


Case Report

Rare case of carcinoma in situ originated in right retrocaval ureter successfully managed with laparoscopic procedure

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Abbreviations & Acronyms

CIS = carcinoma in situ
CT = computed tomography
IVC = inferior vena cava
LRNU = laparoscopic radical nephroureterectomy
RCU = retrocaval ureter
UC = urothelial carcinoma

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Introduction: A retrocaval ureter is a rare congenital abnormality characterized by the persistence of the posterior subcardinal vein on the right, which causes the proximal ureter to deviate medially behind the inferior vena cava. The presence of retrocaval ureter is usually found because of the development of progressive hydronephrosis, but many cases are clinically silent. In addition, an urothelial malignancy associated with retrocaval ureter is very rare.

Case presentation: Herein we report a very rare case of a 57-year-old male with retrocaval ureter and carcinoma in situ diagnosed by ureteroscopy. In spite of strong adhesion in the area of the ureter behind the inferior vena cava, dissection was successfully managed by a laparoscopic procedure after cutting the ureter and separating it into two segments.

Conclusion: Early histopathological diagnosis and radical laparoscopic surgery based on the results of ureteroscopy may lead to a good outcome, even in retrocaval ureter cases with a carcinoma in situ.

Key words: carcinoma in situ, hydronephrosis, laparoscopic surgery, retrocaval ureter, ureteroscopy.

Keynote message

We report a case of CIS originating in the right RCU. Laparoscopic surgery based on the results of ureteroscopy may result in a good outcome.

Introduction

RCU is a rare congenital abnormality due to the impaired development of IVC.^{1–3} This abnormality manifests as a segment of compressed and obstructed ureter posterior to the IVC, therefore, progressively inducing hydronephrosis and renal dysfunction. However, urothelial malignancies in RCU are very rare, without any established surgical procedure described in the literature. Herein we describe a case of CIS associated with a right RCU that was successfully managed by radical laparoscopic surgery based on the results of ureteroscopy.

Case presentation

A 57-year-old, previously healthy, Asian male consulted a nearby clinic complaining of leg edema and difficulty in walking. CT showed right hydronephrosis and a ureter that traversed behind the IVC, resulting in compression and obstruction posterior to the IVC (Fig. 1a–c). Retrograde pyelography showed right hydronephrosis and a typical S-shaped deformity of the ureter; therefore, a ureteral stent was placed. Urine cytology of the right renal pelvis was suspicious positive, and the patient was referred to our hospital. On physical examination, the abdomen was soft and flat, and a slight pitting edema was observed in both lower limbs. Urine cytology was repeatedly suspicious positive therefore, ureteroscopy was performed, but the lesion was barely accessible just before the renal pelvis and ureteral junction, even using a flexible ureteroscope (Fig. 1d). Apparently, obvious abnormal findings were not found,

