Primary Adenocarcinoma of the Renal Pelvis: a case report and review of the literature

CHING HSUEH¹ PEI-RU WU² CHIA-BANG CHEN¹ SHANG-YUN HO¹ KWOWHEI LEE¹ CHIH-WEI LEE¹

Department of Medical Imaging¹, Department of Pathology², Changhua Christian Hospital, Changhua, Taiwan

ABSTRACT

Neoplasms of the renal pelvis account for approximately 7-8% of all renal malignancies, and most such neoplasms are urothelial in origin. Among these malignancies, the most common type is urothelial carcinoma, followed by squamous cell carcinoma. Primary adenocarcinoma of the renal pelvis is an extremely rare type, with only a few case reports in the literature. It may occur as a result of metaplasia of the renal pelvic transitional epithelium into glandular epithelium due to longstanding obstruction and infection and chronic irritation by urolithiasis, which then undergoes a malignant transformation. Herein, we report a case and the image findings of primary adenocarcinoma of the renal pelvis in a 50-year-old woman with history of renal calculi and end-stage renal disease.

CASE REPORT

A 51-year-old female experienced abdominal pain, nausea, and vomiting for 2 days and then visited our emergency department. She had histories of urolithiasis, end-stage renal disease under continuous ambulatory peritoneal dialysis (CAPD), hypertension, and gastric ulcer.

On physical examination, the patient experienced left flank knocking pain. A catheter for CAPD was placed. Otherwise, there was no fever. Laboratory investigations revealed elevated leucocyte count (26,200/μL), with neutrophilic predominance (94.1%). High serum creatinine level (5.69 mg/dL) indicated non-functioning kidneys. There was no obvious malignant cell to be detected in the patient’s voided urine. The serum levels of tumor markers (CA125, CEA, and CA19-9) were not measured before operation.

KUB radiography (Fig. 1) showed an enlarged left renal shadow with multiple curvilinear calcifications. Additionally, right renal calculi were suspected. Ultrasonography of the left kidney (Fig. 2) revealed marked dilatation of the pelvicalyceal system with presence of protruding mass lesion and several calculi. Computed tomography (CT) of the abdomen (Fig. 3) showed a large left kidney, measuring 21.1x15.9x15.7 cm³, with prominent calcifications.

Correspondence Author to: Chih-Wei Lee
Department of Medical Imaging, Changhua Christian Hospital, Changhua, Taiwan
No. 135, Nan-Xiao Street, Changhua 500, Taiwan
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dilatation of the collecting system without obvious residual normal renal parenchyma. The collecting system revealed extensive marginal calcifications. There were multiple enhancing papillary projections on the surface of the renal pelvis and calyces. Mild perinephric fat infiltration was also noted. In addition, some small (short-axis diameter <1 cm) lymph nodes appeared in the paraaortic and interaortocaval regions. No evidence of metastatic lesion nor other malignancy was demonstrated in this CT study.

Under the impression of renal pelvicalyceal tumors, the patient underwent radical nephroureterectomy with bladder cuff excision. On gross pathological examination (Fig. 4), the resected kidney was measured 21.0x14.5x5.0 cm³ and weighed 690 grams. The renal capsule was difficult to strip focally. The renal parenchyma showed prominent atrophy. The pelvicalyceal system displayed marked dilatation and was filled with gelatinous material. Multiple brownish and elastic-to-soft papillary nodules were found on the surface of the renal pelvis and calyces. Microscopically, sections of the papillary tumors demonstrated a picture of adenocarcinoma of intestinal type with villoglandular, complex papillary, or focal cribriform patterns. Immunohistochemically, the tumor cells showed CK7 (+, diffuse) and CK20 (+, diffuse). Also, the mucosa elsewhere still showed intestinal metaplasia with flattened or micropapillary pattern. In summary, the pathologic diagnosis was renal adenocarcinoma of intestinal type and the tumor had invaded into the muscularis propria (T2).

After surgery, the serum levels of tumor markers were measured, and they showed higher levels (CA125: 44.4 U/ml, CEA: 10.2 ng/ml, and CA19-9: 154.7 U/ml). On the follow-up CT image obtained two years after surgery, there was no recurrent tumor in this patient.

DISCUSSION

The most common malignant tumor of the renal pelvis is urothelial carcinoma, accounting for 90% of renal pelvic tumors, followed by squamous cell carcinoma (SCC). Primary adenocarcinoma of the renal pelvis is extremely rare, accounting for less than 1% of renal pelvic tumors [2]. There are several cases of these tumors that have been reported, mainly from India and Asia. It may suggest an environmental or dietary etiology [5]. There was no gender preponderance found in these cases [6]. Most of these malignancies occurred in adults, but pediatric cases have also been reported [7].

In the published literature, renal pelvic adenocarcinomas can be further subdivided into tubulovillous, mucinous, and papillary nonintestinal categories histopathologically. The first two categories are more common and morphologically similar to intestinal adenocarcinomas, accounting for 93% of cases [8]. In our patient, the histologic type of the neoplasm was intestinal type.

