Anatomic abnormalities associated with the opening of the pancreas and biliary systems into the upper gastrointestinal tract have been increasingly recognized since endoscopic retrograde cholangiopancreatography (ERCP) has come into wide use in clinical practice [1].

A 38-year-old Chinese woman presented with a 4-month history of dull abdominal pain. She had no history of endoscopic intervention (e.g., ERCP or sphincterotomy) or surgery. Physical examination revealed mild abdominal tenderness in the right upper quadrant. Serum levels of triglycerides, calcium, carcinoembryonic antigen, and amylase were normal. Serum carbohydrate antigen 19–9 was moderately elevated (202.60 U/mL, reference value: 0–37 U/mL). Computed tomography (CT) of the abdomen revealed diffuse pneumatosis of the intrahepatic (Fig. 1) and common bile ducts. CT also found dilatation of the pancreatic duct and a high-density shadow in the head of the pancreas (Fig. 2). Magnetic resonance cholangiopancreatography showed dilatation of the pancreatic duct and bile duct (Fig. 3).

ERCP was performed to clear the main pancreatic duct. During the procedure, it became evident that the major duodenal papilla of the patient was absent. Instead, there were two orifices (Fig. 4), one on the right for the pancreatic duct and one on the left for the common bile duct. They were subsequently visualized by cholangiopancreatography. The bile duct was irregularly dilated and contrast emptying was delayed (Fig. 5). ERCP also revealed a dilated, tortuous main pancreatic duct containing stones (Fig. 6).

The etiology of this anomalous opening is unknown. In patients with this defect, duodenal content could easily pass through the biliary tract and pancreatic duct (duodenobiliary reflux). It has been reported that after endoscopic sphincterotomy – which is essentially an artificial opening defect – the occurrence of cholangitis is generally about 3% [2, 3]. Chronic pancreatitis is a very rare complication after sphincterotomy [4, 5]. Long-term absence of the duodenal papilla can lead to chronic pancreatitis and cholangitis.

**Competing interests:** None
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