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Agenesis of gallbladder and multiple anomalies of the biliary tree in a patient with portal thrombosis: A case report

Safra kesesi yokluğu ve çoklu safra yolları anomalisi ile birlikte portal ven trombozu: Olgu sunumu

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A 55-year-old male patient was admitted to hospital because of splenomegaly. Abdominal ultrasonography and computed tomography failed to demonstrate the gallbladder. The diagnosis of right portal vein thrombosis was established by Doppler ultrasonography, splenoportography and computed tomography angiography . To investigate the biliary tree and evaluate the effect on the biliary tree of portal changes, endoscopic retrograde cholangiopancreatography was performed. Endoscopic retrograde cholangiopancreatography study revealed the absence of gallbladder, cystic duct and common bile duct together with the junction of the right and left hepatic ducts at the pancreatic head, with predominant left hepatic duct. To our knowledge, this is the first reported case of multiple congenital anomalies of the extrahepatic biliary tree associated with right portal vein thrombosis. The presence of these rare pathologies in two viscera running together in a patient with right portal vein thrombosis is a very rare condition.

Key words: Congenital anomaly, gallbladder agenesis, absence of choledochus, portal vein thrombosis

55 yaşında erkek hasta splenomegali nedeniyle yatırıldı. Abdominal sonografi ve bilgisayarlı tomografide safra kesesi görüntülenemedi. Doppler sonografi ve splenoportografi tetkikinde ise portal vende trombüs olduğunu saptandı. Safra kesesini incelemek ve trombüsün koledoğa etkisini göstermek amacıyla ERKP yapıldı. Bu tetkikte safra kesesinin, sistik kanalın ve anatomik anlamda koledokun olmadığı dikkati çekti. Pankreas başında sağ ve sol hepatik kanalların birleştiği ve sol kanalın baskın olduğu görüldü. Bizim bilgilerimize göre çoklu safra sistemindeki anomalilere safra kesesi agenesisinin eşlik ettiği ilk olgudur.

Anahtar kelimeler: Kongenital anomali, safrakesesi yokluğu, koledok yokluğu, portal ven trombozu

INTRODUCTION

Congenital anomalies of the extrahepatic biliary tree are rare and different types have been reported either alone or in combinations in the literature. Agenesis of gallbladder without extrahepatic biliary atresia is a rare congenital anomaly, occurring in 13 to 65 per 100000, and probably results from failure of the gallbladder to develop or vacuolize in utero (1, 2). Congenital variants of the pancreaticobiliary union in which the junction is outside the duodenal wall are also rare anomalies (3). Extrahepatic biliary atresia is the leading cause of obstructive jaundice during infancy, whereas anomalies of the extrahepatic biliary tree rarely cause symptoms during adulthood. Although most adults with agenetic gallbladder as well as those with variations of pancreaticobiliary union are asymptomatic (3, 4), the attempted laparoscopic cholecystectomy in those patients with symptoms may be unexpectedly complicated. Here we report a case with multiple congenital extrahepatic biliary anomalies such as absence of gallbladder, a predominant left hepatic duct unifying with the right hepatic duct at the pancreatic head to form common hepatic duct, and associated right portal vein thrombosis (PVT).

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CASE REPORT

A 55-year-old male patient was admitted to the hospital because of splenomegaly. His medical history had been unremarkable except for cigarette (1/2 pack/day) and whiskey (1 bottle/month) consumption for 15 years. During his admission to our hospital, physical examination revealed massive splenomegaly 9 cm below the left costal margin on palpation. The laboratory findings were as follows: hemoglobin 10.4 g/dl (12-18 g/dl); white blood cells 3,200/ml (3,6000-10,000/ml); thrombocytes 46,000/ml (150,000-450,000/ml), alanine transaminase 30 U/L (5-40 U/L); aspartate transaminase 45 U/L (8-33 U/L); alkaline phosphatase 284 U/L (91-258 U/L); gamma-glutamyl-transpeptidase 127 U/L (5-40 U/L); total/direct bilirubin 2.27/1.31 mg/dl (0.1-1.2/0.0-0.3 mg/dl); glucose 290 mg/dl (70-110 mg/dl); albumin ranging between 3.2–4.1 g/dl (3.2-4.8 g/dl); and international normalized ratio 1.58 (0.75-1.5). The serum electrolytes, blood urea nitrogen and creatinine were all within normal ranges. The serology for hepatitis B and C viruses, anti-dsDNA antibody, anti-nuclear antibody, anti-smooth muscle antibody, anti-liver-kidney microsomal antibody and anti-mitochondrial antibody were negative. Other laboratory measures were serum ferritin 38 ng/ml (6-159 ng/ml); ceruloplasmin 24.2 mg/dl (20-60 mg/dl); alpha-fetoprotein 4.01 ng/ml (0-6.65 ng/ml); thyroid-stimulating hormone (TSH) 4.28 uU/ml (0.35-4.94 uU/ml); C_3 74.2 mg/dl (90-180 mg/dl); C4 8.08 mg/dl (10-40 mg/dl); vitamine B_{12} 550 pg/ml (160-800 pg/ml) and folate 14.6 ng/ml (3-17 ng/ml).

