Multiple cystic lesions around the bile duct

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A previously healthy 33-year-old man visited the emergency room complaining of fever, abdominal pain, and jaundice. He was immediately hospitalized based on a diagnosis of acute cholangitis and sepsis. Blood tests showed a highly inflammatory state, high concentrations of hepatobiliary enzymes, and predominantly direct bilirubin. Magnetic resonance imaging after admission (Figs. 1-A, 1-B) indicated dilated intrahepatic and extrahepatic bile ducts and multiple cystic lesions present diffusely throughout the liver. Based on clinical symptoms and blood biochemistry findings, acute gallstone cholangitis was considered. Especially, high ALP (702 IU / L) on biochemical findings seem to suggest cholestasis caused by bile duct obstruction. Imaging findings (dilatation of intrahepatic and extra bile ducts, and calculi in the extrahepatic bile ducts) also implicated acute gallstone cholangitis. Immediately after admission, infusion of sulbactam/cefoperazone was started. Furthermore, we performed endoscopic retrograde cholangiography (ERC). Markedly dilated bile ducts with stones were revealed. No biliary stricture nor any communication between bile duct and cystic lesions was found from ERC results. Endoscopic stone removal was performed using a balloon catheter after sphincterotomy. After treatment, the cholangitis improved over time. The patient was discharged with improved symptoms. Magnetic resonance imaging conducted 3 months later revealed no evidence of intrahepatic or extrahepatic bile duct dilatation. Multiple cystic lesions in the liver had all disappeared (Fig. 1-C). As a differential disease of this case, findings of multiple cystic lesions in the liver from magnetic resonance imaging suggested Caroli’s disease, with von Meyenburg’s complex as an underlying pathology [1,2]. Caroli’s disease involves congenital cystic dilatation of the peripheral intrahepatic bile ducts. Diffuse lesions occur in bilateral liver lobes in many cases. Caroli’s disease is a rare autosomal recessive disorder that is known to be complicated by autosomal recessive polycystic kidney disease. Nevertheless, no renal lesion was found in this case. In contrast, von Meyenburg’s complex is characterized by bile duct hamartomas containing innumerable diffuse multiple
cystic lesions. Both of those findings are similar to findings obtained for this patient, but they are not conditions that improve with bile duct drainage. Multiple liver abscesses were also differentiated. However, multiple cystic lesions disappeared immediately after stone removal. This clinical course was regarded as atypical for the course of treatment for liver abscess. For this case, peripheral bile ducts in the liver were considered to show cystic dilatation due to biliary obstruction. According to the classifications of intrahepatic cystic dilatation and related diseases, these findings consistent with dilatation of the intrahepatic biliary ducts without the Meyenburg complex [3]. To date, no case of acute cholangitis showing similar imaging has been reported in the literature. It is considered a very rare condition. In fact, it remains unclear why bile ducts exhibit such peculiar morphology. However, the pathological findings in this case should be understood as a differential diagnosis of multiple intrahepatic cystic lesions.

References:
Figure 1. 1-A Magnetic resonance cholangiopancreatography findings at admission. Dilated intrahepatic and extrahepatic bile ducts and multiple cystic lesions are present diffusely throughout the liver. 1-B T2-weighted images show diffuse multiple cystic lesions and stone formation in the common bile duct. 1-C Magnetic resonance cholangiopancreatography findings obtained at 3 months after discharge. Bile duct dilation and multiple cystic lesions have disappeared.