


## Case Report

# A case of pyelocalyceal diverticulum urothelial carcinoma that was difficult to distinguish from cystic renal cell carcinoma preoperatively

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### Abbreviations & Acronyms

CT = computed tomography  
MRI = magnetic resonance imaging  
RP = retrograde pyelography  
UC = urothelial carcinoma

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**Introduction:** Pyelocalyceal diverticulum is a rare disease sometimes difficult to distinguish from cysts. We report a case of urothelial carcinoma originating from a pyelocalyceal diverticulum, difficult to distinguish from cystic renal cell carcinoma preoperatively.

**Case presentation:** A 51-year-old Japanese man complained of gross hematuria. Computed tomography revealed a solid mass in one of the many cystic lesions in the left kidney. He was diagnosed with left cystic renal cell carcinoma and underwent retroperitoneal laparoscopic nephrectomy. Pathological examination revealed high-grade invasive urothelial carcinoma arising within the renal pyelocalyceal diverticulum. The definitive diagnosis was high-grade invasive urothelial carcinoma (pT3). In retrospect, the retrograde pyelography findings indicated the cyst and urinary tract connection. Residual ureterectomy and adjuvant chemotherapy were later performed. The patient has since been recurrence-free.

**Conclusion:** Whether cystic renal cell carcinoma is suspected on imaging, pyelocalyceal diverticulum should be considered a differential diagnosis, though unlikely to be encountered in daily practice.

**Key words:** cysts, diverticulum, renal pelvis, urography, urothelial carcinoma.

## Keynote message

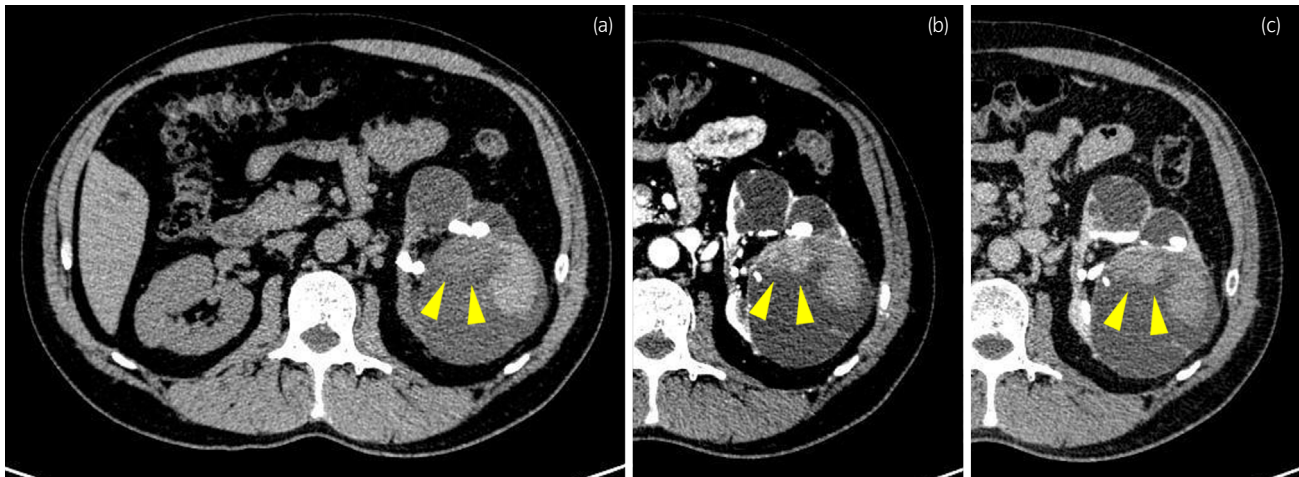
Urothelial carcinoma originating from the pyelocalyceal diverticulum is a rare disease. It is sometimes difficult to distinguish it from cystic renal cell carcinoma. Retrograde pyelography and urinary cytology may be useful in the differential diagnosis of these diseases.

## Introduction

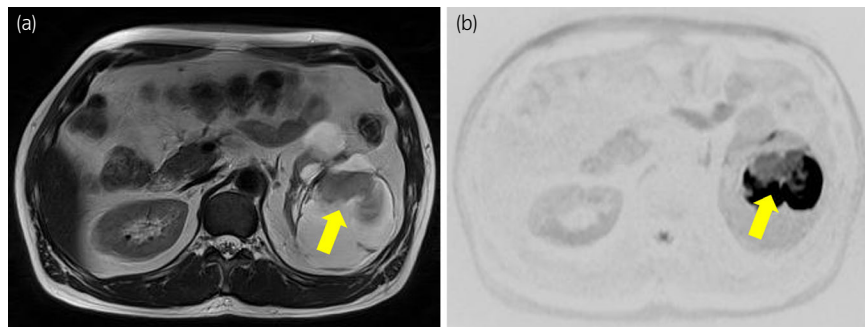
Pyelocalyceal diverticulum is a rare condition.<sup>1,2</sup> Furthermore, urothelial carcinoma developing in a pyelocalyceal diverticulum is extremely rare, and only 11 cases have been reported in Japan.<sup>3–13</sup> Here, we report a rare case of urothelial carcinoma originating from a pyelocalyceal diverticulum, which was difficult to distinguish from cystic renal cell carcinoma.

