Internal Medicine
The Japanese Society of Internal Medicine
doi: 10.2169/internalmedicine.8644-21
Intern Med 61: 2079-2080, 2022
http://internmed.jp

【 PICTURES IN CLINICAL MEDICINE 】

Numerous Pancreatic Cysts Associated with von Hippel-Lindau Disease

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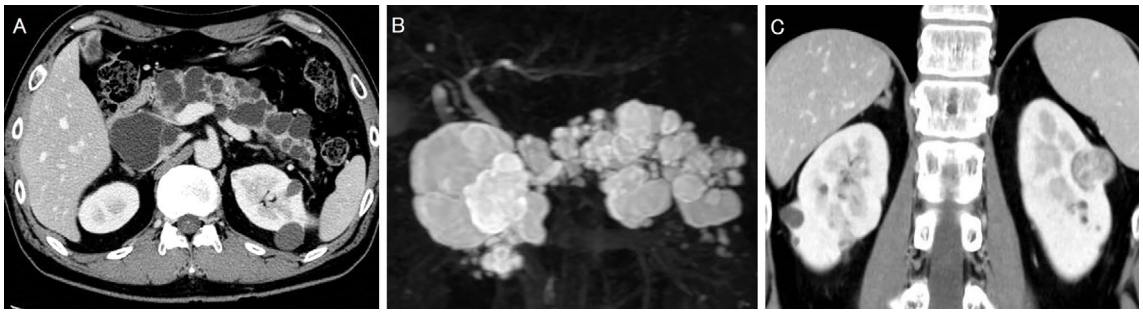
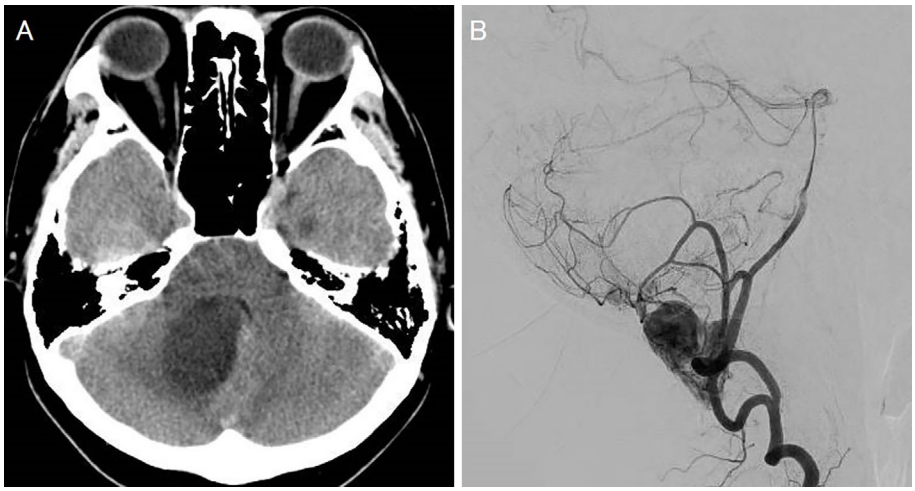
Key words: von Hippel-Lindau disease, serous cystic neoplasm, pancreatic cyst

(Intern Med 61: 2079-2080, 2022)
(DOI: 10.2169/internalmedicine.8644-21)



Picture 1.



Picture 2.

A 32-year-old man visited an outpatient clinic with complaints of abdominal discomfort. His mother has a history of renal cell carcinoma. Abdominal enhanced computed tomography (CT) and magnetic resonance cholangiopancreatography revealed numerous cysts throughout the pancreas. A 21-mm left kidney tumor was also detected by enhanced CT, raising suspicion of von Hippel-Lindau disease (VHL) (Picture 1). Brain CT showed a 48-mm tumor in the right cerebellum, and angiography demonstrated a hypervascular lesion. Craniotomy was performed, and the patient was diagnosed with hemangioblastoma (Picture 2). In addition, partial nephrectomy was performed for the left kidney tu-

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Received: September 8, 2021; Accepted: October 21, 2021; Advance Publication by J-STAGE: December 4, 2021
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mor, which was pathologically diagnosed as clear cell carcinoma.

VHL is an autosomal dominant hereditary disease that occurs in about 1 family per 1 million people (1). It can present with cerebrospinal hemangioblastomas (60-80%), retinal hemangiomas (40-70%), renal cell carcinomas (25-50%), adrenal pheochromocytomas (10-20%), pancreatic cysts (both serous cystic neoplasms and simple cysts; 17-61%), and pancreatic neuroendocrine tumors (8-17%) (1, 2). When multiple cystic lesions develop throughout the pancreas, VHL should be considered.

The authors state that they have no Conflict of Interest (COI).

References

1. Lonser RR, Glenn GM, Walther MC, et al. von Hippel-Lindau disease. Lancet **36**: 2059-2067, 2003.

2. Neumann HP, Dinkel E, Brambs H, et al. Pancreatic lesions in the von Hippel-Lindau syndrome. Gastroenterology **101**: 465-471, 1991.