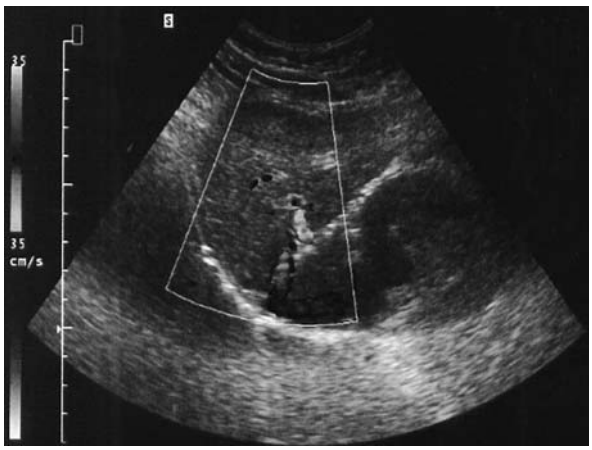

Figure 1. The Doppler ultrasonography revealed the patent vena portalis

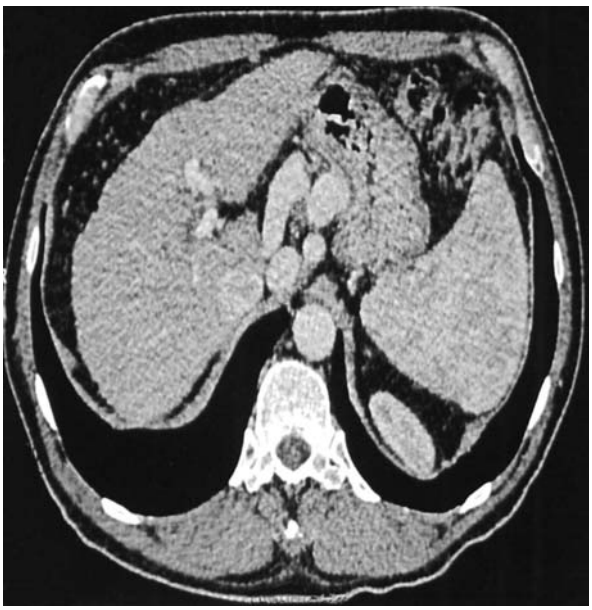

Figure 2. The computed tomography angiography demonstrated the thrombosis at the right vena portalis

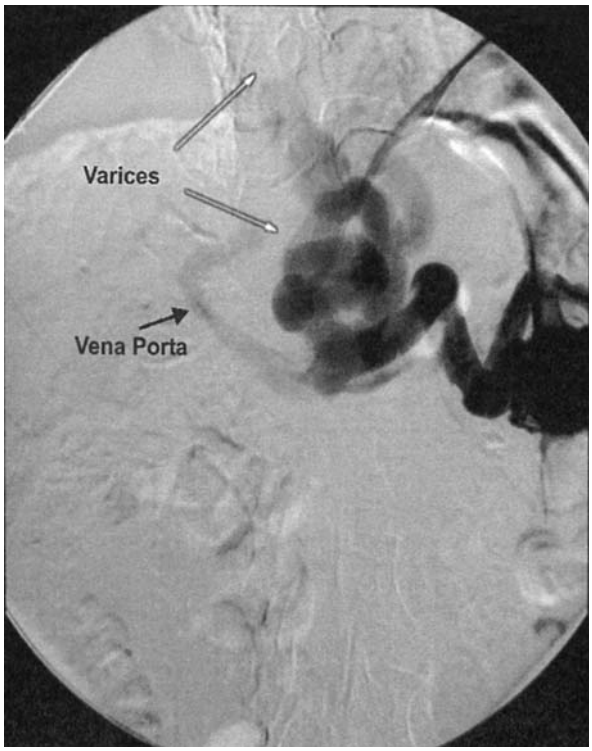

Figure 3. Thinner-than-normal vena portalis and its left branch (black arrow) with hepatofugal flow, thrombosis of the right branch of vena portalis and esophageal varices (white arrows) were present on splenoportography

Factor VIII, IX, XI and fibrinogen levels were within normal ranges. Among the anticoagulant proteins, serum protein C activity was 33% (70-130%), protein S activity was 19% (70-130%) and anti-thrombin III activity was 48% (80-120%). Abdominal ultrasonography demonstrated absence of the gallbladder, spleen 158 cm in length, and right PVT. The findings of abdominal computed tomography, of Doppler ultrasonography (Figure 1) and of computed tomography angiography (CTA) (Figure 2) supported ultrasonographic findings. In order to confirm the diagnosis of PVT, splenoportography was performed, which demonstrated thinner-than-normal vena portalis and its left branch with hepatofugal flow, thrombosis at the right branch of vena portalis and esophageal varices (Figure 3). The endoscopic retrograde cholangiopancreatography (ERCP) demonstrated that the confluence of the right and the left hepatic ducts was just above the papilla of Vater, and the left intrahepatic biliary system was predominant (Figure 4). Neither gallbladder nor cystic ductus could be visualized by ERCP, so there was no choledochus in anatomical mean. Pancreatic ductus was normal. The esophageal varices were present at upper gastrointestinal endoscopy.

