ECRONICON OPEN ACCESS

EC GASTROENTEROLOGY AND DIGESTIVE SYSTEM

Case Report

A Rare Case of Infected Isolated Polycystic Liver Disease Presenting Jaundice and Ascites

W Hliwa, I Jbara*, F El Rhaoussi, M Tahiri, F Haddad, A Bellabah and W Badre

Department of Gastroenterology, Ibn Rochd Hospital, Medical University of Casablanca, Morocco

***Corresponding Author:** I Jbara, Department of Gastroenterology, Ibn Rochd Hospital, Medical University of Casablanca, Morocco.

Received: April 04, 2023; **Published:** May 31, 2023

Abstract

Isolated polycystic liver disease is a rare disease. Patients with isolated polycystic liver disease are usually asymptomatic and the liver cysts are detected incidentally on imaging studies performed for other purposes. Patients rarely develop symptoms, which are most often secondary to cyst enlargement and hepatomegaly. Management of symptomatic PCLD focuses on symptom relief and treatment of complications.

We report the case of a 39-year-old man who presented with ascites and febrile jaundice, whose laboratory tests showed an infectious syndrome, cholestasis and negative hydatid and viral serologies. The CT scan showed a hepatomegaly with multiple cysts of variable size, ascites and normal kidney structure. The diagnosis of isolated polycystic hepatitis was retained. The patient was put on antiobiotherapy with albumin infusion, the patient's clinical and biological condition has improved evolution.

Keywords: *Isolated Polycystic Liver Disease; Jaundice; Ascites; Infectious Hepatic Cyst*

Abbreviations

PKH: Polycystic Liver Disease; ADPKD: Isolated Polycystic Liver Disease Autosomal Dominant ADPKD: Polycystic Kidney Disease Autosomal Dominant Polycystic Kidney Disease; PCLD: Isolated Polycystic Liver Disease; PRKCSH: Protein Kinase C Substrate 80K-H

Introduction

Polycystic liver disease (PKH) is a rare hereditary disease characterized by the presence of multiple diffuse liver cysts. It can be isolated (isolated PKH) or, most often, associated with either autosomal dominant polycystic kidney disease (ADPKD), or autosomal recessive polycystic kidney disease (PKRAR), which is more rare [1]. The isolated polycystic liver disease (PCLD) is an autosomal dominant disease with clinical and genetic heterogeneity. A small proportion of patients develop acute liver-cyst-related complications and/or massive cystic liver enlargement, causing morbidity and mortality. We present a case of a -man with family history of polycystic liver disease presented by recent onset of ascites and febrile jaundice.

Case Report

A 39-year-old patient, no past history of medical importance or history of taking hepatotoxic drugs who had a family history of his sister and his mother with polycystic liver disease. He was admitted to the gastroenterology unit for progressive abdominal distension, chronic abdominal pain in the upper right quadrant and four days onset of febrile jaundice. The clinical examination showed febrile jaundice at

38°, bilateral lower limb edema, ascites and enlarged liver at 28 cm. Routine blood tests showed normochromic normocytic anemia at 11.2 g/dl, normal white blood cell and platelet count. CRP (C reactive protein) and PCT were high at 270 mg/l and 1.09 respectively with cytolysis and conjugated hyperbilirubinemia at 16,1mg/l. He also had a low prothrombin level at 48% and low factor V at 45%. Renal function tests were within normal levels. Hydatid, viral and HIV serology were negative and CA19-9 was slightly elevated at 60.

The abdominal CT scan revealed significant hepatomegaly of 34 cm in crania-caudal diameter; 29 cm in transverse diameter and 20 cm in anteroposterior diameter, multiple cysts of variable size with thin and regular walls laminating the hepatic parenchyma and minimal ascites but no dilation of the bile ducts and normal kidneys (Figure 1). The ascitic fluid was analyzed and showed a transudative type with a low number of neutrophils (PNN) and no malignant cells. The fluid from the liver cysts was aspirated and examined, showing a sterile serous fluid. Therefore, the case was diagnosed as an isolated polycystic liver disease complicated by jaundice, hydrops and probably a superinfected cyst.

