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Signet ring cell carcinoma of the ampulla of vater: Report of a case and a review of the literature

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ABSTRACT

INTRODUCTION: Signet ring cell carcinoma (SRCC) of the ampulla of vater is a very rare tumor that is reported infrequently in the literature.
PRESENTATION OF CASE: A 59-year-old woman visited our hospital for evaluation of elevated transaminase levels. On laboratory examination of tumor marker levels, carcinoembryonic antigen (CEA) and carbohydrate antigen (CA) 19–9 levels were normal, and DUPAN-2 was elevated. Computed tomography (CT) confirmed a 2 cm, enhanced mass in the periampullary region, with marked common bile duct dilatation. Endoscopic retrograde cholangiopancreatography (ERCP) showed a swollen papilla of vater, with a reddish, erosive mucosa. Histological examination of biopsy samples from the ampulla of vater showed signet ring cell carcinoma (SRCC). The patient underwent radical pancreatoduodenectomy. Pathological examination showed that the SRCC had infiltrated into the duodenal muscularis propria and pancreatic parenchyma, and lymph node metastases were identified around the abdominal aorta and common hepatic artery. Based on the immunohistochemical staining patterns of the positive results for CDX2 and MUC2, the tumor cells in the present case appeared to have an intestinal type origin. The ampullary cancer was diagnosed as T3bN1M1, Stage IV according to the International Union Against Cancer TNM classification (UICC). After undergoing adjuvant chemotherapy with cisplatin–gemcitabine chemotherapy for 6 months, the patient has remained disease-free in the 7 months since surgery.
DISCUSSION: SRCC of intestinal-type origin is associated with a favorable outcome.
CONCLUSION: Investigation to confirm the histological origin of SRCC by immunohistochemical staining might inform the treatment strategy and identify patients with ampullary SRCC who may have a good prognosis.

1. Introduction

Most tumors of the ampulla of vater are well-differentiated adenocarcinomas. Signet ring cell carcinoma (SRCC) at this site is uncommon [1], and only 26 resected cases have been previously reported in the English literature. A rare case of a 59-year-old woman with SRCC and paraaortic lymph node metastases is presented.

2. Case presentation

A 59-year-old woman visited our hospital for evaluation of elevated transaminase levels. She had no symptoms, and her past history was unremarkable. The physical examination findings were also unremarkable, with no abdominal tenderness or palpable masses. Routine laboratory test results were: aspartate aminotransferase (AST) 5 IU/L (normal 10–33 IU/L); alanine aminotransferase (ALT) 121 IU/L (normal 6–35 IU/L); alkaline phosphatase 884 IU/L (normal 120–340 IU/L); γ-glutamyltransferase 684 IU/L (normal 8–60 IU/L); total bilirubin (T-bil) 0.5 mg/dL (normal 0.4–1.4 mg/dL); amylase 87 IU/L (normal 31–106 IU/L); lipase 49 IU/L (normal 8–46 IU/L); and normal inflammatory markers. Laboratory test results for tumor markers were: carcinoembryonic antigen (CEA) 2.9 ng/mL (normal 0–5.0 ng/mL); carbohydrate antigen (CA) 19–9 2 U/mL (normal 0–37 U/mL); DUPAN-2 940 U/mL

Abbreviations: SRCC, signet ring cell carcinoma; PD, pancreatoduodenectomy; PPPD, pylorus-preserving pancreatoduodenectomy.

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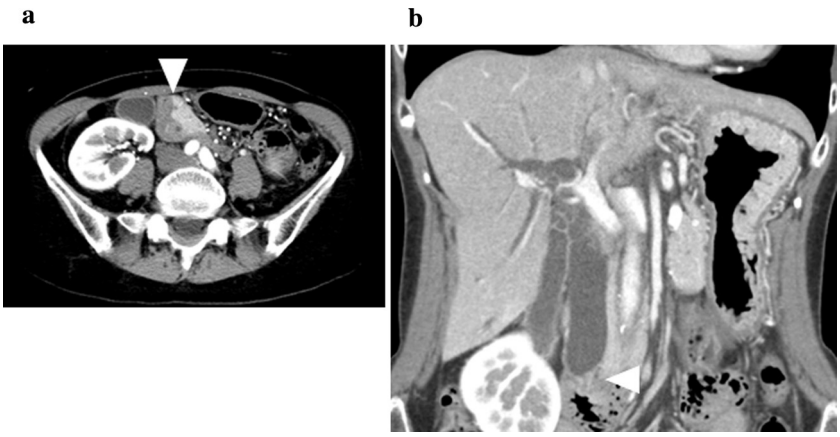

Fig. 1. Computed tomography: CT scan confirms a 2 cm, enhanced mass in the periampullary region (arrowhead) and shows marked common bile duct dilatation.