## Case presentation

A 51-year-old Japanese man developed gross hematuria. He visited a local hospital where he underwent abdominal computed tomography, which revealed many cysts with calcification inside the left kidney. He was then referred to our hospital for further examination. A blood test showed no abnormal findings. Urinary cytology yielded a pseudo-positive result (class 3). However, dynamic contrast-enhanced computed tomography revealed a mass, which showed enhancement in the early phase and appeared washed out in the late phase, in a cyst at the upper pole of the left kidney (Fig. 1). Magnetic resonance imaging revealed a tumor with an abnormal signal on a diffusion-weighted image (Fig. 2). Retrograde pyelography showed no wall irregularity at the left



**Fig. 1** Dynamic computed tomography images ((a) plain, (b) early phase, and (c) late phase). Polycystic lesions with calcification are found in the left kidney. The mass shadow (yellow arrowheads) in the cyst is stained in the early phase of contrast and shows mild washout in the late phase. Hematoma is observed as a high-density component besides the enhanced lesion. No cystic lesions are found in the right kidney.



**Fig. 2** Magnetic resonance imaging ((a) T2-weighted image and (b) diffusion-weighted image). An irregular mass shadow is observed inside the cyst (yellow arrow). Diffusion-weighted image shows a significant abnormal signal on the mass.

renal pelvis, and urinary cytology of samples from the left pelvis and urinary tract yielded negative results. He was diagnosed with left cystic renal cell carcinoma (cT1N0M0) and underwent retroperitoneal laparoscopic nephrectomy. The surgical specimen showed a cystic lesion filled with papillary formation (Fig. 3). Microstones and brownish liquid retention were also observed inside the cystic lesion. Pathological examination

revealed that the wall of the cystic lesion was covered with urothelial cells and high-grade urothelial carcinoma with renal parenchymal invasion. In immunohistochemical staining, GATA3, p63, and p40 were positive and PAX8 was negative. The definitive pathological diagnosis was urothelial carcinoma originating from the renal pyelocalyceal diverticulum, invasive urothelial carcinoma, high-grade (G3), and pT3. An additional residual ureterectomy and two courses of gemcitabine and cisplatin adjuvant chemotherapy were performed. Pathological examination showed no malignant findings of the residual ureter, and no recurrence was observed during the 12-month follow-up.



**Fig. 3** Surgical specimen. Neoplastic lesions inside the cyst (red circle).

## Discussion

A pyelocalyceal diverticulum is a cystic cavity connected to the renal calyx, and it is composed of urothelial cells. It is a relatively rare disease with a reported incidence of only 0.21–0.6% on retrograde pyelography. Urothelial carcinoma developing in the pyelocalyceal diverticulum, as in the present case, is a very rare disease, with only 11 cases reported in Japan (Table 1).<sup>3–13</sup> Of these 11 cases, four were diagnosed as cystic renal cell carcinoma on preoperative examination,

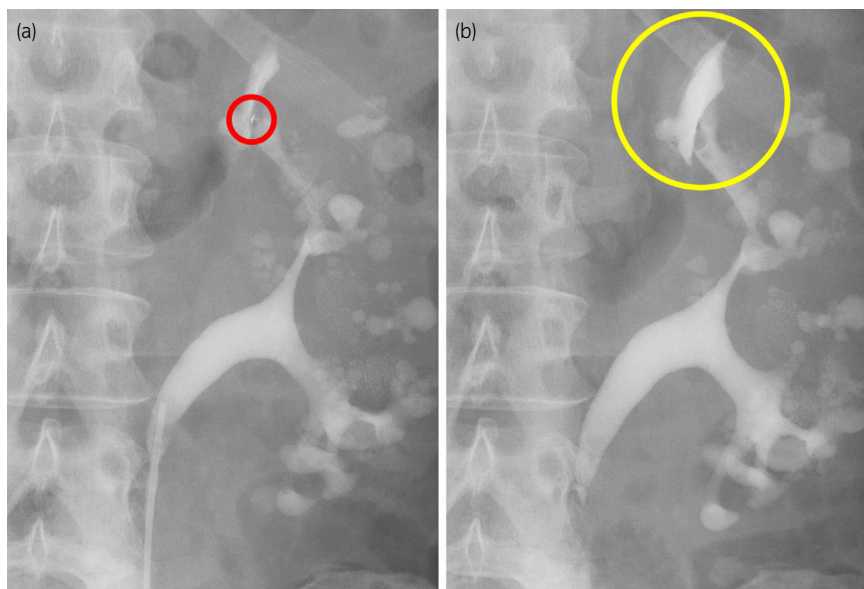
**Table 1** Cases of cancer in the pyelocalyceal diverticulum in Japan (previous cases and ours)

