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Acquired Cystic Disease-Associated Renal Cell Carcinoma Extending to the Renal Pelvis Mimicking Urothelial Carcinoma on Computed Tomography (CT): Two Case Reports

Authors' Contribution:
Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

ABCDEF 1 Hiromi Edo
ADEF 1 Yohsuke Suyama
AEF 1 Hiroaki Sugiura
EF 2 Kenichiro Ojima
ABEF 2 Keiichi Ito
BDEF 3 Kosuke Miyai
CDEF 3 Susumu Matsukuma
ABCDEF 1 Hiroshi Shinmoto

1 Department of Radiology, National Defense Medical College, Tokorozawa, Saitama, Japan
2 Department of Urology, National Defense Medical College, Tokorozawa, Saitama, Japan
3 Department of Pathology and Laboratory Medicine, National Defense Medical College, Tokorozawa, Saitama, Japan

Corresponding Author: Hiromi Edo, e-mail: hedo@ndmc.ac.jp
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Case series
Patients: Male, 66-year-old • Male, 67-year-old
Final Diagnosis: Acquired cystic disease-associated renal cell carcinoma
Symptoms: Hematuria
Medication: —
Clinical Procedure: —
Specialty: Oncology • Radiology

Objective: Mistake in diagnosis

Background: Acquired cystic disease-associated renal cell carcinoma (ACD-associated RCC), which was added to the 2016 World Health Organization classification, is the most common subtype of RCC in patients undergoing long-term dialysis. ACD-associated RCC is underrecognized and reports of computed tomography (CT) and magnetic resonance imaging findings for the lesion are sparse. Similar to urothelial carcinoma, ACD-associated RCC is poorly to slightly enhanced on dynamic CT. Here, we report 2 cases of ACD-associated RCC filling the renal pelvis and mimicking urothelial carcinoma.

Case Reports: We describe 2 cases of ACD-associated RCC filling the left renal pelvis in patients undergoing dialysis for more than 10 years. In both cases, the patient's chief complaint was hematuria, and a left renal pelvic mass with poor enhancement was seen on dynamic CT. In both cases, the preoperative diagnosis was urothelial carcinoma of the left renal pelvis. Total nephroureterectomy was performed, and the final diagnosis was ACD-associated RCC.

Conclusions: ACD-associated RCC is a common tumor in patients undergoing long-term dialysis. When ACD-associated RCC is located in the renal pelvis, the imaging findings are similar to those of urothelial carcinoma. Therefore, it is important for radiologists to include ACD-associated RCC in the differential diagnosis.

MeSH Keywords: Carcinoma, Renal Cell • Dialysis • Kidney Failure, Chronic • Tomography Scanners, X-Ray Computed

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Background

Previously, renal cell carcinoma (RCC) in patients undergoing long-term dialysis was diagnosed as papillary RCC or clear cell RCC, the same histological types of tumors as those that occur in patients who do not receive dialysis [1]. However, in 2006, Tickoo et al. [2] reported a specific histological type of RCC that frequently occurs in the kidneys of patients undergoing long-term dialysis. Accordingly, the 2016 revision of the World Health Organization (WHO) classification (henceforth the 2016 WHO classification) listed ACD-associated RCC as the most common and characteristic renal tumor in patients undergoing long-term dialysis [3].

Despite being the most common histological subtype of RCC in patients undergoing long-term dialysis [4], ACD-associated RCC is not widely known to clinicians such as radiologists and urologists. Because it is underrecognized, there are few reports on its features on CT and magnetic resonance imaging (MRI). ACD-associated RCC demonstrates poor to slight enhancement on dynamic CT [5,6], which is a pattern similar to that of urothelial carcinoma.

In general, RCC and urothelial carcinoma respond to different treatment methods. Patients with RCC typically undergo total nephrectomy, whereas those with urothelial carcinoma undergo total nephroureterectomy. For this reason, the preoperative diagnosis of renal tumors is important. Herein, we report 2 cases of ACD-associated RCC extending to the renal pelvis and mimicking urothelial carcinoma on CT.

Case Reports

Consent

Informed consent was obtained in writing from both patients.

