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Mixed adenoneuroendocrine carcinoma derived from the cystic duct: A case report

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ABSTRACT

INTRODUCTION: Mixed adenoneuroendocrine carcinomas (MANECs) derived from cystic duct are extremely rare.
PRESENTATION OF CASE: An 80-year-old woman was admitted to the department of surgery, Onomichi general hospital with abnormal liver function and jaundice. Enhanced abdominal computed tomography (CT) detected a well-enhanced papillary tumor in the cystic duct, which protruded into the common bile duct. The intrahepatic bile duct was dilated due to tumor obstruction. The entire tumor showed high intensity in T2-weighted magnetic resonance imaging (MRI) imaging. Endoscopic retrograde cholangiopancreatography (ERCP) showed that the tumor ranged from part of communication of three ducts (cystic, common hepatic and common bile duct), to the middle of common bile duct. Biliary cytology determined a class V malignancy (adenocarcinoma). Endoscopic ultrasound determined that the tumor was primarily at the cystic duct with heterogeneous echoic pattern, which extended into the common bile duct. The preoperative diagnosis was cystic duct carcinoma (T3N0M0, StageIIIA). An extended cholecystectomy with regional lymph nodes dissection was performed. Histologically, the tumor had components of both well-differentiated tubular adenocarcinoma and neuroendocrine carcinoma, which is classified as MANECs according to the 2010 WHO classification of endocrine tumors. Eight months after surgery, multiple liver metastases were discovered, and treatment with adjuvant chemotherapy was initiated.
DISSCUSION: We present a rare case of MANECs derived from cystic duct. Until now, an established adjuvant systemic chemotherapy has not emerged, and curative resection, with poor long-term prognosis, remains the only treatment option.
CONCLUSION: Though standards of treatment for MANECs have not been established,multidisciplinary theraphy is necessary to improve outcome.

1. Background

This work has been reported in line with the SCARE criteria. R.A. Agha, A.J. Fowler, A. Saeta, I. Barai, S. Rajmohan, D.P. Orgill, The SCARE Statement: Consensus-based surgical case report guidelines, Int. J. Surg. (2016).

According to the 2010 World Health Organization (WHO) classification of endocrine tumors, at least 30% of neoplasms are defined as mixed adenoneuroendocrine carcinomas (MANECs) [1]. These tumors include: composite carcinoid, mucin-producing carcinoid, Argentaffin cell adenocarcinoma, goblet cell carcinoid, adenocarcinoid, and small cell undifferentiated carcinoma. Neuroendocrine tumors (NETs) commonly occur in the gastrointestinal tract, pancreas, and lung, but bile duct origin of NETs is uncommon [2]. Some cases of MANEC derived from the bile duct have been reported [3–6]. Although NECs of the bile duct are frequently associated with liver or lymph node metastases in the preoperative state, aggressive radical operative therapy and adjuvant chemotherapy contributes to long-term outcomes [7]. Herein, we report a rare case of MANEC that originated from the cystic duct following curative intent surgery.

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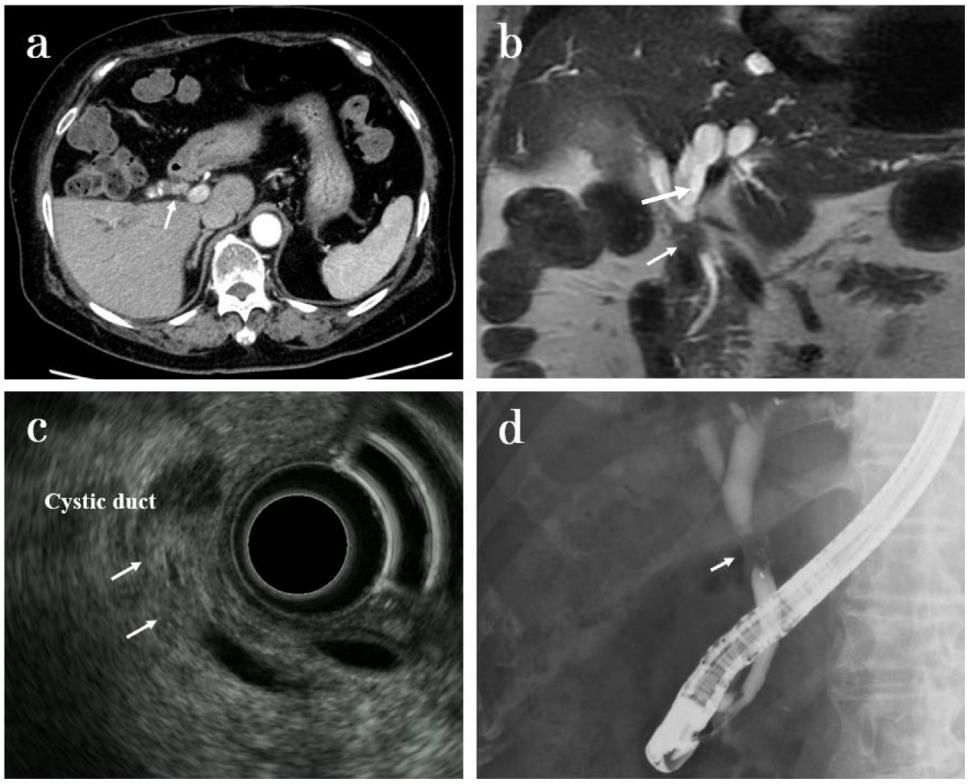

