

Image Quiz for Surgeons

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The MELD Score Exception for Polycystic Liver Disease

G. Smira¹, E. Matei², L. Gheorghe¹, I. Popescu²

¹Center of Gastroenterology and Hepatology, Fundeni Clinical Institute, Bucharest, Romania

²Center of General Surgery and Liver Transplantation, Fundeni Clinical Institute, Bucharest, Romania

Rezumat

Boala polichistică hepatică - excepție de la scorul MELD

Boala polichistică hepatică este o afecțiune progresivă rară caracterizată prin creșterea volumului hepatic secundară unor chisturi multiple, cu simptome datorate dimensiunii ficatului precum și compresiei organelor învecinate. Cei mai mulți pacienți nu necesită tratament medical sau chirurgical. Pe de altă parte, hepatomegalia gigantă cu simptome severe care nu pot tratate medicamentos, necesită tratament chirurgical. Transplantul hepatic oferă singura opțiune curativă cu dispariția simptomelor legate de creșterea chisturilor și compresia viscerală secundară. Prezentăm cazul unui pacient tânăr cu boală polichistică hepatică izolată intens simptomatică al cărui volum hepatic total era de 23.200 cm³ și care provoca un handicap major fizic și social, și la care am considerat ca hepatectomia totală și transplantul hepatic pot oferi o șansă definitivă de tratament. În corelație cu datele din literatură, acesta este cea mai mare piesă de hepatectomie a unui pacient transplantat.

Cuvinte cheie: boala polichistică hepatică, excepție de la scorul MELD, transplant hepatic

Abstract

Polycystic Liver Disease (PLD) is a rare progressive disease characterized by increased liver volume due to many cysts, with symptoms related mainly to the size of the liver and the compression on adjacent organs. Most patients who have PLD require no medical or surgical intervention. On the other hand, massive hepatomegaly with severe symptoms which cannot be managed conservatively requires surgical procedures. Liver transplantation (LT) offers the only curative option for the relief of symptoms arising from cyst enlargement and compression of abdominal structures. We presented a rare case of a young man with highly symptomatic isolated PLD due to liver volume - 23,200 cm³, which provokes severe physical and social handicaps and we considered that only total hepatectomy and LT provides a chance of definitive treatment. To our knowledge this is the largest specimen from a PLD patient who was transplanted.

Key words: polycystic liver disease, MELD score exception, liver transplantation

Corresponding author:

Dr. Gabriela Smira
Center of Gastroenterology and Hepatology
Fundeni Clinical Institute, Bucharest
Sos Fundeni 258, 022328, Romania
E-mail: drsmira@yahoo.com

Introduction

Polycystic Liver Disease (PLD) is an inherited autosomal dominant condition characterized by the presence of multiple scattered cysts of biliary origin in the liver parenchyma (1). PLD often occurs in association with autosomal dominant polycystic kidney disease (ADPKD), but also exists as a distinct genetic entity in 10% of cases (2).

The first suggestions of an isolated form of polycystic

liver disease were made in the mid-1980's and confirmation that there was a distinct autosomal dominant polycystic liver disease (PLD) occurred in the late 1990's.

Most patients with PLD are asymptomatic. Despite impressive physical examination and radiologic findings, only a minority of patients with PLD will progress to advanced liver disease or develop complications as a result of massive hepatomegaly.

However, this minority of patients may become incapacitated by the disease, with symptoms of severe abdominal pain, massive abdominal distension, early satiety, supine dyspnea and chronic wasting with malnutrition.

They generally have preserved liver function and normal Model for End Stage Liver Disease (MELD) scores if they do not have renal involvement (3).

Liver transplantation (LT) is the final treatment for PLD.

Case report

A 39-year-old man with isolated polycystic liver disease was diagnosed in October 2009. The personal history of the patient included the appearance of a voluminous abdominal mass associated with weakness, early satiety and minimum anemia. He experienced gradual increase in abdominal fullness and pain and he was referred to our center six months later for transplant evaluation because of progressive symptoms due to cyst enlargement and compression. He has no remarkable familial history of PLD.

Clinical examination showed pale skin and mucosa, supine dyspnea, massive abdominal distention with severe abdominal pain, and advanced malnutrition.

The abdominal ultrasound examination showed massive hepatomegaly with multiple cysts of varying size throughout all segments of the liver, moderate ascites and normal kidney structure.

Further contrast-enhanced CT scan evaluation demonstrated diffuse polycystic liver with total hepatic volume of 23,320 cm³ and confirmed normal kidney aspect.

According to the Schnelldorfer classification (3) the patient was type D PLD with absence of preserved parenchymal sector, diffuse enlarged cysts and severe symptoms - fig 1 and 2.

The biological results showed mild anemia with normal liver function tests: Hb = 9 g/dl; Ht = 30%; ALT = 12,2 U/L; AST = 16,5 U/L; TBIL = 0.97 mg/dl; GGT = 42 U/L; ALP = 100 U/L; CHOL = 178; ALB = 4,5; PT = 86,76%; INR = 1,1 and normal renal function tests: CREAT = 0,86; BUN = 36 mg/dl. The Model for End Stage Liver Disease (MELD) Score calculated at that moment was normal - 7.

During hospitalization, the patient was completely investigated for LT with normal cardiovascular, neurological and endoscopic screening.

We considered that orthotopic LT represents the only curative option in our patient because massive hepatomegaly produced severe physical and social handicap, fatigue, clinically advanced malnutrition and poor quality of life leading to disability.