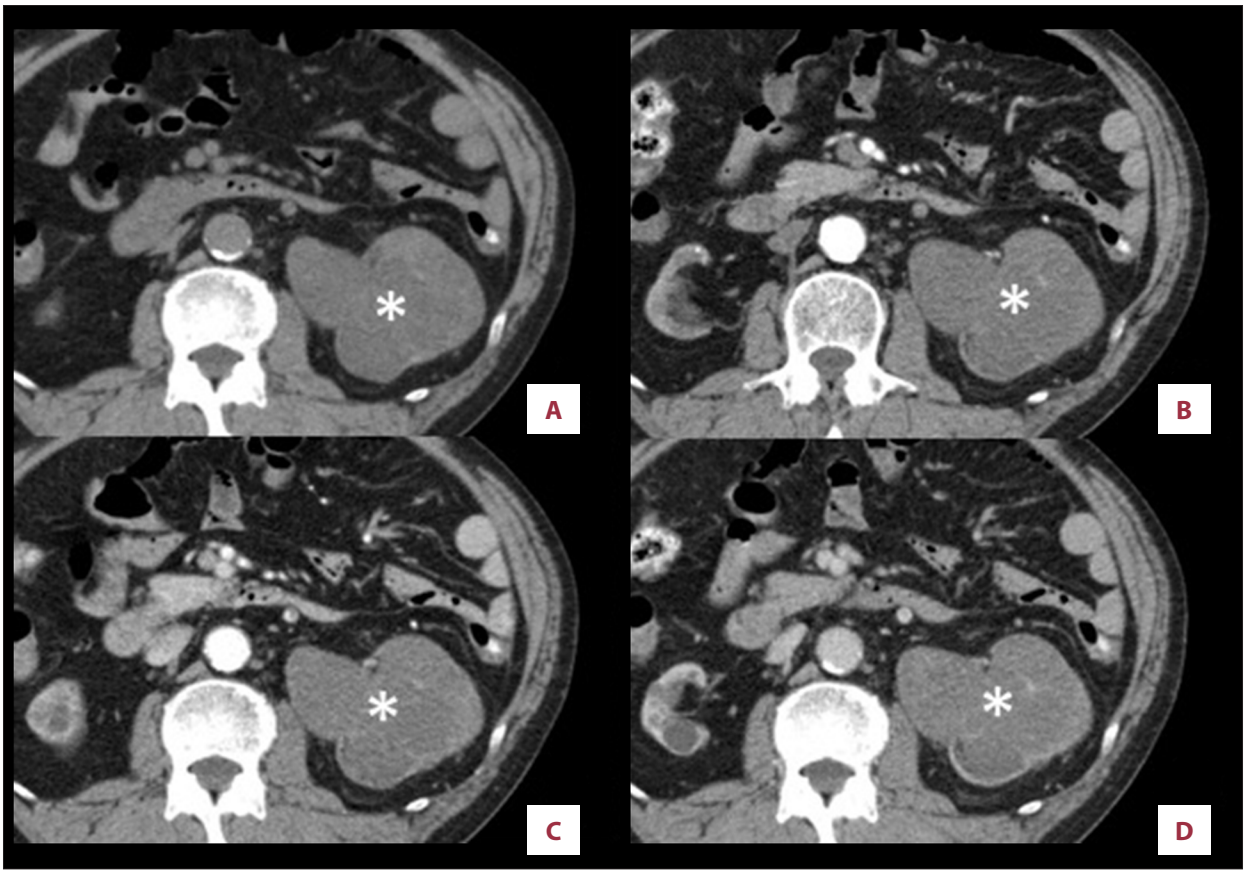

Figure 1. Computed tomography (CT) images of an acquired cystic disease-associated renal cell carcinoma in the 66-year-old man in Case Report 1. The left renal parenchyma is markedly thin and the dilated pelvis and calyx are filled with masses (*) without obvious enhancement in either phase (**A–D**). For the region of interest of a part of the lesion, the CT value of each phase was measured. (**A**) Plane phase (30 Hounsfield units [HUs]). (**B**) Corticomedullary phase (41 HUs). (**C**) Nephrographic phase (55 HUs). (**D**) Early excretory phase (42 HUs). There was no obvious calcification in the mass.

