

von Hippel - Lindau : 1

von Hippel - Lindau

von Hippel - Lindau 1

von Hippel - Lindau(VHL) (autosomal dominant) (Fig. 6).

35,000 - 40,000 1
(1, 2). 가

VHL

VHL

3

가

가

(1, 2, 4 - 6). VHL

35,000 - 40,000 1

(1, 2). VHL

40 가
(computed tomography, CT)

(computed 가

(Fig. 1).

, VHL

(1 - 6).

(magnetic resonance imaging, MRI)

VHL

, VHL

가

(Fig. 2),

가

,

(

,

,

가

MRI

,

, 2

2 cm

가

T1 -

(Fig. (1, 2).

VHL

3).

가

2.3 cm
2 mm

VHL

가

(Fig. 4).

VHL

가

(Fig. 5),

가

(1, 2).

CT

10

MRI CT

1

2001 5 29

2001 9 24

가 (1).

VHL
가
가
가
MRI가 가 (1, 2).
VHL
VHL 50 - 70%
가
VHL 70% 가

VHL
CT 1
(renal parenchymal sparing surgery)
(1 - 5).
가
CT MRI
2 cm 2 mm
VHL 가

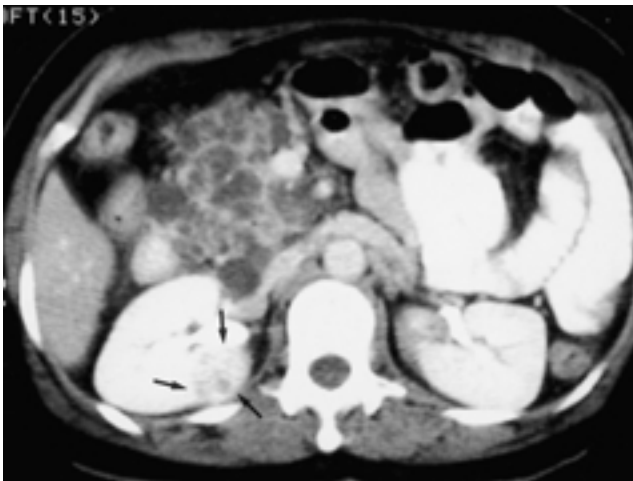


Fig. 1. Contrast-enhanced CT scan shows a 2 cm sized solid mass (arrows) on right kidney. Multiple variable sized cysts are also noted throughout pancreas.



Fig. 2. Gd-DTPA enhanced T1-weighted image (400/12) shows a cerebellar hemangioblastoma as a cystic tumor with solid enhancing mural nodule (arrow).

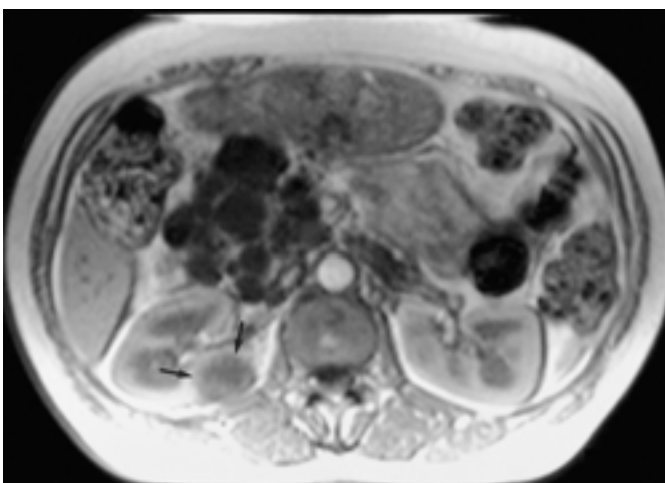
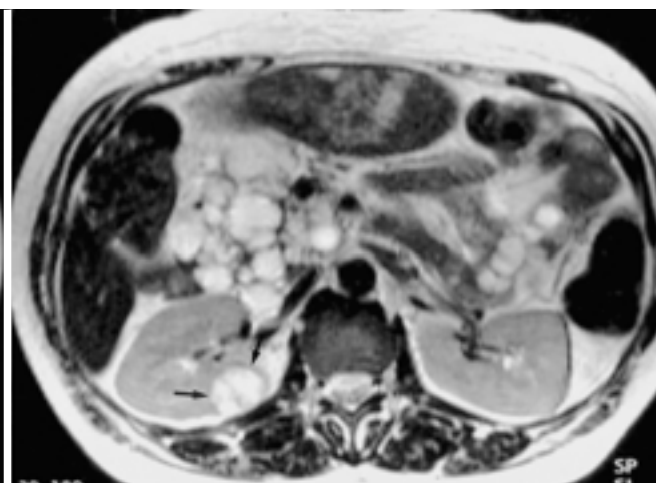


