Adult polycyst liver disease

Raúl Carrillo Esper,* Jorge Raúl Carrillo Córdova,** Luis Daniel Carrillo Córdova***

69 years old woman without antecedents of hepatic disease who was admitted to Intensive Care Unit by severe pneumonia. Clinical exploration with nodular hepatomegaly without ascitis or splenomegaly. Liver function test with total proteins 7 g/dL, albumin 3.6 g/dL, Globulins 3.4 g/dL, total bilirrubins 0.62 mg/dL, Direct bilirrubin 0.15 mg/dL, Indirect bilirrubin 0.47 mg/dL, ALT 31 U/L, AST 40 U/L, AP 85 U/L, GGT 34U/L, LDH 188 U/L. Creatinine 0.8 mg/dL, BUN 13.3 mg/dL. Computed tomography shows multiple hepatic and renal cysts (Figure1). Hepatic Ultrasound shows multiple hepatic cysts and a normal portal system, anterograde portal flow, suprahepatic veins and biliary tract without abnormalities or obstruction.

Autosomal dominant polycyst liver disease is a rare disorder usually associated with autosomal dominant kidney polycyst disease with an increasing prevalence associated with age and female gender. Patients with autosomal-dominant polycystic disease present a wide spectrum of hepatic and renal involvement. Some patients have renal cysts only; others have liver cysts only; while others show variable degrees of both kidney and liver involvement. Although usually asymptomatic, patients with Adult Policyst Liver Disease (ALDP) may present with abdominal pain or swelling. Liver function is not usually compromised and computed tomography or abdominal ultrasonography is the most useful investigations. The complications

