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Sharp liver excision under hepatic vascular exclusion in case of liver transplant for large polycystic disease. Case report of a new surgical technique☆

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ABSTRACT

INTRODUCTION: Polycystic liver disease is observed in 75–90% of patients with autosomal dominant polycystic kidney disease (ADPKD). ADPKD has a high prevalence of 1/1000. Hepatomegaly severely reduces quality of life and liver transplantation seems to be method of choice for many patients. Because of the rarity of this disease and the small number of symptomatic patients with massive hepatomegaly indicated for the transplantation, there is no standard approach for explantation of the liver.
CASE PRESENTATION: In our case, 57-year-old woman with massive hepatomegaly was treated with simultaneous split liver and kidney transplantation with bilateral nephrectomy.
DISCUSSION: For the native liver excision we used unique surgical approach – sharp liver transection under hepatic vascular exclusion. Because we experienced some cases with massive bleeding during the polycystic liver explantation, we decided to change the surgical approach. The technique offers limited blood loss and comfortable operation field exposure.
CONCLUSION: The giant polycystic liver could safely be explanted only using sharp transection hepatectomy under hepatic vascular exclusion. There is significant difference between blood loss in patients treated with or without hepatic vascular exclusion.

1. Introduction

Autosomal dominant polycystic kidney disease (ADPKD) is a multisystem and progressive disorder characterized by cyst formation and enlargement of kidney and other organs (liver, pancreas, spleen). Up to 50% of patients with ADPKD require renal replacement therapy by 60 years of age. Polycystic liver disease (PLD) is observed in 75–90% of patient suffering from ADPKD. Such patients generally do not suffer from hepatic insufficiency, their score of the "Model for End-Stage Liver Disease" (MELD) is normal or near normal. Pain in the abdomen, flank or back is the most common initial complaint and it is almost universally present in patients with ADPKD. Patients with advanced PLD may suffer from abdominal pain, hepatomegaly, inferior vena cava compression, anorexia, cyst infections, dyspnea, portal hypertension (rarely), and severely reduced quality of life in general. Patients may even die as the consequence of malnutrition [2]. Orthotopic liver transplantation (LTx) has been the only radical approach to cure the disease. LTx usually leads to excellent symptomatic relief, but it is a costly and an invasive procedure. Since some patients with combined polycystic kidney and liver disease also have renal insufficiency, combined liver-kidney transplantation is the preferred treatment option [3]. According to the evidence, the palliative disease-directed interventions (hepatic resection-fenestration procedure) foregoing liver transplantation are associated with increased risks of perioperative morbidity and mortality [4]. Almost 6000 LTx per year are currently performed in Europe, a number similar to that of the United States. However, benign liver tumors (mainly polycystic disease) represent less than 1% of indications in Europe [5]. The procedure was made in community hospital – Institute for Clinical and Experimental Medicine in Prague, Czech Republic. The work has been reported in line with the SCARE criteria [6].

2. Surgical technique

Our case was a 57-year-old Czech woman (body mass index 24 kg/m^2) with end-stage renal disease due to ADPKD and func-

Abbreviations: ADPKD, autosomal dominant polycystic kidney disease; LTx, liver transplantation; HVE, hepatic vascular exclusion; MELD, model for end-stage liver disease; PLD, polycystic liver disease; TVI, total vascular isolation; UCLA, University of California Los Angeles; IVC, inferior vena cava.

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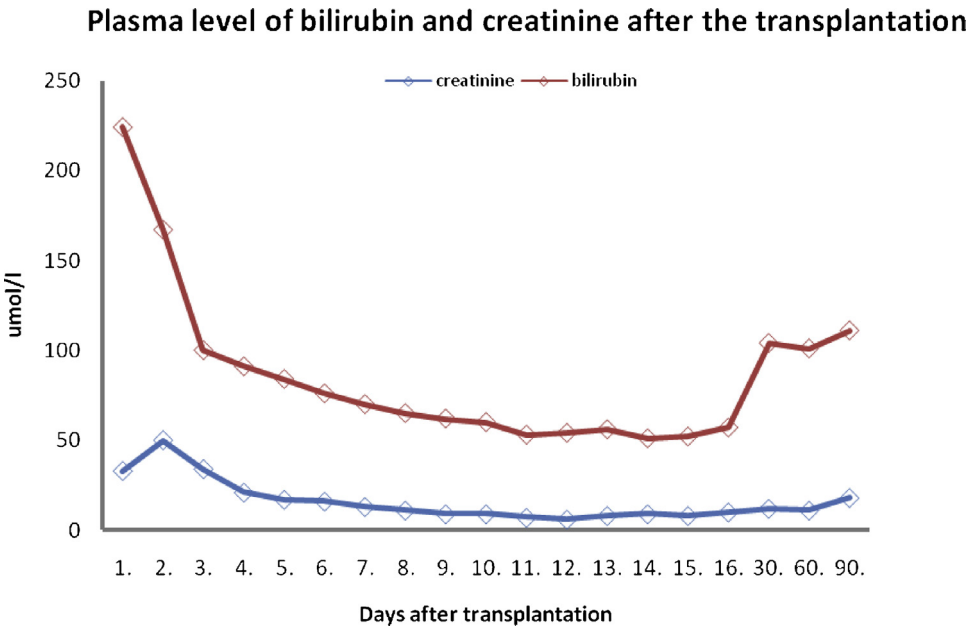

