A 77-year-old woman attended our hospital with a 10-day history of epigastric and right upper quadrant abdominal pain. Her past medical history included diabetes mellitus and hypertension, and she had undergone open cholecystectomy for gallstones 10 years previously. Physical examination revealed tenderness in the right upper quadrant of the abdomen. Laboratory findings were as follows: white blood cell count 4802/μL, total bilirubin 0.58 mg/dL, γ-glutamyltransferase 17 IU/L, and alkaline phosphatase 207 IU/L. Abdominal computed tomography (CT) showed multiple stones with diffuse dilatation of the intrahepatic and extrahepatic biliary tree (Fig. 1). Endoscopic retrograde cholangiopancreatography (ERCP) showed a markedly dilated extrahepatic bile duct with multiple bile duct stones (Fig. 2). After endoscopic sphincterotomy followed by endoscopic papillary large balloon dilation, bile duct stones were removed using a Dormia basket and retrieval balloon, after which endoscopic retrograde biliary drainage was placed. At 3 months after discharge, the patient returned to our emergency department with fever and abdominal pain. Abdominal CT on this occasion showed a multiseptated, low-density lesion in the right lobe of the liver, about 6 cm in size, suggestive of abscess formation, and stone remnants in the biliary tree (Fig. 3). Percutaneous drainage of the hepatic abscess was performed. Magnetic resonance cholangiopancreatography showed extrahepatic bile duct duplication with choledocholithiasis. ERCP confirmed the bile duct duplication complicated by common bile duct stones, and the stone remnants in the right extrahepatic bile duct were removed (Fig. 4). Congenital extrahepatic bile duct duplication is extremely rare, and to the best of our knowledge there have only been two cases [1, 2] of type Va extrahepatic bile duct duplication according to the modified classification of Choi et al. [1]. We report a case of extrahepatic bile duct duplication associated with multiple bile duct stones without a communicating channel between the two extrahepatic bile ducts.

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Fig. 4 ERCP demonstrated extrahepatic bile duct duplication without communication between the two separate bile ducts.

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