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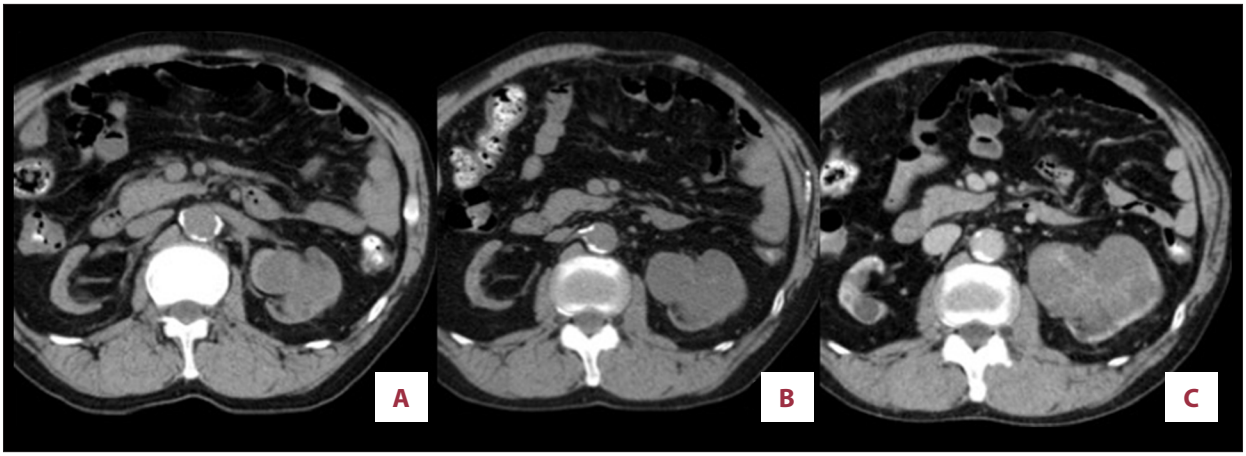

Figure 2. Comparison with computed tomography (CT) images of acquired cystic disease-associated renal cell carcinoma in the 66-year-old man in Case Report 1. (**A**) CT image 2½ years before total nephroureterectomy. (**B**) CT image 1½ years before total nephroureterectomy. (**C**) CT image 3 months before total nephroureterectomy. The mass filling the left renal pelvis and calyx slowly increased in size over 2½ years (**A–C**).

