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A long-term recurrence-free survival of a patient with the mixed adeno-neuroendocrine bile duct carcinoma: A case report and review of the literature

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ABSTRACT

INTRODUCTION: Neuroendocrine tumors arising primarily in the bile duct are rare. And among these tumors, mixed adeno-neuroendocrine carcinoma (MANEC) is quite uncommon. We report a patient with MANEC who achieved long-term recurrence-free survival. And our case report includes analysis previous case reports.

PRESENTATION OF CASE: A 66-year-old man underwent investigation for persistent anorexia and fatigue. Laboratory tests showed that the values of hepatobiliary enzymes were increased. On CT, a 10 mm × 8 mm hypervascular tumor was observed in the distal bile duct and the proximal bile duct was markedly dilated. Endoscopic retrograde cholangiography (ERC) also showed a stenosis with a long diameter of 10 mm. Examination of a biopsy specimen obtained from the narrow site of the bile duct at the time of ERC revealed tubular adenocarcinoma. Therefore, pylorus-preserving pancreaticoduodenectomy was performed under a preoperative diagnosis of distal bile duct carcinoma. Postoperative pathologic examination revealed alveolar structures and a mixture of moderately differentiated adenocarcinoma with synaptophysin-positive and chromogranin-A-positive neuroendocrine carcinoma. Therefore, the final diagnosis was MANEC, pT3, pN1, M0, pStage II B (TNM classification of the UICC). Curative resection was achieved and there has been no recurrence after 30 months.

DISCUSSION: In the previous reports, only five patients (14.7%) survived for 24 months or longer. Median survival was longer (14 months) in the curative resection group and shorter (6 months) in the non-curative resection group.

CONCLUSION: Curative resection is essential to achieve long-term survival in patients with bile duct MANEC, even if these patients have lymph node metastasis.

1. Introduction

Neuroendocrine tumors arising primarily in the bile duct are rare. Among these tumors, mixed adeno-neuroendocrine carcinoma (MANEC) is quite uncommon and its prognosis is thought to be poor. We report a patient with MANEC arising primarily in the bile duct who achieved long-term recurrence-free survival.

2. Presentation of case

A 66-year-old man underwent investigation for persistent anorexia and fatigue which had persisted for one month. On his past history and family history, there was nothing noteworthy, including no evidence of multiple endocrine neoplasia type1or von Hippel-Lindau disease. And he was not a smoker or a drinker. On his occupational history, he had not worked in printing industry where incidence of the bile duct carcinoma is known to be high. On his physical examination, the bulbar conjunctiva was yellow. Laboratory tests showed that the values of hepatobiliary enzymes were increased (T-Bil was 8.9 mg/dl, D-Bil was 5.6 mg/dl, AST was 377 U/L, ALT was 653 U/L, ALP was 1105 U/l, and γ-GTP was 2291 U/l). CEA and CA19-9 were within normal limits. On abdominopelvic contrast-enhanced CT, an enhancing 10 mm × 8 mm hypervascular tumor was observed in the distal bile duct and the proximal bile duct was markedly dilated (Fig. 1).

Abbreviations: CT, computed tomography; ERC, endoscopic retrograde cholangiography; MANEC, mixed adeno-neuroendocrine carcinoma; MRI, magnetic resonance imaging; NEC, neuroendocrine carcinoma; NEN, neuroendocrine neoplasm; NETs, neuroendocrine tumors.

