CASE REPORT

TJG
The Turkish Journal of Gastroenterology

Unusual cholangiographic findings in a patient with primary sclerosing cholangitis: Cystic dilatation

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A 45-year-old woman with the diagnosis of primary sclerosing cholangitis and ulcerative colitis admitted with the complaints of pruritus and jaundice. Endoscopic retrograde cholangiography revealed entirely narrow, irregular common bile duct and common hepatic duct and unusual cystic dilations in the common hepatic duct and left hepatic duct. Balloon dilation of the common bile duct was performed, and a 10 F double pigtail stent was inserted into the cyst. Three months after the endoscopic retrograde cholangiography, cystic dilatations had completely resolved. Primary sclerosing cholangitis may present with cystic dilatations up to a level that it may resemble Caroli disease.

Key words: Primary sclerosing cholangitis, cyst, dilatation

PSC'de olağan dışı bir kolangiografik bulgu: Kistik dilatasyon

Primer sklerozan kolanjit ve ülseratif kolit tanıları ile takip edilen 45 yaşında bir bayan hasta kaşıntı ve sarılık yakınmaları ile başvurdu. Endoskopik retrograd kolanjiyopankreatografide koledok ve ortak hepatik kanal boylu boyunca daralmış, irregüler olup ortak hepatik kanal ile sol hepatik kanalda olağan dışı kistik dilatasyonlar izlendi. Ortak hepatik kanala balon dilatasyonu yapılıp, kist içine 10 F double pigtail stent konuldu. Endoskopik retrograd kolanjiyopankreatografiden üç ay sonra kistik dilatasyonların tamamen düzeldiği gözlendi. Primer sklerozan kolanjit Caroli hastalığını anımsatacak düzeyde kistik dilatasyona neden olabilir.

Anahtar kelimeler: Primer sklerozan kolanjit, kist, dilatasyon

INTRODUCTION

Primary sclerosing cholangitis (PSC) is a chronic, cholestatic disorder characterized by inflammation, fibrosis, and stricturing of the intrahepatic and extrahepatic bile ducts (1). Progressive destruction of the bile ducts leads to biliary cirrhosis, hepatic failure, and cholangiocarcinoma. Diagnosis is established by demonstrating multifocal strictures with intervening segments of normal or slightly dilated intrahepatic and/or extrahepatic bile ducts on cholangiography. Herein, we present a case of PSC with unusual cystic dilatation of the common hepatic and left hepatic ducts up to a level that is seen in patients with Caroli disease.

CASE REPORT

A 45-year-old woman was admitted to the Gastroenterology Department of another hospital in 2009 with the chief complaints of pruritus and jaundice. Laboratory tests at that time revealed an increase in cholestatic liver tests and Ca 19-9 level. Magnetic resonance cholangiopancreatography revealed dilated intrahepatic bile ducts and

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Manuscript received: 20.01.2012 **Accepted:** 20.02.2012

Turk J Gastroenterol 2012; 23 (6): 792-794
doi: 10.4318/tjg.2012.0496

a normal common bile duct. The patient underwent a surgical exploration with the diagnosis of cholangiocarcinoma. In the exploration, no malignancy was observed. The liver was found to be cirrhotic and histopathologic examination of the biopsy specimen obtained during surgery revealed PSC and biliary cirrhosis (Figure 1). Ulcerative colitis was diagnosed later by colonoscopic examination and biopsy. The patient was placed on ursodeoxycholic acid and mesalazine treatment.

Approximately two years after the first presentation, the patient admitted to our department with the complaints of gradually increasing jaundice and pruritus. Physical examination was normal other than an icteric sclera and body. Laboratory evaluations revealed increase in total bilirubin (15 mg/dl), direct bilirubin (11 mg/dl), alkaline phosphatase (299 U/L), gamma- glutamyl transpeptidase (68 U/L), alanine aminotransferase (77 U/L), and aspartate aminotransferase (123 U/L) levels. An endoscopic retrograde cholangiopancreatography (ERCP) was carried out. The papilla was slightly retracted. Common bile duct and common hepatic duct were entirely narrow and irregular. The appearance of intrahepatic bile ducts was compatible with PSC. There were cystic dilatations in the common hepatic duct and left hepatic duct resembling Caroli disease (Figure 2). Endoscopic sphincterotomy and balloon dilation of the common bile duct were performed. A 10 F double pigtail stent was inserted into the cyst. Biliary brush cytology revealed normal biliary epithelium. Three months after the first ERCP, a second one was performed. Cystic dilatations had completely resolved (Figure 3). The stent was withdrawn. Balloon dilation of the common bile duct was performed. The patient remained well on mesalazine and ursodeoxycholic acid treatment.

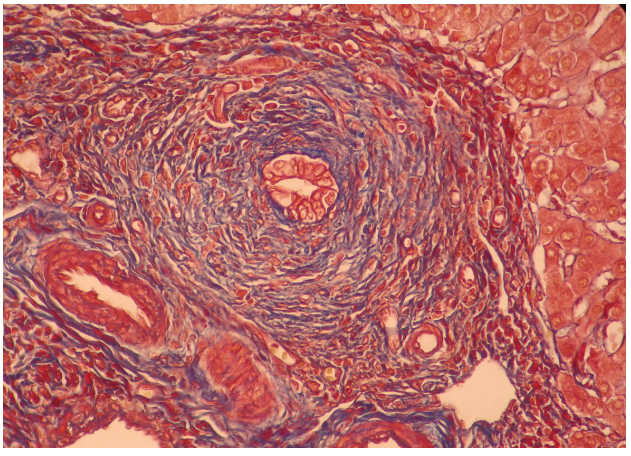

Figure 1. 'Onion skin' fibrosis around the bile duct in the portal area (Masson's trichrome stain).