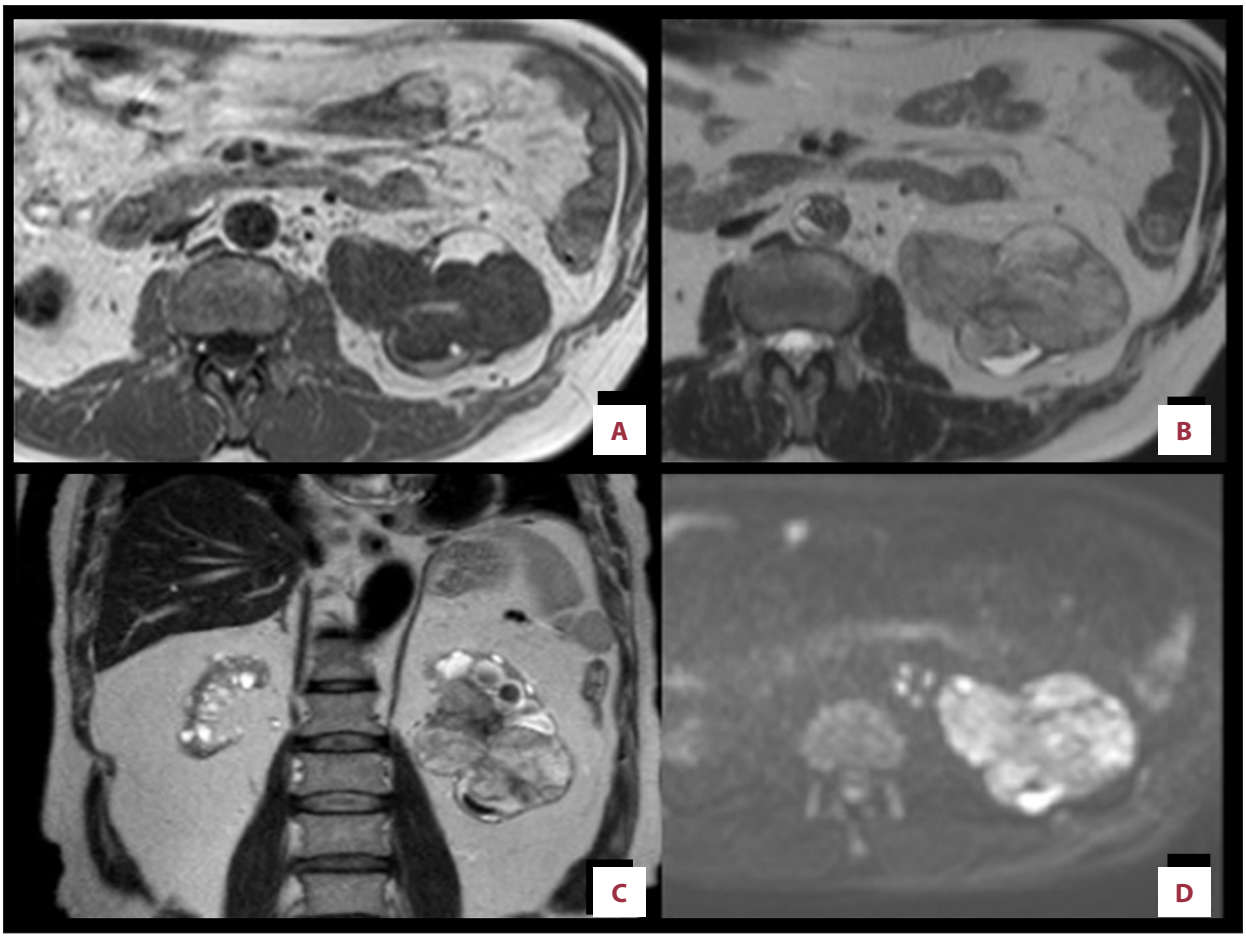

Figure 3. Magnetic resonance imaging of an acquired cystic disease-associated renal cell carcinoma in the 66-year-old man in Case Report 1. (**A**) An axial T1-weighted image shows a left pelvic tumor with heterogeneous low to high signal intensity, which suggests hemorrhage in the tumor. (**B, C**) Axial (**B**) and coronal (**C**) T2-weighted images show a left pelvic tumor with heterogeneous low to slightly high signal intensity. (**D**) A diffusion-weighted image shows a left pelvic tumor with heterogeneous high signal intensity.

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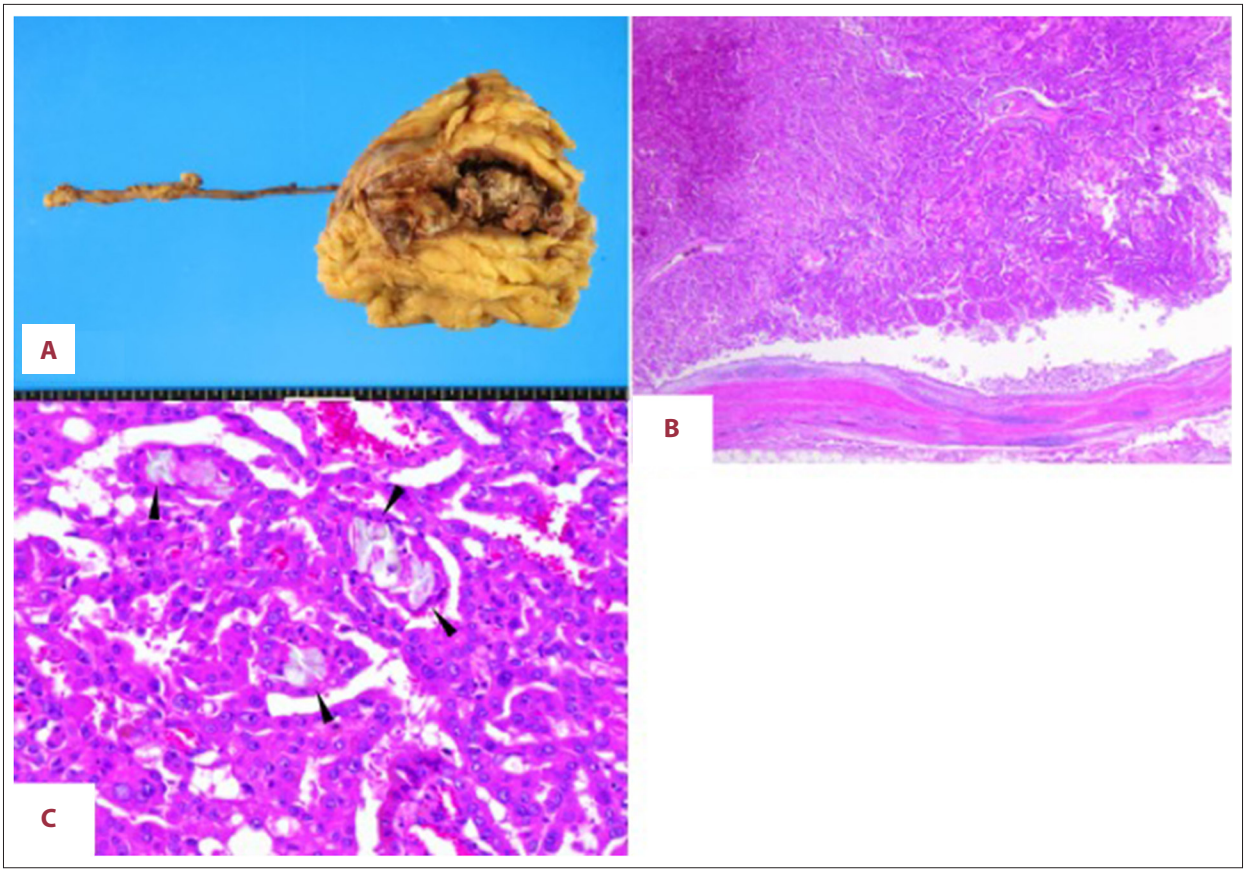