Case	Symptom	Urinary cytology	Connection with the urinary tract	Treatment	Histopathological type	Outcome (observation period)
56 year M	Gross hematuria	Unknown	Unknown	Nephrectomy	Urothelial carcinoma	Unknown
Unknown	Unknown	Unknown	+	Unknown	Urothelial carcinoma	Unknown
42 year M	Gross hematuria	-	+	Total nephroureterectomy	Urothelial carcinoma	No evidence of disease (4 months)
53 year F	Gross hematuria	Unknown	Unknown	Nephrectomy	Urothelial carcinoma	No evidence of disease (2 years)
58 year M	Microhematuria	-	+	Nephrectomy	Urothelial carcinoma	Unknown
61 year M	Gross hematuria	+	-	Total nephroureterectomy + Radiation therapy	Urothelial carcinoma	Recurrence (6 months)
63 year M	Gross hematuria	-	+	Total nephroureterectomy	Urothelial carcinoma	Unknown
66 year M	Gross hematuria	+	-	Total nephroureterectomy	Urothelial carcinoma	Recurrence (10 months)
78 year F	Gross hematuria	+	+	Total nephroureterectomy	Urothelial carcinoma	No evidence of disease (14 months)
70 year M	Lesion pointed out by CT	+	-	Total nephroureterectomy	Urothelial carcinoma	No evidence of disease (12 months)
64 year M	Microhematuria	-	-	Nephrectomy → Residual ureterectomy	Urothelial carcinoma	No evidence of disease (6 months)
51 year M (our case)	Gross hematuria	± (pseudo-positive)	+	Nephrectomy → Residual ureterectomy	Urothelial carcinoma	No evidence of disease (18 months)

and these cases were difficult to distinguish from those of cystic kidney cancer on preoperative examination, as in the present case.

However, a subsequent reanalysis of the preoperative retrograde pyelography findings revealed a connection between

the urinary tract and the pyelocalyceal diverticulum (Fig. 4). In all previous reports, patients preoperatively diagnosed with urothelial carcinoma of the pyelocalyceal diverticulum had positive urinary cytology results or findings of a connection with the urinary tract. These examinations may be useful for



**Fig. 4** Retrograde pyelography on the left side ((a) early phase and (b) late phase). The connection between the urinary tract and the diverticulum (red circle). Initially, we missed the connection and only cystic exclusion is considered to exist in the left renal pelvis (yellow circle).

diagnosis. There have been few reports of urothelial carcinoma of the pyelocaliceal diverticulum, and the characteristics of the disease are not clear. However, some cases have reported progress without recurrence after surgical treatment. We think that conventional treatment may be as effective as urothelial carcinoma.

Although not detected in the current case, the presence of stones is useful in differentiating the pyelocalyceal diverticulum from cysts. A pyelocalyceal diverticulum is often known to be associated with stones, but it is also sometimes associated with “milk of calcium.”<sup>10,12,14,15</sup> Milk of calcium is a fluid shadow of microstones composed of calcium, which could be detected as a calcified shadow like a half-moon along the horizontal plane on imaging performed in the standing or supine position. In this case, we could not obtain the findings of milk of calcium. Nevertheless, if the findings of milk of calcium are obtained, physicians should proceed with preoperative examinations considering the possibility of a pyelocalyceal diverticulum.

## Conclusions

We experienced a case of urothelial carcinoma originating from the pyelocalyceal diverticulum, which was difficult to distinguish from cystic renal cell carcinoma preoperatively. Although it is unlikely to be encountered this disease in daily practice, even if cystic renal cell carcinoma is diagnosed based on preoperative imaging findings, the retrograde pyelography findings should be reanalyzed to distinguish it from urothelial carcinoma in the pyelocalyceal diverticulum particularly when urinary cytology yields false-positive or positive results.

Moreover, if we obtain findings suggestive of stones such as milk of calcium, a pyelocalyceal diverticulum should be positively considered as a differential diagnosis.

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## Authors' contributions

IM drafted the manuscript. TM, TK, and HT were involved in the management of the patient. NK, YM, and RH analyzed the pathological specimens. YM performed the surgery and the clinical follow-up. IM, YM, and MS were responsible for the conception and interpretation of the data, and critical revision of the manuscript. All authors have read and approved the final manuscript.

## Conflict of interest

The authors declare no conflict of interest.

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

Not applicable.

## Informed consent

We have obtained informed consent from the patient to publish this case report and save the data in electronic records.

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

Not applicable.

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