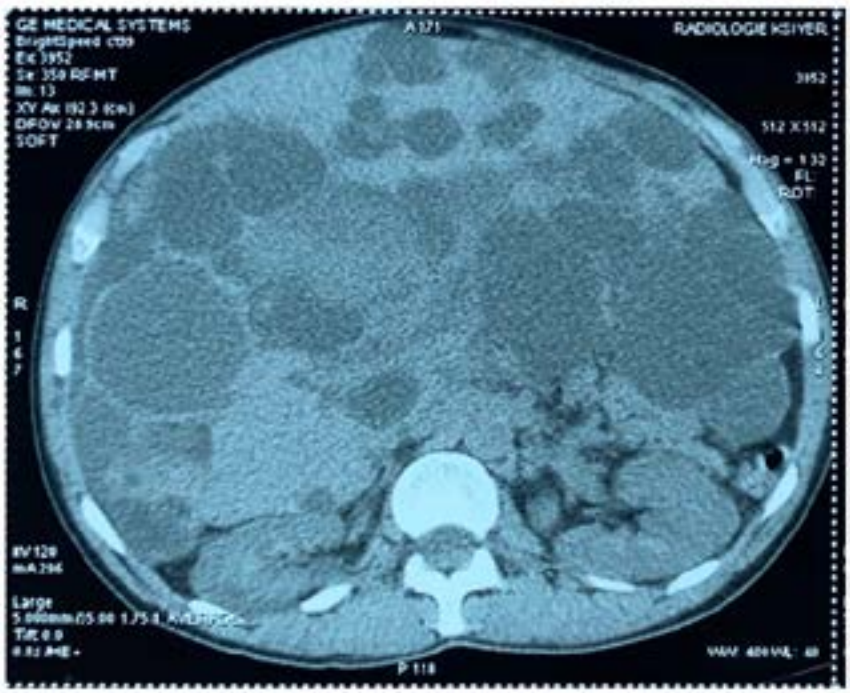

Figure 1: *Scanning image of polycystic liver disease.*

The patient was treated with intravenous metronidazole and 3rd generation cephalosporins for 10 days, followed by oral treatment for four weeks and albumin infusion. The patient improved clinically with the progressive disappearance of jaundice, ascites and microbiological signs of infection.

Discussion

Isolated PKH is a rare disease with an estimated prevalence of 1/100,000 to 1/1000,000 [2]. It is autosomal dominant [1], mutations in the PRKCSH or SEC63 gene in (about 20 - 30%) [3,4], but in a large number of cases the pathogenic gene cannot be found [5]. Women have

a more frequent and earlier onset of massive polycystic liver disease than men [6] is strongly correlated with the number of pregnancies [6]. Oral contraception significantly increases the risk of cysts [6], and hormone replacement therapy during menopause increases their volume [6].

The majority (> 80%) of PCLD patients are clinically asymptomatic [7]. Patients who develop symptoms are usually secondary to the increased size and mass effect of liver cysts.

They may present by abdominal distension, chronic abdominal pain, early satiety, or dyspnea. Compression of the inferior vena cava or, more rarely, the biliary system may lead to edema of lower limbs s and jaundice [8]. In addition to these manifestations due to the cystic mass effect, non-cystic liver parenchyma may be altered due to obstruction of the suprahepatic venous outflow, leading to portal hypertension, which may be complicated by ascites, variceal haemorrhage, or hypersplenism [1].

This polycystic liver disease, especially if the cysts are large, can be complicated by either cystic rupture or intracystic hemorrhage. Superinfection of the cyst is also a described complication [9], rare but serious, with a reported mortality rate of 2% [9]. Its incidence is 0.1%/patient/year), the diagnosis is confirmed by puncturing the cyst, revealing enterobacteria and an increase in neutrophils in the liquid [9]. But most often the diagnosis is probably made on the basis of a fever of 38°C, a biological inflammatory syndrome, a significant increase in CA19-9 [1] and the absence of intracystic bleeding on CT scan [9]. The most frequently liver abnormalities observed in polycystic liver disease is a slight elevation of gamma glutamyl transferase and alkaline phosphatase [10].

