Annals of Surgical Case Reports

Case Report
Published: 12 May, 2022

Signet Ring Cell Carcinoma of the Ampulla of Vater: Report of a Case with Long-Term Survival

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Abstract

Ampullary adenocarcinomas are extremely rare tumors and constitute approximately 0.2% of all gastrointestinal malignancies. Signet ring cell variant of ampullary tumors is much rarer and has a poor prognosis, and chemotherapy has been reported to have no effect on survival. In this study, a 68-year-old male patient with Signet Ring cell Ampullary Adenocancer (SRCA) is presented. Radiologically, an ampullary mass was detected in the patient admitted to the emergency department with abdominal pain, nausea, vomiting, and jaundice.

Histopathological examination revealed ampullary signet ring cell carcinoma. There was no metastasis and the tumor was resectable in radiological evaluation, so the patient underwent Pylorus Preserving Pancreaticoduodenectomy (PPPD). The patient was discharged without complications on the 12th postoperative day. Histopathological evaluation revealed that the tumor was T4N1, and adjuvant chemotherapy was recommended to the patient, but the patient refused chemotherapy. No recurrence was detected in the postoperative 4 years follow-up.

Keywords: Ampulla of Vater; General surgery; Hepatobiliary; Signet ring cell carcinoma

Introduction

The incidence of malignancy in the ampulla, which has a complex pancreaticobiliary and intestinal histological structure, is reported to be 2 to 6 per million in the general population [1,2]. Ampullar signet ring cell cancers are very rare types, and only case reports and case series are reported in the literature. SRCA, which is usually encountered in the stomach, can also be seen in the gastrointestinal tract, hepatopancreaticobiliary system, and urogenital system [3]. A SRCA account for approximately 15% to 30% of stomach tumors and is characterized by more than 50% mucin-secreting signet ring cells [4]. SRCA has a lower response to chemotherapy and has a worse prognosis compared to other types of ampullary tumors [5,6]. In the literature review, 41 cases were reported in the pairing of signet ring cell carcinoma and ampulla vatery. In this study, a 68-year-old male patient with T4N1 histopathology according to the TNM classification, who did not receive adjuvant chemotherapy and has no recurrence in long-term follow-up (4 years), is presented.

Case Presentation

A 68 year-old man with upper abdominal pain, nausea, vomiting and jaundice was admitted to emergency room. The laboratory results were; ALT 269 U/L, ALP 889 U/L, GGT 1117 U/L, amylase 1753 U/L, total bilirubin 5.93 mg/dl, and direct bilirubin 4.63 mg/dl. The patient was admitted to General Surgery clinic, diagnosed with acute biliary pancreatitis. CA 19-9 level was normal while CEA minimally increased (CEA: 5.36 ng/ml). Multiphase abdominal computed tomography showed ampullary mass without periampullary vascular invasion. Magnetic resonance cholangiopancreatography showed dilated choledoc-abrupted bluntly at periampullary region (Figure 1). Endoscopic Retrograde Cholangiopancreatography (ERCP) revealed the ampulla was infiltrated with mass and endoscopic biopsy showed poorly diffentiated ampullary adenocarcinoma comprising signed ring cell. There was no metastatic disease or vasculary invasion at preoperative radiologic examinations and the patient underwent Pylorus Preserving Pancreaticoduodenectomy (PPPD). Microscopic examination of the tumor revealed ampullary signet ring cell carcinoma with pancreatic tissue invasion. Tumor size was 1.8 cm × 1.6 cm, located at ampullary bile duct region. The minimum tumor free margin was 3.5 cm. 18 lymph nodes were removed two of them were metastatic (Figure 2). Perineural invasion was reported. Immunohistochemically CK20, Cdx2,

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Received Date: *19 Apr 2022*
Accepted Date: *05 May 2022*
Published Date: *12 May 2022*

Citation:
Büyüker F, Seneldir H, Ilgin Erol C, Leblebici M, Sait Ozsoy M, Bas G, et al. Signet Ring Cell Carcinoma of the Ampulla of Vater: Report of a Case with Long-Term Survival. Ann Surg Case Rep. 2022; 5(1): 1056.

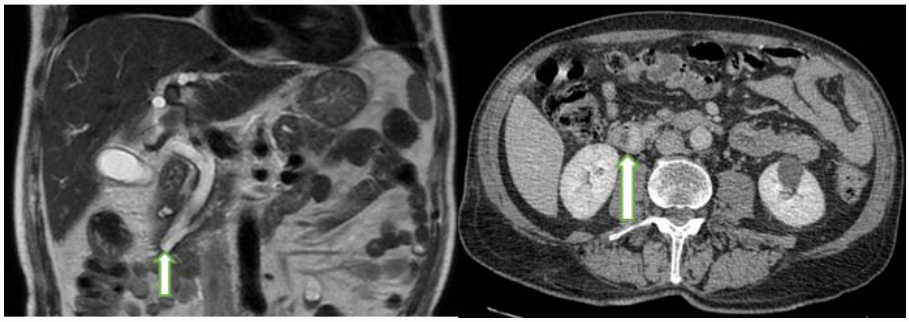

Figure 1: Preoperative radiological images of tumor; A) MRCP and B) CT imaging.
1a) Magnetic resonance cholangiopancreatography shows abrupt cessation of common hepatic duct, and dilatation of extrahepatic bile ducts. 1b) Contrast enhanced computer tomography shows mass in ampullary region.