Fig. 1. The plasma level of bilirubin and creatinine after the transplantation (1st–16thday – hospital, 16thday – discharged, 30th, 60th, 90th day – out-patient visits).

tional abdominal pain syndrome caused by PLD. She was diagnosed with ADPKD with 39 years of age. The first sign was the abdomen pain. According to the family history, the disease was inherited from her mother. She has two children. One of them is also the carrier of the mutation. The patient was treated by symptomatic therapy only (antihypertensive therapy, calcium, calcidiol and allopurinol). After 18 years she was waitlisted for bilateral nephrectomy and simultaneous liver-kidney transplantation.

2.1. Phase 1–the liver explantation

The bilateral nephrectomy and simultaneous liver-kidney transplantation procedure was performed by the Head of Transplant Surgery Department (Institute for Clinical and Experimental Medicine) via full size midline incision. Fig. 2A. The liver manipulation was not easy because of the massive polycystic disease. The first step was preparation of the structures in the liver hilum; all structures except portal vein were tied off and transected. After that, inferior vena cava (IVC) above and below the liver was dissected, portal vein was transected and IVC above and below the liver finally cross-clamped. In such situation of total vascular liver exclusion, the liver was ready for excision without the risk of large blood loss [7]. Use of the ordinary scalpel, the liver was sharply transected into two parts along the right side of IVC. For safety during cut of liver parenchyma, we used cotton band behind the liver along the right hand side of IVC. Fig. 2B. The band led us with the liver transection. Fig. 2C. After removing the right lobe Fig. 2E. we excised the left lobe and over sewed the retro hepatic veins including left mid and right hepatic vein. Fig. 2D. The liver weighed 6 862 g, whole retro hepatic IVC was preserved. Fig. 2F. The anesthesia used was standard (combined anesthesia – sufentanil, propofol and rocuronium bromide). During the explantation it was necessary to administer low dose of the norepinephrine (0.05–0.1 ug/kg/min).

2.2. Phase 2–the liver transplantation

We used right hepatic lobe from classic split liver procedure (extended right hepatic lobe) and the piggyback technique – the recipient vena cava was preserved in situ and transplantation occurred with side-to-side cavo-caval-anastomosis [8]. After the IVC, the portal vein, hepatic artery and bile duct anastomosis were done.

2.3. Phase 3–nephrectomy of polycystic kidneys

After the liver transplantation, both of polycystic kidneys were removed from retroperitoneal space. The left one weighed 940 g and the right one 886 g.

2.4. Phase 4–the kidney transplantation

The kidney graft form the same deceased donor had simplex anatomy. The kidney transplant was performed trans-peritoneally, using external iliac vessels on right hand side and also L-shape peritoneum flap to fix the kidney graft in the retroperitoenum. The uretero-cysto-anastomosis was made using the double-J stent.

2.5. Phase 5–transitory closure

In many combined liver and kidney transplant cases, because of the risk of hematoma formation, we tend to pack the abdomen with swabs and close only the skin for 24–48 h. We did the same in this case.

2.6. Phase 6–second look

Next day, we did second look with extraction of the surgical drapes and definitive wound closure.

Plasma levels of creatinine and bilirubin were continuously falling, patient received conventional triple immunosuppression including steroids, calcineurin inhibitor and antimetabolite. Fig. 1. Protocol ultrasound and Doppler of kidney and liver grafts have proven optimal flows in all the graft vessels. Our patient was discharged home on day 16 after the surgery with normal laboratory markers and excellent functions of both grafts. The pathologic examination showed polycystic tissue of liver and kidney without malignancy. The patient is having been followed up at Out-patient department of the Institute for Clinical and Experimental Medicine with no signs of rejection or the organs failure.

