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Case Report

Re-Transplantation in a Pediatric Patient with Hepatorenal Syndrome: A Case Report

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Abstract

Hepatorenal syndrome is common in patients with decompensated cirrhosis with severe ascites. Liver transplantation (LT) is the only viable option. Combined Liver-Kidney transplantation (CLKT) is recommended in cases with irreversible renal damage. Recovery of the renal functions following LT is common. We aimed to present a pediatric patient with biliary atresia with full recovery of renal functions after retransplantation for chronic rejection and biliary obstruction.

Living donor LT (LDLT) was performed on a 4-year-old female patient due to biliary atresia 6 years before the admission to our department. The patient suffered chronic anastomotic stenosis, cholangitis, and subsequent chronic rejection. On admission, the patient was severely icteric with massive ascites. The renal function was poor with no urine output and required hemodialysis. CLKT was planned but only LDLT could be performed due to deterioration of the patient's condition. However, the renal function improved dramatically in the postoperative period and the kidney transplant was not performed. The patient had an uneventful postoperative period.

Hepatorenal syndrome, can resolve following LT. The patients who have a high risk of chronic liver failure should be determined before the LT procedure to plant a CLKT and to allocate insufficient organ resources.

Keywords: Hepatorenal syndrome, liver transplantation, retransplantation.

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Cirrhosis is the result of irreversible damage to the liver tissue that causes changes in the liver microarchitecture that is defined by extensive fibrosis. The only cure for end-stage liver disease is liver transplantation.[1] Hepatorenal syndrome (HRS) is a complication of severe decompensated cirrhosis. The physiopathological mechanisms that lead to HRS include splanchnic vasodilation, reduced systemic vascular resistance and central hypovolemia, inflammation, altered cardiovascular reflexes, and intraabdominal hypertension related to severe ascites. The prognosis of HRS is poor and mortality reaches 80%.[1,2]

Diagnosis depends on clinical suspicion since no definitive clinical and radiologic diagnostic test exists. HRS can be suspected in a patient with cirrhosis with elevated serum creatinine, oliguria, and low urinary sodium concentration.[4,5] There are no proteinuria or histologic changes and for this reason, the renal failure is reversible.[4,5]

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Medical therapy of HRS includes treatment with a combination of albumin and posterior pituitary lobe hormone analogs such as terlipressin or vasopressin to obtain potent vasoconstriction. Medical treatment is a bridging therapy until definitive therapeutic options are performed. Definitive treatment includes liver transplantation (LT) or combined kidney and liver transplantation (CKLT).[2,5] Spontaneous recovery of the renal function following LT is common.[6] We need criteria to predict whether patients can recover from HRS after LT. This is very important for allocating scarce resources of deceased donor organs to patients who need them.

In the present study, we report a case of a 10-year-old female patient with a prior history of LT due to biliary atresia. She lost the graft due to chronic rejection and had HRS that required hemodialysis at the time of retransplantation. Following the procedure, the renal function of the patient recovered completely.

Case Report

A 10-year-old girl was admitted to our department with abdominal distension, decreased urine output, and deterioration of her general health. The patient had undergone a Kasai procedure for biliary atresia and received a living donor liver transplantation when she was 4 years old. She suffered from stenosis of the biliary anastomosis for which multiple sessions of percutaneous transhepatic biliary catheterization were performed. She suffered from ascending cholangitis and received a broad spectrum of antibiotics. During these episodes, the immunosuppressive therapy was discontinued and she gradually developed graft rejection. Nevertheless, all attempts failed and the patient gradually developed fibrosis of the graft due to poor bile drainage and chronic rejection of the liver graft.

The physical examination upon admission showed that the condition of the patient was deteriorating, and she was severely icteric. The abdomen was distended due to severe ascites and caput medusae were observed. She showed severe sarcopenia and was almost immobile. The patient had severe renal failure with no urine output. She had to receive regular intervals of hemodialysis.

The computerized tomography scan showed that the bile ducts were severely dilated forming lacunar changes, and the liver parenchyma was destroyed (Fig. 1). The laboratory values at admission showed that she had severe renal failure (Table 1). A liver biopsy showed severe fibrosis and destruction of the microarchitecture of the liver parenchyma.

