Recurrent Biloptisis and Severe Hemobilia Due to Frequent Intrahepatic Infections in a Patient with Caroli's Disease: Unreported Morbidities of the Disease

SIK İNTRAHEPATİK ENFEKSİYONLARA BAĞLI TEKRARLAYAN BİLOPTİZİ VE CİDDİ HEMOBİLİASI OLAN KAROLİ HASTALIĞI: HASTALIĞIN BİLDİRİLMEMİŞ MORBİDİTELERİ

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Summary

Caroli's disease is a congenital disease of multiple cystic or saccular dilatations of the intrahepatic bile ducts. Frequent complications in Caroli's disease are intraductal lithiasis, recurent cholangitis and liver abscess. Effective decompression of the biliary tract in Caroli's disease is the main stem of therapy. However, partially controllable attacks of intrahepatic infections in Caroli's disease may result in dramatic complications. Herein we report a patient with adult-onset Caroli's disease and was complaining of recurrent biloptisis and severe hemobilia.

Key Words: Caroli's disease, Biloptisis, Hemobilia

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Özet

Caroli hastalığı konjenital kökenli intraheaptik safra yollarının çok sayıda kistik/sakküler dilatasyonları ile karakterize bir patolojik tablodur. Bu hastalık ile ilgili sık olarak saptanan infeksiyöz komplikasyonlar intraduktal litiazis, tekrarlayan kolnajit atakları ve karaciğer abse oluşumudur. Ana tedavi şekli safra yollarının etkin bir şekilde drenajıdır. Çeşitli tedavi yöntemleri ile kısmi olarak kontrol altına alınabilen intrahepatik infeksiyon atakları ciddi komplikasyonlara neden olabilmektedir. Bu olgu ile kroli hastalığında tekrarlayan biloptizis ve hemobili ile kliniğe başvuran bir hastanın sunumunu yapmaktayız.

Anahtar Kelimeler: Caroli hastalığı, Biloptizis, Hemobili

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Caroli's disease is a rare condition characterized by saccular dilatations of intrahepatic biliary tree (1). There is stasis in these dilated ducts which are liable to become infected and lead to intrahepatic lithiasis and recurrent hepatic infections (2). Though these are known complications of the disease that are commonly encountered, bronchobiliary fistula and hemobilia associated with recurrent intrahepatic infections in Caroli's disease have not been reported to date. Here we report a patient who had adult-onset Caroli's disease and, was admitted with recurrent attacks of biloptisis and severe hemobilia.

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Case Report

A 37-year- old man was admitted to our department with the symptoms of right upper quadrant pain, pruritis, low grade fever, malaise and fatigue. Past medical history was consistent with a diagnosis of Caroli's disease that had been made in another facility, in 1989. Thereafter, he had been followed up regularly at the same hospital for recurrent attacks of intrahepatic biliary infection. A percutaneous liver biopsy that was taken in 1994 excluded the association of congenital hepatic fibrosis. Thus, the simple form of Caroli's disease was diagnosed. As far as we learn from hospital records, on many occasions, long term prophylactic antibiotics had been given to control recurrent bil-

iary infections and, endoscopic retrograde cholangiographic (ERC) interventions had been performed to clean the choledochus from residue stone and debris. In March 1997, a choledochoduodenostomy operation had been performed in the same hospital. Thereafter, his general condition improved and he was able to gain weight.

