Single Case

Primary Duodenal Carcinoma with Embryonal Carcinoma Features in a Young Man

Naoto Yamamoto^a Kota Washimi^b Masaaki Murakawa^a
Mariko Kamiya^a Yuto Kamioka^a Makoto Ueno^c Takeshi Kishida^d
Yasushi Rino^e Munetaka Masuda^e Soichiro Morinaga^a

^aDepartment of Gastrointestinal Surgery, Kanagawa Cancer Center, Yokohama, Japan;

^bDepartment of Pathology, Kanagawa Cancer Center, Yokohama, Japan; ^cDepartment of Hepatobiliary and Pancreatic Medical Oncology, Kanagawa Cancer Center, Yokohama, Japan;

^dDepartment of Urology, Kanagawa Cancer Center, Yokohama, Japan;

^eDepartment of Surgery, Yokohama City University Hospital, Yokohama, Japan

Keywords

Duodenal cancer · Embryonal carcinoma · Germ cell tumor · Burned-out tumor · Pancreaticoduodenectomy

Abstract

We present the case of a 35-year-old man with intractable nausea, vomiting, and severe anemia. A computed tomography (CT) scan of the chest, abdomen, and pelvis showed a circumferential lesion thickening of up to 3.5 cm at the level of the third portion of the duodenum. No aortocaval, retroperitoneal lymphadenopathy, nor secondary lesion was observed. Esophagogastroduodenoscopy (EGD) revealed a circumferential mass within the third portion of the duodenum. Histopathology of biopsy materials from the duodenal mass showed it most likely to be a poorly differentiated adenocarcinoma. The patient underwent a subtotal stomach-preserving pancreaticoduodenectomy with regional lymph node dissection. Histologically, tumor cells with basophilic cytoplasm and pleomorphic nuclei showed a solid pattern, and expressed CD30 and SALL4 immunohistochemically, leading to a diagnosis of embryonal carcinoma-like tumor. No other primary tumor could be identified, and the location of the tumor, mainly on

the mucosal surface, suggested a duodenal origin. The UICC TNM staging was T3N2M0, stage IIB. This is a rare case of primary duodenal carcinoma with features of embryonal carcinoma.

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Introduction

Primary duodenal cancer (PDC), which arises from the region of duodenum, excluding ampullary regions of the Vater, is rare and accounts for less than 1% of all carcinomas of the gastrointestinal tract [1, 2]. Histologically, adenocarcinoma and gastrointestinal stromal tumor (GIST) were the most common types of PDC. Other histological types included neuroendocrine carcinoma and adeno-squamous carcinoma [3]. Surgical approaches to cancer of the duodenum can vary and are highly dependent on the location of the tumor. Tumors that occur in the first, second, or third part of the duodenum usually require pancreaticoduodenectomy, but tumors that occur in the fourth part may be better suited for partial duodenal resection.

Embryonal carcinoma is a malignant germ cell tumor (GCT) composed of primitive epithelial tumor cells that recapitulate early stages of embryonic development and show specific immunohistochemical expression of CD30 [4]. Testicular GCTs make up 95% of testicular cancers and are the commonest solid malignancies in young men [5]. It should be noted that about 5% of patients of GCTs may present with a metastatic localization on the digestive tract [6]. Extragonadal GCTs are rare and account for only 1–5% of all GCTs [7]. They most commonly occur in the mediastinum or sacrococcygeal region; however, they can occur at various other locations. Here, we report a case of a rare primary duodenal tumor with features of an embryonal carcinoma curatively resected with pancreaticoduodenectomy.

Case Presentation

A 35-year-old man presented to our hospital with intractable nausea and vomiting complaints. His medical history and family history were unremarkable. Clinical examination showed that the patient had a body mass index of 20.3 kg/m². Examination of the abdomen revealed epigastric distention without any abdominal mass.

Biologically, the patient presented with hemoglobin at 7.5 g/dL in relation to iron deficiency anemia (serum iron: 16 µg/dL, mean corpuscular volume: 65.6 fL). For this reason, he underwent transfusion with 6 units of packed red blood cells after he was admitted to our department. There was no excessive inflammatory syndrome (leukocytes: 4,400/µL and C-reactive protein: 2.35 mg/dL). Electrolytes and hepatic function were within normal limits. Tumor markers were measured as follows: carcinoembryonic antigen, 0.9 ng/mL; carbohydrate antigen, 19–9. 41.5 U/mL; carbohydrate antigen, 125: 12.1 U/mL.

A computed tomography (CT) scan of the chest, abdomen, and pelvis showed a circumferential lesion thickening of up to 3.5 cm at the level of the third portion of the duodenum (Fig. 1). Although up to 20 mm, several lymphadenopathies were noted around the duodenum, no aortocaval, retroperitoneal lymphadenopathy, or secondary lesion was observed.

An esophagogastroduodenoscopy (EGD) was performed to further investigate his anemia and vomiting, which revealed a circumferential mass within the third portion of the duodenum (Fig. 2). Histopathology of biopsy materials from the duodenal mass showed it most

likely to be a poorly differentiated adenocarcinoma with positive immunoexpression for keratin (AE1/AE3) and negative for CD3, CD20, and p40.