Initially, CKLT was planned. Her mother was a suitable candidate to be a partial liver donor. Her grandmother was a suitable kidney donor. The Child Pugh-Turcot score of the patient was 10 and the PELD score was 23. The perioperative of the patient deteriorated rapidly and our team decided to go through first with the liver transplantation and postpone the renal transplant after the condition of the patient improved. A left lateral lobe transplantation was successfully performed, and the patient required five sessions of hemodialysis in the postoperative period and renal

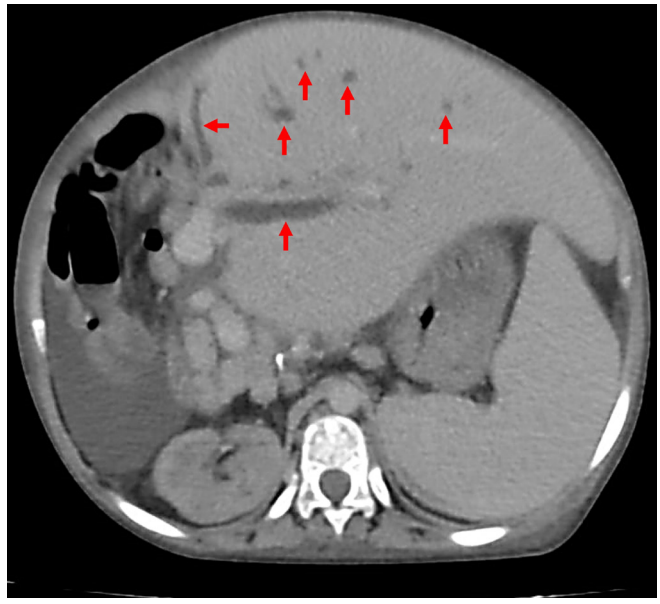

Figure 1. The computerized tomography sections of the liver showing dilated bile ducts and destruction of the normal architecture of the liver (The red arrows show the dilated bile ducts). The contours of the liver are rough indicating chronic liver disease.

Table 1. Laboratory values of the patient at admission and postoperative 9th month.

Laboratory values	Admission	Posttransplant 9th month
ALT	26	32
AST	37	19
GGT	127	29
ALP	194	245
T.Bil	14.8	1.1
D.Bil	11.3	0.22
Alb	2.9	3.9
PTT	15.7	14
INR	1.43	0.99
BUN	100	36
Cr	3.71	0.4

Abbreviations (normal value ranges) of the laboratory parameters: ALT: Alanine aminotransferase (1-40 IU/mL); AST: Aspartate aminotransferase (1-40 IU/mL); GGT: Gamma Glutamyl transferase (1-36 IU/mL); ALP: Alkaline Phosphatase (100-400 IU/mL); T.Bil: Total Bilirubin (<1.2 mg/dL); D.Bil: Direct Bilirubin (<0.2 mg/dL); Alb: Albumin (3-5 g/dL); PTT: Prothrombin Time (>12 sec); INR: International normalized ratio (0.8-1.5); BUN: Blood urea nitrogen (8-36 mg/dL); Cr: Creatinine (0.5-1.1 mg/dL).

functions gradually improved and urine output restored. Our team decided not to perform kidney transplantation. Her postoperative follow-up was uneventful and laboratory values on the postoperative 9th month are summarized in Table 1 showing full recovery of the renal functions. She is currently in the postoperative first year and there is no problem in her post-transplant controls.

Discussion

HRS is a serious complication of severely decompensated cirrhosis. If there is a strong suspicion of HRS, medical treatment should be started rapidly.[7,8] The interval between the diagnosis and LT is a risk factor for renal dysfunction. Prolonged periods of HRS that require renal replacement therapy may indicate permanent kidney damage.[7] LT is the gold-standard treatment for HRs, regardless of the response to medical therapy.[2,5,7] Impaired renal function while awaiting transplantation has been shown to increase morbidity and mortality after liver transplantation because ongoing renal dysfunction following LT is common in this case.[6-9] The risk factors for refractory HRS are resistance to medical therapy and the presence of underlying chronic renal disease that may be suspected in patients with diabetes, hypertension, anatomic problems in the kidneys observed during imaging, and the presence of proteinuria (>2 g/day).[10] Recovery of the renal functions following liver transplantation may be prolonged to a year after the procedure.[6,11] Determination of the patients that will recover is important for the allocation of the limited source of organs and also the prevention of unnecessary renal transplant procedures.

In the present study, our patient suffered from liver failure due to chronic rejection and she was also taking tacrolimus as an immunosuppressive therapy. A common side effect of tacrolimus is renal toxicity. Both liver failure and the use of tacrolimus played an important role in the development of HRS in our patient. We have performed a thorough literature search and have seen no information about concomitant chronic rejection and HRS in pediatric patients undergoing living donor liver transplantation. Our case report is unique because we report a pediatric patient suffering chronic rejection and HRS that resolved following liver retransplantation.

In conclusion, LT is the gold standard treatment for patients with end-stage liver failure and HRS. If the duration of HRS is prolonged, patients may suffer from irreversible renal damage. These patients require CLKT. The patients with refractory HRS should be determined before the LT procedure to plan the necessary therapeutic option.

Disclosures

Informed consent: Written informed consent was obtained from the patient for the publication of the case report and the accompanying images.

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