On admission to our hospital in December 1997, an abdominal ultrasonography demonstrated multiple hyperechogenities in the liver parenchyma which were cystic dilatations of the intrahepatic biliary tree that were filled with stones and sludge. On ERC through choledochoduodenostomy orifice, we observed mild dilatation of the common bile duct due to stones and debris inside. There were also multiple cystic dilatations of intrahepatic bile ducts containing stone and sludge material (Figure 1). We partially succeeded in baloon extraction for drainage of biliary sludge through choledochoduodenostomy orifice. Then, a sphincterotomy and balloon extraction of residue stones and debris were also successfully applicated. He was given ciprofloxacin and ursodeoxycholic acid to suppress intrahepatic infection and relieve the pruritis. The patient was hospitalized on many other occasions, and biliary sludge and stone retrieval was performed by extraction balloons or basket catheters during ERC procedure. In November 1998, the patient's main complaints were biloptisis, fever, severe cough and malaise. A P-A chest-X-ray showed an elevated right dome of diaphragm with a prominent right pleural effusion. Abdominal computerized tomography (CT) demonstrated multiple intrahepatic cystic dilatations (the largest measuring 3.5x 2.2 cm) filled with sludge and small stones and, air-fluid levels in the subdiaphragmatic area that were consistent with abscess formation (Figure 2). On thoracal sections of the CT scan, subdiaphragmatic collections were observed to extend into the pulmonary parenchyma and nearby main bronchus. These findings were confirmed on abdomino-thoracal magnetic resonance (MR) examination (Figure 3). Though the patient's symptom of biloptisis suggested the presence of bronchobiliary fistula, neither technetium 99m IDA cholescintigraphy nor ERC could detect a fistulous tract. During ERC procedure, balloon extraction of biliary sludge, debris and small stones was performed successfully and then, a nasobiliary tube was inserted to help seal off the suspected bronchobiliary fistula.The nasobiliary drainage tube, which stayed in place persistently for 2 months, provided biloptisis to decrease and resolve completely within 2 weeks of its placement. However, at the end of two months of silent period, the biloptisis recurred soon after the symptoms of a new attack of cholangitis started. Neither a repeat ERC procedure nor biliary scanning demonstrated a bonchobiliary fistula, which indeed remained a clinical diagnosis. There were filling defects and air within the bullous intrahepatic bile ducts, and the cleaning of the common bile duct was performed again exactly same as the previous trials. Later, a percutaneous drainage catheter was placed into the right hepatic bile duct to help close the biliary bronchial connection. This effectively decompressed the biliary tree and resolved the biloptisis completely. Two months later, the patient was admitted with severe hematemesis, dizziness and syncope. He was rehospitalized once again, and after conservative measures were attempted, an emergent upper endoscopy was performed. This revealed blood oozing from the orifice of the ampulla of Vater and choledochoduo-

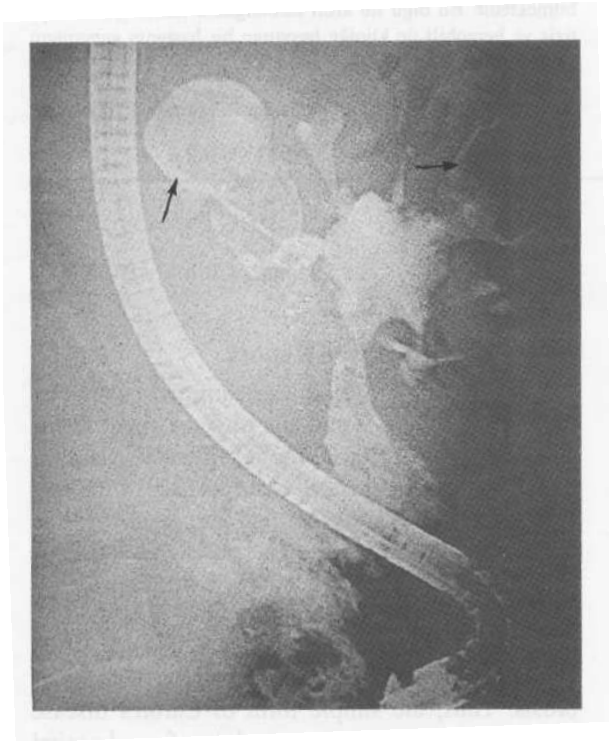

Figure 1. Endoscopic retrograde cholangiography shows bullous dilatations of intrahepatic biliary tree that contain sludge and stones (arrows).