(normal 0–150 U/mL); and SPAN-1 16 U/mL (normal 0–30 U/mL). Abdominal ultrasonography demonstrated gross dilatation of the common bile and pancreatic ducts. Subsequent computed tomography (CT) confirmed a 2 cm, enhanced mass in the periampullary region, marked common bile duct dilatation, and no evidence of lymphadenopathy or distant metastases (Fig. 1). Positron emission tomography with 2 [18 F]-fluoro-2-deoxy-D-glucose (FDG) showed a mass with an SUVmax of 4.2, consistent with the CT scan findings. Endoscopic retrograde cholangiopancreatography (ERCP) revealed a swollen papilla of vater with a reddish, erosive mucosa (Fig. 2). Cholangiography demonstrated an abrupt obstruction of the lower common bile duct. Histological examination of the biopsy samples from the ampulla of vater showed SRCC. To decompress the biliary system, an endoscopic retrospective biliary drainage tube (8.5 Fr) was inserted before surgery. The patient underwent surgery based on a diagnosis of SRCC of the ampulla of vater in September 2014. Surgery revealed no liver metastases or peritoneal dissemination, and a radical pancreatoduodenectomy was performed. Microscopic examination of the ampullary tumor revealed a poorly differentiated adenocarcinoma, of signet ring cell type that infiltrated the duodenal wall and adjacent pancreas (Fig. 3). There was lymphatic and vascular invasion. Lymph node metastases were identified around the abdominal aorta and common hepatic artery. Immunohistochemical staining was performed, and the signet ring cells were positive for CK7, CK19, CK20, CDX2, and MUC2, but negative for MUC5AC and E-cadherin (Fig. 4). The ampullary cancer was diagnosed as T3bN1M1, Stage IV according to the International Union Against Cancer TNM classification (UICC). After undergoing adjuvant chemotherapy with cisplatin-gemcitabine chemotherapy for 6 months, the patient has remained disease-free in the 7 months since surgery.

3. Discussion

Adenocarcinoma of the ampulla of vater is rare, with an incidence of less than 6 cases per million persons annually. It represents 0.2% of all gastrointestinal malignancies, and accounts for only 6% of all cancers developing in the periampullary region [1]. SRCC in the ampulla of vater is extremely uncommon, and in a PubMed search of the English literature using the key words SRCC and ampulla of vater; it was found that only 26 such well-documented; resected cases have been previously described (Table 1) [2–19].

Including the present case, the 26 cases consisted of 12 men and 14 women. The median age at diagnosis was 61 years (range, 38–83 years). The median tumor diameter was 20 mm (mean, 26 mm; range, 10–95 mm). Six cases had UICC T2 (duodenal invasion) tumors, twelve cases had T3 (pancreatic invasion) tumors, and three cases had T4 tumors. Previous reports of ampullary SRCCs

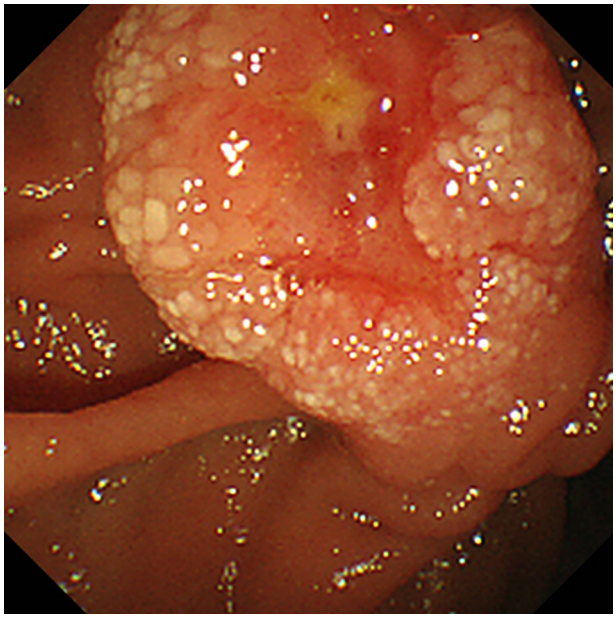

Fig. 2. Endoscopic retrograde cholangiopancreatography: ERCP shows a swollen papilla of vater with a reddish, erosive mucosa.

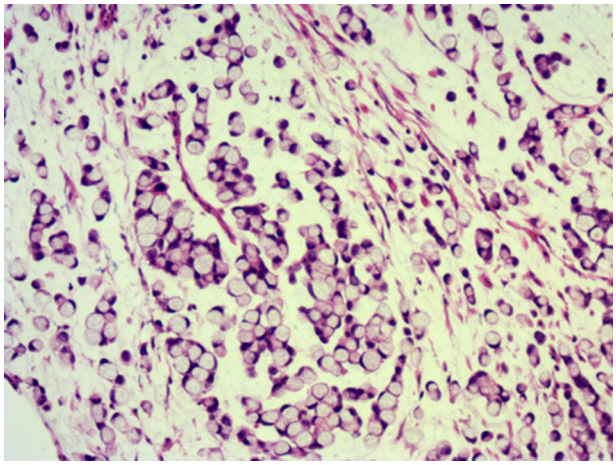

Fig. 3. Microscopic examination: the ampullary tumor is a poorly differentiated adenocarcinoma of signet ring cell type.