After discussion at a multidisciplinary tumor board, he was diagnosed with severe bowel obstruction and anemia caused by primary duodenal cancer, and pancreaticoduodenectomy was performed. Intraoperatively, examination of the abdominal cavity did not reveal any peritoneal carcinomatosis or any other tumoral lesions. A subtotal stomach preserving pancreaticoduodenectomy with regional lymph node dissection was performed. His postoperative course was uneventful.

Macroscopically, there was a papillary elevated lesion measuring 65 × 57 mm in size, centering caudal to the duodenal papilla, and growing along the duodenum. Microscopically, cells with large, pleomorphic vesicular nuclei displaying single or multiple macronucleoli, amphophilic cytoplasm, and prominent mitotic figures grew in a solid pattern. There was no ductal architecture or squamous differentiation. Immunohistologically CD30 and SALL4 were positive, CK7 was partially positive, and CDX-2, c-kit, and PALP were only partially positive in a small fraction. On the other hand, CK20, S100, melanA, AFP, synaptophysin, chromogranin A, and HCGβ were negative. The Ki-67 labeling index was over 50%. The tumor was diagnosed as a carcinoma with embryonal carcinoma features. There was mild lymphovascular invasion. The tumor cells spread mainly on the duodenal mucosal surface and partially invaded the subserosal layer. There was no evidence of infiltration into the pancreas. The UICC TNM staging was T3N2M0, stage IIIA (Fig. 3).

There was a small area of nuclear stratification in the duodenal mucosa around the tumor, the tumor spread mainly on the mucosal surfaces, and no other primary lesion could be identified on CT imaging, suggesting a duodenal origin. Testicular ultrasound revealed only slight calcification in his testis. Histologic confirmation was not made because the patient refused orchiectomy or any other procedure. The patient is being closely followed without adjuvant therapy because of his refusal. Currently, at 12 months of follow-up, there is no evidence of active disease, with the levels of AFP and HCG-beta being within normal limits.

Discussion

This is a case of primary duodenal cancer with unusual characteristics of embryonal carcinoma. Embryonal carcinoma of the duodenum is rare and is not often reported as a primary tumor [8]. In most cases of metastatic duodenal germ cell tumors (GCTs), the images appear to be clustered with swollen lymph nodes of the retroperitoneum. Local extension from the retroperitoneal lymph node into the GI tract is a common method of spread [9].

In the case of metastatic GCTs, ultrasonography and histological examination with orchiectomy are performed to search for the primary lesion. When searching for primary lesions, “burned-out” tumors must also be considered [10–12]. In our case, the primary lesion was searched and a calcified lesion suspected to be a “burned-out tumor” was found. However, due to the patient’s refusal, no systematic diagnosis with orchiectomy has been performed, and the possibility of metastatic disease cannot be ruled out. However, there are few cases where the duodenal lumen side is the main site of the lesion, and without clustered masses of swollen lymph nodes of the retroperitoneum, as in our case. This is one of the reasons to support this case as a primary lesion.

If GCT is diagnosed preoperatively, even if no obvious primary can be pointed out, chemotherapy will be preceded according to the guidelines [13]. Chemotherapy was markedly

effective for metastatic duodenal GCTs, and there are several reports of progress without surgery [14–16]. It is not clear whether chemotherapy is successful for primary duodenal cancers with the characteristics of an embryonal carcinoma, as in our case. Therefore, it is considered appropriate to select surgical treatment for resectable lesions.

Similarly, with regard to postoperative adjuvant therapy, it has been reported that recurrence of nonseminomatous testicular germ cell tumors of stage IA and IB is significantly reduced by performing adjuvant chemotherapy [17]. However, in this case, there is no clear evidence of the effectiveness of adjuvant therapy when gross curative resection is performed. Therefore, in consideration of the possibility of a metastatic tumor in this case, it is necessary to continue to carry out careful follow-up by performing CT from the chest to the pelvis, ultrasonography of the testes, and regular measurement of tumor markers.

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Statement of Ethics

We have reported this case in compliance with the Declaration of Helsinki. Informed written consent was obtained from the patient for publication of this case report and any accompanying images.

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

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Author Contributions

N. Yamamoto contributed the design of the report. N. Yamamoto, K. Washimi, and T. Kishida drafted the manuscript. M. Murakawa, M. Kamiya, and Y. Kamioka collected the data. Y. Rino, M. Masuda, and S. Morinaga revised the manuscript. All authors read and approved the final version of the manuscript.

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Fig. 1. Abdominal CT showed a circumferential lesion thickening of up to 3.5 cm at the level of the third portion of the duodenum. No aortocaval nor retroperitoneal lymphadenopathy was observed.

Fig. 2. EGD showed a circumferential mass within the third portion of the duodenum.

Fig. 3. Histological findings of the operative specimen showing the features of an embryonal carcinoma: the cells with large, pleomorphic vesicular nuclei displaying single or multiple macronucleoli, amphophilic cytoplasm, prominent mitotic figures grow solid pattern (a). Immunohistologically CD30 (b) and SALL4 (c) were positive, CK7 (d) was partially positive, and CDX-2 (e), c-kit (f), and PALP (g) were only partially positive in a small fraction. On the other hand, CK20 (h), S100 (i), melanA (j), AFP (k), synaptophysin (l), chromogranin A (m), and HCGβ (n) were negative. The Ki-67 labeling index was over 50% (o). Original magnification $\times 400$.