In the patients with primary adenocarcinoma of the renal pelvis, renal calculi, hydronephrosis, pyelonephritis, and nonfunction are the common associated clinical findings, and these features are also frequently associated with SCC of the renal pelvis [3]. Primary adenocarcinoma
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Figure 3. Unenhanced axial a, contrast-enhanced axial b, and reconstructed coronal c, d. CT images depicted a large left kidney with prominent dilatation of the collecting system (star) without obvious residual normal renal parenchyma. The collecting system revealed extensive marginal calcifications (thin arrows). There were multiple enhancing papillary projections (thick arrows) on the surface of the renal pelvis and calyces.

of the renal pelvis leading to pseudomyxoma peritonei has been only reported in a single case [9]. Even though several theories about the etiology of primary adenocarcinoma of the renal pelvis have been proposed, the exact etiology remains unknown [10]. So far, the most acceptable theory is that the transitional epithelium of the renal pelvis first undergoes glandular metaplasia and then malignant transformation [6]. It may be attributed to longstanding chronic inflammation, sometimes secondary to renal calculi. However, some authors have postulated that formation of the calculi might be the result of oversecretion of glycoproteins by the tumor and binding of that with cations such as sodium, calcium, and magnesium, forming large calculi. Thus, calculi may be the result rather than the cause of the neoplasm [8, 11]. Although it is difficult to correlate renal calculi and malignancy, our patient had past history of renal calculi, hydronephrosis, and end-stage renal disease for a long time. Whether the renal calculi are the cause of malignancy or they are just formed by the tumor, it cannot be well recognized in the imaging study of our patient.
As to the diagnosis, a preoperative diagnosis of primary renal pelvic adenocarcinoma is difficult to make. Most of these tumors could not be diagnosed clinically. Final diagnosis is made from surgically resected specimen [4]. If there is any suspicion of renal pelvic adenocarcinoma clinically, it is first essential to exclude the possibility of metastatic neoplasm before labeling the neoplasm as primary in origin. However, there was no any malignant history in the patient.

Carbohydrate antigen 125 (CA125), carcinoembryonic antigen (CEA), and carbohydrate antigen 19-9 (CA19-9) are frequently used as the biomarkers of the gynecologic or gastrointestinal tract tumors. Elevation of these biomarkers had been reported in some patients with renal pelvic adenocarcinomas [12], but they were not elevated in most of patients with urothelial tumors. However, the serum CA125, CEA, and CA19-9 levels of our patient were not measured before operation, but they did show higher levels a week after surgery.

Regarding the preoperative radiologic investigation, most of the renal pelvic adenocarcinomas could not be diagnosed on the imaging studies in published reports. Renal calculi and hydronephrosis are the common findings of ultrasonography, but the tumor entity is often difficult to recognize. Even on CT, seldom has a specific image feature suggesting this tumor. In the CT of our patient, most tumors could be recognized on CT, but they showed exophytic papillary growth pattern. Even though most tumors could be recognized on CT, it was difficult to differentiate from urothelial

Figure 4. a. Gross specimen revealed a markedly atrophic kidney with multifocal papillary nodules (arrows) on the dilated pelvis mucosa. b. The cut surface showed exophytic papillary growth pattern of these tumors (arrows). c. Photomicrography (original magnification, x20; hematoxylin-eosin stain) showed enteric-type dysplastic tumor cells arranged in complex papillary architecture (arrows). Adenocarcinoma of intestinal type was confirmed.
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Carcinoma and thus preoperative diagnosis is difficult. Generally speaking, urothelial carcinoma is commonly encountered in the urinary bladder. Five percent of urothelial carcinomas arise from the ureter or the renal pelvis or calices. On CT imaging, early renal urothelial carcinoma is typically manifested as a small filling defect, a mass lesion, or circumferential thickening in urothelial wall during the excretory phase. Advanced tumor can extend into the renal parenchyma in an infiltrating pattern that destroys the normal architecture [13, 14]. Primary SCC of the renal pelvis is rare because it represents only 0.5% to 0.8% of malignant renal tumors. Radiologically, renal pelvic SCC may appear as a large, necrotic, and ulcerated mass with gross invasion of the renal parenchyma or as a renal pelvic infiltrative lesion without evidence of a distinct mass. The radiological findings of SCC are variable. Most renal pelvic SCCs present at an advanced stage [15, 16].

As to treatment, most patients reported in the literature were subjected to radical nephroureterectomy, including our patient. In the absence of significant diagnostic radiographical features in favor of this diagnosis, peroperative diagnosis with frozen section may be helpful to confirm the diagnosis and planning of an appropriate surgery [2]. There is no established regimen for chemotherapy for these tumors. Onishi et al reported that four cycles of TJ (paclitaxel/carboplatin) regimen resulted in complete disappearance of the primary tumor in the ureter and paraaortic lymph node swelling without severe side effect [12]. From the published data, the prognosis appears to be poor, with about 1/2 of these patients dying within two years after surgery. However, neither recurrent tumor nor other malignancy was found in our patient on the follow-up CT image obtained two years after surgery.

CONCLUSION

Primary adenocarcinomas of the renal pelvis are extremely rare neoplasms, with only a few case reports in the literature. A preoperative diagnosis of the neoplasm is usually difficult to make. There is no a specific imaging feature about this tumor to be shown in the published reports. In this article, we have shown the imaging feature of the tumor on CT. Although these tumors are extremely rare, the possibility of primary renal pelvic adenocarcinoma should be kept in mind for patients who have longstanding renal calculi and hydronephrosis with an irregular renal pelvic wall.

REFERENCES