



Hepatorenal Polycystosis Complicated By Hepatic Cirrhosis: A Case Report

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Abstract

Polycystic liver disease is most commonly associated with autosomal dominant polycystic kidney disease. Hepatic cysts are the most common extrarenal manifestation of autosomal dominant polycystic kidney disease. The progression to cirrhosis remains rare, and the data is sparse, the only curative treatment is liver transplantation. We report the case of a young patient with hepato-renal polycystosis at the stage of cirrhosis.

Keyword: Cirrhosis, hepato-renal polycystics, portal hypertension, hematemesis, liver transplantation, case report.

Introduction

Polycystic liver disease is the most common benign extra-renal manifestation of polycystic kidney disease. Liver cysts usually develop after kidney cysts and have an annual growth rate of around 0.9% to 3.2%, most of studies have shown that exposure to estrogen, growth factor (IGF-1), and Ciliary dysfunction can influence the growth of cysts. The progression to cirrhosis is rare and late, presenting especially in the elderly ^[1]. We report the case of a young patient, follow-up from polycystic hepatic and renal disease complicated by cirrhosis at the stage of portal hypertension revealed by massive digestive hemorrhage.

Patient information

An 18-year-old patient, diagnosed with polycystosis hepatic and renal since the age of 3, referred to our unit for the management of upper gastrointestinal bleeding made of 2 episodes of abundant hematemesis, in a context of hemodynamic instability requiring initial management in an intensive care unit.

Clinical findings

Clinical examination on admission showed a mucocutaneous pallor and asthenia, no sign of hepato-cellular insufficiency found in our patient, the abdominal examination has shown a splenomegaly reaching up to the umbilicus, rectal examination and anal margin didn't notice any abnormalities.

Diagnostic assessment

A routine blood count gave the following results: Leucopenia : white blood cell count = 1210 / μ L, Anemia : hemoglobin = 7.3 g / dL, low plaquet level at 54,000 / mm³, correct renal function with creatinine at 8.6, urea at 0.3, a low prothrombin level at 41.9%, Albuminemia at 21 g / l, Bilirubinemia at 11.3 μ mol / L, no cytolysis or cholestasis was found in our patient, the rest of the blood panel was normal including the etiological investigation for auto-immune, viral hepatitis B and C and hepatic overload. On endoscopic exploration: eso-gastro-duodenal fibros copy found oesophageal varices grade III mammillated with red sign, osogastric varices type I without red sign, hypertensive gastropathy. Abdominal imaging including computed tomography (CT) and ultrasound revealed a dysmorphic liver with irregular contours, segment IV atrophy and segment I hypertrophy, portal trunk retracted, collateral venous circulation of the splenic hilum and splenomegaly associated with hepato-renal polycystosis (**Figure 1**). On fibro-scan, the liver fibrosis was staged at 20 Kpa. The diagnosis of cirrhosis was retained based on clinical, biological, endoscopic, and scanno-graphic criteria. Genetic counseling was suggested for his siblings.

Therapeutic intervention and follow-up:

5 sessions of endoscopic variceal ligation were performed, and put on beta blockers with good tolerance and efficiency, annual endoscopic surveillance is planned as well as a liver transplant.

