Primary liver transplantation for biliary atresia after percutaneous transhepatic cholangio-drainage

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A B S T R A C T

We herein report a case of primary living-donor liver transplantation (LDLT) for delayed diagnosis of biliary atresia under percutaneous transhepatic cholangio-drainage (PTCD). The case was a 10-months old female infant who was born in other Asian country and had been treated for cytomegalovirus hepatitis. After she moved to Japan at 9 months of age, she visited the referring hospital because of high fever. Severe jaundice and liver dysfunction were detected, and she was transferred to our hospital. Because of intrahepatic bile duct dilatation detected by computed tomography (CT), a PTCD tube was inserted to reduce jaundice, and liver biopsy was performed under general anesthesia. CT and pathologic findings led to the diagnosis of biliary atresia with liver cirrhosis. The patient was scheduled to undergo primary LDLT without Kasai portoenterostomy because of the progression of irreversible liver cirrhosis. While waiting for improvement of the severe fatty liver in her mother, a candidate donor, she underwent bile juice re-administration via the nasogastric tube. During this period, the total bilirubin did not increase above that at admission, and the albumin gradually improved. She underwent LDLT at 15 months of age, and her postoperative course was uneventful.

Rhu et al. [1] reported that a >60-day delayed diagnosis of biliary atresia (BA) is a potential risk for a bad prognosis involving the native liver. Chardot et al. [2] also reported that performance of Kasai portoenterostomy (PE) after 3 months of age is justified in only select patients without advanced liver disease for whom liver transplantation should not be delayed. Thus, in general, in extremely late-diagnosed patients, primary living-donor liver transplantation (LDLT) can be considered if the liver is already cirrhotic [3].

Some reports concluded that donors with severe fatty liver are not good liver donor candidates because of high complication rates [4–6]. Recently, Oshita et al. [7] reported that the use of diet-treated donors with steatotic livers for LDLT is feasible with respect to donor safety and recipient outcome.

We herein report the first case of successful primary LDLT without Kasai PE at the age of 15 months after percutaneous transhepatic cholangio-drainage (PTCD). In this case, the patient’s age at diagnosis for BA with cirrhosis was 9 months. While the donor’s fatty liver was being improved by dietary therapy, successful management of the patient’s nutritional status by re-administration of bile juice and vaccination were performed.

1. Case report

A 10-month-old female infant was admitted to our hospital with jaundice and liver dysfunction. She was born in another Asian country and had been treated for cytomegalovirus (CMV) hepatitis. After she moved to Japan at 9 months of age, she visited the referring hospital because of a high fever. Severe jaundice and liver dysfunction were found, and she was transferred to our hospital for further examination and treatment.

At admission to our hospital, her vital signs were within their normal ranges with a low-grade fever. On physical examination, she had a slightly greenish face and mild hepatosplenomegaly. Defecation was normal, but her stool was always cream- or white-colored with negative fecal bilirubin. Laboratory data showed severe jaundice, markedly elevated liver transaminase [total bilirubin, 8.9 mg/dL; aspartate aminotransferase (AST), 163 IU/L; alanine aminotransferase (ALT), 85 IU/L; and malnutrition (albumin, 2.3 g/dL; cholinesterase, 69 IU/L)]. Coagulation times were slightly prolonged (activated partial thromboplastin time, 56.6 s; prothrombin time, 14.8 s). A CMV test showed a past infectious pattern [CMV
immunoglobulin M (IgM), negative; CMV immunoglobulin G (IgG), positive. Because of intrahepatic bile duct dilatation detected by computed tomography (CT), PTCD tube insertion was planned to reduce jaundice. After verbal and written informed consent was obtained from her parents and safe levels of both coagulation parameters and platelet counts were confirmed, PTCD drainage and liver biopsy were performed under general anesthesia. The laboratory findings and pathologic findings, including complete bile duct obstruction at the hepatic portal lesion as revealed by cholangiography (Fig. 1A), no evidence of a space-occupying lesion at the hepatoduodenal ligament on CT (Fig. 1B), and progression of hepatic fibrosis, led to the diagnosis of BA with liver cirrhosis (Fig. 1C).

She was scheduled to undergo primary LDLT without Kasai PE because of the progression of irreversible liver cirrhosis and under the consideration that there are insufficient brain-dead donors in Japan. Oral and written informed consent was obtained from her parents. However, her mother, who was the only candidate liver donor in her family, was not matched because of severe fatty liver at the time of PTCD tube insertion. We proposed to the patient’s family that her mother would try to improve her fatty liver by dietary and exercise therapy for a while. The patient had been undergoing bile juice re-administration from the PTCD using a nasogastric tube prior to the LDLT.

The time course of laboratory data from admission to LDLT is shown in Fig. 2. The mother's fatty liver improved as a result of dietary and exercise therapy for half a year. The total bilirubin did not increase above that at admission, and the albumin gradually improved. There was no worsening in the total bilirubin level, Pediatric End-Stage Liver Disease (PELD score), or prothrombin time-international normalized ratio (PT-INR) during this period. She had also undergone vaccination during this period, including that against mumps, measles, varicella, hemophilus influenza, and pneumococcus. The patient underwent LDLT at the age of 15 months. Intraoperative cholangiography through the gallbladder revealed type 3 BA (Fig. 1D). The pathological findings of the resected liver, namely the progression of hepatic fibrosis, were similar to those of the previous liver biopsy at 6 months before LDLT.

After LDLT, her jaundice was dramatically improved with no evident rejection of the liver allograft. Postoperative graft function was excellent, and no major complications occurred.

2. Discussion

There are three potential advantages of PTCD tube insertion for delayed diagnosis of BA with irreversible cirrhosis, as in the present case. First, in the present case, judging from the pathological findings of the liver at the time of PTCD insertion and at LDLT, PTCD tube insertion had a possible suppressive effect on the progression of liver cirrhosis. This suppressive effect provided her with a window of opportunity for vaccination before performing LDLT. Moreover, although Japan is lacking in brain-dead donors, this additional time allowed her mother (donor) to improve her fatty liver before transplantation. Second, after insertion of the PTCD tube, the patient’s nutritional status was improved step by step with the re-administration of bile juice via the nasogastric tube. Her albumin level had been elevated and maintained at >3.0 g/dl from 1 month after the PTCD insertion. Some authors have reported that nutritional support has a beneficial effect on preventing perioperative infections associated with liver transplantation [8,9]. In this case,
the perioperative course was uneventful and without infectious complications. Third, in this case, primary LDLT without Kasai PE has the advantage of no surgery-related intra-abdominal adhesion operation compared with LDLT after Kasai PE. Sandler et al. [10] reported that liver transplantation following Kasai PE was associated with a higher blood transfusion volume at the time of the operation and a higher rate of postoperative bowel perforation compared with liver transplantation without Kasai PE. Sugawara et al. also reported that if the number of previous PEs was twice or more before the LDLT for BA, the operation time tended to be longer, the volume of blood loss tended to be greater, and vascular complications tended to be more frequent because of severe intra-abdominal adhesions at the time of LDLT [11].

3. Conclusion

Based on these results and those of a similar case (concomitant with intrahepatic bile duct dilatation in BA), PTCD insertion without PE during the waiting period for LDLT might be an acceptable therapeutic option because of the above-described advantages, especially in countries such as Japan, in which the ability to perform a liver transplantation with a brain-dead donor is lacking.

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