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CASE REPORT

Rare histological subtype of invasive micropapillary carcinoma in the ampulla of Vater: A case report

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Author contributions: Noguchi H performed the pathological analysis, and wrote the initial draft of the manuscript; Higashi M and Tanimoto A were responsible for the revision of the manuscript for important intellectual content; Idichi T, Kurahara H, Mataka Y and Ohtsuka T were the patient’s surgeons, reviewed the literature and contributed to manuscript drafting; Tasaki T and Kitazono I performed the pathological examination, reviewed the literature and contributed to manuscript drafting; all authors issued final approval for the version to be submitted.

Informed consent statement: Informed written consent was obtained from the patient’s daughter for publication of this report and any accompanying images.

Conflict-of-interest statement: The

Abstract

BACKGROUND
Carcinoma of the ampulla of Vater is an uncommon ampullo-pancreatobiliary neoplasm, and the most common histological type is adenocarcinoma with a tubular growth pattern. Invasive micropapillary carcinoma (IMPC) is an aggressive variant of adenocarcinoma in several organs that is associated with lymph node metastasis and poor prognosis. IMPC was first described as a histological subtype of breast cancer; however, IMPC of the ampulla of Vater is extremely rare, with only three articles reported in the English literature.

CASE SUMMARY
We have reported a case of IMPC of the ampulla of Vater in an 80-year-old man. Microscopically, the surface area of the carcinoma was composed of tubulopapillary structures mimicking intra-ampullary papillary-tubular neoplasm, and the deep invasive front area exhibited a pattern of IMPC. The carcinoma showed lymphatic invasion and extensive lymph node metastasis. The immunohistochemical study revealed mixed intestinal and gastric/pancreatobiliary phenotypes.

CONCLUSION
This rare subtype tumor in the ampulla of Vater showed a histologically mixed phenotype and exhibited aggressive behavior.

Key Words: Micropapillary carcinoma; Rare histological subtype; Ampulla of Vater; Ampullo-pancreatobiliary region; Intra-ampullary papillary-tubular neoplasm; Case report
The patient's symptoms started 3 mo prior to the admission. History of present illness

An 80-year-old man was referred to our hospital with symptoms such as anorexia, malaise, and jaundice.

History of present illness

The patient's symptoms started 3 mo prior to the admission.
History of past illness
The patient’s medical history included hypertension and chronic pancreatitis. He was found to have dilatation of the main pancreatic duct incidentally on abdominal echography during medical examination 2 years ago. He used to consume 350 mL of beer per day for over 55 years, but he had quit 5 years ago. He smoked cigarettes briefly at the age of 20.

Personal and family history
The patient did not have any family history.

Physical examination
Upon admission, his vital signs showed a heart rate of 50/min, blood pressure of 147/77 mmHg, and respiratory rate of 27/min. Body weight was 40 kg, height was 144 cm, and body surface area (BSA) was 1.27 m². There were no physical findings to note.

Laboratory examinations
The results of the laboratory tests revealed a total bilirubin level of 4.3 mg/dL (normal range, 0.2-1.2 mg/dL), an aspartate aminotransferase level of 108 IU/L (normal range, 8-38 IU/L), an alanine aminotransferase level of 282 IU/L (normal range, 6-43 IU/L), a carcinoembryonic antigen (CEA) level of 7 ng/mL (normal range, 0-5 ng/mL), and a carbohydrate antigen 19-9 level of 62 U/mL (normal range, 0-37 U/mL).

Imaging examinations
Magnetic resonance cholangiopancreatography showed a 7 mm × 7 mm shadow defect in the duodenal papilla with a rim-enhancing lesion in the periamputillary region and slight dilatation of the main pancreatic duct. Endoscopic retrograde cholangiopancreatography revealed a 7-mm lesion deficit of the contrast medium from the duodenal papilla to the distal bile duct. A stent was placed in the pancreatic duct and the common bile duct to decompress the pancreas and bile duct. A biopsy specimen revealed well-differentiated tubular adenocarcinoma. Based on the diagnosis of adenocarcinoma of the ampulla of Vater, the patient underwent subtotal stomach-preserving pancreateoduodenectomy and was discharged after an uneventful recovery.