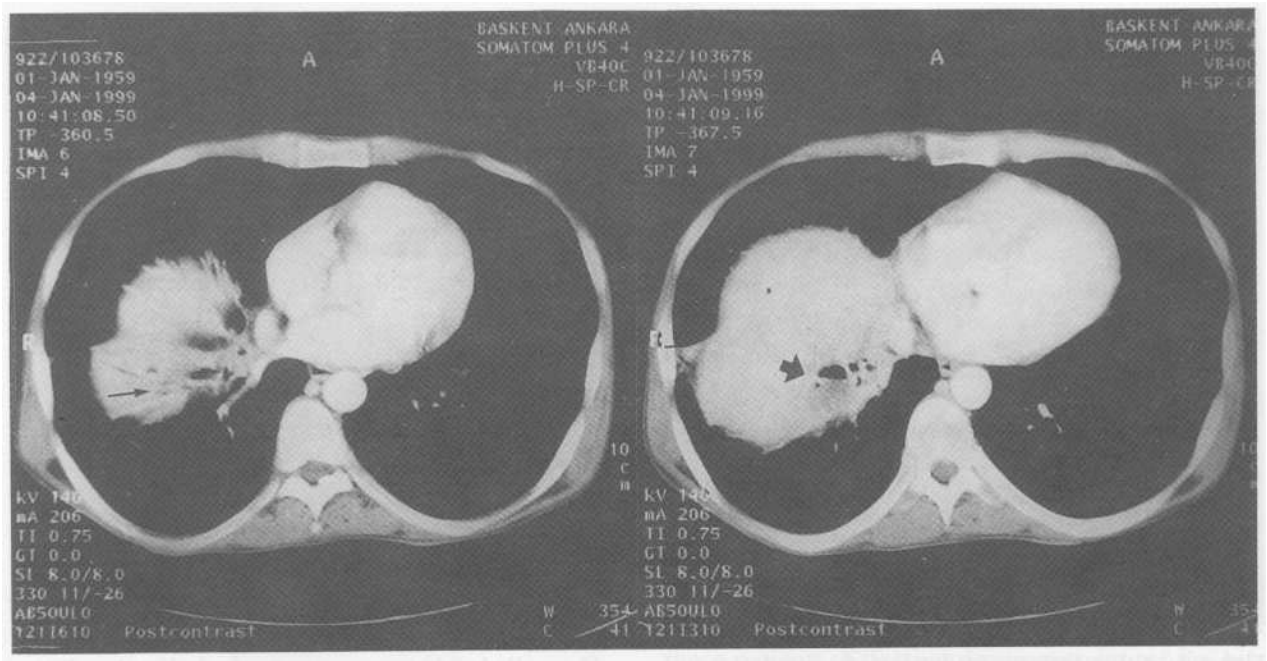

Figure 2. Abdominal CT axial images demonstrate subdiaphragmatic (thick arrow) and supradiaphragmatic abscess and atelectasis of the right pulmonary basal segment (thin arrow).

denostomy orifice. Hemobilia was diagnosed and celiac angiography was done immediately. During celiac angiography, selective common hepatic artery injection demonstrated a pseudoaneurysm (PA) of the left hepatic artery branch, measuring approximately 12x14 mm in diameters. This artery was selectively catheterized by a microcatheter and selective injection revealed a fistula between PA and left bile duct. Since the patient had Caroli's disease and had dilated biliary ducts, the contrast filled the enlarged left hepatic duct and the choledochus. The PA and the small branch of the left hepatic artery were embolized by n-butyl cyanoacrylate. Postembolization arteriography showed complete occlusion of the PA and the small hepatic artery branch (Figure 4a-c). This halted the bleeding, which never recurred thereafter.

Discussion

Caroli's disease is characterized by multifocal berry-like dilatations of the intrahepatic biliary ducts that communicate with the extrahepatic biliary tree (1). It is a probably congenital condition which inherently abnormal ductal wall development is the primary pathology that occurs during embryogenesis (3). It was proposed that unequal proliferation of biliary duct epithelial cells more active proximally than distally leads to abnormally dilated biliary tree proximally and normal tree distally (4).

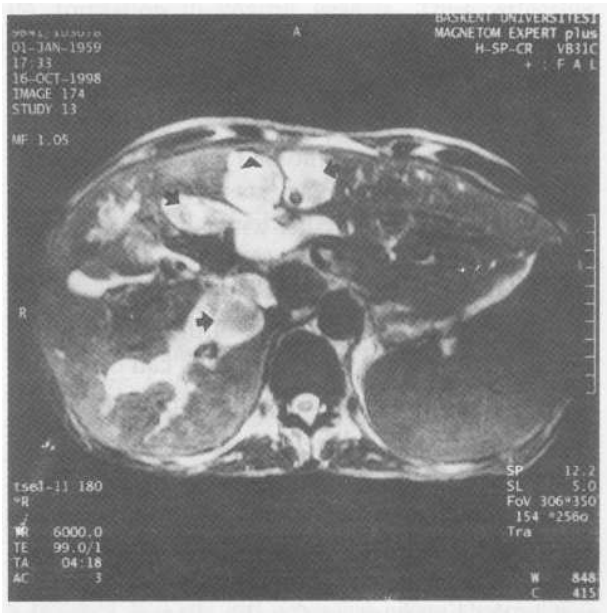

Figure 3. Abdominal MR axial examination on T2-weighted image reveals cystic and irregular dilatations of the intrahepatic bile ducts which includes air (arrowhead) and filling defects that represent biliary sludge and stones (arrows).

