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Von Hippel-Lindau Syndrome: GI Involvement

Linda Pantongrag-Brown, M.D.

Von Hippel-Lindau syndrome (VHL) is a rare, but well-known, autosomal dominant inherited multisystem disorder[1]. It is characterized by abnormal growth of the vessels. Dominant symptoms are related to retinal and CNS involvement[2], secondary to retinal angiomas/hemangioblastomas or cerebellar and spinal cord hemangioblastomas. Renal and adrenal gland involvement are also common. Renal cell carcinomas,

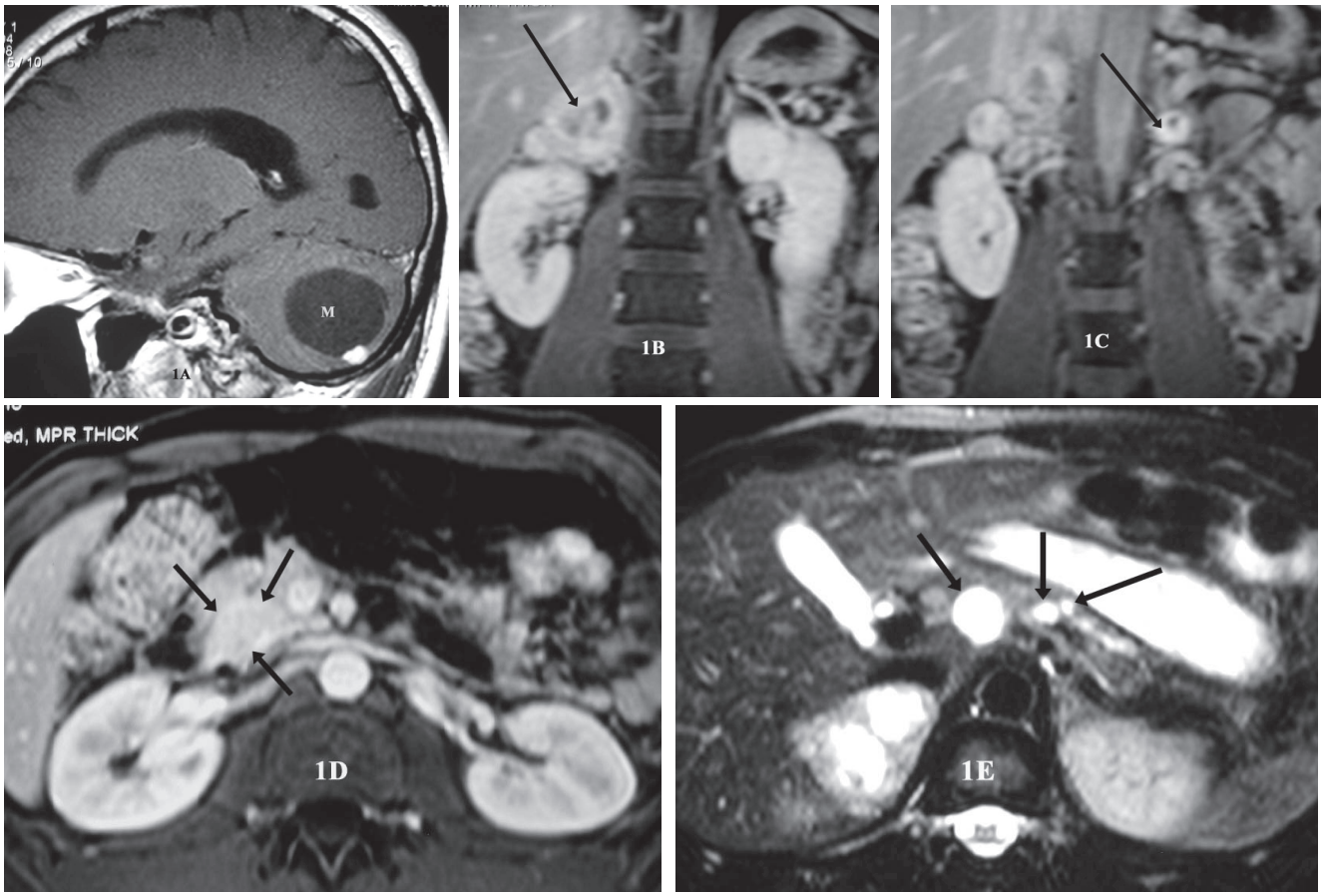

Figure 1 Von Hippel-Lindau syndrome in a 25-year-old male with cerebellar hemangioblastoma, bilateral pheochromocytomas, pancreatic cysts and pancreatic neuroendocrine tumors.