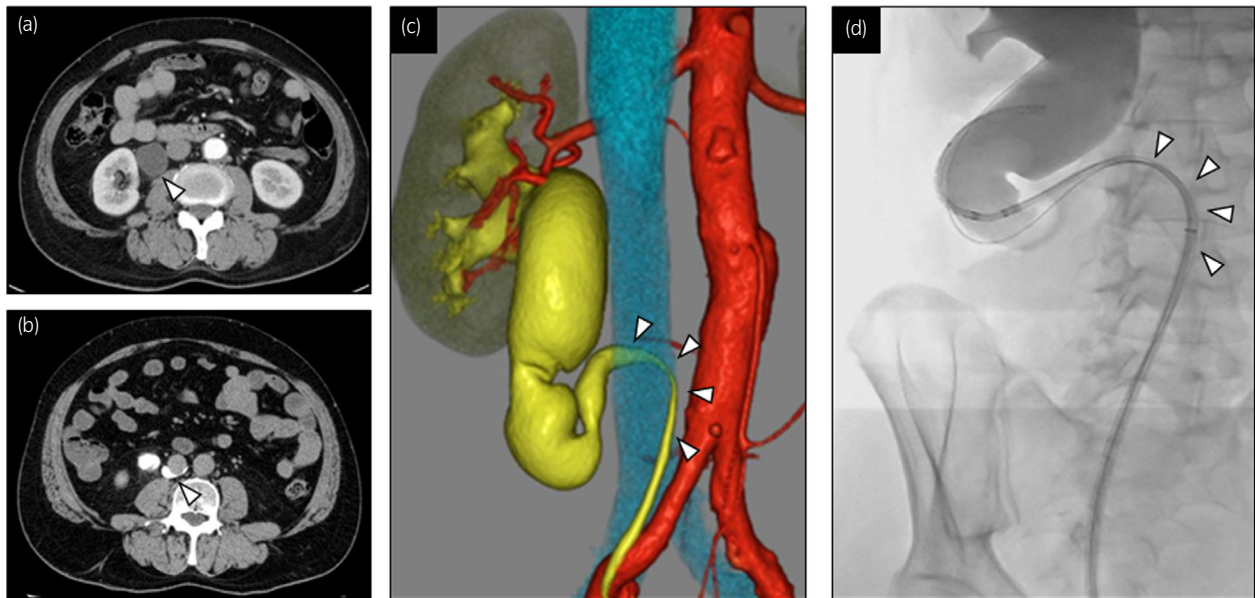


Fig. 1 (a–d) Abdominal enhanced CT early phase (a) and CT urography (b) (white arrowhead: ureter). Contrast-enhanced three-dimensional CT shows a ureter running behind the IVC (white arrowheads) (c). Fluoroscopy of the ureteroscopy (d).

therefore, slightly red lesions of the mucosa was biopsied. Pathological findings revealed tissue lined by atypical cells with hyperchromatic nuclei, abnormal mitosis, and irregular nuclei leading to a diagnosis of CIS (Fig. 2c). After informed consent, LRNU was performed. After a renal arteriovenous

amputation, detachment around the IVC was undertaken; the ureter was confirmed to be behind the IVC as shown in Figure 2a. The area between the ureter and IVC was highly adhesive. Therefore, the ureter was clipped by hem-o-lok clips and cut after sealing using Ligasure[®] (Medtronic,

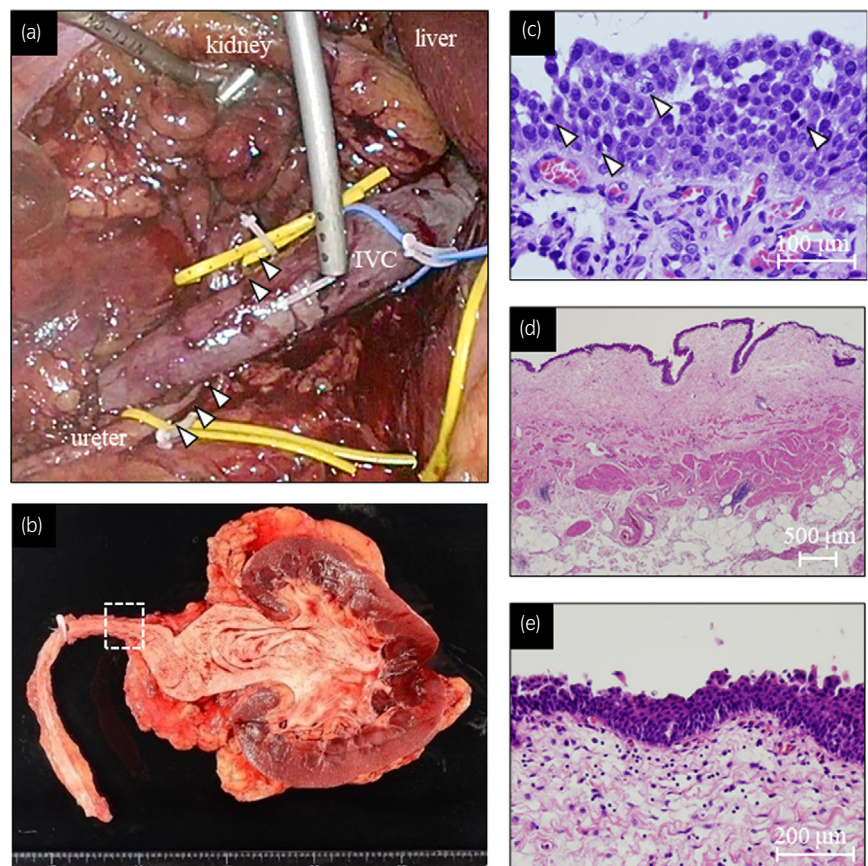


Fig. 2 (a) An intraoperative image of the ureter and IVC. The white arrowheads indicate the ureter. (b) Macroscopic finding of the resected specimen. CIS was recognized only in the enclosed part of dotted line. (c–e) Hematoxylin and eosin stains of the tumor specimen. The white arrowheads indicate the cells of karyokinesis.