The indication for OLT was primarily related to abdominal



Figure 1. Preoperative aspect of the patient

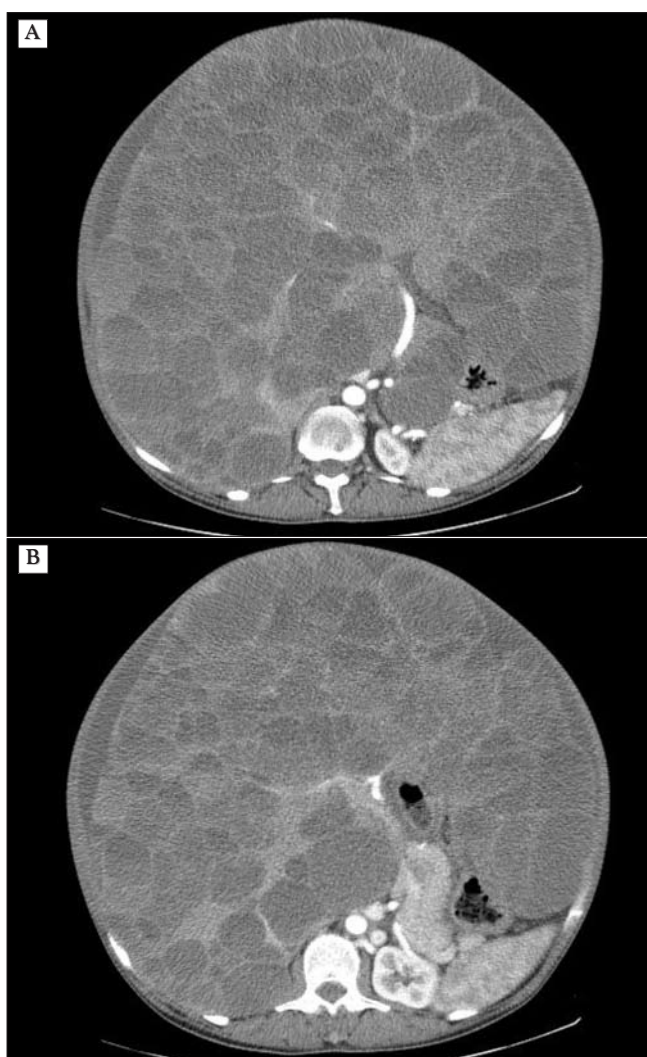


Figure 2. CT scan aspect showing a massive cystic liver enlargement



Figure 3. Gross appearance of the polycystic liver



Figure 4. Postoperative aspect of the patient

distention and pain due to massive organomegaly and not to liver failure that would increase the MELD score.

Due to the massive hepatomegaly, the hepatectomy was difficult. The retrohepatic inferior vena cava (IVC) was embedded in the liver so that the hepatectomy was performed en-bloc with the IVC according to the Starzl's method (4). No venous by-pass was required.

The caval continuity was performed by supra- and infra-hepatic cava-cavostomies followed by portal, arterial and biliary anastomoses.

The specimen weighed 23.2 kg. – fig. 3.

In the postoperative day 1 the patient was reexplored for peritoneal bleeding without obvious source. After this, the recovery was uneventful – fig. 4.

The patient was discharged after one month.

Discussion

Polycystic live disease is a rare autosomal dominant disease with a heterozygous germ line mutation characterized by an increased liver volume due to many (> 20) fluid-filled cysts. The development of PCLD appears to correlate with age, female gender and gravidity (4).

There are important differences in disease severity between men and women who are more severely affected. It seems that estrogens play a role in cyst development.

The patients are diagnosed between 30 and 40 years of age when they start to suffer from the increased liver volume and the pressure it exerts on other organs.

Most patients do not require treatment, but life-threatening complications related to massive hepatomegaly may occur including cyst infection, hepatic vein outflow obstruction, portal hypertension, cyst carcinoma and most importantly pain and/or chronic wasting with malnutrition (5).

Depending on the patient and the size and number of cysts, hepatic resection, cyst fenestration, or cyst aspiration with sclerosing agent instillation can be performed (6). However, these treatments are only palliative.

LT represents the complete and definitive treatment for highly symptomatic PLD; LT eliminates the disease and its complications and provides long-lasting relief of symptoms (7).

The indications for OLT are related to severe symptoms. However, timing of listing patients for LT is difficult to address. There is no definite sign or symptom that will trigger placing a patient on the liver waiting list.

Patients with PLD typically do not have any parenchymal disease that would increase the MELD score. A low MELD score puts them at a disadvantage for receiving a liver in a timely manner.

During the waiting period, these patients experience progression of symptoms and malnutrition, with decrease in quality of life and increase morbidity at transplantation.

Concerning this patient without family history, the progression of their disease was very accelerated with severe incapacitation.

The massive abdominal distension caused a marked social and professional impact. The liver volume represented 33 % of his body weight.

The largest series of PLD patients undergoing LT published by Kirchner et al. (8) comprises 36 patients, the largest specimen weighing 20 kg. This is the biggest polycystic liver according to the published data.

During LT there was no need for venous by-pass due to the secondary portal hypertension, and the embedded IVC required en-bloc resection.

Conclusion

Liver transplantation is the only curative treatment for selected patients with massive diffuse PLD.

In this time of organ shortage, using the current MELD system, exception scores should be considered for timely transplantation.

We presented a rare case of a male patient with the biggest polycystic liver volume which was successfully transplanted.

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