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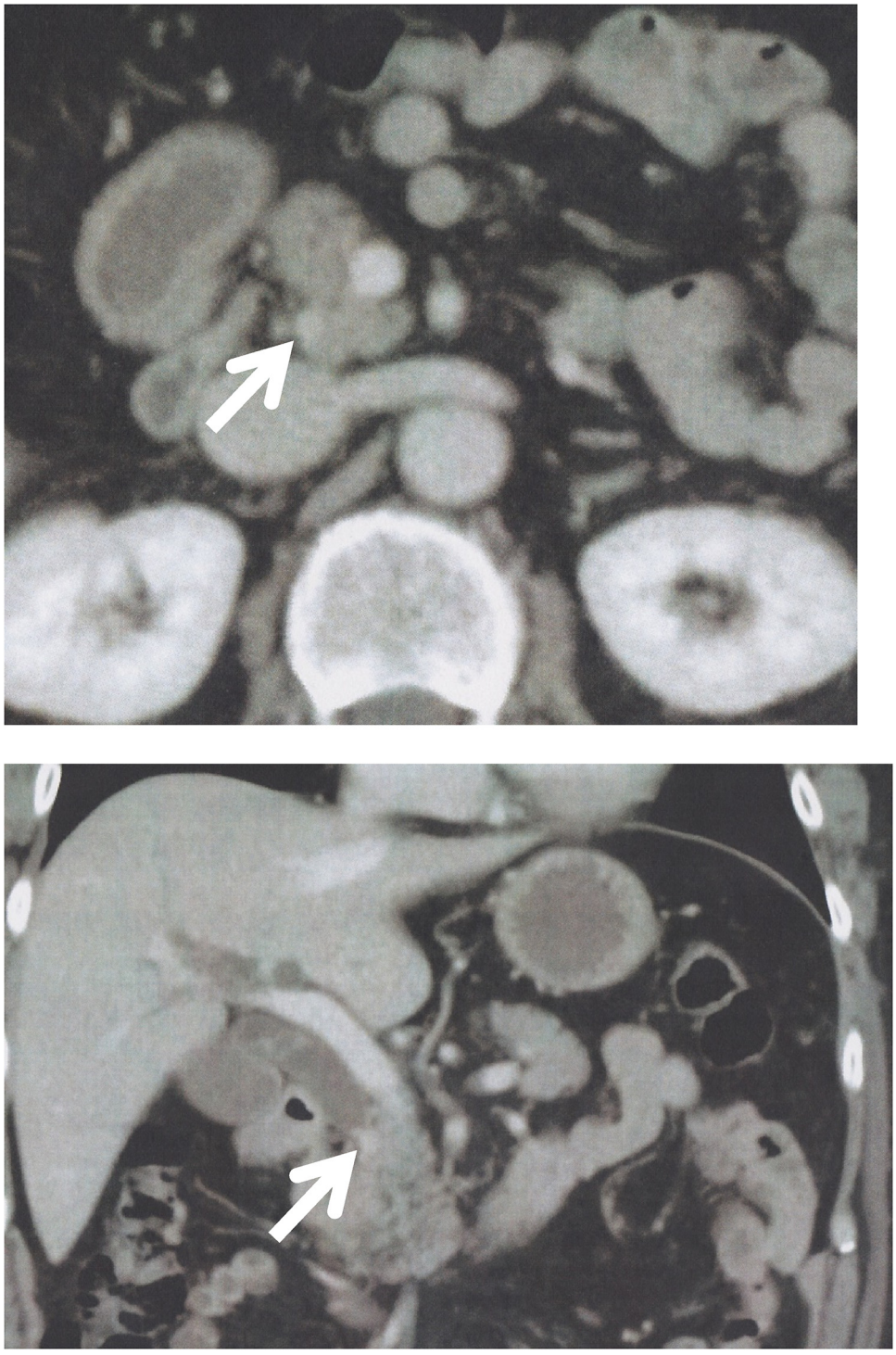

Fig. 1. Abdominal contrast-enhanced CT scan: an enhancing tumor of 10 mm × 8 mm in size was seen in the distal bile duct.

MRCP demonstrated a filling defect spanning 10 mm in the maximal diameter in the distal bile duct with markedly dilatation of the proximal bile duct. There was no evidence of pancreatobiliary maljunction (Fig. 2). Endoscopic retrograde cholangiography (ERC) also showed a stenosis of 10 mm in the maximal diameter was observed in the distal bile duct (Fig. 3). Examination of a biopsy specimen obtained from the narrow site of the bile duct at the time of ERC revealed moderately differentiated tubular adenocarcinoma. With a preoperative diagnosis of distal bile duct cancer, pylorus-preserving pancreaticoduodenectomy was performed. On surgical findings, a median incision was made in the upper abdomen and observation of the abdominal cavity did not reveal tumor dissemination or liver metastasis. The tumor was palpable as an induration in the distal bile duct. Pylorus-preserving pancreaticoduodenectomy (Child II-A) was performed. The operating time was 343 min and blood loss was 533 mlMacroscopic findings (Fig. 4) shows a white nodular infiltrating tumor of 10 mm in the maximal diameter was observed in the distal bile duct. Histopathological findings (Fig. 5) shows the tumor composed of cells in solid clusters and cells forming tubular structures with abundant fibrous