Figure 4. Findings from histopathological analysis of an acquired cystic disease-associated renal cell carcinoma in the 66-year-old man in Case Report 1. (**A**) The gross image shows a yellow solid tumor with bleeding that fills the left renal pelvis. (**B**) The tumor exhibits a wide spectrum of histological characteristics, including microcystic, papillary, tubular, and cribriform architecture. (**C**) Histologically, the tumor has eosinophilic and prominent nucleoli. The arrowhead indicates calcium oxalate crystal deposition.

Case 1

A 66-year-old man had been diagnosed with a left renal mass 2 years ago and was being followed up in another hospital. Because the mass had increased in size, he was referred to our hospital. He had a 10-year history of hemodialysis due to nephritis and had persistent hematuria for 4 years. No malignant cells were detected in urine cytology.

Dynamic CT was performed (Figure 1) and the images demonstrated a left renal pelvic mass without obvious enhancement in either phase. It was larger than on the CT done 2 1/2 years before (Figure 2). On non-enhanced MRI, the mass had low and high signal intensity on T1-weighted imaging (Figure 3A) and heterogeneous mild high signal intensity on T2-weighted imaging (Figure 3B, 3C). Diffusion-weighted imaging (DWI) demonstrated heterogeneous high signal intensity (Figure 3D). There was no apparent invasion surrounding the renal pelvis or extra-ureteral tissue.

Clinically, urothelial carcinoma of the left pelvis was suspected, and total nephroureterectomy was performed. A yellowish solid tumor involved the left renal pelvis and histology revealed a mixture of papillary, acinar, and/or microcystic proliferation (Figure 4A, 4B). The tumor cells consisted of polygonal or rounded cells with chiefly eosinophilic cytoplasm and swollen nuclei containing prominent nucleoli (Figure 4C). Calcium oxalate crystals were scattered throughout the tumor (Figure 4C). The pathological diagnosis was ACD-associated RCC (pT2a). The patient did not require any additional treatment and follow-up CT scanning was performed every 3 months. No recurrence or metastases were observed at a follow-up visit 1 year after surgery.

Case 2

A 67-year-old man had a 15-year history of hemodialysis and a 1-year history of hematuria. CT was performed to determine the cause of the hematuria and a left renal mass was found. No malignant cells were detected on urine cytology. Dynamic

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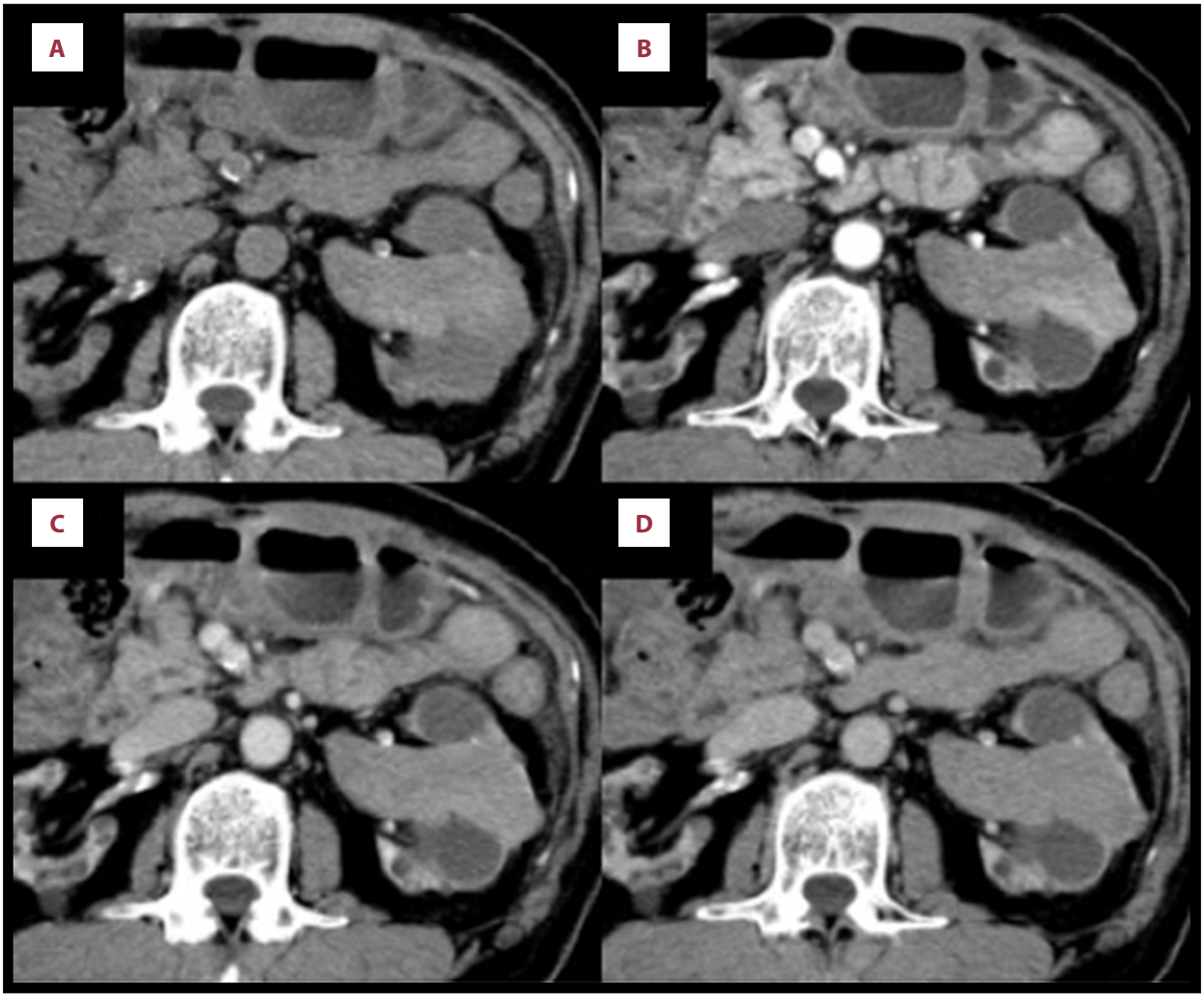

