Hepatectomy for a Patient with Polycystic Liver Disease Associated with Cystobiliary Communication: A Case Report

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Abstract

Cystobiliary communication is a rare complication of polycystic liver disease. We present the case of a 45-year-old woman with frequent epigastric discomfort. Liver function test results revealed an elevated serum total bilirubin and elevated serum alkaline phosphatase. Computed tomography scan showed multiple cysts over left lobe liver, with an 8-cm cyst located at the hepatic hilum with downward displacement of the hepatic hilar structures. An endoscopic retrograde cholangiopancreatography revealed segmental narrowing with extrinsic compression of the bile ducts over the medial segment of the left lobe of the liver with communication of the biliary tree with the large cyst near the porta hepatic. Under the diagnosis of symptomatic polycystic liver disease with biliary communication, the patient underwent left hepatectomy. Histological examination of the surgical specimen revealed polycystic liver disease with multiple diffuse cystic lesions lined by cuboidal to flat biliary epithelium, without mesenchymal stroma or cellular atypia. After operation, the patient was symptom free with normal liver function tests during follow-up. This case indicates that hepatic resection is the treatment of choice for patients with polycystic liver disease complicated with cystobiliary communication.

Keywords: Polycystic liver disease; Cystobiliary communication; Hepatic resection

Introduction

Polycystic Liver Disease (PLD) is arbitrarily defined as a liver containing >20 cysts [1]. The disease is associated with two genetically distinct diseases: as a primary phenotype in isolated PCLD and as an extrarenal manifestation in Autosomal Dominant Polycystic Kidney Disease (ADPKD) [1]. Hepatic cysts in PCLD are usually asymptomatic. In some patients, the disease may be associated with abdominal tenderness [2]. Cystobiliary communication is a rare complication of PCLD [3]. In 1998, Wind et al. [4] reported the first female patient with PCLD associated with spontaneous communication between liver cyst and bile duct, which was diagnosed by Endoscopic Retrograde Cholangiopancreatography (ERCP) and cystography. The patient received conservative treatment with percutaneous drainage only. We hereby report a case of isolated PCLD complicated with cystobiliary communication detected with ERCP, and was successfully treated with hepatic resection.

Case Presentation

A 45-year-old woman presented with frequent epigastric discomfort. Physical examination revealed a 6 cm hepatomegaly more in the epigastric region. Laboratory findings showed liver transaminases were normal, whereas total bilirubin (2.8 mg/dL, normal: 0.2 to 1.6 mg/dL) and alkaline phosphatase (347 U/L, normal: 10 to 100 U/L) were elevated. Renal function was normal and there was no proteinuria. Computed Tomography (CT) scan showed multiple liver cysts, most of which were located in the left lobe. An 8-cm cyst with water attenuation, located at the hepatic hilum with downward displacement of the hepatic hilar structures (Figure 1, arrow), accompanied by mild dilatation of the intrahepatic bile duct. The kidneys showed no cystic lesions. An ERC revealed segmental narrowing with extrinsic compression of the bile ducts over the medial segment of the left lobe of the liver (Figure 2, arrow). Communication of the biliary tree with the largest cyst near the porta hepatic was noted, with contrast medium filling and opacification of the cystic
content (Figure 2, arrowheads). Under the diagnosis of symptomatic PCLD with cystobiliary communication, the patient underwent left hepatectomy. At resection, the left hepatic lobe was sectioned to reveal multiple cysts with a large cyst containing serous fluid and a thick fibrous wall, but without an identifiable lining (Figure 3). Histological examination of the surgical specimen revealed PCLD with multiple diffuse cystic lesions lined by cuboidal to flat biliary epithelium, without mesenchymal stroma or cellular atypia. The postoperative course was uneventful, and the results of liver function tests returned to normal two weeks after operation. The patient was symptom free with normal liver function test results during follow-up.

Discussion

Cystobiliary communication occurs in 14% to 37% of patients with hepatic hydatid disease but is a rare complication of polycystic liver disease [5,6]. In both circumstances, the communication occurs in the cysts located in the segments close to the hilum. The frequency of bile duct distribution is among the predisposing factors. It can also be due to the fact that the compression effect of the cyst over the bile ducts is high at the level of the hilum than at the peripheral location [4]. With the presence of cystobiliary communication, careful selection of treatment is essential for effective symptom relief [7]. Simple aspiration and drainage of the cysts can be the primary treatment option but usually results in recurrence [8]. Communication of the cyst with the biliary tree precludes treatment with alcohol sclerotherapy because of the risk of sclerosing cholangitis [4]. Laparoscopic internal drainage with unroofing of the cyst intraperitoneally is not suitable if the case is complicated with biliary communication [9]. In our case, resection of the hepatic cyst with cystobiliary communication with partial hepatectomy resulted in a favorable outcome.

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References