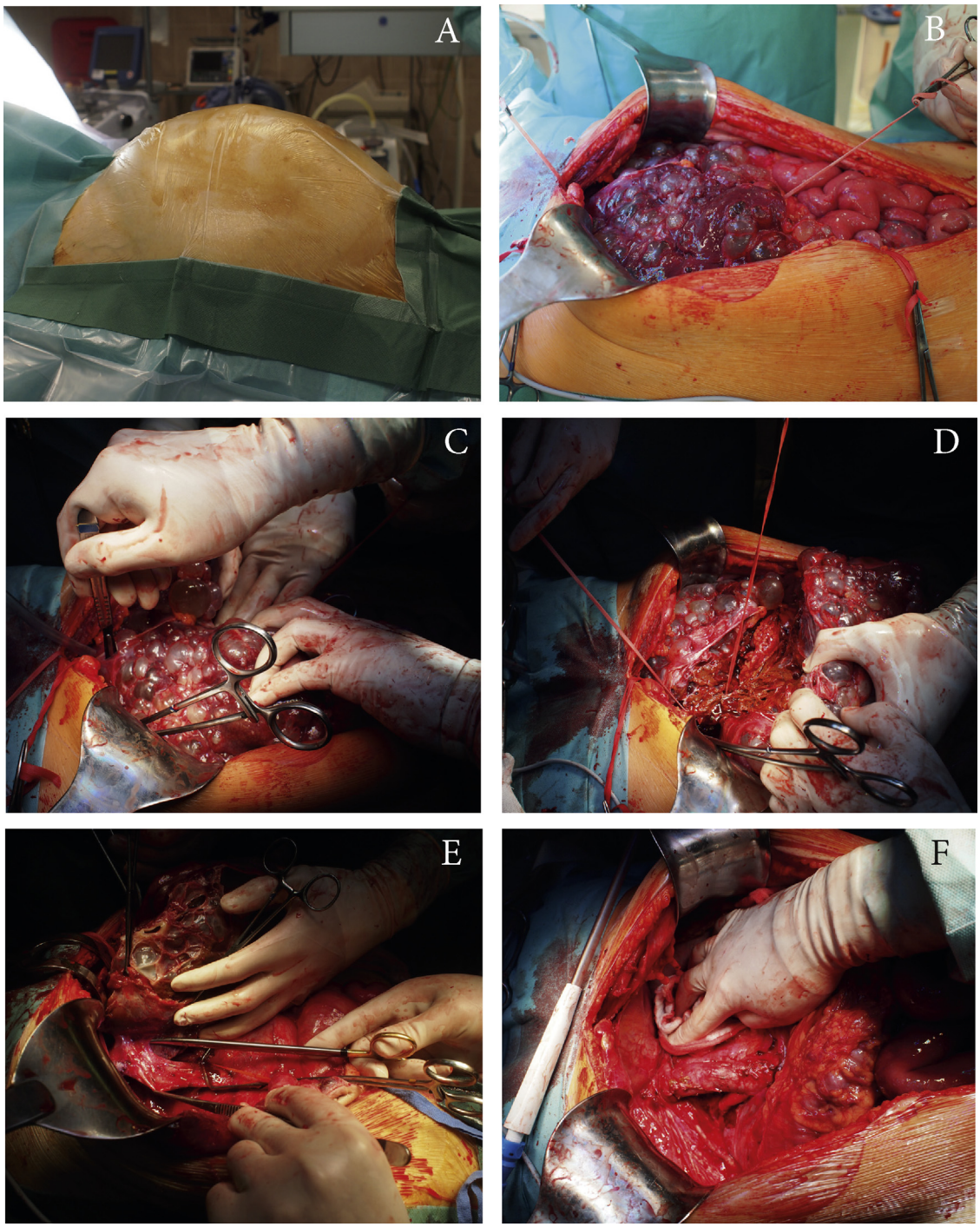

Fig. 2. A – The huge abdomen full of polycystic liver and kidneys; B – The cotton band behind the liver along the Vena Cava Inferior; C – The hepatic vascular exclusion with clamps and sharp liver transection; D – The rest of hepatic tissue above the cotton band; E – The middle and left hepatic vein preparation; F – The abdominal cavity after the liver explantation and preserved Vena Cava Inferior.

3. Discussion

We report here the case of 57-year-old woman with massive hepatomegaly and renal failure, who was treated with simultaneous split liver and kidney transplantation with bilateral nephrectomy. For the large polycystic liver excision we used unique surgical approach – sharp liver transection under hepatic vascular exclusion. The first simultaneous liver and kidney transplantation was conducted by the UCLA (University of California, Los Angeles) Medical center in 1998 [9]. It was performed in a middle-aged woman having adult polycystic liver and kidney disease. In 1990, the Starzlís group was the first to succeed liver transplantation on 4 female patients with severe complications of hepatomegaly caused by PLD [10]. Hepatomegaly severely reduces quality of life and orthotopic liver transplantation has been the only approach to cure. The palliative disease-directed interventions (hepatic resection-fenestration procedure) are just transitional solution and if foregoing the liver transplantation they are associated with increased risks of perioperative morbidity and mortality [4].