Fig. 1. a: Abdominal computed tomography (CT) indicates mass formation at the part of communication of cystic, common hepatic and common bile duct (white arrow).
b: Magnetic resonance cholangiopancreatography (MRCP) reveals dilation of the common bile duct and signal defects. (white arrow).
c: Endoscopic ultrasonography (EUS) reveals a mass in the cystic duct extending to common bile duct (white arrow).
d: Endoscopic retrograde cholangiopancreatography (ERCP) shows a filling defects in the communication of cystic, common hepatic and common bile duct (white arrow).

2. Case presentation

An 80-year-old woman was admitted at the department of surgery, Onomichi general hospital, with liver function abnormalities and jaundice. The patient had a medical history of hypertension treated with calcium blocker and angiotensin receptor blocker.

Laboratory data revealed an elevation of total bilirubin (11.18 mg/dL), aspartate aminotransferase (162 U/L), alanine amino transferase (269 U/L), alkaline phosphatase (1482 U/L), and γ-glutamyltransferase (1156 U/L). There was no elevation in serum tumor markers (carcinoembryonic antigen, carbohydrate antigen 19-9). The patient's Child-Pugh score grade was B, and neither hepatitis virus B nor hepatitis virus C infection were detected. Abdominal contrast enhanced computed tomography (CT) imaging revealed a tumor located in the communication of three ducts (the cystic, common hepatic, and common bile ducts), as well as dilation of the common hepatic and intrahepatic bile ducts (Fig. 1-a). On magnetic resonance imaging (MRI), the entire tumor showed low signal in T1-weighted images and enhanced signal in T2-weighted images (Fig. 1-b). The tumor was 1.0 × 0.8 cm in size. The CT showed that the tumor was highly enhanced in the arterial phase, and this enhancement pattern was prolonged in the delay phase. An endoscopic ultrasound showed that the tumor had a heterogeneous echoic pattern, which was mainly located in the cystic duct. Endoscopic retrograde cholangiopancreatography showed that the dilation of the common hepatic and the intrahepatic bile ducts was due to tumor obstruction. In addition, the cystic and common bile ducts' communication showed severe stenosis. No portal vein or right hepatic artery invasion was observed. The range of horizontal extension of the tumor was considered to be from the part of communication of the main hepatic duct to the middle bile duct (Fig. 1-c and d). Biliary cytology was determined to be class V (adenocarcinoma). The preoperative diagnosis was a cystic duct carcinoma, T3N0M0 Stage IIIA, according to 7th edition Union for International Cancer Control (UICC) TNM system. An extended cholecystectomy, extrahepatic bile duct resection, regional lymph nodes dissection, and choledochojejunostomy were performed; with a blood loss of 240 mL and an operative time of 304 min. Post operation, an intraabdominal abscess at the hepatic bed presented 8 days after surgery. This was treated by percutaneous drainage, which was graded as IIIA in according to the Clavien-Dindo classification [8].

Macroscopically, the tumor was mainly located from the cystic duct to the common biliary duct (Fig. 2-a). On microscopic examination, tumor cells extended from the cystic duct to the common bile duct. In the mucosal lesion, well-differentiated tubular adenocarcinoma was observed. In the invasive area, the tumor cells with hyperchromatic nuclei and scant cytoplasm were spread (Fig. 2-b and c). Immunohistochemically, the tumor cells were diffusely positive for chromogranin A, synaptophysin, and CD56. The Ki-67 labeling index was 80.0%. These findings were consistent with neuroendocrine carcinoma according to the WHO criteria for the clinicopathologic classification of neuroendocrine tumors (Fig. 3). Massive venous invasion of neuroendocrine carcinoma cells was detected. Neuroendocrine tumor cells extended into the subserosal layer (SS). The final pathological diagnosis was mixed adenoneuroendocrine carcinoma (MANEC) of cystic duct, with tumor dimensions of 13 × 13 × 7 mm, Neuroendocrine carcinoma ≫ Well differentiated tubular adenocarcinoma, pT3, N0, M0, pStage IIIA according to 7th edition UICC TNM system. Eight months after surgery, multiple liver metastases were discovered, and treatment with adjuvant chemotherapy was initiated.