Pathological examinations
Gross examination showed an irregular papillary projection at the enlarged papilla of Vater measuring 23 mm × 10 mm. The papillary tumor had spread in the dilated ampullary channel and had an exophytic growth pattern (Figure 1A). The cut surface of the tumor was gray to white in color (Figure 1B). Histopathological examination revealed that the surface of the tumor consisted of an adenomatous component with a low-to-high grade dysplastic area (Figure 2A and B), while the deep part of the tumor consisted of an adenocarcinoma showing an invasive micropapillary pattern (Figure 2C and D). The micropapillary component comprised 60% of the tumor. Numerous neutrophilic infiltrations were observed in the tumor nests and stroma, and they formed focal intraepithelial microabscesses (Figure 2E). The carcinoma invaded the duodenum, pancreas, and intrapancreatic bile duct. Metastasis was seen in five of the 13 peripancreatic lymph nodes. Immunohistochemical analysis showed the tubular and micropapillary areas were positive for CK7, a caudal-related homeobox transcription factor-2 (CDX2) (Figure 3A-D), and MUC014E. The tubular component on the surface was positive for CK20 (Figure 3E) and focally positive for MUC2, MUC5, and MUC6. The micropapillary component was focally positive for MUC1 (Figure 3F); no tumor cells expressed MUC4.

FINAL DIAGNOSIS
The final diagnosis of the presented case is IMPC of the ampulla of Vater.

TREATMENT
The patient underwent subtotal stomach-preserving pancreateoduodenectomy with D2 lymphadenectomy. He received chemotherapy after surgery with oral administration of S-1 (also known as TS-1; Taiho Pharmaceutical Co. Ltd.; Tokyo, Japan). The initial
Ampullary invasive micropapillary carcinoma

Figure 1 Macroscopic appearance. A: Macroscopic appearance of the resected specimen showing an irregular papillary surface at the enlarged papilla of Vater measuring 23 mm × 10 mm. The tumor is seen extending into the dilated ampullary channel with an exophytic growth pattern; B: The cut surface of the tumor is grey to white in color.

Figure 2 Microscopic findings. A: The section shows that the surface of the tumor is composed of tubulo-papillary proliferation [hematoxylin and eosin (H&E) stain, magnification 40 ×]; B: Significant architectural and nuclear atypia, including loss of polarity (H&E stain, magnification 200 ×); C: Transition from the tubular structure to the invasive micropapillary pattern (H&E stain, magnification 40 ×); D: The invasion edge of the tumor composed of moderately differentiated adenocarcinoma and micropapillary carcinoma (H&E stain, magnification 200 ×); E: Neutrophilic infiltration forming intraepithelial microabscesses in the tumor nest (H&E stain, magnification 400 ×).

dose of S-1 was determined according to the BSA and administered at 80 mg per day. A 2 wk administration followed by a week rest was continued as postoperative adjuvant chemotherapy for a year.

OUTCOME AND FOLLOW-UP

The postoperative course showed abdominal pain caused by pancreatic fistula, which was successfully treated with fluid replacement and fasting for 7 d. The patient was discharged 21 d after surgery. The patient received postoperative chemotherapy as described above and was monitored at regular intervals through clinical examinations, biochemical investigations, and imaging studies.

Serum tumor marker levels of CEA gradually increased monthly after surgery, whereas CA19-9 levels remained normal (Figure 4). However, there was no abnormality observed on the clinical examination performed each month, and no evidence of recurrence was noted on routine 6 mo follow-up computed tomography.
Figure 3 Immunohistochemical findings. A and B: Immunohistochemical staining for CK7 revealing positivity in both tubular and micropapillary carcinoma (immunostain, magnification 200 ×); C and D: Tumor cells in both tubular and micropapillary portions showing positivity for caudal-related homeobox transcription factor-2 (immunostain, magnification 200 ×); E: Tumor cells on the surface area showing positivity for CK20 (immunostain, magnification 200 ×); F: The micropapillary component showing focally positivity for mucin1 (immunostain, magnification 400 ×). CDX2: Caudal-related homeobox transcription factor-2; MUC1: Mucin1.

Figure 4 Changes in carcinoembryonic antigen and carbohydrate antigen 19-9 serum levels from the perioperative period to the last follow-up. Serum tumor marker levels of carcinoembryonic antigen gradually elevated to 4.1 ng/mL, 7.3 ng/mL, 8.9 ng/mL, 9.4 ng/mL, and 9.8 ng/mL every 20-40 d after the surgery. However, carbohydrate antigen 19-9 serum levels remained within normal limits after the surgery. CEA: Carcinoembryonic antigen; POD: Postoperative day; CA19-9: Carbohydrate antigen 19-9.

The patient currently remains in a stable condition without recurrence of the disease for 6 mo since surgery and continues to receive treatment with S-1.