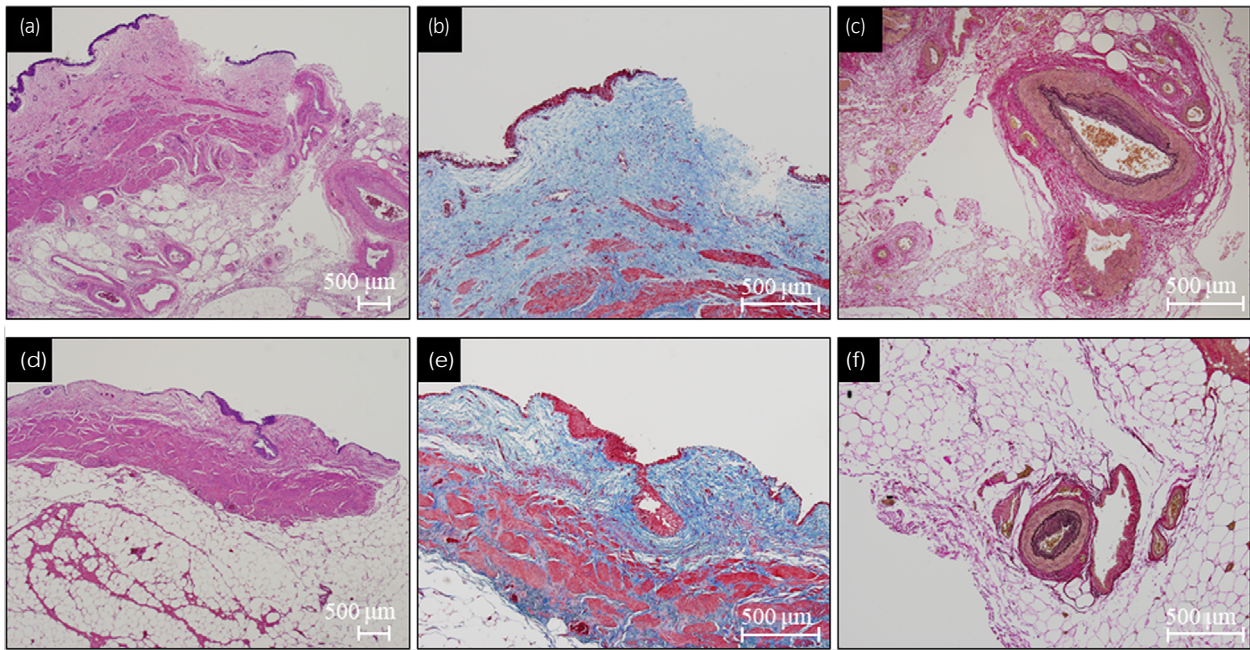


Fig. 3 (a–c) Normal ureter as a control. (d–e) Histopathological analysis of the hydroureters in this case. (a,d) Hematoxylin and eosin staining of each specimen. (b,e) Masson's trichrome staining of each specimen. (c,f) Elastica van Gieson staining of each specimen.

Dublin, Ireland); the specimen was separated into two segments. Finally, the lower ureter and bladder cuff were excised by a lower midline incision. Pathological findings revealed that urothelial cells exhibited full-thickness cytological and architectural disorder, a loss of cell polarity, and were crowded with hyperchromatic nuclei (Fig. 2b,d,e). The final diagnosis was CIS originating in the right RCU with no metastatic sites. The patient has shown no sign of recurrence and kidney dysfunction 6 months after the operation.

Discussion

RCU is rare congenital anomaly, with an autopsy incidence of about 1 per 1000 that is three times more common in males.^{1–3} Several variants in anatomic and topographic presentations of RCU have been described in the literature.⁴ The chief complaints of RCU patients are flank pain, hematuria, and repetitive urinary infections in their 30s or 40s. As typified by our patient, an urothelial malignancy in RCU is very rare, with about 20 cases described in the literature.^{5,6} In addition, reports describing a case with CIS in RCU are nonexistent. In this case, because of the results of urine cytology, ureteroscopy was performed, which contributed to the early diagnosis of CIS and surgical intervention. With this case in mind, we should rule out urothelial malignancies when a patient is considered having RCU. Based on accumulated data, including our case, the risk factors relating to the concomitant of UC in RCU need to be elucidated in future.