Figure 5. Computed tomography (CT) images of an acquired cystic disease-associated renal cell carcinoma in the 67-year-old man in Case Report 2. A left renal mass is seen filling the left renal pelvis with poor to slight enhancement on dynamic CT (**A–D**). For the region of interest of a part of the lesion, the CT value of each phase was measured. (**A**) Plane phase (57 Hounsfield units [HUs]). (**B**) Corticomedullary phase (92 HUs) (**C**) Nephrographic phase (77 HUs). (**D**) Early excretory phase (70 HUs).

CT was performed and a hypovascular mass that filled the left renal pelvis was noted (Figure 5). Visually, a part of the tumor showed mild enhancement, but most of it had no noticeable enhancement. There was no apparent invasion surrounding the renal pelvis or extra-ureteral tissue. On non-enhanced MRI, the mass had low signal intensity on T1-weighted imaging (Figure 6A) and heterogeneous low to slightly high signal intensity on T2-weighted imaging (Figure 6B, 6C). DWI demonstrated heterogeneous low signal intensity (Figure 6D).

Clinically, urothelial carcinoma of the renal pelvis was suspected, and a total nephroureterectomy was performed. Macroscopically, a yellow solid tumor filled the left renal pelvis (Figure 7A). Histologically, the tumor cells had a mixture of papillary, acinar, and/or microcystic characteristics, with abundant eosinophilic cytoplasm and prominent nucleoli (Figure 7B). The final diagnosis was ACD-associated RCC (pT1b) This patient did not require any additional treatment and follow-up imaging was performed least once a year. No recurrence or metastases were observed at a follow-up visit 10 years after the surgery.

Discussion

ACD-associated RCC, which was added to the 2016 WHO classification [3], is the most common subtype of RCC in patients undergoing long-term dialysis [7]. Many renal tumors that occur in such patients were previously diagnosed as papillary RCCs or clear cell RCCs but are now diagnosed as ACD-associated RCCs based on the 2016 WHO classification. However, there is poor recognition of this subtype of RCC by radiologists, and few reports exist of imaging findings for ACD-associated RCC.