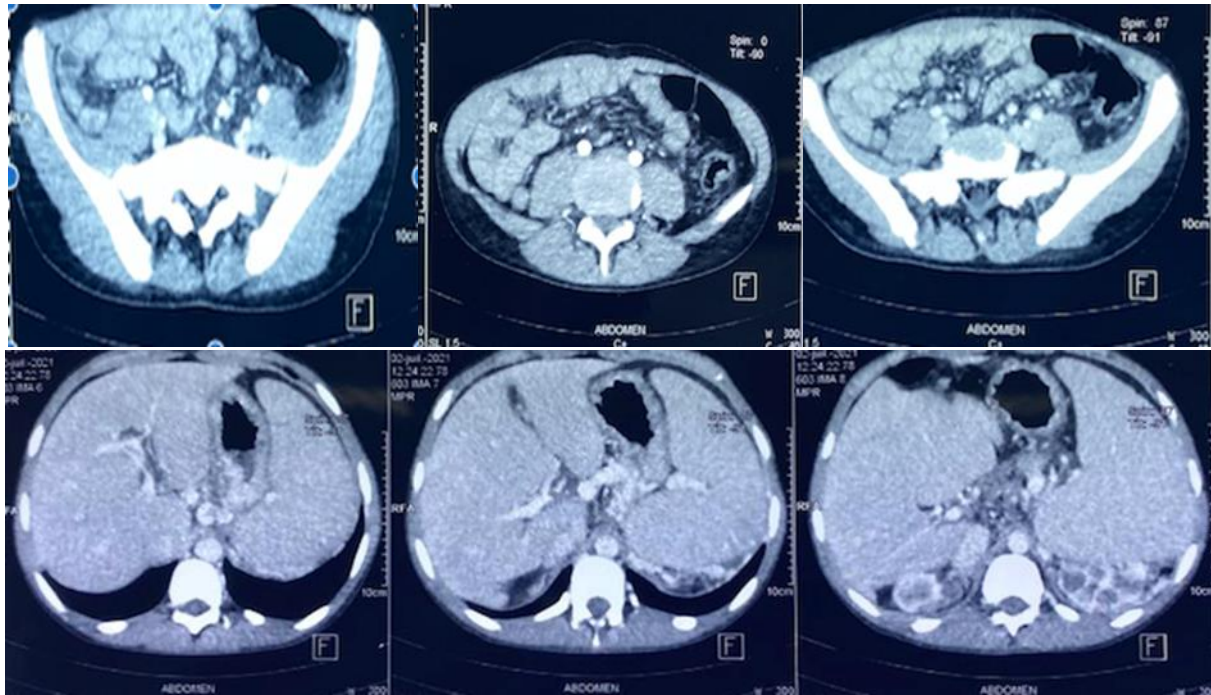


Figure 1: CT appearance in favor of a dysmorphic liver with irregular contours, with atrophy of segment IV and hypertrophy of segment I, retracted PT, CVC of the splenic hilum and splenomegaly associated with hepato-renal polycystosis

Discussion

Polycystic liver disease results from an embryonic malformation of the ductal plaque of the intrahepatic bile tree [2]. The prevalence ranges between 100 and 1/1000 [3]. The phenotype consists of numerous cysts distributed throughout the usually silent hepatic parenchyma. Women generally have a more severe liver phenotype, especially for those with a history of multiple pregnancies and prolonged exposure to estrogens [4]. In advanced stages, hepatic cysts can cause compromised portal venous flow, leading to the development of hepatic fibrosis resulting in hepatic cirrhosis which remains a rare event. It is symptomatic in isolated polycystic liver disease or as part of autosomal dominant polycystic kidney disease, often these cysts are found incidentally and clinically insignificant.

The symptoms are diverse ranging from abdominal pain or abdominal distension, to a masse effect by the compression of adjacent tissues leading to hepatomegaly, splenomegaly, portal hypertension syndrome due to reduced flow of the hepatic veins or portal vein influx which may be compressed due to the volume effect of hepatic cysts, ascites either secondary to progression to cirrhosis or to lymphatic leakage and chronic renal failure, jaundice by compression of the bile duct, gastroesophageal reflux and deterioration of the general condition. Liver function is preserved for a long time [2]. Abdominal imaging is the first instrument to assess the hepatic phenotype, it allows the macroscopic detection of hepatic and renal cysts without or with contrast. On ultrasound cysts appear as homogeneous round spaces filled with anechoic fluid. Magnetic resonance imaging is superior to ultrasound and computed tomography and allows better characterization of small cysts in young people [5,6].

In familial forms the diagnosis is based on the detection of hepatorenal cytogenesis and on the other hand, on a positive family history compatible with an autosomal dominant or recessive pattern of inheritance [4]. The main goal of treatment is to reduce symptoms by reducing the development of liver cysts. The treatment of choice is motivated by individual complaints. The assessment of symptoms associated with quality of life is one item

to focus on. Therapeutic interventions are not justified in asymptomatic patients. Stopping oral contraceptives remains an important step in preventive treatment, the use of intrauterine slides may be an acceptable alternative. Analgesic management is the first-line treatment in patients with acute or chronic abdominal pain and tenderness. Different invasive approaches with possible beneficial results in independent studies include aspiration sclerotherapy, laparoscopic cyst removal, or liver transplantation. The treatment of patients in the stage of cirrhosis is no different from that of other causes of cirrhosis, some authors suggest stents to relieve the temporal symptoms of portal hypertension and ascites. A portosystemic shunt may be indicated in the presence of acute thrombosis or vascular compression to establish hepatic and portal venous flow. The main goal is to improve symptoms by decompressing the cysts, this is often achieved by end-treatment strategies, including liver resection or liver transplantation. [7-16].

Conclusion

Polycystic hepato-renal disease is a relatively rare inherited condition. Genetics and environmental factors contribute to the progression of the disease. They have resulted in several symptoms which can be serious if not treated in time. The progression to cirrhosis is exceptional, but it should not be overlooked that the assessment of symptoms and quality of life is necessary to decide on adequate and beneficial management.

