



Primary Extrarenal Type 2 Papillary Renal Cell Carcinoma: A Case Report

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Type 2 papillary renal cell carcinoma (PRCC) is a malignant tumor originated from renal tubular epithelial cells. Here we present a rare case of extrarenal type 2 PRCC which is located at the right lateral side of the retroperitoneal inferior vena cava. No suspicious lesions were detected in the bilateral kidneys. The diagnosis was confirmed by histopathological examination after complete excision of the tumor. No recurrence was observed in 5 months of follow-up after surgery. UROLOGY 123: e1–e3, 2019. © 2018 Elsevier Inc.

Papillary renal cell carcinoma (PRCC) which was first reported by Mancilla and Stanley in 1976¹ is a malignant tumor originated from renal tubular epithelial cells. PRCC is classified into type 1 and type 2 according to histological morphology.² The prognosis of type 2 PRCC is worse than that of type 1.^{3,4} Nearly half of the patients with type 2 PRCC are diagnosed due to healthy examination. A small number of patients are diagnosed due to symptoms such as hematuria, backache, mass, and metastasis. Radical nephrectomy is one of the main strategies for localized type 2 PRCC, although recurrence occurs in about 40% of patients undergoing surgical resection.⁵ In advanced stages, immunotherapy and cytotoxic chemotherapy have been widely used in the past, although with limited efficacy.⁶ Targeted drug therapy is a therapeutic option for delaying postoperative recurrence and metastasis.⁷ Ectopic renal cell carcinoma is extremely rare without any report. This case report presents a case of type 2 PRCC located in the right lateral side of inferior vena cava without any suspicious lesions in both kidneys. To the best of our knowledge, this is the first case of extrarenal type 2 PRCC.

CASE REPORT

A 63-year-old male was hospitalized due to a right retroperitoneal mass detected in a health examination without any other complaint and specific past history. After hospitalization, regular physical examination and routine tests showed no obvious abnormality. Abdominal computer tomography (CT) suggested that round irregular soft tissue

masses were observed on the right lateral side of the retroperitoneal inferior vena cava with uneven internal density and clear boundary. The maximum diameter of the mass was 5.11 cm. Contrast-enhanced CT scan showed slight enhancement of the mass and local compression of the inferior vena cava (Fig. 1A). CT angiography showed the mass was supplied by a small branch of the right renal artery (Fig. 1B-D). A 2.4 cm cyst was detected in the right kidney while no abnormality was detected in the left kidney. Tests of 24 hours urine catecholamine, serum cortisol, plasma adrenocorticotropic hormone, plasma renin angiotensin, and serum aldosterone showed no abnormality. Chest CT did not show obvious abnormality.

Robot assisted laparoscopic right retroperitoneal mass resection was performed. Intraoperative exploration revealed that the mass was located on the right lateral side of the inferior vena cava and was about 6.0 cm in diameter. The mass was supplied by a small branch of the renal artery without any adhesion to the right kidney. After ligation of the supply artery, the mass was removed completely. The specimens were examined by histopathology and immunohistochemistry. The specimen is an 8 cm × 7 cm × 5 cm nonplastic tissue with a capsule in parts of the surface. Under the microscope, the tumor tissue showed papillary arrangement, and the nucleolus could be seen (Fig. 2). Immunohistochemical results showed that the tumor tissue expressed CD10, CAIX, E-CAD, CK7, P504S, TFE3, TFEB, SDHB, CK20, Ki 67 (about 3%), PAX-8 and PAX-2, and was negative for Vim, CD117, Ksp-Cadherin (Fig. 3). According to the histological morphology and immunohistochemical results, the mass was pathologically diagnosed with type 2 PRCC, with World Health Organization (WHO)/International Society of Urological Pathology (ISUP) classification of level 2-3/4. No tumor thrombus was observed in the vessel and no cancer tissue was involved in the nerve. The margin was negative.

Five months following surgery, no recurrence was detected based on the abdominal CT scanning.

