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Polycystic liver disease

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CASE REPORT

A 54-year-old man had an asymptomatic elevation of cytolysis-related enzymes, GGT, and alkaline phosphatase, detected at a routine laboratory study. The patient reported no alcohol abuse, no history of liver disease, and no constitutional syndrome. Physical examination showed a good general status, normal colored skin and mucosas, and no stigmata of chronic liver disease. The abdomen was soft, depressible, and slightly tender in the right hypochondrium with hepatomegaly of three finger-widths. There were no signs or symptoms of ascites. A laboratory study for liver disease was also negative. An initial ultrasound study showed the presence of multiple, well-defined anechoic lesions with posterior acoustic enhancement, distributed across the hepatic parenchyma; the kidneys were free of lesions. The study was completed with a liver MRI, which showed hypointense lesions resembling cerebrospinal fluid (CSF) on T1-weighted images, whereas in T2-weighted images with fat saturation lesions were hyperintense (Figs. 1 and 2).

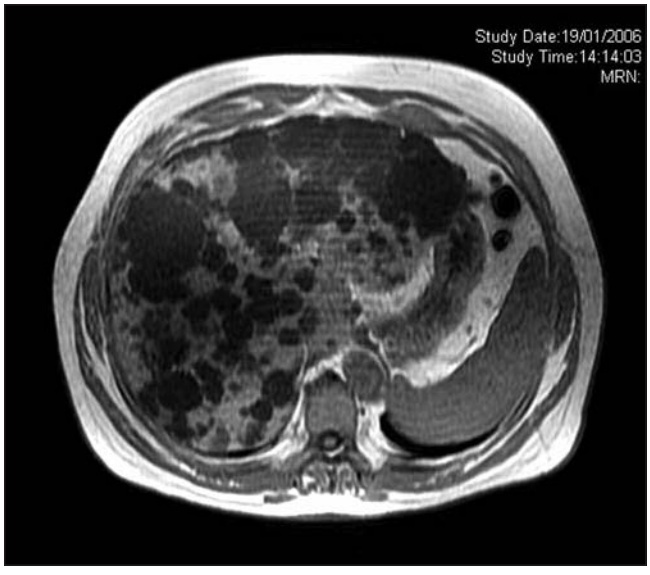

Fig. 1. Gadolinium-enhanced images showing hypointense lesions similar to CSF and without gadolinium enhancement.
Imágenes con potenciación T1 y administración de gadolinio en la que se observan lesiones hipointensas similar a LCR y sin realce de gadolinio.

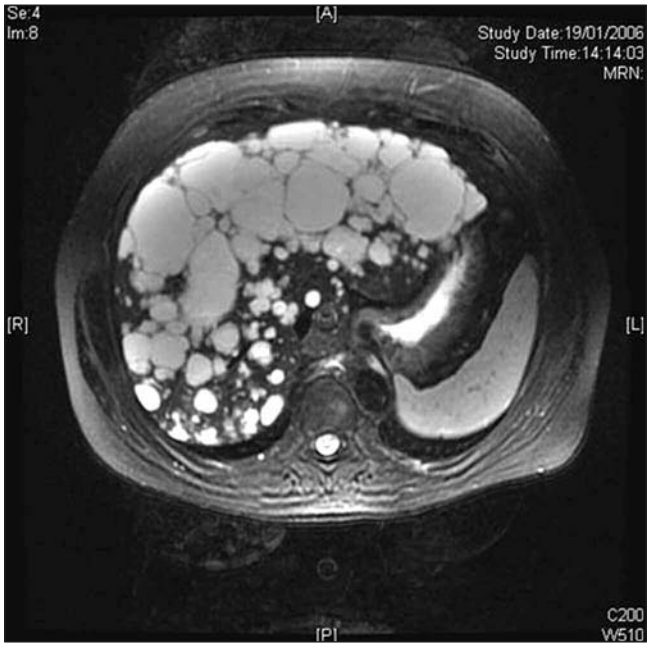

Fig. 2. T2-weighted sequence with fat saturation in which the lesions are hyperintense, similar to CSF.
Secuencia T2 con saturación grasa en la que las lesiones presentan un comportamiento hiperintenso similar al LCR.

DISCUSSION

Polycystic liver disease is an autosomal dominant hereditary disorder characterized by the presence of multiple disperse cysts of biliary origin in the liver. Liver cysts often present during the fourth decade of life. A gene related to this disease has been localized on chromosome 19. Certain mutations in the PRKCSH gene, which codes for the substrate protein 80K-H of protein kinase C, have recently been reported to be associated with this disease (1). Most patients are asymptomatic and the diagnosis is usually incidental. When this is not the case, typical symptoms include painful hepatomegaly, abdominal distension, a feeling of fullness, or lumbar pain (2). Very occasionally symptoms derive from cyst complications (rupture, infection, malignancy) (3). Treatment is symptomatic. Surgery is required for patients with cysts larger than 10 cm or with complications, such as intracystic hemorrhage, infection not susceptible to interventional radiology, or in cases of suspected or confirmed cystic malignancy (3).

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