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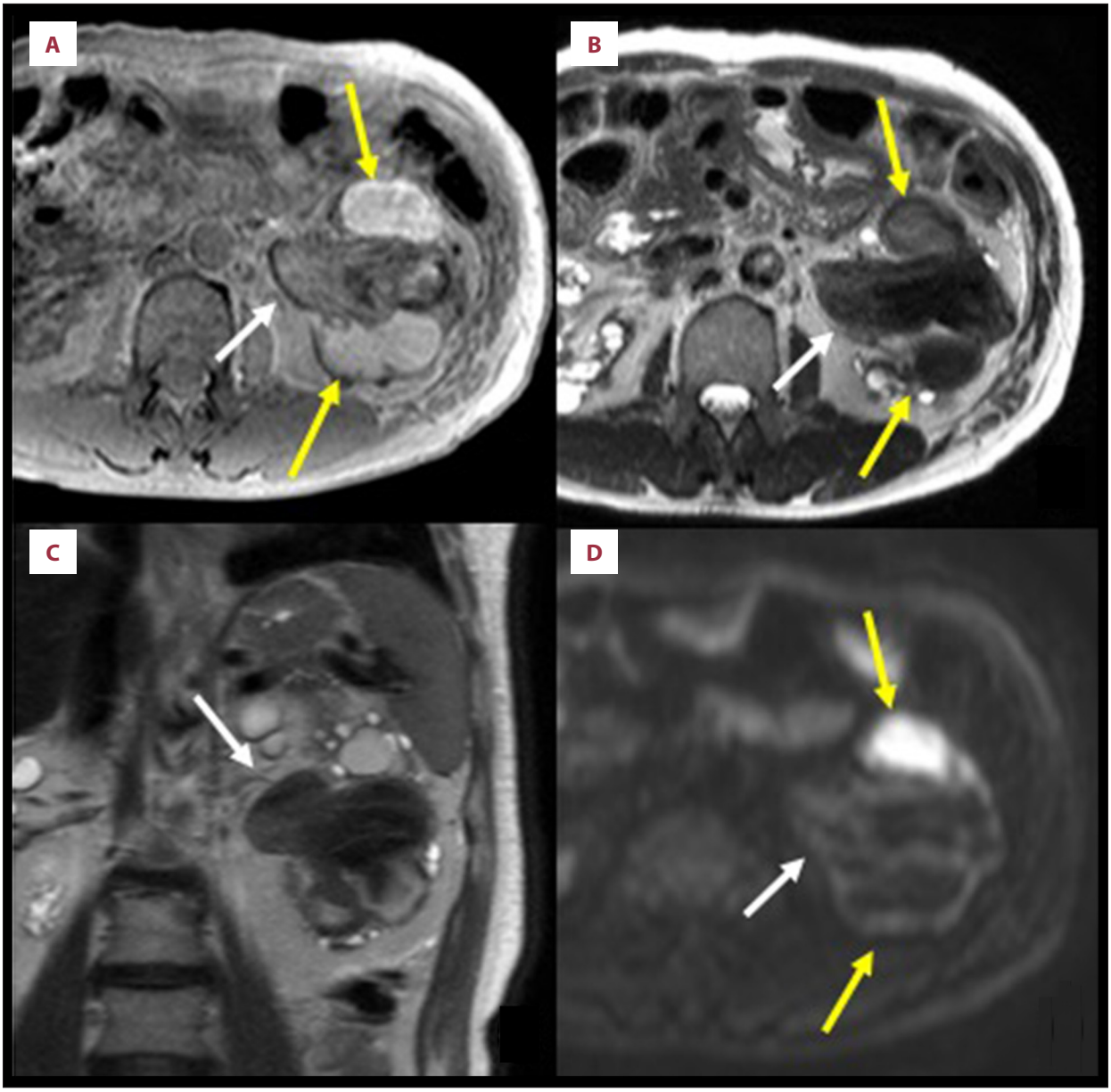

Figure 6. Magnetic resonance imaging of an acquired cystic disease-associated renal cell carcinoma in the 67-year-old man in Case Report 2. (**A**) An axial T1-weighted image shows a left pelvic tumor with heterogeneous low signal intensity (white arrow). (**B** ,**C**) Axial (**B**) and coronal (**C**) T2-weighted images show a left pelvic tumor with heterogeneous low to slightly high signal intensity, as indicated by the white arrow. (**D**) A diffusion-weighted image (DWI) demonstrates a left pelvic tumor with renal parenchyma-equivalent signal intensity (white arrow). The yellow arrow indicates acquired renal cysts, which have different signals on T1- and T2-weighted images and DWIs, depending on their contents, including hemorrhage and abundant protein.

To the best of our knowledge, there is no previously published literature on ACD-associated RCC extending to the renal pelvis and mimicking urothelial carcinoma.

ACD-associated RCC arises from a cyst in the kidney acquired as a result of long-term dialysis [2,8,9]. Therefore, it commonly presents as an intracystic growth and/or as a solid mass that surrounds cysts [5,6,10]. In the present cases, it was presumed that a carcinoma derived from the acquired cyst extended to the renal pelvis and was recognized as a renal pelvic tumor, but no clear findings were observed on imaging because the renal parenchyma was thin. Focusing on the contrast pattern of dynamic CT, our patients had hypovascular masses, which is typical for ACD-associated RCC [5,6].

