Hepatic Cirrhosis Caused by Caroli Disease


INTRODUCTION

Caroli’s disease is a congenital intrahepatic biliary dilatation (a choledochal cyst). The dilated ducts connect with the main duct system and are liable to become infected and contain stones. Two distinct diseases associated with Caroli’s disease have been recognized as a simple type associated with medullary sponge kidney in 60-80% of cases and periportal fibrosis type associated with congenital hepatic fibrosis, cirrhosis, portal hypertension, and esophageal varices. Caroli’s disease usually presents in childhood or early adult life as abdominal pain, hepatomegaly, and fever. Caroli’s syndrome is applied to coexistence of Caroli’s disease and congenital hepatic fibrosis. Both are result from malformations of the embryonic ductal plate at different levels of the biliary tree. Presentation may be as abdominal pain and cholangitis or as bleeding esophageal varices. The hemorrhage usually precedes the cholangitis by a mean of 10 years. In infants, the cyst may be diagnosed by hepatobiliary scintigraphy or ultrasound, which allows diagnosis in utero or after delivery. In older children and adults, ultrasound and CT scan may be diagnostic. Diagnosis is usually confirmed by endoscopic cholangiography.1,2,3

CASE ILLUSTRATION

A 32 year old man was admitted to Cipto Mangunkusumo hospital because of blood vomiting and liquid black stools one day prior to admission. He had suffered from this bleeding episode since he was 25 years old. He used to be admitted at Husada hospital where he was diagnosed Caroli’s disease. The diagnosis was made by endoscopic retrograde cholangiopancreatography (ERCP). When he was 15 years old, he had nausea, slight abdominal pain, and was admitted in hospital for three weeks. He was told that he suffered from cholangitis. Since then, he always had the same symptoms every year until 24 years old. In 1989 the patient was consulted to the University of Hannover and Munster (West Germany) and was considered whether the patient could get liver transplantation or not. But the problem was they could not get the organ donor, so they decided to give only conservative therapy.

In 1995, he complained of blood vomiting and black stools for the first time, and he was admitted in the hospital for few days. He was examined by an internist with endoscopic procedure, and the result that he had varices of the esophagus grade II-III (figure 1). After he was recovered from the illness, he was discharged from the hospital and continued with conservative therapy.

Since that year, he had to admitted to be the hospital every three or four months for undergoing band ligation for his varices.

On physical examination, he looked pale but fully alert. The blood pressure was 105/75 mmHg, pulse 106 times/minute. From the lung and heart examination, there was no abnormalities. From the abdomen examination was found ascites, splenomegaly, but the liver was not palpable. From the extremity examination was found palmar erythema, but there was no edema.

Laboratory findings showed a level of Hb 7,8 g/dl; haematocrit 32 vol %; white blood count 8000 /uL; and platelet count was 85.000/uL. The renal function and the blood glucose was normal.

Ultrasound examination showed hepatomegaly with rough structure, multiple cysts at the right lobe of the liver, splenomegaly, and ascites. The CT-scan abdomen

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* Department of Internal Medicine, Faculty of Medicine, University of Indonesia-Dr. Cipto Mangunkusumo Hospital, Jakarta. ** Division of Hematology-Medical Oncology. *** Division of Geriatrics. **** Division of Hepatology, Department of Internal Medicine, Faculty of Medicine, University of Indonesia-Dr. Cipto Mangunkusumo Hospital, Jakarta.
revealed hepatosplenomegaly, multiple intrahepatic cysts, and ascites. Liver biopsy showed congenital hepatic fibrosis and chronic cholangitis.

**DISSCUSSION**

Caroli disease is a rare congenital disorder (an autosomal recessive disease) of the intrahepatic ducts (figure 2 and 3). It is characterized by intrahepatic dilatation of the biliary tree, thought to be the result of a pathologic developmental process known as a ductal plate malformation (DPM). In this case, the patient had Caroli’s disease based on the major complaint such as blood vomiting and liquid black stools since he was young. The gastrointestinal bleeding was caused by hepatic cirrhosis, which resulted in portal hypertension (Caroli’s disease type 2). Other data that supported the diagnosis of hepatic cirrhosis was the presence of splenomegaly, ascites, palmar erythema on physical examination. All these data were confirmed by ultrasound examination, CT-scan of the abdomen (figure 4), and ERCP revealed hepatosplenomegaly, ascites, multiple cysts on the right liver lobe, and the dilated of the bile duct (figure 5).4,5,6,7

At the first time, recurrent fever, right upper quadrant abdominal pain, and jaundice that could be a cholangitis were caused by bile stasis as the most common presenting symptoms as the literature said.3,6

The kidney function in this patient was still good, as mentioned in the literature that precursor of the intrahepatic biliary tree is double-layered sleeve of
ductal plate. Ductal plate arises first from hepatocyte precursors surrounding hilar portal vein vessels on the eight week of gestation. Because the kidney process was the same type as hepatic process, the patient could have polycystic kidney too.\textsuperscript{7,9,10,11}

The treatment of Caroli disease include reducing the risk of blood loss such as: fluid replacement, blood transfusion, gastric lavage, and pharmacological therapy like sucralfate, H2 blocker or proton pump inhibitor. And to reduce the portal pressure is by pharmacological therapy (such as beta blocker and nitrates) and non-pharmacological therapy. If the cholangitis were present, the treatment should be by broad spectrum antibiotics.\textsuperscript{4,6,9,11}

In the minority of patients who have intrahepatic ductal ectasia like Caroli syndrome, the prognosis is determined largely by the frequency and severity of episodes of cholangitis, which may cause sepsis or death. Progressive liver failure may also develop, possibly requiring liver transplantation.\textsuperscript{4}

**REFERENCES**