, preoperative diagnosis is challenging. To the best of our knowledge, this is only the fourth reported case of EMP of the ureter.

Introduction
Solitary plasmacytoma is defined as a localized mass of neoplastic monoclonal plasma cells. EMP arises from soft tissue throughout the body. An EMP often occurs in the upper respiratory or gastrointestinal tract, accounting for approximately 80% of cases. The ureter is a rare location for an EMP. Here, we report a case of a primary EMP of the ureter.

Case presentation
A 77-year-old man who presented with abdominal pain was referred to our hospital because an ultrasound sonogram revealed left hydronephrosis. The patient’s height was 165.5 cm and his body weight was 55.45 kg. The patient’s medical history included constipation, hyperuricemia, benign prostatic hyperplasia, auditory disturbance, and lower back pain. The patient’s regular medications included elobixibat hydrate, febuxostat, silodosin, loxoprofen sodium hydrate, and senna. Regarding family history, the patient’s father died of lung cancer and his mother died of unknown causes. The patient had smoked 20 cigarettes per day for 10 years. CT revealed a tumor in the left ureter with a diameter of approximately 2.5 cm (Fig. 1). Tumor invasion into the periureteric fat was suspected. Distant metastasis and regional node involvement were not found on CT. Urine cytology showed atypical cells whose nuclei were hyperchromatic (class IIIb). UC was suspected. The left ureteral tumor was diagnosed as UC (cT3N0M0) preoperatively.

An increased level of creatinine was observed (1.96 mg/dL). Laboratory findings are shown in Table 1. This renal impairment was partly due to atrophy of the right kidney. Segmental ureterectomy was considered to be challenging because the diameter of the left ureteral tumor was approximately 2.5 cm and tumor invasion into the periureteric fat was suspected. Subsequently, we performed laparoscopic radical nephroureterectomy with bladder cuff excision.

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Although the serum creatinine level increased to 3.75 mg/dL after surgery, hemodialysis was not required. He was discharged on postoperative day 13. Macroscopically, a solid tumor (20 × 15 mm) was observed in the middle of the left ureter. Histopathological findings revealed inflammatory cell infiltration and atypical cells, which included numerous eosinophil granules (Fig. 2). No UC component was observed in the tumor. Immunohistochemical staining revealed that atypical cells were positive for CD138 and CD79a, characteristic markers of B cells or plasma cells. In addition, we detected light chain restriction (Igκ > Igγ). Therefore, the left ureter tumor was pathologically diagnosed as a plasmacytoma.

After the diagnosis of EMP, the patient was referred to a hematologist. A postoperative positron emission tomography scan did not show abnormal uptake, and bone marrow aspiration findings were normal. No monoclonal protein was detected in the patient’s blood and urine. Therefore, we diagnosed the left ureter tumor as a primary EMP of the ureter. The patient survived without renal replacement therapy and showed no evidence of multiple myeloma, local recurrence, or distant metastasis until 22 months after surgery.

Discussion

Solitary plasmacytoma is defined as a localized mass of neoplastic monoclonal plasma cells. SBP is characterized by a sole lesion of bone. EMP arises from soft tissue throughout the body. The International Myeloma Working Group defines EMP using the following criteria: (i) a tumor comprising of monoclonal plasma cells in a single extramedullary site, (ii) no lesion in the whole-body bone, (iii) no lesion in the bone marrow, (iv) no involvement of organs, and (v) no monoclonal Ig in serum or urine. Since our patient met the above criteria, the left ureter tumor was diagnosed as a primary EMP of the ureter. An EMP usually occurs in the upper respiratory or gastrointestinal tract, accounting for approximately 80% of cases. The ureter is a rare location for an EMP. To the best of our knowledge, this is only the fourth reported case of EMP of the ureter (Table 2). Compared with EMP, SBP has a significantly higher risk for progression to multiple myeloma than EMP. Therefore, SBP has poor prognosis in comparison with EMP. Symptoms of EMP vary depending on the tumor occurrence site. Alexiou et al. reported approximately 65% of EMP had no recurrence and did not progress to multiple myeloma after treatment. Younger age was reported as a good independent prognostic factor. In contrast, anaplastic type plasmacytoma, a higher histologic grade, and a high level of angiogenesis were reported as poor prognostic factors. Solitary plasmacytoma is highly radiosensitive. It has been reported that a local control rate of 94% was achieved by doses over 40 Gy. Radiotherapy should encompass the primary tumor with a margin of at least 2 cm. Adjuvant chemotherapy might be considered for the tumor larger than 5 cm and/or of a high histological grade.