DISCUSSION

We presented herein a PSC patient with an unusual dilatation of the common hepatic and left hepatic ducts large enough to resemble Caroli disea-

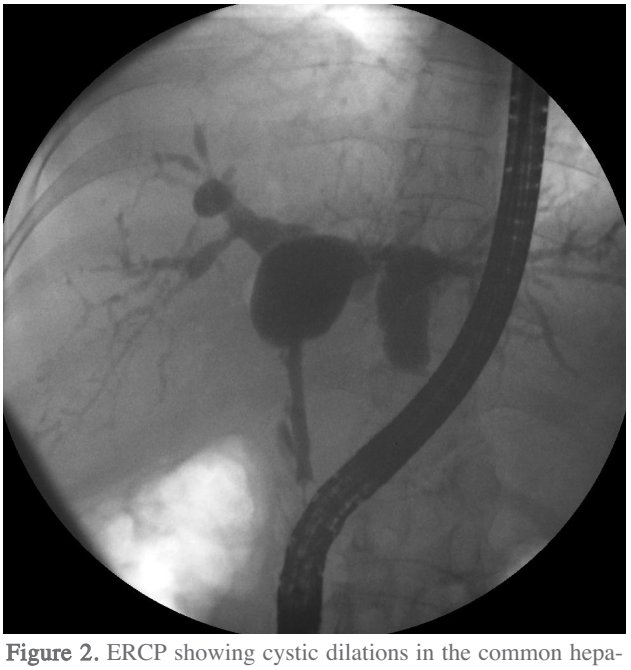

Figure 2. ERCP showing cystic dilations in the common hepatic and left main ducts resembling Caroli disease.

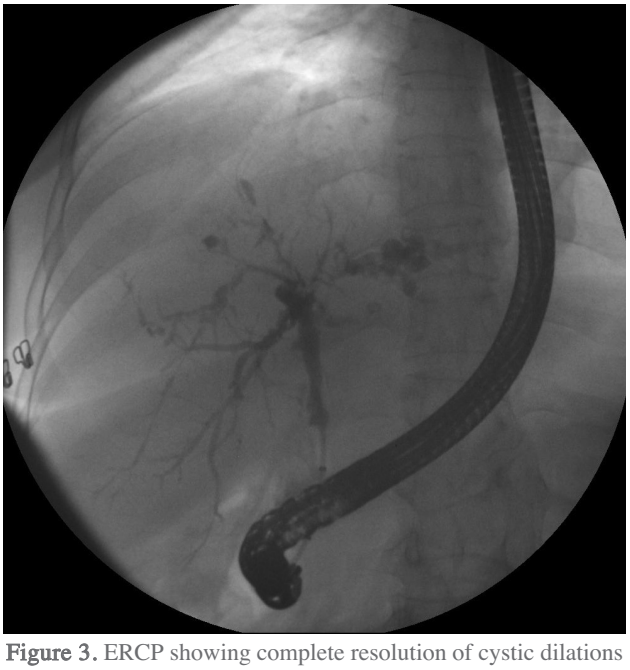

Figure 3. ERCP showing complete resolution of cystic dilations three months after stent insertion.

se. The patient had a histologically proven PSC, and total resolution of biliary cysts after biliary drainage led to exclusion of the possible association with Caroli disease, which was not reported in the literature before. The resolution of cystic dilatations after drainage suggested that they were of pre-stenotic origin.

To our knowledge, there are four studies in the literature reporting cystic dilations in patients with PSC. Ludwig et al. (2) was the first to describe the presence of intrahepatic cholangiectases in the explanted liver of patients with PSC. The cholangiectases were much smaller than described in Caroli disease. The shape and distribution (no relation with biliary stenoses) suggested that cholangiectases were not passively dilated ducts. Theilman et al. (3) reported the in situ observation of large intrahepatic cholangiectases in PSC. The contrast material was rapidly flowing off the cyst when the balloon occluding the main intrahepatic bile ducts was deflated. Therefore, they suggested that the dilations were not of pre-stenotic origin. Goldwire et al. (4) reported a patient whose initial findings were suggestive of type IV choledochal cyst and common bile duct stricture. One year after stenting procedures, ERCP revealed total resolution of the cystic dilation and irregular intrahepatic and extrahepatic bile ducts, consistent with the diagnosis of PSC. Genève et al. (5) reported a case with cystic dilatations of the intrahepatic bile ducts simulating Caroli disease. They suggested that cystic dilatation of intrahepatic bile ducts could be included among the radiologic features of the disease.

The typical cholangiographic findings of PSC are multifocal strictures and dilations producing a beaded appearance. Dilatations are slight and may appear as saccular or diverticular outpouchings. Cystic dilation is an unusual finding of PSC. It may become large enough to mimic Caroli disease. They may be pre-stenotic or represent manifestations of the disease process. Whether or not it may cause a predisposition to cholangiocarcinoma remains speculative.

In conclusion, cystic dilatation is an unusual cholangiographic finding of PSC, with an unknown pathogenesis and prognostic importance. It can be included among the cholangiographic findings of the disease.

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