Case Report

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Left hepatic lobectomy in a long-term biliary atresia survivor

Go Miyano, Takuo Hayashi[1], Atsushi Arakawa[1], Geoffrey J. Lane, Tadaharu Okazaki, Yoichi Ishizaki[2], Seiji Kawasaki[2], Atsuyuki Yamataka

ABSTRACT

Surgery was performed on day 43 for biliary atresia. Left lobectomy with preservation of Roux-en-y cholecystojejunostomy was performed for refractory cholangitis after 25 years. Our case is currently awaiting liver transplantation. This is the second report of liver resection in a long-term biliary atresia survivor in the English literature.

Key words: Biliary atresia, lobectomy, long-term survivor

INTRODUCTION

Successful liver resection (segmentectomy) for intrahepatic biliary cyst and intractable recurrent cholangitis in a long-term post-Kasai biliary atresia survivor has been reported in the English literature.[1] We believe our case of liver resection (left hepatic lobectomy) in a long-term biliary atresia survivor is only the second case of liver resection in a long-term biliary atresia survivor reported in the English literature.

CASE REPORT

Our case underwent laparotomy, cholecystojejunostomy with retrocolic Roux-en-Y loop elsewhere on day 43 of life for persistent jaundice, and was diagnosed with type-1 cystic biliary atresia. Postoperative status had been totally uneventful without jaundice until he was 25 years old when he developed refractory cholangitis and was hospitalised. The cholangitis was treated conservatively with antibiotics, but abdominal ultrasonography (US) showed what appeared to be an intrahepatic bile lake that could have been the cause of the recurrent cholangitis. The patient was referred to us for further management of the intrahepatic cystic lesion. On presentation, total bilirubin (t-bil) was 4.42 mg/dl, direct bilirubin (d-bil) was 3.08 mg/dl and AST/ALT was 64/86 IU/l. Diagnostic investigations included abdominal US and dynamic computed tomography, which showed an ovoid cystic lesion with multiple intrahepatic biliary dilatations in the left hepatic lobe [Figure 1]. As symptoms of cholangitis were refractory to conservative antibiotic therapy, percutaneous transhepatic cholangiodrainage (PTCD) was performed. However, symptoms persisted after PTCD insertion with deteriorating liver function (t-bil: 6.87 mg/dl, d-bil: 4.36 mg/dl, AST/ALT: 78/112 IU/l). Technetium Tc 99m-labelled hepatoiminodiacetic acid scintigraphy (Tc-99m) identified satisfactory bile drainage from the right hemiliver into the Roux-en-Y loop and a non-functioning left hepatic lobe with cystic lesion and intrahepatic biliary dilatations [Figure 2]. After informed consent was obtained from the patient, left hepatic lobectomy was performed. On laparotomy, the liver was found to be densely adhesed, and intraoperative US confirmed cystic dilatations in the left lobe. The umbilical portion was initially covered



Figure 1: Preoperative CT. Dilated intrahepatic bile ducts and bile lake in the left lobe

Departments of Pediatric General and Urogenital Surgery, [1]Human Pathology, and [2]Liver and Transplantation, Juntendo University School of Medicine, Tokyo, Japan

Address for correspondence:
Dr. Go Miyano, Department of Pediatric General and Urogenital Surgery, Juntendo University School of Medicine, 2-1-1 Hongo, Bunkyo-ku, Tokyo 113-8421, Japan.
E-mail: go1993@hotmail.co.jp

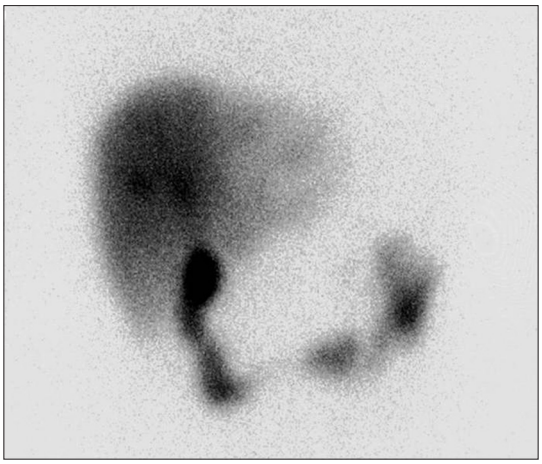


Figure 2: Preoperative Tc-99m. There is satisfactory bile drainage through the cholecystojejunostomy anastomosis to the Roux-en-y loop

