A Case of Primary Neuroendocrine Carcinoma of the Gallbladder Associated with Anomalous Union of the Pancreaticobiliary Duct

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Primary neuroendocrine carcinoma of the gallbladder is extremely rare because normal gallbladder mucosa does not contain neuroendocrine cells. Neuroendocrine cells can be detected at sites of intestinal metaplasia induced by chronic inflammation, which may be the initial step in the development of neuroendocrine tumor of the gallbladder. Anomalous union of the pancreaticobiliary duct (AUPBD) is an uncommon congenital anomaly that is frequently associated with cholelithiasis, cholangitis, pancreatitis, and cancer of the gallbladder or bile duct. In AUPBD, cancers of the gallbladder and bile duct can be induced by chronic inflammation. We report herein a case of large-cell neuroendocrine tumor of the gallbladder associated with AUPBD. (Gut and Liver 2009;3:231-234)

Key Words: Anomalous union; Pancreaticobiliary duct; Neuroendocrine tumor; Gallbladder

INTRODUCTION

Although neuroendocrine tumor can be seen frequently in gastrointestinal tract, primary neuroendocrine carcinoma of the gallbladder is rare and among them, large cell neuroendocrine carcinoma is extremely rare.

Anomalous union of pancreaticobiliary duct (AUPBD) is an uncommon congenital anomaly which defined as a junction of the bile duct and pancreatic duct outside the duodenal wall. This anomaly is frequently associated with cholelithiasis, choledocholithiasis, cholangitis, pancreatitis and malignancy of the gallbladder or bile duct.

We report a case of AUPBD associated with large cell neuroendocrine carcinoma of the gallbladder.

CASE REPORT

A 38-year-old woman was admitted to Chonnam National University Hospital with 10 kg-weight loss for 2 months. Physical examination showed an icteric sclera and a tender palpable mass in the right upper quadrant of the abdomen. The blood chemistry were albumin 4.4 g/dL, AST 162 U/L, ALT 189 U/L, alkaline phosphatase 255 U/L, total bilirubin 7.3 mg/dL, direct bilirubin 4.3 mg/dL, gamma glutamyl transpeptidase 1,098 U/L, amylase 30 U/L, and prothrombin time 10.2/129.9/0.89 sec/INR. The CA 19-9 was 79.9 U/mL.

The chest X-ray was within normal limit. The abdominal ultrasound demonstrated a 5 cm sized mixed echoic mass in S4 segment of the liver and an 8 cm sized mass in the fundus of the gallbladder. Abdominal computerized tomography (CT) showed a 7.8×6.2 cm sized exophytic necrotizing mass in the fundus of the gallbladder. There were multiple conglomerated necrotizing lymphadenopathies in the hepatic hilum which invade confluent portion of the intrahepatic bile ducts, cystic duct, portal vein and hepatic artery (Fig. 1).

In endoscopic retrograde cholangiopancreatography (ERCP), there was a filling defect in the common hepatic duct. When the radiocontrast dye was injected to proximal portion of the common bile duct (CBD), the pancreatic duct was visualized at the same time and the length of common channel was 30 mm (Fig. 2A). After endoscopic biliary sphincterotomy (EST), the endoscopic nasobiliary drainage (ENBD) catheter was inserted. The ENBD
Fig. 1. (A) CT scan showing a 7.8×6.2-cm exophytic necrotizing mass in the fundus of the gallbladder. (B) CT scan showing multiple conglomerated necrotizing lymphadenopathies in the hepatic hilum. The proximal CBD, cystic duct, portal vein, and hepatic artery were invaded. 

CT, computerized tomography; CBD, common bile duct.

Fig. 2. (A) A filling defect in the common hepatic duct with proximal dilatation was noted on ERCP. The length of the common channel was 30 mm. (B) The ENBD cholangiogram showed a long common channel, and the pancreatic duct was visualized simultaneously with the common bile duct. ERCP, endoscopic retrograde cholangiopancreatography; ENBD, endoscopic nasobiliary drainage.

Fig. 3. An endoscopic biliary metal stent was inserted from the first branch of the right intrahepatic bile duct to the mid common bile duct.

The cholangiogram (Fig. 2B) showed long common channel (over 20 mm), and pancreatic duct was visualized with CBD at the same time. The analysis of bile juice showed amylase 33,762 U/L, lipase 185,080 U/L, and CA 19-9 18,100 U/mL.

On hospital day 5, she complained abdominal pain and the blood chemistry were amylase 143 U/L and lipase 641 U/L. After conservative treatment, abdominal pain was relieved and the levels of serum amylase and lipase were normalized. On hospital day 12, a biliary stent (10×40 mm, covered, Wall stent, Endoprosthesis, Boston scientific, Galway, Ireland) was inserted endoscopically (Fig. 3). Biopsy was performed under the ultrasonographic guidance. The tumor showed a monotonous proliferation of small round cells with hyperchromatic nuclei and scanty cytoplasm, resembling neuroendocrine carcinoma. Immunohistochemical staining for chromogranin