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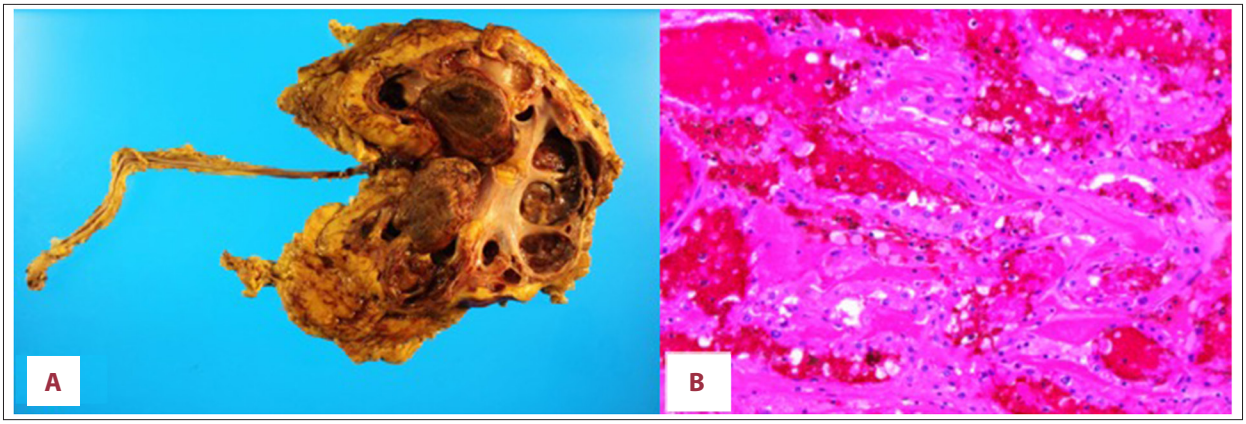

Figure 7. Findings from histopathological analysis of an acquired cystic disease-associated renal cell carcinoma in the 67-year-old man in Case Report 2. (**A**) The gross image shows a yellow solid tumor filling the left renal pelvis. (**B**) Microscopically, the tumor cells have abundant eosinophilic cytoplasm and prominent nucleoli.

The other differential diagnoses for hypovascular renal masses are usually considered to be urothelial carcinoma, papillary RCC, and mucinous tubular and spindle cell carcinoma [11,12]. Because the enhanced pattern on dynamic CT is similar to that for urothelial carcinoma, ACD-associated RCC extending to the renal pelvis is difficult to distinguish from urothelial carcinoma of the renal pelvis. There is no comprehensive report of MRI images from ACD-associated RCC, and one previous report mentioned MRI findings that were nonspecific, as in our patients [13]. However, the iso signal intensity of the tumor compared with the renal parenchyma on the DWI seen in Case 2 is uncommon for urothelial carcinoma, and therefore, could suggest a diagnosis of RCC.

Focusing on patients undergoing long-term dialysis, it has been reported that the incidence of malignant tumors in these individuals is approximately 1.4 times higher than in the general population [14,15]. Moreover, the incidence of RCC in patients on dialysis is approximately 4 to 5 times higher than in the general population [14, 15]. Kondo et al. [7] reported that the incidences of histological subtypes of RCC in patients undergoing dialysis for more or less than 10 years were 37.8% to 50.7% *vs.* 7.6% for ACD-associated RCC, 21.6% to 38.1% *vs.* 76.1% for clear cell RCC, and 9.8% to 20.7% *vs.* 8.5% for papillary RCC, respectively. Because tumor frequencies vary in patients who have a dialysis history of more than 10 years, it is important to consider the possibility of ACD-associated RCC for hypovascular tumors in the renal pelvis in these individuals.

To summarize, ACD-associated RCC is a common tumor in patients undergoing long-term dialysis. Although it is underrecognized, it is by no means rare and awareness of the tumor's imaging findings may help prevent misdiagnosis. When ACD-associated RCC is located in the renal pelvis, the imaging findings are similar to those of urothelial carcinoma. Accurate diagnosis is of paramount importance because surgery tailored to the nature of the mass is associated with better patient outcomes.

Conclusions

ACD-associated RCC is a renal tumor commonly seen in patients undergoing long-term dialysis. If it extends to the renal pelvis, it can be mistaken for ureteral carcinoma because the imaging findings, including the contrast pattern on dynamic CT, are similar. Therefore, it is important for radiologists to include ACD-associated RCC in their differential diagnosis.

Acknowledgements

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Conflicts of interest

None.

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