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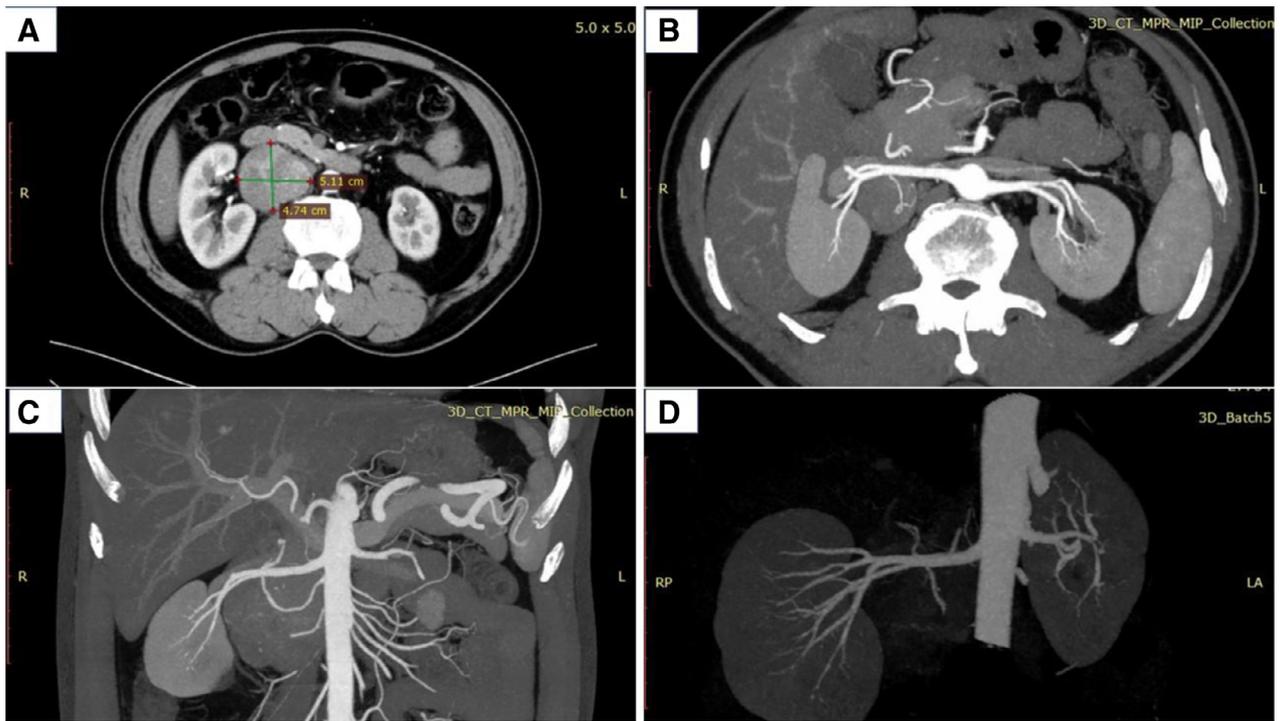


Figure 1. (A) Abdominal CT suggested that round irregular soft tissue masses were observed on the right lateral side of the retroperitoneal inferior vena cava with uneven internal density and clear boundary. The maximum diameter of the mass was 5.11 cm. Contrast-enhanced CT scan showed slight enhancement of the mass. (B-D) CT renal artery imaging showed the right retroperitoneal mass was supplied by a small branch of the right renal artery.

DISCUSSION

PRCC is the second most common subtype of renal cell carcinoma (RCC), accounting 10%-15% of RCC.⁸ Pseudo-stratification of tumor cells, high nuclear pleomorphism, and rich cytoplasm of eosinophilic cells are the histological features of type 2 PRCC while a cubic tumor cell monolayer without pseudo-stratification and a large number of foam macrophages are the histological features of type 1 PRCC.⁹ Primary rarely detected in extrarenal tissue. Till now, only 2 articles have reported primary extrarenal clear cell carcinoma.^{10,11} Extrarenal RCC refers to the occurrence of RCC in locations other than the normal native kidneys, likely arising from mesonephric remnants.¹⁰ To the best of our knowledge, this is the first case

