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VHL综合征伴发双侧肾癌个案分析

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Case Analysis of Von Hippel－Lindau Syndrome Complicated by Bilateral Renal Cancer

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摘要

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全文: PDF (1424 KB) HTML (1 KB) 输出: BibTeX | EndNote (RIS) 背景资料

摘要 Von Hippel-Lindau（VHL）综合征为临床十分罕见的家族性常染色体显性遗传性肿瘤综合征，表现为多发、多器官的良、恶性肿瘤征候群， 包括中枢神经系统血管母细胞瘤、内脏肿瘤和囊肿等。发病率约1/3.6万，平均发病年龄为26.3～30.9岁。预后不良，平均生存年龄<49岁，其主要死亡原因是中枢神经系统血管母细胞瘤破裂出血、肾细胞癌和嗜铬细胞引起的恶性高血压。现介绍1例经浙江湖州师范学院附属第一医院泌尿外科收治的中枢神经系统血管母细胞瘤伴发双侧肾癌，附睾、肝、胰腺多发囊肿病例。该患者经左侧保留肾单位的肾癌切除术，索尼替尼分子靶向治疗后效果较佳，随访30个月，生存良好。

关键词： VHL综合症 泌尿系统 中枢神经系统血管母细胞瘤 肾肿瘤

Abstract: Von Hippel－Lindau disease is a rare, autosomal dominant neoplastic syndrome characterized by multiple benign and malignant tumors that affect multiple organs, including the central nervous system (CNS) hemangioblastomas, visceral tumor, and cysts. The average age of onset of Von Hippel－Lindau disease is between 26.3 years old and 30.9 years old, with an incidence rate of 1:36 000. This disease has a poor prognosis, and the average life expectancy is below 49 years old. The main causes of death is usually rupture and bleeding of CNS hemangioblastomas, renal cell carcinoma, and malignant hypertension caused by pheochromocytoma. We present a case of Von Hippel－Lindau disease with CNS hemangioblastoma, complicated by bilateral renal cancer, and multiple cysts in the epididymis, liver, and pancreas in the First Affiliated Hospital of Huzhou Teachers College. The patient underwent nephron - sparing surgery of the left kidney and he was given sunitinib malate as molecule－targeted therapy. Patient followed-up consistently for 30 months, and the patient survived and is living well.

Key words: Von Hippel－Lindau disease Urinary system Central nervous system hemangioblastoma Renal tumor

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