Surg. Gastroenterol. Oncol. 2019;24(2):122-126 DOI: 10.21614/sgo-24-2-122

Multiple Organ Cystic Lesions: A Possible Imaging Target Sign in Von Hippel Lindau Disease - Case Report

Adrian Dumitru Dijmărescu[1,2], Ioana Gabriela Lupescu[1,2]
[1]Radiology and Medical Imaging Department, Fundeni Clinical Institute
[2]"Carol Davila" University of Medicine and Pharmacy, Bucharest, Romania

Abstract

Introduction: Von Hippel Lindau disease (VHLD) is a rare autosomal-dominant disorder. Multi-organ cysts and low histological grade tumors with various localizations are characteristic for VHLD. Imaging studies are important for the evaluation of VHLD patients.

Case report: We report the case of a 53 years old woman who was admitted for weight loss, chronic abdominal pain and mild jaundice. The patient was known with a cerebral tumor previously removed. Family history was suggestive for VHLD. US showed voluminous cystic lesions developed on the kidneys and in the interhepatopancreatic region with dilated intrahepatic bile ducts. Subsequently contrast enhanced CT and MRI with MRCP were performed. CT showed disorganized pancreatic parenchyma, inhomogeneous by the presence of numerous hypodense lesions, most of them with fluid content, thin nonenhancing walls and small intramural calcifications. Both kidneys had irregular contour, with multiple bilateral cortical cysts and a heterogeneous fast enhancing arterial lesion in the left kidney suggestive for a malignant tumor. MRI showed the same aspect as CT, but also that the lesions exerted a mass effect on the pancreatic tissue and on the Wirsung duct, with no visible communications.

Conclusions: Imaging aspects plays a key role together with the family history for establishing or at least suspecting the diagnosis of VHLD, even though genetic testing is not highly available in all regions. A multidisciplinary team approach is important in the diagnostic and management of VHLD.

Key words: Von Hippel Lindau Disease, pancreatic cysts, renal cysts, solid renal tumor, magnetic resonance imaging, computed tomography