PCLD is diagnosed by imaging studies, including ultrasound, CT scan and magnetic resonance imaging (MRI). The diagnosis of PKH is usually made when the number of liver cysts is greater than 20, or greater than 4 in a patient with a family history of isolated PKHAD [11]. These cysts vary in size but are often supra-centimetric and have all the characteristics of simple cysts. When the number of cysts is very large, the architecture of the liver is altered, leading to a loss of the usual anatomical and vascular landmarks [12]. Ultrasound is generally preferred because of its low cost, accessibility and lack of radiation exposure. However, CT and MRI are more sensitive and accurate in detecting the presence and size of liver cysts. In addition to this, MRI is most useful in the case of complications, to visualise a T1 hyperintense signal, a sign of intracystic haemorrhage [12].

All these radiological examinations also explore the kidneys of these patients, it is necessary to search for a cystic renal dysmorphism to differentiate an autosomal dominant hepatorenal polycystosis from an isolated hepatic polycystosis.

The presence of clinical features such as voluminous hepatomegaly, febrile jaundice and lower limb oedema associated with radiological findings of PCLD, elevated CRP, elevated CA19-9 and liver function tests, as in the present case, may help to make the diagnosis of infected PCLD in the absence of other causes of infection.

Asymptomatic polycystic liver disease is the most common and does not require treatment [13,14]. Only symptomatic or complicated polycystic liver disease requires therapeutic discussion that ranges from therapeutic abstention to liver transplantation, including medical treatment (somatostatin analogues, ursodeoxycholic acid), percutaneous treatment (aspiration-sclerosis) and surgical treatment (fenestration, liver resection) [1]. The choice between all these treatments will depend on the number, size and location of the cysts and local expertise.

Conclusion

PKH is a rare, autosomal dominant disease that predominates in women. It is slowly progressive and remains asymptomatic for years. Only symptomatic or complicated forms need to be treated.

This case shows that polycystic liver disease should also be considered in the presence of hepatomegaly, jaundice and ascites.

Conflict of Interest

The authors declare no conflict of interest. The article is not sponsored.

Bibliography

1. Vincent DI Martino. "Maladies kystiques hépatiques non infectieuses". *POST'U* (2022).
2. WR Cnossen and JPH Drenth. "Polycystic liver disease: An overview of pathogenesis, clinical manifestations and management". *Orphanet Journal of Rare Diseases* 9.1 (2014): 69.
3. Li S Davila., *et al*. "Mutations in PRKCSH cause isolated autosomal dominant polycystic liver disease". *American Journal of Human Genetics* 72.3 (2003): 691-703.
4. S Davila., *et al*. "Mutations in SEC63 cause autosomal dominant polycystic liver disease". *Nature Genetics* 36.6 (2004): 575-577.
5. Boerrigter Masyuk AI., *et al*. "Autophagy Promotes Hepatic Cystogenesis in Polycystic Liver Disease via Depletion of Cholangiocyte Ciliogenic Proteins". *Hepatology* (2022).
6. Ludovic ROSENFELD., *et al*. "Les polykystoses hépatiques Principales complications et prise en charge". *Gastroentérologie Clinique et Biologique* 26 (2002): 1097-1106.
7. Qian Q., *et al*. "Sirolimus reduces polycystic liver volume in ADPKD patients". *Journal of the American Society of Nephrology* 19.3 (2008): 631-638.
8. Dmitrewski J., *et al*. "Case report: obstructive jaundice associated with polycystic liver disease". *HPB Surgery* 10 (1996): 117-120.
9. Gelu-Simeon M and Samuel D. "Aspects hépatiques de la polykystose hépato-rénale". *Postu* (2014).
10. Abascal J., *et al*. "Infection of hepatic cysts in polycystic disease". *World Journal of Surgery* 8.3 (1984): 424-425.
11. Zhang ZY., *et al*. "Polycystic liver disease: Classification, diagnosis, treatment process, and clinical management". *World Journal of Hepatology* 12 (2020): 72-83.
12. Vilgrain V. "Lésions kystiques du foie". *Gastroentérologie Clinique et Biologique* 25 (2001): B167- B177.
13. Debenes B., *et al*. "Kyste solitaire et polykystose hépatique de l'adulte. Cystadénome hépatique". Editions Techniques - Encycl Med Chir (Paris - France)". *Hépatologie* 7039.A18 (1992): 6.
14. Danaci M., *et al*. "The prevalence of seminal vesicle cysts in autosomal dominant polycystic kidney disease". *Nephrology Dialysis Transplantation* 13 (1998): 2825-2828.

Volume 10 Issue 4 April 2023