We describe five patients diagnosed with von Hippel-Lindau disease who complained of abdominal distension, pain and discomfort for a long time. All patients underwent ultrasonography, CT scan and MRI, which showed huge pancreas filled with multiple cysts. Additionally, extrapancreatic findings such as cerebellar hemangioblastoma (3 patients), retinal hemangioblastoma (2), renal cell carcinoma (3), renal adenoma (1), renal cysts (4), and splenic cyst (1) helped to reach the right diagnosis. One patient who had no known associated pathology had a family history of von Hippel-Lindau disease. Pancreatic cysts detected on imaging may be a clue to the diagnosis of von Hippel-Lindau disease. In all patients with multiple pancreatic cysts, this disease should be included in the differential diagnosis. [Indian J Gastroenterol 2006;25:90-91]

Von Hippel-Lindau disease (VHLD) is a rare autosomal dominant familial tumor syndrome associated with retinal hemangioblastoma (59%), cerebellar hemangioblastoma (59%), spinal cord hemangioblastomas (13%); renal cysts (50%-75%), renal cell carcinoma (28%-45%), pheochromocytomas, pancreatic cysts, pancreatic serous cystadenomas, and pancreatic neuroendocrine tumors (15%-77%). VHLD is associated with inactivation of a tumor suppressor gene, p25-p26, in the short arm of chromosome 3.

Abdominal CT may depict a pancreatic lesion, which has remained asymptomatic. In fact, this condition has been reported to present usually with pancreatic cystic lesions. We describe five cases presenting with abdominal symptoms due to huge pancreatic cystic lesions who were finally diagnosed with VHLD.

Discussion
VHLD is a rare cause of pancreatic cysts. The frequency of pancreatic involvement in VHLD varies from 0% to 72%, with a mean incidence of approxi-
mately 50%. The other two systemic diseases that commonly manifest with multiple pancreatic cysts are polycystic kidney disease (PKD) and cystic fibrosis (CF). Renal involvement in our patients was not compatible with PKD. CF presents with dilatation of acini and cyst formation due to obstruction by protein plugs, and the pancreatic cysts are very small, leading to atrophic pancreas. In our patients pancreatic cysts were not small and the pancreas was enlarged prominently.

Calcification is observed in 40% of VHLD patients with pancreatic cysts. This is generally thin and peripheral and differs from that seen in chronic pancreatitis. One of our cases had calcification located at the periphery of the cysts.

Abdominal ultrasonography has poor sensitivity and negative predictive value for pancreatic lesions. Dynamic intravenous contrast-enhanced CT is an accurate technique for detecting and characterizing a pancreatic mass. Contrast-enhanced MRI is superior to CT in detecting septa formations and enhancing components of the lesion, which is typical in microcystic adenomas. Noninvasive treatment and radiological follow up is acceptable if these lesions ever become symptomatic. The following schedule is recommended: USG yearly, beginning at 11 years of age; CT yearly or every 2 years, beginning at 20 years of age; MRI as indicated. Repeated radiological examination discloses change in size. Morbidity and mortality due to pancreatic disease is not severe, as in renal and central nervous system involvement.

In conclusion, pancreatic involvement occurs in most patients with VHLD. Pancreatic cysts detected by abdominal imaging may be the first clue to VHLD. Extrapancreatic findings and detailed medical history help with the right diagnosis. Although VHLD is not a common cause of pancreatic cystic disease, it should be considered in any patient who presents with multiple pancreatic cysts.

References

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