Declaration

Conflicting interest

The authors declare no conflict of interest.

Contributions from the authors

All authors have contributed to this study since conception, reading and have approved the final version of the manuscript

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Reference

- [1] Christina Awad, Grand Gardner Gallimore, Internal medicine, keesler medical center, biloxi, Mississippi, USA BMJ Case rep. 2018
- [2] Wybrich R Cnossen and Joost PH Drenth, Polycystic liver disease : an overview of pathogenesis, clinical manifestations and management Orphanet J rare dis 2014, 9 :69
- [3] Van Keimpema L, De Koning DB, Van Hoek B, Van Den Berg AP, Van Oijen MG, De Man RA, Nevens F, Drenth JP. Patients with isolated polycystic liver disease referred to liver centres: clinical characterization of 137 cases. *Liver Int.* 2011;31:92–98. [PubMed] [Google Scholar]
- [4] Miranda Henriques MS, de Morais Villar EJ, The Liver and polycystic kidney disease Department of internal medicine USA 2015, Chapter 17
- [5] Mortele KJ, Ros PR. Cystic focal liver lesions in adult: Differential CT and MR imaging features. *Radiographics.* 2001 ;21 :895-910
- [6] Bae KT, Zhu F, Chapman AB, Torres VE, Grantham JJ, Guay-Woodford LM and All. Consortium for radiologic imaging studies of polycystic kidney disease (CRISP) Magnetic resonance imaging evaluation of hepatic cysts in early autosomal-dominant polycystic kidney disease : The consortium for radiologic imaging studies of polycystic kidney disease cohort. *Clin J Am soc Nephrol.* 2006 ;1 :64-69
- [7] Temmerman F, Missiaen L, Bammens B, Laleman W, Cassiman D, Verslype C, Van Pelt J, Nevens F. Systematic review: the pathophysiology and management of polycystic liver disease. *Aliment Pharmacol Ther.* 2011;34:702–713. [PubMed] [Google Scholar]
- [8] Macutkiewicz C, Plastow R, Chrispijn M, Filobos R, Ammori BA, Sherlock DJ, Drenth JP, O'Reilly DA. Complications arising in simple and polycystic liver cysts. *World J Hepatol.* 2012;4:406–411.
- [9] Uddin W, Ramage JK, Portmann B, Wilson P, Benjamin I, Tan KC, Williams R. Hepatic venous outflow obstruction in patients with polycystic liver disease: pathogenesis and treatment. *Gut.* 1995;36:142–145.[PMC free article] [PubMed] [Google Scholar]
- [10] Torres VE, Rastogi S, King BF, Stanson AW, Gross JB Jr, Norgorney DM. Hepatic venous outflow obstruction in autosomal dominant polycystic kidney disease. *J Am Soc Nephrol.* 1994;5:1186–1192. [PubMed] [Google Scholar]
- [11] Chauveau D, Grunfeld JP, Durand F, Belghiti J. Ascites in a polycystic patient. *Nephrol Dial Transplant.* 1997;12:228–230. [PubMed] [Google Scholar]
- [12] Van Keimpema L, Nevens F, Adam R, Porte RJ, Fikatas P, Becker T, Kirkegaard P, Metselaar HJ, Drenth JP, European L. Intestine Transplant A: Excellent survival after liver transplantation for isolated polycystic liver disease: an European Liver Transplant Registry study. *Transpl Int.* 2011;24:1239–1245.[PubMed] [Google Scholar]
- [13] Morgan DE, Lockhart ME, Canon CL, Holcombe MP, Bynon JS. Polycystic liver disease: multimodality imaging for complications and transplant evaluation. *Radiographics.* 2006;26:1655–1668. quiz 1655.
- [14] Gevers TJ, Drenth JP. Diagnosis and management of polycystic liver disease. *Nat Rev Gastroenterol Hepatol.* 2013;10:101–108.[PubMed] [Google Scholar]
- [15] Wijnands TF, Neijenhuis MK, Kievit W, Nevens F, Hogan MC, Torres VE, Gevers TJ, Drenth JP. Evaluating health-related quality of life in patients with polycystic liver disease and determining the impact of symptoms and liver volume. *Liver Int.* 2013. doi:10.1111/liv.12430. [PubMed]
- [16] Drenth JP, Chrispijn M, Nagorney DM, Kamath PS, Torres VE. Medical and surgical treatment options for polycystic liver disease. *Hepatology.* 2010;52:2223–2230. [PubMed] [Google Scholar]



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