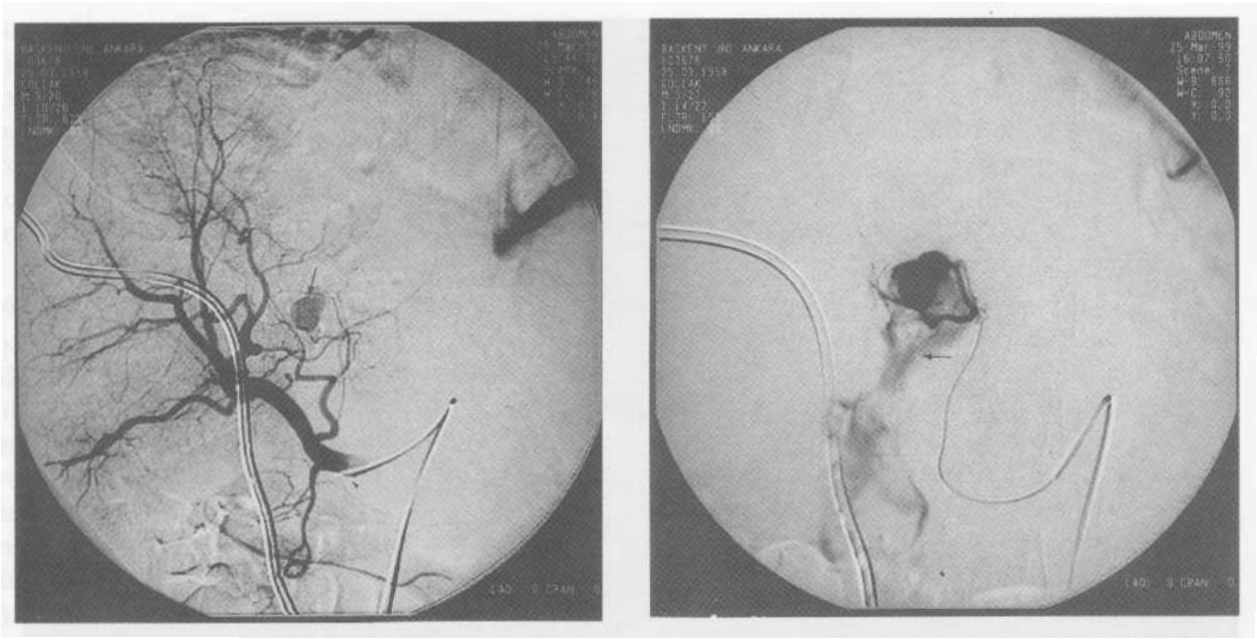

Figure 4a. Celiac angiography demonstrates pseudoaneurysm of the left hepatic artery. A percutaneously placed biliary catheter tip is also visible in the right side of the biliary system (arrow).

Figure 4b. Left hepatic artery was selectively catheterised and selective injection revealed fistula between the PA and the left duct of the biliary system (arrow).

Patients with Caroli's disease are prone to develop several complications, including bacterial cholangitis, biliary sludge build-up, calculi formation, and cholangiocarcinoma (5). These problems are mostly related to biliary stasis within the intrahepatic biliary tree, together with a high resting bile flow arisen from the cysts (6). This high resting bile flow leads to excessive biliary drainage, so much that spontaneous rupture of the common bile duct and consequent bile peritonitis can occur (7).

Pyogenic liver abscess and fatal gram (-) septicemia may complicate uncontrolled intrahepatic infection in Caroli's disease (1,8). Partially suppressed infection may not lead to septicemia, but, as important as septicemia, intrahepatic infection may extend to the peripheral organs. Only one report in the literature on Caroli's disease notes the development of subphrenic extension of a liver abscess, within the cystic biliary cavities (9). The suphrenic space is especially important for these kind of infection, since hepatic infection can cross the diaphragm and lead to pleuropulmonary infection. Another potential end result is the formation of a fistula between the biliary tree and bronchi, as occurred in our patient. In our case, the clinical diagnosis was clear since the patient's sputum contained bile, a pathognomonic finding for bronchobiliary fistula. This type of fistula occurs in 4%

