CLINICAL IMAGES

Pancreatic Cysts in Von Hippel-Lindau

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A 30-year-old woman presented with 3 months of epigastric pain. Three years prior, she had a suboccipital craniotomy and resection of a hemangioblastoma. Family history was notable for brain tumors in her father and paternal uncle. Examination revealed a tender irregular mass in the epigastrium. Abdominal MRI showed innumerable pancreatic cysts. The largest one measured 22 mm with a solid component (Fig. 1). Genetic testing confirmed Von Hippel-Lindau (VHL) disease.

This autosomal-dominant condition presents with an assortment of benign and malignant tumors and cysts. It is caused by germline mutations of the VHL tumor suppressor gene. The mean age of presentation is 26 years and most commonly presents with retinal tumors. Other common tumors include central nervous system hemangioblastomas, renal cell carcinomas and pheochromocytomas, middle ear tumors, and cystadenomas of the epididymis and adnexa. Ophthalmologic examination was normal in this patient but may reveal retinal hemangioblastomas or capillary hemangiomas. Pancreatic involvement occurs in 65–77% of patients, and may be the sole manifestation in 8%. The majority of pancreatic lesions are simple cysts. Rarely serous cystadenomas or neuroendocrine tumors occur. Most lesions are asymptomatic, but occasionally they present with pain, obstructive jaundice, or pancreatic insufficiency.

Starting as infants, patients require frequent screening tests such as blood pressure and pulse monitoring, ophthalmology and audiology exams, urine (or plasma) metanephrines, and central nervous system and abdominal imaging.

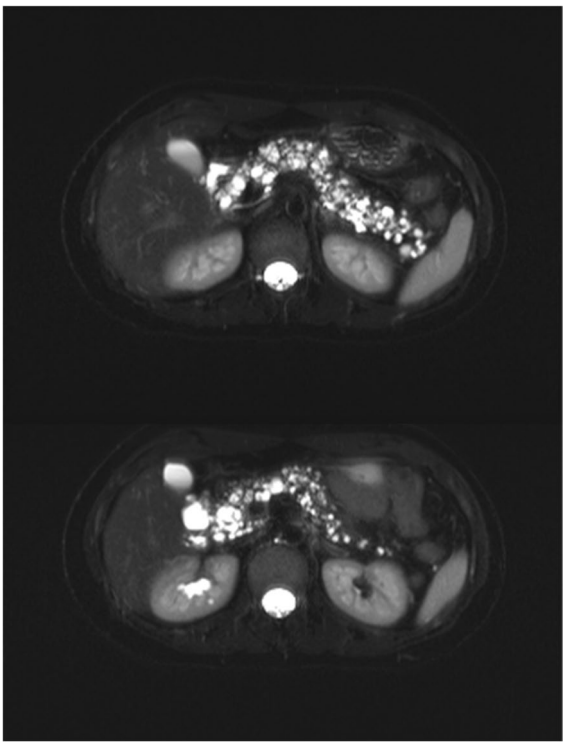

Fig. 1 Multiple pancreatic cysts visualized on abdominal MRI.

Declarations

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