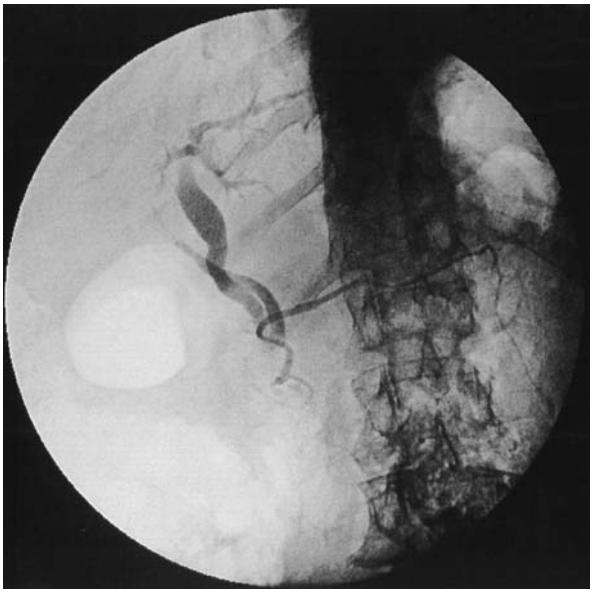

Figure 4. The confluence of the right and the left hepatic ducts was just above the papilla of Vater and the left intrahepatic biliary system was predominant on ERCP. Neither gallbladder nor cystic ductus could be visualized, so there was no choledochus in anatomical mean. Pancreatic ductus was normal

DISCUSSION

Agenesis of the gallbladder and cystic duct without extrahepatic biliary atresia occurs in 1 in every 6,000 childbirths (1, 2). Those children with associated multiple fetal anomalies (13%) die in less than a month of causes related to their nonbiliary anomalies. The rest of the children either remain asymptomatic during their entire life (54%), or have symptoms (33%) compatible with hepatobiliary dysfunction such as right upper quadrant pain, nausea and vomiting that lead to exploratory surgery (4). Since abdominal ultrasonography may not distinguish an absent gallbladder from one shrunken by inflammation, ultrasonography is not enough for the diagnosis. Furthermore, abnormal locations of the gallbladder have been described as intrahepatic, left-sided within lesser omentum, retroperitoneal, retrohepatic, within falciform ligament, retroduodenal, and in retropancreatic areas (5). For these reasons, Frey et al. (6) considered gallblader agenesis proven after explorative laparotomy as meeting the following two criteria: 1. Operative findings confirmed by cholangiogram, 2. The biliary tract visualized by dissecting it from the duodenum to the bifurcation of the hepatic ducts. On the other hand, it has been suggested that ultrasonography combined with ERCP may be adequate and a relatively noninvasive mean for the diagnosis of extrahepatic biliary agenesis (4, 7). Neither ultrasonography nor ERCP visualized the gallbladder in our patient. ERCP demonstrated the predominent left hepatic duct joining the right hepatic duct at the pancreatic head as well as absence of gallbladder and cystic duct.

The right PVT was confirmed by Doppler ultrasonography, CTA and splenoportography in our patient. Numerous conditions may cause PVT, such as hypercoagulable states, infectious and inflammatory diseases, medical interventions to hepatobiliary system or chronic liver dieases. The deficiency of the anti-coagulant proteins might be the possible cause of the right PVT in this patient and since his hepatic function did not acutely deteriorate during his three-year follow-up period, right PVT seems to be a chronic event. Another reason for PVT in the patients with extrahepatic biliary atresia is the ascending cholangitis after portoenterostomy surgery (8, 9). Although our patient had multiple extrahepatic biliary anomalies he never underwent biliary intervention, so ascending cholangitis was an unlikely cause for right PVT.

Portal vein thrombosis with extrahepatic biliary anomalies has not been reported in the literature except our previously reported patient with chronic PVT, absence of common bile duct and junction of the cystic duct with the left hepatic duct (10). Here we report another case of PVT associated with extrahepatic biliary anomalies in an adult patient. To the best of our knowledge, the possible association between PVT and congenital extrahepatic biliary anomalies, according to findings of our two cases, is described for the first time in the literature.

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