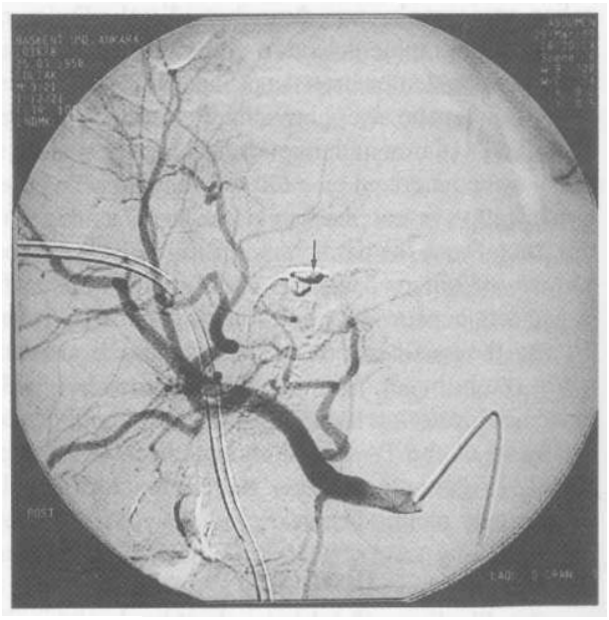

Figure 4c. Postemboliziation angiography showed complete occlusion of the PA (arrow).

of patients with hepatic abscess, and 10.5% of those with subphrenic abscess (10). The diagnosis of bronchobiliary fistula mostly rests on endoscopic, percutaneous or T tube cholangiography, or cholescintigraphy (10,11). In our patient, all of these methods failed to show a fistulous connection between the bronchial system and the biliary tree. One of the reasons for this might have been the dispersion of radiocontrast or scintigraphic material within the large cystic biliary tree. Another reason may have been the chronic inflammation with adhesion formation, which could have prevented the radiographic, or scintigraphic agent from reaching the cyst that led to the fistulous tract. This case shows the diagnostic difficulties in demonstrating a bronchobiliary fistula in patients with Caroli's disease.

The second important morbidity in our patient was severe hemobilia two months after percutaneous 10 F internal-external drainage catheter placement into the biliary tree via a small peripheral branch of the right biliary tract. Biliary catheter drainage can be complicated by hemobilia for several reasons, including arterial injury, hepatic artery PA and/or hepatic artery portal vein fistula formation, and a rare occurence of varix along the tube tract (12). Generally, intrahepatic vascular lesions following percutaneous biliary drainage catheterization have been reported in up to 33% of patients in the literature, although 6% were found to have clinical evidence of hemorrhage (13). It has been suggested that the incidence of hepatic artery injury including PA and or hepatic artery portal vein fistula formation has been related to the technique during initial catheter placement (13). In our patient's case, peripheral puncture of the bile ducts was performed to avoid large hepatic arteries close to the liver hilus. It was a single-pass procedure done under ultrasound guidance. Moreover, the celiac angiographic findings indicated PA formation in the left hepatic artery, which was far away from the path of needle passage. Thus, the possibility of catheter induced PA formation was nearly excluded. Neither was there the possibility of hemobilia due to previously placed percutaneous drainage catheters, since long term biliary drainage was reported to be necessary for PA formation in such cases (12). This was not the situation with our patient. Instead, we used long term nasobiliary drainage in this patient to decrease biloptisis. A final possibility is mycotic arteritis secondary to chronic infection in biliary cystic cavities (12). Pseudoaneurysm formation in relation with a large choledochal cyst was described in one case report (14). The authors suggested the possible mechanisms of the hepatic artery PA formation in relation with a choledochal cyst on twofold. One is mycotic arteritis due to the chronic inflammation within the cyst. Another is the pressure effect of the cyst on hepatic arteries. We propose that the same mechanisms may have occurred in our case.

We presented this case to demonstrate the progression and unreported complications of intrahepatic infection in Caroli's disease. Bronchobiliary fistulous tract visualization may not be possible in these patients. Effective decompression of the biliary system provide resolution of biloptisis. Severe intrahepatic infection due to stasis or lithiasis in cystic cavities of Caroli's disease may play role in PA formation in nearby vessels. Thus, effective control of hepatic infection carries great importance to prevent hepatic vascular events, pulmonary extension and bronchial connection of intrahepatic infection.

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