with dense adhesions. After careful dissection, although the left hepatic duct was not clearly identified, the left hepatic artery and left portal vein were divided. The usual line of demarcation in left lobectomy cases was indistinct, but the liver was divided, preserving the caudate lobe, and the left hepatic vein ligated. The adjacent Roux-en-Y loop was preserved. Left lobectomy took 8.5 hours, blood loss was 20 g and no blood transfusion was required. Histological examination showed purulent inflammation of the biliary passages around the portal region [Figure 3]. Some bile ducts were dilated and filled with mucopurulent fluid. These findings were consistent with suppurative cholangitis. Additionally, the liver parenchyma showed diffuse periportal fibrosis with bridging fibrosis in some areas. There were no obstructed intrahepatic bile ducts. Some portal tracts had concentric periductal fibrosis around intrahepatic bile ducts with reactive changes. There were no major postoperative complications, and he was discharged from hospital 14 days after surgery, with improved liver function (t-bil: 1.20 mg/dl, d-bil: 0.75 mg/dl, AST/ALT: 50/61 IU/l). However, 7 months later, he represented with symptoms of cholangitis. Antibiotics, ursodeoxycholic acid and steroids were commenced, but Tc-99m scanning showed poor bile drainage from the right hemiliver into the Roux-en-Y loop, and 10 months after surgery, t-bil was 4.38 mg/dl, d-bil was 2.30 mg/dl, AST/ALT was 81/136 IU/L, with abdominal tenderness and persistent high fever. Currently, he is awaiting liver transplantation.

DISCUSSION

The prognosis of type-1 BA is usually much better than that of other types, such as type-2 or type-3.[2] However, there are reports of type-1 patients developing severe complications more than 20 years after corrective surgery, and it would appear that the incidence of late cholangitis is considerably higher in type-1 patients.[3] Although the intrahepatic biliary system is severely destroyed in the perinatal period or early infancy in type-3 patients, type-1 patients would appear to have minimal damage, which could be the cause of complications in the long-term.[3] In fact, in our case, there was focal intrahepatic bile duct damage without anatomic predilection. In addition, histopathology of the portal tracts in our case, that is, periductal fibrosis surrounding affected bile ducts was similar to "onion-skin fibrosis" usually seen in patients with primary sclerosing cholangitis. The mechanism for the destruction of intrahepatic bile ducts in BA remains unclear, but from our case, periductal fibrosis with onion-skin appearance was present and this mechanism may be involved in type-1 patients developing liver dysfunction in the long-term. However, the same "onion-skin fibrosis" could have been a secondary change due to suppurative cholangitis as well.

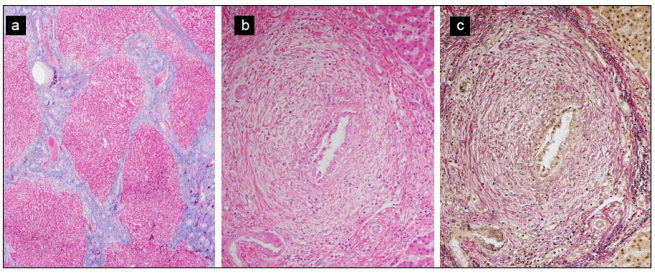


Figure 3: Histopathology of the resected liver specimen. In the low-power view, there is bridging fibrosis in the liver parenchyma (a; Azan stain), and an intrahepatic bile duct undergoing degeneration with concentric "onion-skin" fibrosis. (b; Haematoxylin and Eosin stain, c; Elastic van Gieson stain)

In summary, liver resection in a long-term biliary atresia survivor for refractive cholangitis was initially successful, but now, liver transplantation is required for deteriorating liver function. To the best of our knowledge, this is only the second report of such surgery in the English literature.

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