Fig. 3. A. T1-weighted MR image shows a intermediate signal intensity renal mass (arrows) at right kidney and multiple cysts at the head of pancreas.



B. Gd-DTPA enhanced T1-weighted MR image shows homogenous enhancement of the right renal mass (arrows).

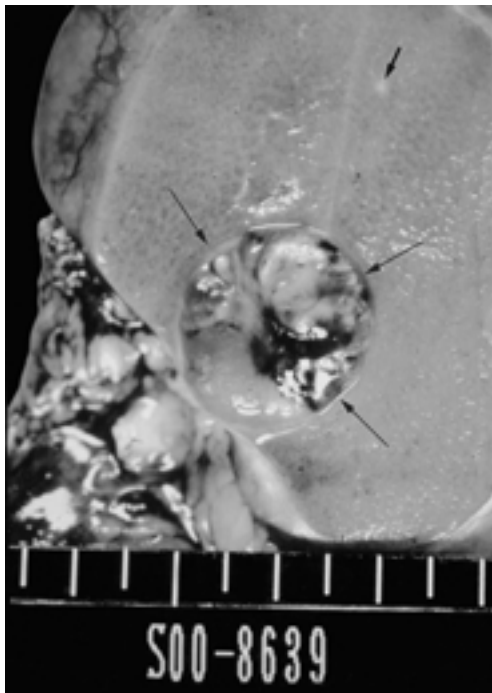


Fig. 4. Gross specimen of right kidney shows a 2.3 cm sized solid tumor, which was detected on CT and MR (long arrows). In addition, there is another small nodule (arrow), which was not detected on CT and MR.



Fig. 6. On the contrast-enhanced CT scan, the patient's son shows multiple pancreatic cysts.

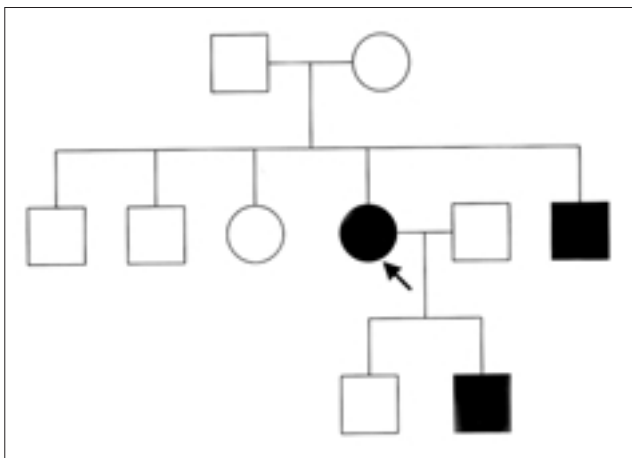


Fig. 5. Pedigree. Squares are male, circles are female and affected individuals are designated in black. The arrow indicates the patient of this case.

VHL
15 가 가
10 가 CT MRI가
(1, 2, 4, 6).
VHL 7 - 18%
VHL

가 (1, 2, 4).
VHL
(4).
MRI (2, 7),
CT MRI 가 (1 - 6).
VHL 가

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Von Hippel-Lindau disease: A Case Report¹

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Von Hippel-Lindau disease is an autosomal dominant disorder characterized by the presence of multiple benign and malignant tumors including hemangioblastomas of the retina and central nervous system, pancreatic cysts and tumors, renal cell carcinomas, pheochromocytomas and epididymal cystadenomas. We report the radiologic findings of a case of von Hippel-Lindau disease, describing the family history.

Index words : Pancreas, cysts
Kidney neoplasms, MR
Brain neoplasms, MR

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