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CASE REPORT | BILIARY

A Case of an Ectopic Ampulla of Vater in the Pyloric Channel

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Abstract
A 51-year-old male presented with abdominal pain and jaundice. He was subsequently diagnosed with cholestatic jaundice and cholangitis. A side-viewing duodenoscope failed to identify the ampulla of Vater in the second portion of duodenum. A regular gastroscope was used, and an ectopic ampulla of Vater was identified in the pyloric channel.

Introduction
The ampulla of Vater encompasses the openings of both the common bile duct and pancreatic duct. It is typically located within the wall of the duodenum, surrounded by the small circular and longitudinal muscular segments that comprise the sphincter of Oddi. We report the case of a man who presented with cholestatic jaundice and cholangitis, and was found to have an ectopic ampulla of Vater in the pyloric channel.

Case Report
A 51-year-old male presented with complaints of intermittent abdominal pain and chills for a few days prior to admission. He was afebrile with normal vital signs. His physical examination was significant for scleral icterus and a benign abdominal exam. Laboratory data were significant for an aspartate transaminase (AST) of 153 IU/L, alanine transaminase (ALT) of 177 IU/L, alkaline phosphatase of 169 IU/L, total bilirubin of 5.5 mg/dL, and a direct bilirubin of 3.5 mg/dL.

Computed tomography (CT) suggested possible annular pancreas with pneumobilia. Right upper quadrant ultrasound revealed choledolithiasis, pneumobilia, and a common bile duct diameter of 8 mm. Magnetic resonance cholangiopancreatography (MRCP) showed a possible annular pancreas with hepatic and pancreatic duct dilation. On attempted endoscopic retrograde cholangiopancreatography (ERCP), the ampulla of Vater was not visualized in the second portion of the duodenum. A regular gastroscope was used for visualization and revealed a 2-cm gastric ulcer in the distal antrum with significant surrounding inflammation (Video 1). Distal to the ulcer in the pyloric channel, two orifices were identified. One of them was draining bile and the other one was draining clear liquid. This was consistent with the biliary orifice and pancreatic orifice, respectively (Figure 1). A sphincterotome with a 0.035 wire was used to enter the orifice that was draining bile, which resulted in deep common bile duct cannulation.

A cholangiogram showed mild intrahepatic duct dilation, common bile duct dilation (to 1 cm), and a “hook-shaped” configuration of the distal common bile duct. These findings were consistent with an ectopic ampulla within the pylorus. A balloon occlusion cholangiogram showed a distal common bile duct diameter of 7 mm and a round filling defect consistent with a bile duct stone (Figure 2). A 3-mm limited sphincterotomy was performed at the ampullary orifice, followed by balloon dilation to 10 mm for 45 seconds under endoscopic and fluoroscopic guidance.
Most of the reported cases suggest the presence of the major papilla in the stomach. The minor papilla is reportedly found in the distal duodenum. Based on few published reports, there appears to be an increased risk for choledocholithiasis due to anomalous bile drainage and lack of sphincter control mechanisms. Pancreatobiliary secretions can lead to mucosal damage with ulcer formation. The anomalous location in the pylorus can facilitate reflux of gastric contents into the biliary tree, predisposing to biliary tree injury and cholangitis.

Endoscopists who perform ERCP should be aware of this rare anomaly. In cases where the ampulla cannot be seen in a normal anatomical location, an ectopic ampulla should be suspected, and the endoscopist must carefully examine the stomach and the duodenum. As in our patient, a regular, forward-viewing gastroscope can be used to better visualize the stomach. A large sphincterotomy should be avoided due to increased risk of perforation. Ampullary balloon dilation is recommended to extract large bile duct stones.

Discussion

Ectopic location of ampulla in the stomach or first, third, or fourth part of the duodenum is a very rare congenital anomaly. The reported incidence of anomalous termination of bile duct is 5.6–23%; the wide range is due to the limited number of cases, but this condition is being reported more frequently due the extensive availability of ERCP. The exact etiology of this congenital anomaly remains unclear, and is believed to be an abnormality during the embryonic developmental period. The liver and the biliary tract are formed from the hepatic diverticulum; the cranial part (pars hepatica) gives rise to the intrahepatic and common hepatic ducts. It has been proposed that an earlier subdivision of the hepatic diverticulum during embryogenesis could cause the common bile duct to empty into ectopic locations.

Ectopic biliary drainage into the stomach is most commonly reported in the body of the stomach, followed by the antrum and cardia. Most of the reported cases suggest the presence of the major papilla in the stomach. The minor papilla is reportedly found in the distal duodenum. Based on few published reports, there appears to be an increased risk for choledocholithiasis due to anomalous bile drainage and lack of sphincter control mechanisms. Pancreatobiliary secretions can lead to mucosal damage with ulcer formation. The anomalous location in the pylorus can facilitate reflux of gastric contents into the biliary tree, predisposing to biliary tree injury and cholangitis.
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