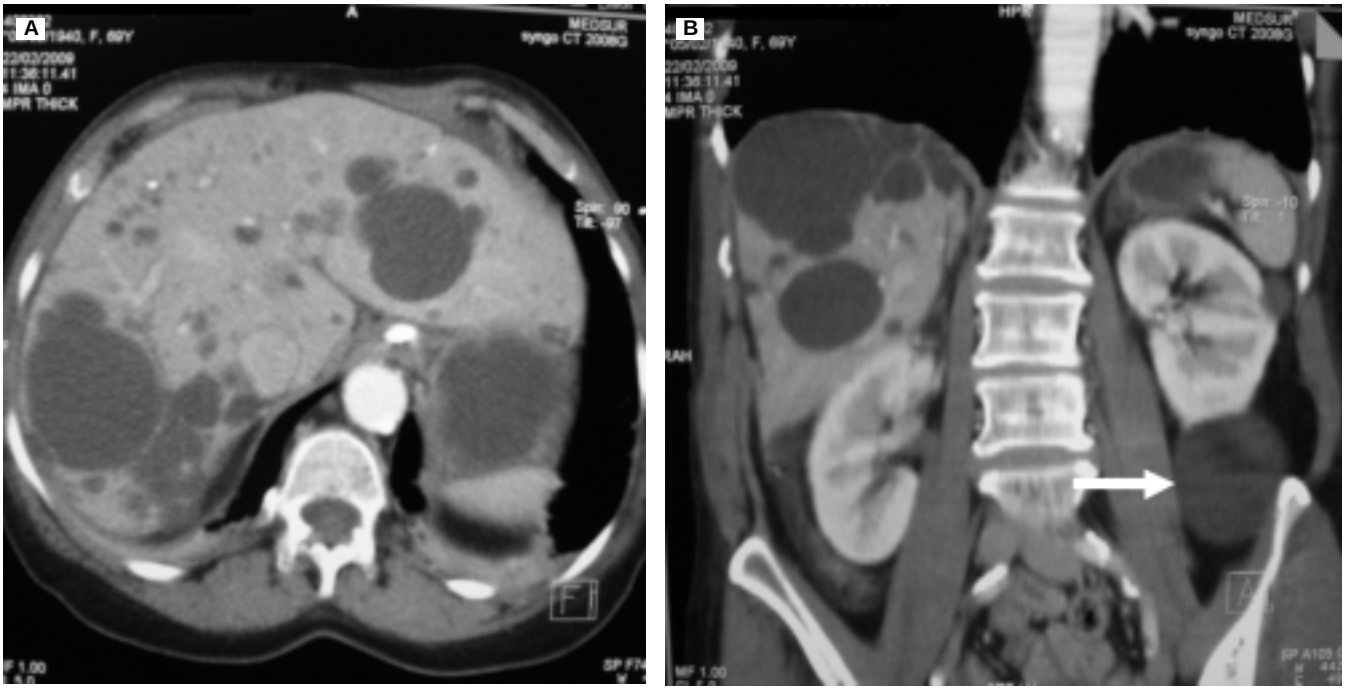

Figure 1. A. CT with different size liver cysts. **B.** Liver and left kidney cysts (arrows).

* Academia Nacional de Medicina. Academia Mexicana de Cirugía. Jefe de UTI Fundación Clínica Médica Sur.
** Interno de Pregrado. Facultad de Medicina. Grupo NUCE. UNAM.
*** Pregrado. Facultad de Medicina. Grupo NUCE. UNAM

are cyst rupture, infection, portal hypertension, obstructive jaundice, liver failure, cholangiocarcinoma and compression of surrounding structures.[1-3]

On histologic examination, the liver cysts are usually lined by cuboidal epithelium, similar in appearance to that lining the bile ducts. It is generally believed that the cysts develop from cystic dilatation of aberrant bile ducts (Meyenburg complexes) that fail to establish connection embryologically with the larger interlobular bile ducts in the portal tracts. The cysts do not communicate with the biliary system, but the lining cells gradually secrete fluid into the cysts. Cyst growth is modulated by cAMP, which stimulates cholangiocyte proliferation and cyst fluid secretion, a process which is inhibited by somatostatin.[4-6]

Most cases of Adult Policyst Liver Disease are asymptomatic and do not require surgical treatment. Surgical treatment remains controversial, other options are cyst puncture, open or laparoscopic fenestration with or without hepatic resection, and liver transplantation.[7,8] By the other hand, octreotide, can reduce the cyst volume.[9]

REFERENCES

1. D'Agata ID, Jonas MM, Perez-Atayde AR, Guay-Woodford LM. Combined cystic disease of the liver and kidney. *Semin Liver Dis* 1994; 14: 215-28.
2. Vauthey JN, Maddern GJ, Blumgart LH. Adult polycystic disease of the liver. *Br J Surg* 1991; 78: 524-7.
3. Levine E, Cook LT, Grantham JJ. Liver cysts in autosomal-dominant polycystic kidney disease: Clinical and computed tomographic study. *Am J Roentgenol* 1985; 145: 229-33.
4. Ramos A, Torres VE, Holley KE, Offord KP, Rakela J, Ludwig J. The liver in autosomal dominant polycystic kidney disease. Implications for pathogenesis. *Arch Pathol Lab Med* 1990; 114: 180-84.
5. Patterson M, Gonzalez-Vitale JC, Fagan CJ. Polycystic liver disease: A study of cyst fluid constituents. *Hepatology* 1982; 2: 475-8.
6. Alvaro D, Gigliozzi A, Attili AF. Regulation and deregulation of cholangiocyte proliferation. *J Hepatol* 2000; 33: 333-40.
7. Russell RT, Pinson CW. Surgical management of polycystic liver disease. *World J Gastroenterol* 2007; 13: 5052-9.
8. Li TJ, Zhang HB, Lu JH, Zhao J, Yang N, Yang GS. Treatment of polycystic liver disease with resection-fenestration and a new classification. *World J Gastroenterol* 2008; 14: 5066-72.
9. Van Keimpema L, de Man RA, Drenth JP. Somatostatin analogues reduce liver volume in polycystic liver disease. *Gut* 2008; 57:1338-9.

Correspondence:

Dr. Raúl Carrillo Esper
Unidad de Terapia Intensiva
Fundación Clínica Médica Sur
Puente de Piedra No. 150
Col. Toriello Guerra. Tlalpan
Tel.: 5424-7200
E-mail: rcarrillo@medicasur.org.mx