

# Bilateral Renal Cell Carcinoma in a Patient with Autosomal Dominant Polycystic Kidney Disease

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Renal cell carcinoma (RCC) in autosomal dominant polycystic kidney disease (ADPKD) is very rare. Only 11 cases of bilateral RCC in ADPKD have been reported since 1954. Herein, we present a 58-year-old male who received laparoscopic bilateral radical nephrectomy for bilateral RCC with different cell variants in ADPKD and end-stage renal disease under regular hemodialysis. [*J Chin Med Assoc* 2007;70(9):403–405]

**Key Words:** autosomal dominant polycystic kidney disease, conventional renal cell carcinoma, papillary renal cell carcinoma

## Introduction

Renal cell carcinoma (RCC) in autosomal dominant polycystic kidney disease (ADPKD) is very rare. Only a total of about 50 cases of ADPKD-associated RCCs have been reported since 1934,<sup>1,2</sup> and only 11 cases of bilateral RCC in ADPKD have been reported since 1954.<sup>2</sup> Herein, we present a case of bilateral RCC with different cell variants in ADPKD.

## Case Report

This 58-year-old man had ADPKD with end-stage renal disease under regular hemodialysis since September 1988. Bilateral multiple renal tumors were noted incidentally on ultrasonography in April 2004 during annual follow-up. Computed tomography (CT) of the abdomen was performed, and autosomal dominant polycystic disease with bilateral renal malignancy was suspected (Figure 1).

Under the impression of ADPKD with bilateral renal tumors, the patient received laparoscopic bilateral radical nephrectomy in May 2004. Two tumor masses, 8 × 6 × 6 cm and 2 × 1 × 1 cm, were noted in the right kidney, and the pathologic reports showed papillary RCC, eosinophilic variant, pT2 and pT1 (Figure 2A). Two other tumor masses, 5 × 4 × 4 cm

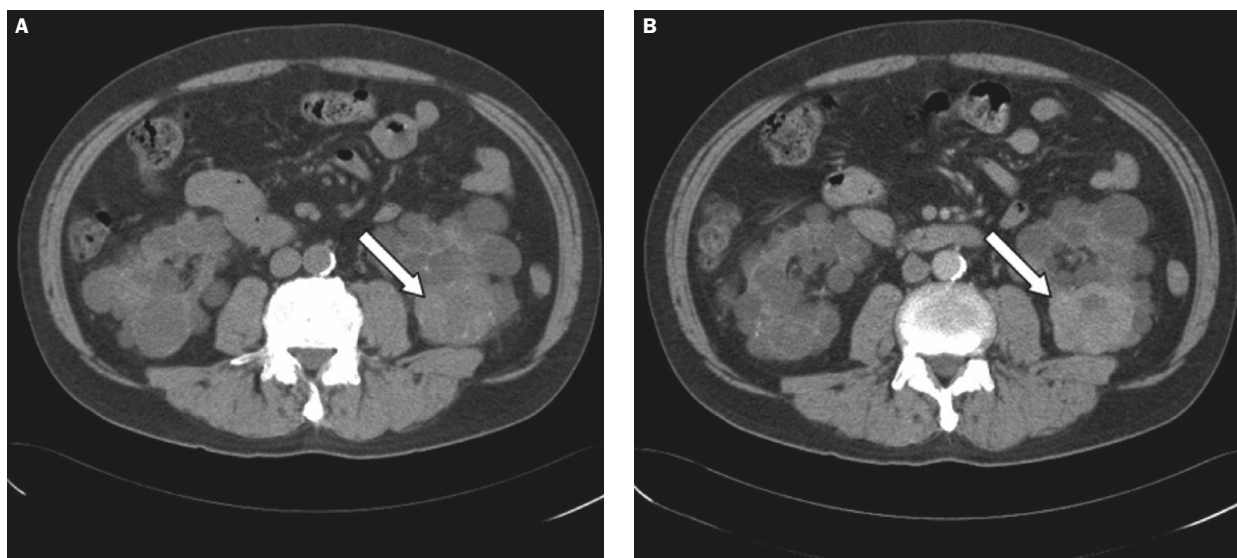
and 4 × 4 × 3.5 cm, were noted in the left kidney, and the pathologic reports showed conventional RCC, pT1 with cystic necrosis in 1 mass and pT1 without necrosis in the other (Figure 2B). The non-neoplastic parts of both kidneys were confirmed as adult polycystic kidney disease by the pathologist.

Postoperative recovery was smooth, and the patient was discharged and then regularly followed-up at the outpatient department (OPD). There has been no evidence of local recurrence or distant metastasis till now.