DISCUSSION

Ampullary carcinoma accounts for 0.2% of all gastrointestinal malignancies, and IMPC is an extremely rare subtype of adenocarcinoma of the ampulla of Vater. Only 10 cases of IMPC in the ampullary region have been reported in the English literature; the clinicopathological findings are summarized in Table 1. The ratio of male to female IMPC is 10 to 1, and the median age is 69 years (range, 41-80 years). True ampullary carcinoma has a better prognosis than pancreatic carcinoma and bile duct cancer. However, in all cases including the present case, the median overall survival time was 11 mo (range, 6-25 mo), with a generally worse prognosis than other subtypes of...
Table 1 Clinicopathological findings of reported case of invasive micropapillary carcinoma of the ampulla of Vater

<table>
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<tr>
<th>Case</th>
<th>Ref.</th>
<th>Age (yr)</th>
<th>Sex</th>
<th>Micropapillary component (%)</th>
<th>Lymph node metastasis</th>
<th>MUC1 staining</th>
<th>Survival (mo)</th>
<th>Status</th>
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<td>79</td>
<td>M</td>
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<tr>
<td>2</td>
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<td>71</td>
<td>M</td>
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<td>3</td>
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<td>41</td>
<td>M</td>
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<tr>
<td>4</td>
<td></td>
<td>71</td>
<td>M</td>
<td>&gt; 50</td>
<td>2/9</td>
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<tr>
<td>5</td>
<td></td>
<td>62</td>
<td>M</td>
<td>&gt; 50</td>
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<td>M</td>
<td>Almost area</td>
<td>13/16</td>
<td>N/A</td>
<td>20</td>
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<tr>
<td>10</td>
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<td>71</td>
<td>M</td>
<td>60</td>
<td>&lt; 4</td>
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<tr>
<td>11</td>
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<td>80</td>
<td>M</td>
<td>60</td>
<td>5/13</td>
<td>+</td>
<td>6</td>
<td>Alive</td>
</tr>
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N/A: Not applicable; MUC1: Mucin1.

Most cases of ampullary adenocarcinoma develop sporadically. Cigarette smoking and chronic infection by liver fluke are considered risk factors[10]; however, our patient reported smoking cigarettes at the age of 20, although very briefly. Neurofibromatosis type I, familial adenomatous polyposis, and Muir–Torre syndrome seem to be predisposing factors for ampullary carcinomas, but these syndromes were not observed in the present case[9-11].

Ampullary carcinomas frequently show a superficial papillary growth pattern; however, they demonstrate poor differentiation at the invasive front[9,10]. In our case, the superficial part of the carcinoma was arranged in a papillary structure, which was composed of the intestinal mucosa, and the invasive area showed a micropapillary growth pattern. Most cases of IMPC showed an approximately 50% area of micropapillary components, similar to that observed in our case[9]. The frequency of lymph node metastasis and its prognosis are not related to the amount of micropapillary components. Since the presence of micropapillary components, even in small amounts, is related to a worse prognosis, IMPC is defined as a carcinoma with >20% of the tumor comprising a micropapillary pattern.

Although most ampullary tumors are adenocarcinomas of INT, GPB, or mixed type, ampullary cancer has been proven to be of highly heterogeneous types[11,21,22]. Ampullo-pancreatobiliary cancers are classified into nine distinct histological subtypes according to the World Health Organization classification[11]. Among them, IMPC is a special and rare subtype of ampullary carcinoma with high malignant potential. The classification of ampullary adenocarcinoma is based on immunohistochemical staining findings[11,12]. The INT immunophenotype is characterized by the expression of MUC2 and CDX2, while the GPB immunophenotype is characterized by the expression of MUC1, MUC5AC, and MUC6[22]. Expression of both CK7 and CK20 were observed in more than half of the cases[22]. In our case, the tumor cells were diffusely positive for CDX2 and CK7. In addition, the surface tubular part of the tumor was positive for MUC2, and the micropapillary part was positive for MUC1. Thus, the present case was classified as a mixed subtype. However, since the ampulla is a transitional area covered with several epithelial components, it is occasionally difficult to clearly subdivide ampullary adenocarcinomas into INT or GPB types. These cases account for half of all the ampullary carcinomas and should be classified as tubular adenocarcinomas of mixed type[11,21].

The differential diagnosis of micropapillary lesions in the ampulla of Vater includes undifferentiated carcinoma with osteoclast-like giant cells and metastatic carcinomas. In our case, the tumor nests mimicked giant cells; however, the tumor cells had high-grade cytologic atypia and were positive for cytokeratin. Metastasis in the ampulla of Vater has been described for various cancers, including breast cancer, colon cancer, and sarcoma. However, in our case, clinical investigation did not detect any primary tumors outside the ampullary region of Vater.
CONCLUSION

We have reported a rare case of IMPC in the ampullary region. A diagnostic criterion to confirm this tumor has not been established thus far. This rare variant of ampullary adenocarcinoma has a poor prognosis and limited information is available regarding its pathogenesis and treatment. Further investigation with other methods, including genetic analysis, would be necessary to differentiate IMPC of the ampulla of Vater.

REFERENCES


