Liver Transplantation for Caroli Disease

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ABSTRACT

Caroli disease is a rare congenital disorder characterized by multifocal, segmental dilatation of intrahepatic bile ducts. Patients with Caroli disease who have recurrent bouts of biliary infection, particularly those who also have complications related to portal hypertension may require liver transplantation. In liver transplant ward of Shiraz University of Medical Science we had 4 patients with Caroli disease who were transplanted. Herein, we describe the demographic characteristics and post-transplant course of the patients.

These patients presented with liver failure, recurrent cholangitis and portal hypertension sequelae unresponsive to medical treatment. The mean age of patients was 24.5 (range: 18–36) years, the mean MELD score was 17.5 (range: 11–23), three patients were female; one was male. All of the patients had good post-transplantation course except for one patient who developed post-operative biliary stricture for whom biliary reconstruction was done.

KEYWORDS: Caroli Disease; Transplantation; Bile Ducts, Intrahepatic; Hypertension, Portal; liver failure; Cholangitis

INTRODUCTION

First described by Jacques Caroli in 1958, Caroli disease is a rare congenital condition characterized by non-obstructive saccular or multifocal segmental dilatation of the intrahepatic bile ducts [1]. The complex form of Caroli disease, the so-called “Caroli syndrome,” is associated with congenital hepatic fibrosis and is less common than Caroli disease [2].

The clinical features of Caroli syndrome in-
The objective of the present study was to assess the demographic characteristics and post-operative course of patients with Caroli disease who underwent liver transplantation in Shiraz University of Medical Sciences transplantation ward.

MATERIALS AND METHODS

During a period of 15 years from 1995 to 2010, 943 liver transplantation were done in our center. A retrospective study was performed on all liver transplantation patients who had undergone operation from 1995 to 2010 to find the cases of Caroli disease. Among the patients we found four who had Caroli disease; they were evaluated for the complications of Caroli disease. After routine laboratory investigations, abdominal sonography and abdominal computed tomography were performed to assess the evidence of hepatobiliary anatomic abnormalities. In cases that the final diagnosis of Caroli disease was not approved by the above investigations, magnetic resonance cholangiopancreatography (MRCP) was performed. All patients were followed two to four years after liver transplantation.

RESULTS

The prevalence of Caroli disease in our liver transplant recipients was 0.44% (4/900 patients). Among these four patients, three were female.

The patients had a mean±SD age of 24.5±4.3 (range: 16–36) years. Clinical findings are summarized in Table 1. The clinical manifestations were different including pruritus, jaundice, recurrent cholangitis, repeated gastrointestinal bleeding and gall stone.

Three patients had radiologic evidence of Caroli disease in sonography and computed tomography of liver; in the 4th patient the diagnosis was confirmed by MRCP. The sever-
ity of liver disease among these patients were assessed by MELD score; the score varied from 11 to 23 (mean = 17.5). All patients had diffuse form of Caroli disease with portal hypertension (Caroli syndrome). All patients received intravenous methylprednisolone for the first three post-operative days that changed to prednisolone (0.5 mg/kg/d), tacrolimus (0.15 mg/kg/d) and mycophenolate mofetil (20 mg/kg/d) from the fourth post-operative day. Prednisolone was tapered and discontinued during the first six months of transplantation.

During post-operative course, one patient developed liver abscess which was treated with percutaneous drainage and antibiotics (imipenem 500 mg iv, q6h for 2 weeks followed by oral therapy for a total of 6 weeks and vancomycin 1 g iv, q6h for 2 weeks).

Explanted liver pathology documented the diagnosis of Caroli disease with no evidence of malignant transformation. After four years of follow-up, all patients were alive and healthy.

**DISCUSSION**

Caroli disease is a rare congenital condition. It is an autosomal recessive disease, although autosomal dominant mode of inheritance has been reported \[6,7\]. Although drainage procedures with ERCP or PTC and sphincterotomy can aid biliary drainage and stone removal, radiological, endoscopical and surgical biliary drainage techniques are palliative treatments leading to temporarily bile drainage. It is necessary to consider that repeated palliative treatments could result in persistence of the symptoms and increase the risk of malignant transformation (cholangiocarcinoma) and should reasonably be avoided \[4,8,9\]. Liver transplantation represents the only curative treatment for the symptomatic Caroli disease. However, when cholangitis occurs, a large number of patients die within 5–10 years \[5\].

In our study, all patients with Caroli disease were selected and scheduled for liver transplantation early after the attack of cholangitis, so the patients tolerated the surgical intervention well and their post-transplantation course was uneventful. In three patients liver were cirrhotic; the high MELD score recorded were indicative of need for transplantation. However, in the fourth patient who had a low MELD score, repeated attacks of cholangitis was the indication for transplantation.

In conclusion, according to our experience, although Caroli disease is rare, since there is no cure for the disease, liver transplantation must be done early in the course of cholangitis \[10,11\] to reduce the risk of malignancy. All four patients reported were diagnosed as Caroli disease during recent years which may be due to possible misdiagnosis of former cases.

**REFERENCES**