1A Sagittal T1W MRI with IV gadolinium reveals a cystic mass with enhanced mural nodule at the cerebellum (M), characteristic of a hemangioblastoma.

1B-C Coronal T1W MRI with IV gadolinium reveals enhanced right and left suprarenal masses (arrows), which proved to be bilateral pheochromocytomas.

1D Axial T1W MRI with IV gadolinium shows an enhancing mass at the pancreatic head (arrows), which proved to be a neuroendocrine tumor.

1E Axial T2W MRI reveals multiple cysts at the body of the pancreas (arrows).

Department of Radiology, Chulalongkorn University Hospital, Bangkok 10330, Thailand

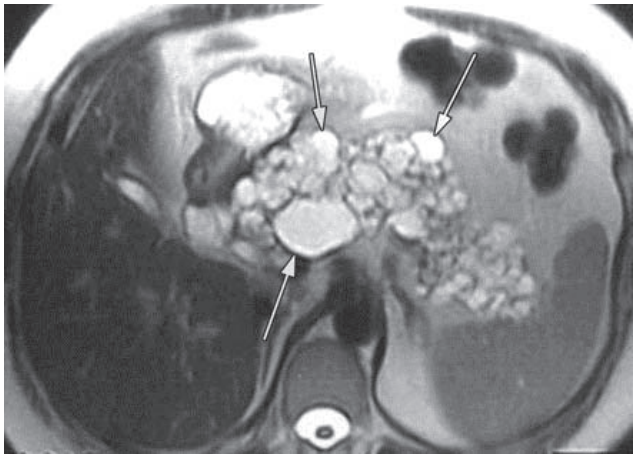

Figure 2 Axial T2W MRI reveals multiple pancreatic cysts (arrows) in a 27-year-old man with Von Hippel-Lindau syndrome.

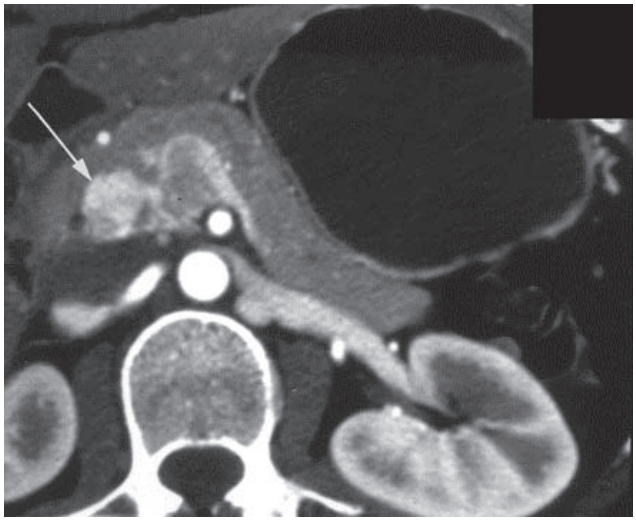

Figure 3 Post IV contrast CT scan reveals an enhancing neuroendocrine tumor (arrow) of the pancreatic head in a 36-year-old female with Von Hippel-Lindau syndrome.

renal hemangioblastomas, and renal cysts are commonly found. Pheochromocytoma of the adrenal gland, either unilateral or bilateral, is usually detected.

GI tract involvement includes pancreas, liver and spleen. Pancreas is among the common organs manifested in VHL. Pancreatic abnormalities are cysts, hemangioblastoma, cystadenoma and neuroendocrine tumors[3-5]. Liver and spleen abnormalities are less common and include cysts, angiomas, and adenomas. Carcinoid tumor of the biliary tract has also been reported.

For follow-up imaging, the affected but asymptomatic individual should have annual renal US examination, MRI or CT scan of the brain every 3 years to age 50 years then every 5 years thereafter, and abdominal CT scan every 3 years.

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