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Simultaneous liver-renal grafting for polycystic disease

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Abstract Our purpose was to describe the first in our country and fourth worldwide simultaneous liver and renal grafting for polycystic disease.

Key words Liver-renal-graft
Polycystic disease

Introduction

Polycystic renal disease is known to be associated with polycystic liver disease in over 75% of cases. Liver cysts increase in number and volume as the subjects grow older, the incidence being higher in females. Renal cysts usually subside following initiation of dialysis, but there is increasing evidence that the opposite is the case in liver cysts, which present with a number of complications [1–3]. Portal hypertension, jaundice, external pressure to the inferior vena cava, cachexia and hepatic insufficiency are eventual consequences of long-term liver cyst expansion [4, 5]. Debilitating disease has also been described in several cases. Owing to the unsatisfactory effects of more conservative surgical procedures, simultaneous renal liver grafting is seen as an alternative solution and has been successfully performed in the USA and UK [6, 7].

Case report

Our case report refers to a 46-year old female suffering from end-stage renal failure due to polycystic disease with recent initiation of dialysis. The liver parenchyma was virtually all replaced by large and small cysts, with blood stasis in the portal axis and splenomegaly. Physically, the subject has marked abdominal distension, manifested extreme muscular weakness and appeared cachectic. A short time later (24 December 1991) the patient was subjected to emergency bilateral nephrectomy (acute abdomen owing to inflammation of the right kidney), elective total hysterectomy (uterine fibromyomas) and appendectomy. The simultaneous organ transplantation was performed on September 1992.

Preoperative laboratory investigation were as follows: Ht = 24%, leucocytes = 3000 cells/μl, PLT = 100 000 cells/μl, creatinine 6 mg%, SGOT = 20 IU, SGPT = 8 IU, γGT = 29 IU, alkaline phosphatase = 76 IU, PT = 12.9″/12.1″, total bilirubin = 0.55 mgr/ml, globulins = 2.9 gr%, albumins = 3.9 gr%, glucose = 105 mg%.

Bone marros microscopy was normal.

Triplex test showed dilatation of the portal vein (diameter ≧ 18 mm)

Blood group: A RH (+)

HLA typing: A1, A9 (W23)
B8 (W44), B15 (W63), BW4, CW4
DR7, DRW53, DGW2

The donor was a 25-year old male, the victim of a road traffic accident, whose brain death ensued from severe brain injury. His data were as follows:

Blood group: 0 RH (+)

HLA typing: A1, A3, B35, B16 (39), CW4, BW6
DR6 (15), DR7, DRW52, DRW53, DQ1, DQ2.

Multiple organ harvest procedure

The heart, liver and kidneys were procured from the same donor. The abdominal harvesting technique was as first described by the

team at the University of Cambridge. For organ irrigation and preservation, the University of Wisconsin solution was utilized.

The recipient's operation

A bilateral subcostal incision with medial extension superiorly was selected. The upper abdominal adhesions were lysed and the liver was mobilized by dividing its ligaments. All elements of the hepatoduodenal ligament were separated from adjacent tissues and then the infra/suprahepatic segments of the inferior vena cava (IVC) were carefully prepared. The right epinephric vein was ligated and divided. The common bile duct (CBD), hepatic artery and portal vein were all divided close to the liver. Craaford clamps were used to clamp the infrahepatic IVC and a Debakey clamp for the suprahepatic IVC. Finally, the IVC was divided and the diseased liver was removed.

The donor's liver was removed from the ice-cold environment and was placed into the abdominal cavity. The suprahepatic IVC anastomosis was started first and that of the portal vein second. Completion of portal vein anastomosis permitted removal of all portal clamps and blood was allowed escape through the infrahepatic IVC. The infrahepatic IVC stump of the graft was clamped again, enabling continuation of liver perfusion.

As soon as the infrahepatic IVC anastomosis was completed, the entire hepatic venous circulation was restored. The hepatic artery stumps and CBD stumps were anastomosed end-to-end. A T-tube was placed in the reconstructed CBD to drain the bile. The liver operation was completed with closure of the surgical incision after haemostasis had been checked and drainages had been placed supra- and infrahepatically on the right side and subphrenically on the left side.

Anastomotic time – liver transplantation

Suprahepatic IVC anastomosis	12 min
Portal vein anastomosis	6 min
Infrahepatic IVC anastomosis	9 min
Hepatic artery anastomosis	13 min
Common bile duct anastomosis	19 min

Anastomosis time – kidney transplantation

Arterial anastomosis	9 min
Venous anastomosis	9 min
Ureterovesical anastomosis	11 min
Total warm ischaemia time	0 min
Total cold ischaemia time	7 h and 30 min
Total operation time	7 h

Intraoperative administration of blood products

Whole blood	22 units
Fresh frozen plasma	13 units
Thrombocells	6 packs
Plasma protein fracture	4 units (200 ml each)

Postoperative course

Following completion of the operation, the patient was transferred to the intensive care unit intubated and she remained on a respirator for 2 days. Continuous monitoring and immediate correction of any parameter concerning cardiac, respiratory, hepatic and renal function was possible. Most important was the direct monitoring of the renal graft function (> 4000 ml/1st 24 h) that enabled a convenient management of fluids and electrolytes. Blood coagulation profile on the 1st postoperative day was: PLT 170000 cells/μl, PT 16.3″/12.1″, aPTT 33.4.

On the 9th postoperative day, the patient manifested symptoms and signs of acute rejection, confirmed histologically by performing needle biopsy examination. Management was possible by means of 1000 mg methylprednisolone per day for 3 days. The protocol of antibiotics consisted of ampicillin (1 g per 4 days), ciprofloxacin (500 mg per 3 days) and metronidazol (500 mg per 3 days). The patient was discharged from hospital on the 28th postoperative day with excellent renal-liver function.

Discussion

The increased survival of patients with polycystic disease that has been the result of dialysis has uncovered the seriousness of polycystic liver complications. Grunfield et al., Van Etrecum et al. and Turnage et al. have recently referred to these complications in detail [1, 8, 9]. Liver function is maintained at a good level until an insult, frequently septic in origin, disturbs it and produces fast aggravation of liver biology and corresponding deterioration in the patient's condition.

Until recently, liver transplantation was not even considered a therapeutic option. However, as survival of liver graft recipients has improved and exceeds 85% in the 1st postoperative year, the option of liver grafting performed early in the course of the disease before the onset of dramatic sequelae has gained significant support. Conservative operations have been shown to result only in temporary relief, at the cost of extensive abdominal adhesions [10]. A later decision to subject these patients to liver grafting should take into account the additional technical difficulties of such an intervention, owing to the multiple adhesions developed.

In some patients the huge enlargement of the liver cysts results in serious respiratory problems that are further aggravated by superimposed infections. Debilitating disease, characterized by physical exhaustion and confinement of physical activity, is not infrequently encountered. In such cases, simultaneous grafting of the liver and kidney to offer a definitive treatment has been proposed. Combined liver-renal grafting for polycystic disease has been performed by the pioneers of organ transplantation, T. Starzl [6] and Sir Roy Y. Calne [7].

In the small series of three cases worldwide, we offer one more that was performed upon a patient in whom all requirements were met. Success of such a procedure enables the patient to overcome all of his/her physical problems and, thus, renders him/her an active member of society again. Our patient is in good health, both clinically and biochemically 1 year postoperatively, and leads a happy life.

In conclusion, despite the limited number of cases worldwide the good results reported in the literature seem to consolidate the role of liver transplantation in the management of complicated polycystic disease.

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