A literature review revealed that many surgical techniques have been used to deal with anatomical anomalies in RCU without a malignancy.⁷ Nowadays, laparoscopic procedures

have led to good outcomes with regard to the removal of symptoms or the prevention of unilateral kidney dysfunction. And, the decision on whether to resect or preserve the retrocaval segment of the ureter has been controversial. Simforoosh *et al.*⁸ reported six cases not involving the excision of the retrocaval segment. However, Zhang *et al.*³ suggested excision if an 8-Fr catheter could not pass through the segment smoothly. No established procedure exists for UC associated with RCU. In laparoscopic procedure for RCU cases, after releasing the kidney and upper ureter, the lower ureter may be able to be passed through the posterior IVC to remove the specimen en bloc. However, in this case, as there was no malignant finding in the lower ureter on ureteroscopy, the ureter was therefore clipped and sealed to prevent dissemination of cancer cells, and the specimen was separated into two segments. Though strong adhesion was present between the ureter and behind the IVC, the dissection was successfully managed in a laparoscopic procedure by this step.

With regard to understanding the mechanisms of adhesion between a hydroureter and RCU, accumulated histopathological data are lacking. Therefore, the hydroureters, in this case, were compared with normal ureters from another LRNU case (Fig. 3a–f). As a result, Masson's trichrome and Elastica van Gieson staining revealed that the ureter in this RCU case had a similar thickness to that of the submucosa (Fig. 3a,d), showed a disruption of muscular continuity (Fig. 3b,e), and had an enlarged arterial diameter (Fig. 3c,f). These results suggest that chronic inflammation existed over a long period of time and that it caused a wide range of fibrosis and vascular proliferation in the ureter around the IVC, resulting in strong adhesions. Nowadays, a few reports have described that robotic reconstruction can easily obtain better outcomes

compared with laparoscopic one in RCU.^{9,10} Therefore, in the future, by robotic-assist, it might be possible to perform LRNU in RCU patients without cutting ureter even in the situation of strong adhesion. However, further investigation of this rare disease is required.

With data from the few cases studied, an RCU patient whose urine cytology is suspicious positive should be suspected of having urothelial malignancies, including CIS. In RCU cases, an early histopathological diagnosis and LRNU based on ureteroscopy results may lead to a good outcome, even with a CIS.

Conclusion

We report here a case of CIS associated with a right RCU where early ureteroscopy contributed to the diagnosis and laparoscopic surgical intervention.

Consent

Written informed consent was obtained for publication of this article.

Conflict of interest

The authors declare no conflict of interest.

Editorial Comment

Editorial Comment to Rare case of carcinoma in situ originated in right retrocaval ureter successfully managed with laparoscopic procedure

Retrocaval ureter (RCU) is a rare congenital abnormality caused by impaired development of the inferior vena cava (IVC). The incidence of RCU has been estimated at around 0.1%.¹ Furthermore, RCU with a urothelial malignancy is very rare, with only 20 cases described in the literature.² Noda *et al.* reported carcinoma in situ (CIS) originating in the right RCU that was successfully managed with a laparoscopic procedure.³


In this case report, ureteroscopy was performed because urine cytology was suspiciously positive for urothelial carcinoma. Pathological findings showed CIS in the ureter. Therefore, the authors performed right laparoscopic radical nephroureterectomy (LRNU). As the authors mentioned, this finding indicates that clinicians should consider the possibility of urothelial malignancy when examining patients with RCU. The risk factors relating to urothelial carcinoma in RCU need to be elucidated.

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Few studies have reported on the treatment of RCU with a urothelial malignancy. It is highly controversial whether to resect or preserve the retrocaval segment of the ureter. In this case, the ureter was clipped by clips and cut with a sealing device to prevent dissemination of cancer cells. Recently, robotic-assisted LRNU has been installed in many centers.⁴ As Noda *et al.* noted, robotic assistance would allow the performance of LRNU in RCU patients without cutting the ureter.³

In this case report, a strong adhesion was found between the ureter and IVC. The authors analyzed the mechanism of this adhesion using special stains. Of note, this staining revealed that chronic inflammation may occur in the area of adhesion and cause a wide range of fibrosis and vascular proliferation. The interesting point of this study is that CIS was found in the area of the adhesion. Although the cause and relationship between CIS and the adhesion in this case are not clear, chronic inflammation may have affected the development of CIS in this patient.

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