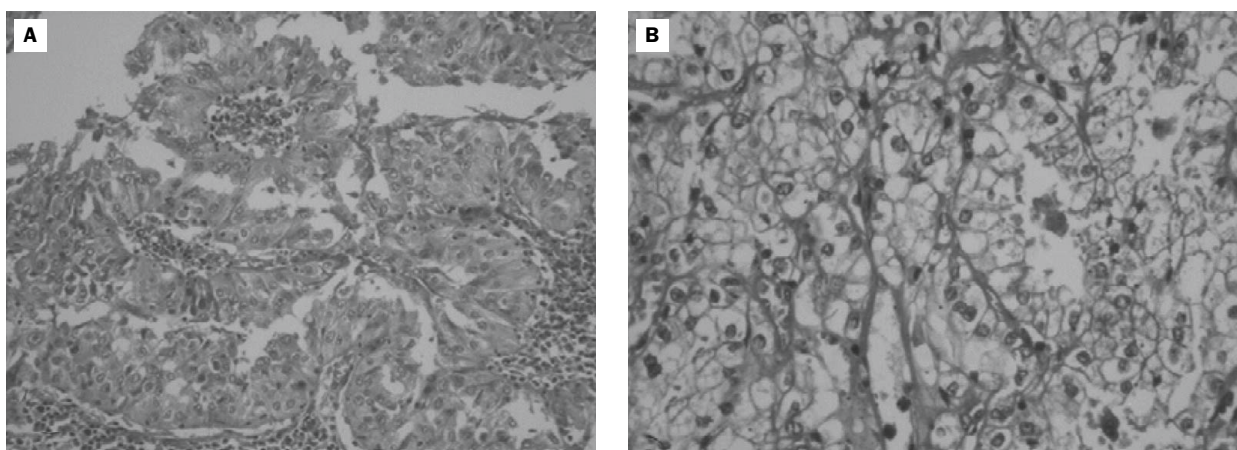
## Discussion

Since 1934, there have only been about 50 cases of RCC in ADPKD,<sup>1,2</sup> and since 1954, only 11 cases of bilateral RCC in ADPKD.<sup>2</sup> The prevalence of the ADPKD gene is 1:1,000 and the age-adjusted incidence of RCC is 5–6/100,000/year in the general population. The chance of both conditions occurring in the same individual is not impossible but is rare.<sup>3</sup> In this case, there were 2 types of cell variants in bilateral polycystic kidneys with papillary RCC, eosinophilic variant and conventional RCC. Though the current data revealed that the incidence of ADPKD-associated RCC is not higher than that of RCC in the general population,<sup>4</sup> some findings of ADPKD-associated RCCs are different from those of RCC in the general

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**Figure 1.** Abdominal computed tomography shows left lower pole renal mass (arrow): (A) pre-contrast, attenuation coefficient is 36.6 HU; (B) post-contrast enhancement, attenuation coefficient is 50.3 HU.



**Figure 2.** Pathologic pictures of renal cell carcinoma (RCC) of the bilateral kidney: (A) papillary RCC of the right kidney (400 $\times$ ); (B) conventional RCC of the left kidney (400 $\times$ ).

population. For example, the age of presentation in ADPKD-associated RCCs is younger than that in the general population (45 years *vs.* 61 years), fever is a more common presenting symptom of RCC in ADPKD (32%) than in the general population (7%), and RCC in ADPKD is more often concurrently bilateral (12% *vs.* 1–5%), multicentric (28% *vs.* 6%), and sarcomatoid in type (33% *vs.* 1–5%) than in the general population.<sup>4</sup>

The different types of kidney cancer have different histologic characteristics and different clinical courses, and they are caused by different genes.<sup>5</sup> The gene for the inherited form of clear RCC associated with the von Hippel-Lindau (VHL) gene has been identified. This gene was found to be a tumor suppressor gene.<sup>5</sup>

The gene of hereditary papillary renal carcinoma was identified to be the proto-oncogene *c-Met*. It was found to be located at chromosome 7q31.1-34 in a 27-cM interval between the 2 anonymous markers D75496 and D751837.<sup>5</sup> Understanding the genetic basis of the renal tumor provides potentially better methods of diagnosis and new forms of therapy. Although the *PKD1* and *PKD2* genes are associated with ADPKD,<sup>6</sup> the associated chromosome or gene changes between RCC and ADPKD are unknown.<sup>1</sup>

The preoperative diagnosis of RCC in the context of ADPKD may be difficult, because the tumor may be masked by the complex cystic background superimposed by bleeding, degenerated blood clot, proteinaceous debris, and infection.<sup>4,7</sup> Accordingly, the

follow-up of ADPKD patients should be very careful. Recently, Lang and Davis<sup>8</sup> reported the diagnostic strategy of RCC in ADPKD by CT. Neoplasms between or in cysts have attenuation coefficients of +30 to +40 Hounsfield Units (HU) in the pre-enhancement phase. In the arterial corticomedullary phase of CT, there is a sharp increase in the attenuation coefficient to +70 to +150 HU. In the parenchymal phase, the attenuation coefficient progressively decreases to +45 to +65 HU. In RCC, however, the attenuation coefficient further decreases to +35 to +50 HU (washout phenomenon). In this case, the pre-enhanced attenuation coefficient, 35.9 HU and 36.6 HU in right and left kidneys, respectively, and the post-enhanced (parenchymal) attenuation coefficient, 47.6 HU and 50.3 HU in right and left kidneys, respectively, were compatible with the report of Lang and Davis. Because it is difficult to find small tumor masses in the cystic background, a high index of suspicion is recommended when we follow ADPKD patients.<sup>2</sup>

The treatment of choice in patients with ADPKD and bilateral renal tumors is the traditional open transabdominal bilateral nephrectomy, which carries significant morbidity.<sup>9</sup> The bilateral hand-assisted radical nephrectomy in a single setting for these patients was proved to be a safe and effective alternative procedure with less morbidity.<sup>10</sup> The prognosis of patients with ADPKD and RCC depends on the stage of the disease

and is not different from that of RCC patients in the general population.

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