Complete resection of EMP is considered equivalent to radiotherapy. Since EMP is a highly radiosensitive tumor, postoperative radiotherapy is recommended for patients with inadequate surgical margins or local recurrence. Our case was diagnosed as a UC before surgery. As the diameter of the left ureteral tumor was approximately 2.5 cm and tumor invasion into the periureteric fat was suspected, segmental ureteral resection was considered challenging; therefore, we performed laparoscopic radical nephroureterectomy with bladder cuff excision. Although Landsmann et al. reported segmental ureter resection and ureterocystostomy for an EMP of the
ureter, Klein et al.\textsuperscript{11} and Nagai et al.\textsuperscript{3} reported that radical nephroureterectomy with bladder cuff excision was performed because UC was suspected before surgery.

The EAU guidelines on upper UTUC define high-risk UTUC by the following criteria: (i) multifocal disease, (ii) tumor size \( \geq 2 \text{ cm} \), (iii) high-grade cytology, (iv) high-grade ureterorenoscopy biopsy, (v) local invasion on CT, (vi) hydronephrosis, (vii) previous radical cystectomy for high-grade bladder cancer, and (viii) variant histology. Our case met three of these high-risk UTUC criteria: tumor size \( \geq 2 \text{ cm} \), local invasion on CT, and hydronephrosis. Per the EAU guidelines on UTUC, radical nephroureterectomy is the standard treatment for high-risk UTUC in the middle or proximal ureter. Therefore, we performed laparoscopic radical nephroureterectomy. Indeed, without a biopsy of the ureteral tumor, preoperative diagnosis of an EMP of the ureter is almost impossible. The differential diagnoses for ureteral tumors include UC, EMP, neuroendocrine tumor, metastatic tumor, paraganglioma, fibroepithelial polyp, and inflammatory myofibroblastic tumor. Among these tumors, EMP is a highly

![Fig. 2](image-url) Hematoxylin and eosin staining showing (a) inflammatory cell infiltration and atypical cells, which include numerous eosinophil granules, and tumor cells positive for (b) CD79a, (c) CD138, (d) Ig\(\lambda\), and (e) Ig\(\kappa\). Staining is more pronounced for Ig\(\kappa\) than for Ig\(\lambda\) (light chain restriction). Magnification, \(\times 100\).

### Table 2. Clinical features of reported cases of EMP of the ureter

| Case number | Authors | Year | Age | Sex | Chief complaint | Preoperative ureteroscopy | Preoperative diagnosis | Treatment | Follow-up (months) | Postoperative dialysis | Outcome |
|-------------|---------|------|-----|-----|-----------------|----------------------------|-----------------------|-----------|---------------------|-----------------------|---------|
| 1           | Landsmann S | 2009 | 80  | Female | Renal colic | Not mentioned | Not mentioned | Segmental ureterectomy (open) | UC (cT2N0M0) | Not mentioned | No evidence of disease |
| 2           | Klein T | 2010 | 82  | Female | Hematuria | Not mentioned | Not mentioned | Nephro-ureterectomy | Nephro-ureterectomy (laparoscopic) | 5 | Not required | No evidence of disease |
| 3           | Nagai T | 2016 | 45  | Male | Hematuria | Not performed | Not performed | UC (cT3N0M0) | Nephro-ureterectomy (laparoscopic) | 22 | Not required | No evidence of disease |
| 4           | Okada S | 2021 | 77  | Male | Abdominal pain | Not performed | Not performed | UC (cT3N0M0) | Nephro-ureterectomy (laparoscopic) | 5 | Not required | No evidence of disease |
radiosensitive tumor.\textsuperscript{12–16} If we had preoperatively diagnosed the ureteral tumor as EMP, we would have been able to choose radiotherapy instead of nephroureterectomy. We did not perform diagnostic ureteroscopy because we considered the left ureteral tumor as a high-risk UTUC before surgery. In addition, the use of diagnostic ureteroscopy has been associated with a higher risk of developing bladder recurrence after radical nephroureterectomy.\textsuperscript{17,18} The possibility of cancer dissemination has also been reported.\textsuperscript{18–20} Segmental ureteral resection is considered as another treatment option.\textsuperscript{21} In segmental ureteral resection for the tumor in the middle or proximal ureter, when end-to-end anastomosis of the ureter is challenging, an ileal-ureteral substitution is required, which is technically challenging and more invasive.

In our case, the tumor size was ≥2 cm and tumor invasion into the periureteric fat was suspected on preoperative CT. Segmental ureteral resection was considered challenging. Therefore, we performed laparoscopic radical nephroureterectomy. However, if we had preoperatively diagnosed the ureteral tumor as EMP, we would have been able to choose radiotherapy instead of nephroureterectomy.

**Conclusion**

We report a rare case of a primary EMP of the ureter. The patient survived without recurrence after laparoscopic radical nephroureterectomy.

**Conflict of interest**

The authors declare no conflict of interest.

**Approval of the research protocol by an Institutional Reviewer Board**

Not applicable.

**Informed consent**

Informed consent for publication was obtained from the patient.

**Registry and the Registration No. of the study/trial**

Not applicable.

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