

同時發生在自體顯性多囊腎病人的兩側腎細胞癌—病歷報告及文獻回顧

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BILATERAL RENAL CELL CARCINOMA IN AUTOSOMAL DOMINANT POLYCYSTIC KIDNEY DISEASE - A CASE REPORT AND LITERATURE REVIEW

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Introduction: Renal cell carcinoma (RCC) in autosomal dominant polycystic kidney disease (ADPKD) is a very rare disease. Since 1994 ADPKD-associated RCCs were reported for a total of about 30 cases. Herein, we will present a rare case of bilateral renal cell carcinoma with different cell variants in autosomal dominant polycystic kidney disease.

Case present: This 58-year-old male was a case of end-stage renal disease under regular hemodialysis since September 1988. Bilateral multiple renal tumors were noted incidentally by ultrasonography in April 2004 during annual follow-up. Then he was referred to our Nephrology out-patient department (OPD) for further survey and management. The computer tomography was performed and autosomal dominant polycystic disease with bilateral renal malignancy was suspected. Under the impression of ADPKD with bilateral renal tumor, he received laparoscopic bilateral radical nephrectomy in May 2004. Two tumor masses, 8x6x6 cm and 2x1x1 cm, were noted over right kidney and the pathologic reports showed papillary renal cell carcinoma, eosinophilic variant, pT2 and pT1. Two tumor masses, 5x4x4 cm and 4x4x3.5 cm, were noted over left kidney and the pathologic reports showed clear cell conventional renal cell carcinoma, pT1 with cystic necrosis in one mass and pT1 without necrosis in the other. The postoperative recovery was smooth and the patient was discharged and regularly followed at OPD.

Discussion: Since 1954 there were 11 cases with bilateral renal cell carcinoma in ADPKD. In our case there were 2 kinds of different cell variant in bilateral polycystic kidneys. The current data revealed that the incidence of ADPKD-associated RCC is similar to that of RCC in the general population. However, ADPKD-associated RCCs were characterized by a younger patient age, no sex preference, frequent simultaneous or sequential bilaterality, frequent multifocality, and a preference for sarcomatoid type. The preoperative diagnosis of RCC in the context of ADPKD may be difficult, because tumor may be masked by the complex cystic background superimposed by bleeding, degenerated blood clot, proteinaceous debris, and infection. So the follow-up of ADPKD patient should be very careful due to above reasons. When we visit a patient with ADPKD, the possibility of RCC in bilateral kidney simultaneously or sequentially should keep in mind.