Carcinosarcoma of Vater’s papilla: case report of a rare neoplasm and review of the literature

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Abstract

Background: Carcinosarcoma is a rare tumor that includes both carcinoma and sarcoma components. It develops commonly in the female reproductive tract, most often in the uterus. However, as there are a small number of similar cases in the English literature, we would like to present a rare case of a carcinosarcoma in Vater’s papilla.

Case presentation: A 76-year-old female patient was preoperatively diagnosed with a papillary adenocarcinoma in Vater’s papilla by endoscopic biopsy. The patient underwent subtotal stomach-preserving pancreaticoduodenectomy, and postoperative pathological examination diagnosed the carcinosarcoma. The patient received adjuvant chemotherapy with gemcitabine, but multiple liver metastases were found 3 months after the operation. Though chemotherapy with gemcitabine and cisplatin was introduced, she died owing to tumor progression 7 months after the operation.

Conclusion: Because carcinosarcoma of Vater’s papilla is a rare disease, a suitable treatment strategy has been unclear. We also present a review of the English literature regarding carcinosarcoma of Vater’s papilla.

Keywords: Carcinosarcoma, Vater’s papilla, Stomach-preserving pancreaticoduodenectomy, Chemotherapy

Background

Carcinosarcoma is a rare tumor that includes both carcinomatous and sarcomatous components [1, 2]. It occurs in the uterus most often and has been reported in lung and breast [1]. It also infrequently develops in gastrointestinal organs such as the liver, bile duct, pancreas, and intestine [3, 4]. Almost all reported cases have been primarily treated by surgical therapy [3, 5]. Even if the surgery achieves curative resection, the prognosis of carcinosarcoma is generally poor [3, 6]. As there were small numbers of similar cases in the English literature, we would like to present a rare case of carcinosarcoma of Vater’s papilla. The histological type of a malignant tumor in Vater’s papilla is usually adenocarcinoma, and its prognosis is usually good [7–9]. However, our reported case showed poor prognosis. We also present a review of the English literature regarding cases of carcinosarcoma that developed in Vater’s papilla.

Case presentation

A 76-year-old-woman was evaluated because of general fatigue, loss of appetite, and jaundice. Laboratory test showed an elevation of total bilirubin (7.7 mg/dL) and hepatobiliary enzyme. An endoscopy showed a 10-mm tumor in Vater’s papilla (Fig. 1A), and endoscopic retrograde biliary drainage (ERBD) was placed for obstructive jaundice. After that, total bilirubin was decreased to 1.9 mg/dL. The pathological diagnosis of endoscopic biopsy of the tumor was a papillary adenocarcinoma. Endoscopic ultrasonography (EUS) revealed that this tumor invaded pancreatic parenchyma (Fig. 1B). A contrast-enhanced computed tomography (CT) revealed a hypovascular mass at Vater’s papilla (Fig. 2A, B). No evidence of distant metastasis was identified. Carcinoembryonic antigen (CEA)

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Fig. 1 Preoperative endoscopy. Endoscopy revealed a 10-mm tumor in Vater's papilla and endoscopic retrograde biliary drainage was placed (A). Endoscopic ultrasonography revealed that the tumor (arrows) invaded pancreatic parenchyma (B). a, common bile duct; b, main pancreatic duct; c, duodenum.

Fig. 2 Preoperative computed tomography. Coronal section (A) and axial section (B) of a contrast-enhanced computed tomography scan revealed a hypovascular tumor (arrows) at Vater's papilla had invaded into the pancreas directly. a, duodenum; b, common bile duct; c, pancreas; d, tumor.
and carbohydrate antigen 19-9 (CA19-9) were not elevated (CEA 1.9 ng/ml, CA19-9 31.5 U/ml). Thus, the patient was preoperatively diagnosed with an adenocarcinoma of Vater’s papilla and underwent an operation. A subtotal stomach-preserving pancreaticoduodenectomy (SSPPD) with D2 lymph node dissection was performed. The pancreas was soft and non-fibrotic. The operation time was 6 h 18 min, and the intraoperative blood loss was 417 g.

The patient developed postoperative pancreatic fistula (grade B) in accordance with the International Study Group for Pancreatic Fistula definition [10]. Appropriate persistent drainage was performed, and the patient recovered immediately and was discharged on the 30th postoperative day.

Macroscopically, a 2.0 × 1.4 cm elastic hard tumor was found at Vater’s papilla (Fig. 3A). The microscopic examination showed that spindle cells that constructed the specimen tissue proliferated with intricate infiltration (Fig. 3B) and growth of tubular adenocarcinoma (Fig. 3C). Two components existed across the transition zone (Fig. 3D). Approximately 30% of the tumor was sarcoma component, and the remainder was carcinoma component. The tumor directly infiltrated into peri-pancreatic fatty tissue, pancreatic parenchyma, and a lymph node. Finally, pathological diagnosis was carcinosarcoma of Vater’s papilla. Resection margins were pathologically negative; thus, curative resection was achieved.

The patient received adjuvant chemotherapy using gemcitabine 1 month after the operation. It was continued without any obvious adverse events; however, an enhanced CT revealed multiple liver metastases at 3 months after the operation. The chemotherapy was changed to gemcitabine plus cisplatin. However, enhanced CT revealed the rapid progression of the metastasis at 6 months after the operation. The patient died at 7 months after the operation due to the continuous tumor progression.