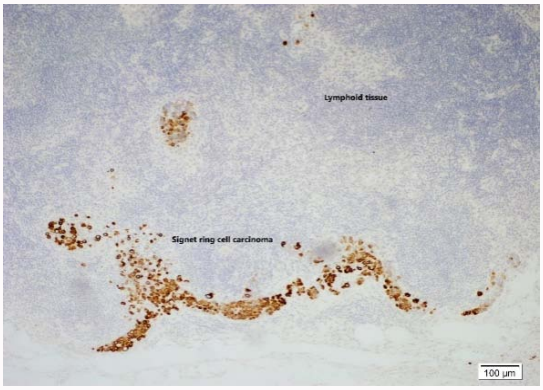

Figure 2: Microscobic image of Signet ring cells infiltrated lymph node (H&E).

MUC2 were positive and the tumor was determined as intestinal type ampullary SRCA. According to the TNM classification of AJCC-2018 tumor was staged as T4N1M0. The patient was discharged without complication on postoperative 12th day. Adjuvant chemotherapy was proposed but the patient rejected the regimen and he is followed 4 years with disease free survival.

Discussion

Ampulla Vateri tumors have more promising results in terms of resectability and prognosis among periampullary tumors [7]. However, there is not enough information about the prognosis of ampullary SRCA variant due to their rarity. After the first SRCA case reported by Gardner et al. in 1990, 39 more cases were reported [5]. The distribution of males and females in the cases was almost similar (20-19, respectively) and the mean age was 58 years. While lymphatic metastasis was detected in 8 of the reported cases (two T2N1, two TxN1, one T3N1, three T4N1), the 9th case is defined in our case. Our case is the fourth T4N1 case is presented according to the TNM classification (Table 1).

Although the pathogenesis of the disease is not well known, Fukuire ported a study suggesting that existing malignant cell mutation results in SRCA [8]. According to the WHO classification, the diagnosis of SRCA is made by the presence of more than 50% mucin-secreting signet ring cells in the tumoral tissue [9]. With immunohistochemical staining, SRCA is examined as Intestinal (I), Pancreaticobiliary (PB), gastric and mixed types [10]. I-type is determined with CK20, MUC2 and Cdx2 expression while K7, CK19, MUC1 expression determine PB-type and MUC5ac, MUC6

Table 1: Reported cases of SRCA, demography, operative procedure and tumor stage.

Author	Year	Sex	Age	Procedure	Stage
Paplomata and Wilfong [15]	2011	Female	45	PPPD	T4N1
Wen et al. (8 cases) [11]	2014	4 Males, 4 Females	40-78	PD	T3 N0–T4 N1
Rahul et al. [16]	2016	Male	53	PD	T4N1
Our case	2021	Male	68	PPPD	T4N1

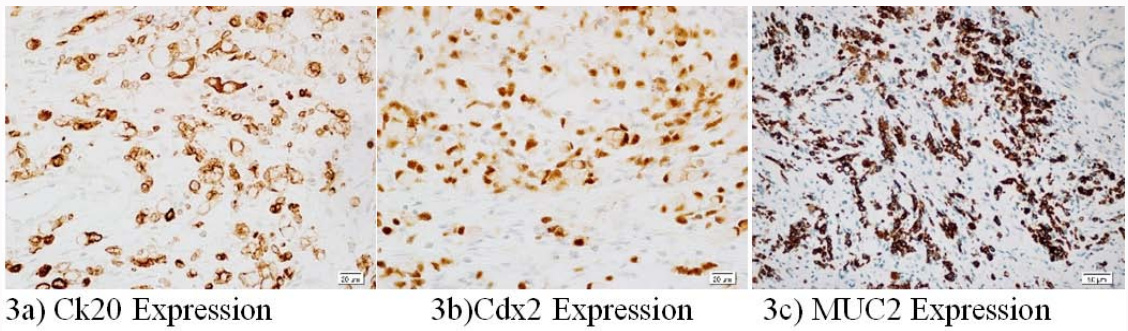

Figure 3: The microscobic image of A) CK20, B) CDX2, C) MUC2.
Diffuse and strong expression of Ck20, Cdx2, MUC2 in tumor specimen determines intestinal type of ampullary SRCA.

are Gastric type markers. The findings in our case were evaluated as compatible with I-type (Figure 3).

Although the Whipple operation is recommended in order to include the infrapyloric lymphatic station in ampullary adenocarcinomas, similar oncological results can be obtained with the pylorus preserving pancreaticoduodenectomy procedure and a more physiological procedure can be performed. PPPD was applied in 5 of the reported ampullary SRCA cases, and the sixth case is reported with our case. As with other periampullary tumors, curative results can only be obtained with surgical resection in the treatment of ampullary SRCA. Although the Dutch guideline does not recommend adjuvant chemotherapy for ampullary tumors there are cases where 5-fluorouracil or gemcitabine-cisplatin combination was applied in reported cases [10-12]. It has been reported that the response to adjuvant chemotherapy is low in gastric and esophageal colonic SRCA cases [6,12]. Due to its rarity, there is still no consensus about chemotherapy in ampullary SRCA cases.

While it has been reported that I-type ampoules SRCA cases have a better prognosis compared to other types, it has been reported that mixed type tumors have the worst prognosis [11-13]. Lymph node involvement was observed to be the most important prognostic factor [12]. While the mean survival was found to be 24.9 months (6 to 132 months) in ampullary SRCA cases, it was reported as 37 months in general ampullary carcinomas [3].

Ethical Approval

Patient's parents provided informed consent for data publication.

Declaration of authorship

FB and CIE conceived and designed the study; all authors acquired the data; all authors acquired the data analyzed and interpreted the data; all authors acquired the data drafted the manuscript; all authors acquired the data critically revised the manuscript for important intellectual content; all authors acquired the data gave approval of the version to be submitted; all authors acquired the data agree to be accountable for all aspects of the work.

Competing interests

All authors have completed the Unified Competing Interest form at www.icmje.org/coi_disclosure.pdf (available on request from the corresponding author).

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