Total vascular isolation (TVI) of the liver is used during parenchymal transection undergoing hepatic resection for large tumors located near hilar structures, hepatic veins, or IVC. The TVI can be achieved by clamping the suprahepatic and infrahepatic VCI and porta hepatis, with or without aortic occlusion. This method, conducted with concomitant occlusion of supraceliac aorta, was first described by Heaney and co-workers in 1966 [7]. Classic hepatic vascular exclusion includes portal triad clamping and clamping of the infra- and suprahepatic VCI [11]. Clamping of the VCI results in major hemodynamic disturbances requiring active intraoperative anesthetic management [7].

The hepatic vascular exclusion (HVE) may be required for patients with parenchymal bleeding, often caused by penetrating juxtahepatic inferior vena cava injuries [12] and those with prior hepatic resection, polycystic liver disease or hepatic hemangiomas [13]. Furthermore, this surgical alternative allows the adequate vascular control in selected cases. For instance, it was used in the new anterior cavoplasty after pediatric liver transplantation [14]. During some of our cases we experienced massive bleeding during the native liver explantation, so we decided to use the HVE as a standard surgical approach with aim to reduce blood loss.

Prior studies showed that 5-year survival after deceased donor liver or combined liver-kidney transplantation is around 80% with good quality of life [3,15]. In the case of severe hepatomegaly and renal failure, the combined transplantation offers the possibility to cure both the ADPKD with PLD without raising the disadvantage (for example infectious complications) of immunosuppression incurred with single organ transplantation [16]. According to a study from Japan, the presence of incidental tumors in removed polycystic kidneys is very rare. On pathologic examination the renal cell carcinoma was found in 2 from 510 patients [17]. To the best of our knowledge, there are no studies which investigated the incidental tumors of removed polycystic liver.

In our center, symptomatic PLD is always indicated to orthotopic liver transplantation. Since some patients with combined polycystic kidney and liver disease also have renal insufficiency, combined liver-kidney transplantation is the preferred treatment option [3]. In our center, there are 120–140 liver transplantations performed per year [18]. Of those, approximately 5 cases per year are indicated for combined simultaneous liver and kidney transplantation for ADPKD with PLD. According to literature, there are currently almost 6000 liver transplantations performed per year in Europe, a number similar to that of the United States. Benign liver tumors (mainly polycystic disease) represent also less than 1% of indications [5].

Because of the rarity of this disease and the small number of symptomatic patients with massive hepatomegaly indicated for the transplantation, there is no standard approach for explantation of the liver. In our experience, the manipulation with enlarged polycystic liver might be difficult and may cause large blood loss during the explantation. In our center, 12 patients with PLD were transplanted from January 2015 to December 2016.The last two patients were treated using HVE. The average blood loss in patients without HVE was 6650 ml including two patients suffering from massive bleeding. One of those patients had disruption of VCI related to difficult native liver manipulation. In this case the IVC injury led to “mors in tabula”. The second patient had portal vein disruption with almost 31000 ml of blood loss. After adopting the HVE technique, the last two patients had the average 500 ml of blood loss and no severe complications during the hospital stay.

We therefore decided to use the described sharp liver transection technique in total vascular exclusion. The described technique has been performed routinely since the mentioned case. The technique offers limited blood loss and comfortable operation field exposure. To the best of our knowledge, we are the first who have used the HVE with sharp transection of hepatic parenchyma in case of liver transplantation for ADPKD with PLD.

4. Conclusions

Polycystic liver disease is frequently associated with adult polycystic kidney disease and renal failure. Symptomatic hepatomegaly is best treated with liver transplantation. In our case, simultaneous kidney and split liver transplantation together with bilateral nephrectomy was performed. The giant polycystic liver could safely be explanted only using sharp transection hepatectomy under hepatic vascular exclusion. There is significant difference between blood loss in patients treated with or without hepatic vascular exclusion.

Conflicts of interests

The authors declare no conflicts of interests.

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Ethical approval

The ethical approval has been exempted as it was not neccessary in this case.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images of transplantation procedure. The manuscript does not contain any personal information. A copy of the written consent is available for review by the Editor-in-Chief of this journal on regest.

Author contribution

Filip Thieme – data collection and interpretation, writing the paper

Jiri Fronek – writing the paper

Guarantor

Ass. Prof. Jiri Fronek MD, PhD.

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