Discussion
Carcinosarcoma is defined as a malignant neoplasm with both epithelial and mesenchymal elements within the same tissue [1]. It is a rare tumor, comprising less than 1% of all malignant neoplasms of the hepatobiliary tract. Carcinosarcoma of Vater’s papilla is extremely rare [3]. We found only five cases (excluding our current case) among current English literatures (Table 1) [7–9, 11, 12]. In all cases, patients had some subjective symptoms, such as jaundice, loss of appetite, and malaise. All patients underwent pancreaticoduodenectomy. Generally, their prognosis of carcinosarcoma of the hepatobiliary tract including Vater’s papilla is poor, as the overall 1-, 3-, and 5-year survival rates for patients with carcinosarcoma of the hepatobiliary tract after surgery are 44.0%, 29.3%, and 27.0%, respectively [3].

Preoperative diagnose of carcinosarcoma is difficult. Recently, several studies showed that 18F-fluorodeoxyglucose(FDG) positron emission tomography-computed tomography (PET-CT) was useful for diagnosing of carcinosarcoma, because it showed intense FDG uptake in a patient with carcinosarcoma [13–15]. In our case, preoperative PET-CT was not performed. If it showed abnormally intense FDG uptake despite being a small tumor, then it may have become an evidence for judging before the operation. However, high SUVmax value of the tumor does not influence the treatment strategy. So, in fact, it is not realistic to perform PET-CT for all patients with tumor of Vater’s papilla.

Only our case received adjuvant chemotherapy using gemcitabine. However, our case developed liver metastasis 3 months after the operation. Thus, the effect of adjuvant chemotherapy and a suitable regimen remains unclear. Because it is a rare tumor, also, the suitable treatment of metastatic carcinosarcoma of the biliary tract, including Vater’s papilla, has not been established. Our patient received gemcitabine plus cisplatin for multiple liver metastases, following treatment strategy for metastatic biliary duct cancer. However, this regimen was not effective in our case. In the gynecological field, there are some reports that chemotherapy using ifosfamide, cisplatin, paclitaxel, or carboplatin are effective for carcinosarcoma [16, 17]. And in the respiratory division, there is a report that nab-paclitaxel plus carboplatin is effective and safe for pulmonary carcinosarcoma [18]. Combination chemotherapy that is effective for both carcinoma and sarcoma might be considered for carcinosarcoma [19].

There were some reports that cancer metastasis or recurrence revealed along the catheter tract of biliary drainage [20], or the patients with ampullary cancer who had preoperative biliary drainage, had poor prognosis [21]. Recently, a report was published that patients who underwent preoperative endoscopic retrograde cholangiopancreatography (ERCP) had a significantly higher rate of early distant metastasis within 1 year, especially in patients with early-stage cancer of Vater’s papilla [22]. And we also think there is a possibility that preoperative biliary drainage may be one of the possible reasons why this patient had an early recurrence. ERBD was placed for obstructive jaundice in this patient. So, it is necessary to keep it in mind that these invasive procedures may cause disruption or dissemination of cancer cells.

Conclusion
Because carcinosarcoma of Vater’s papilla is a rare disease, a suitable treatment strategy has been unclear. Curative resection may contribute to a better prognosis; however, adjuvant chemotherapy and treatment for metastatic disease should be discussed more in the future.
Fig. 3 Pathological findings. Macroscopically, a 2.0 × 1.4 cm elastic hard tumor was found at Vater’s papilla (A). The microscopic examination of the specimen showed that spindle cells, which constructed sarcomatous tissue proliferated with intricate infiltration (B) and growth of tubular adenocarcinoma (C). Two components existed across the transition zone (D). a, sarcomatous component; b, adenocarcinomatous component; c, transition zone.

Table 1 Previous reports about cases of carcinosarcoma of Vater’s papilla

| Author     | Year | Sex | Age | Chief complaint         | Operation | Adjuvant therapy | Lymph node metastasis | Prognosis      |
|------------|------|-----|-----|-------------------------|-----------|------------------|-----------------------|---------------|
| Kench [11] | 1997 | F   | 46  | Melena, fatigue, dysnea | PD        | NA               | Positive              | Died (8 POM)  |
| Kijima [12] | 1999 | M   | 46  | Jaundice, liver dysfunction | PD        | NA               | NA                    | NA            |
| Tanaka [7]  | 2008 | F   | 70  | Jaundice, loss of appetite | PPPD      | NA               | Positive              | Alive (24 POM) |
| Rao [8]     | 2016 | M   | 67  | Abdominal discomfort, weight loss | PD        | None             | Positive              | Alive (5 POM) |
| Izumi [9]   | 2016 | M   | 73  | Jaundice                | SSPPD     | None             | Negative              | Alive (5 POM) |
| This case   |      | F   | 76  | Jaundice, loss of appetite, general malaise | SSPPD      | Gemcitabine       | Positive              | Died (7 POM)  |

NA not available, PD pancreatoduodenectomy, POM postoperative month, PPPD pylorus-preserving pancreatoduodenectomy, SSPPD substomach-preserving pancreatoduodenectomy
The authors declare that they have no competing interests.

**Constitution**

We obtained consent for publication from the patient.

**Consent for publication**

Not applicable.

**Ethics approval and consent to participate**

Revision of the manuscript. BB reviewed it and is responsible for the manuscript. DH, AC, and RI performed the surgery and followed up the patient. YN, TY, NU, HO, KL, HH, and BB participated in the critical analysis during the current study.

**Authors’ contributions**

RI wrote the initial draft of the manuscript. YY supervised the writing of the manuscript. DH, AC, and RI performed the surgery and followed up the patient. YN, TY, NU, SN, HO, KL, HH, and BB participated in the critical revision of the manuscript. BB reviewed it and is responsible for the manuscript. All authors have read and approved the final manuscript.

**Ethics approval and consent to participate**

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