of type 2 PRCC located in extrarenal tissue. Type 2 PRCC is a tumor with deficient blood supply. The enhancement degree of type 2 PRCC in contrast-enhanced CT is usually mild, which is significantly lower than that of RCC.¹² We originally considered the possible diagnosis of this case as an ectopic pheochromocytoma according to the contrast-enhanced CT. However, 24 hours urine catecholamine and adrenocortical hormone levels were within normal range, and the patients had no history of hypertension, dizziness, amaurosis, palpitation, hidrosis, and other discomfort complaints, providing the evidence for excluding the possibility of ectopic pheochromocytoma. No primary lesions were detected in both kidneys and other organs according to the results of

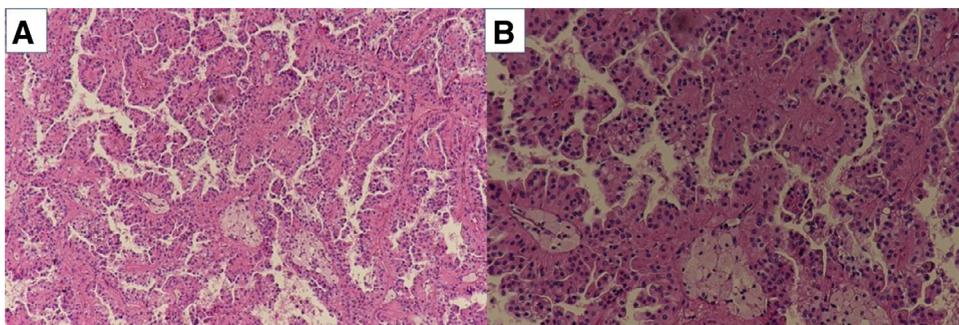


Figure 2. Microscopy demonstrates cytomorphologic features consistent with conventional type 2 PRCC (A) H and E, $\times 100$ (B) H and E, $\times 200$.

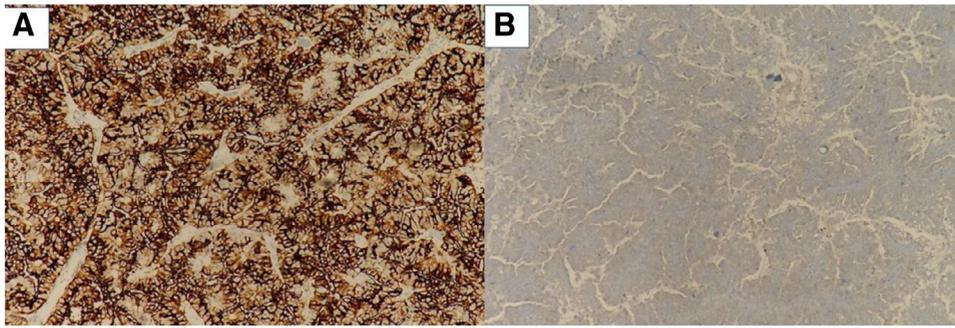


Figure 3. The tumor showed immunoreactivity for (A) CD10, $\times 100$ and (B) PAX2, $\times 100$.

chest and abdomen CT scanning, excluding the possibility of metastasis. Neither imaging nor intraoperative findings showed that the tumor had a separate ureter. So the possibility of an extra kidney could be excluded.¹³ According to the postoperative pathological results, extrarenal type 2 PRCC was diagnosis due to the high expression of CD10 and PAX-2.¹⁴

So far, radical nephrectomy is the first choice of treatment of localized type 2 PRCC. Since this is the first reported case of extrarenal type 2 PRCC, there is no recommendation for surgical resection range. In this case, we only complete resection of the tumor, without resection of surrounding tissue or lymph nodes. Long-term follow-up is definitely needed. At present there is no recognized effective treatment to prevent its recurrence and metastasis. In this case, no evidence of recurrence was found after a 5-month follow-up.

CONCLUSION

This case report presents a rare case of extrarenal type 2 PRCC which is located at the right lateral side of the right retroperitoneal inferior vena cava. No suspicious lesions were found in both kidneys. Surgical resection was the treatment strategy and long-term follow-up are recommended.

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