A Case of Advanced Rectal Adenocarcinoid Tumor with Long-term Survival

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INTRODUCTION

Adenocarcinoid tumor is a rare entity that possesses histological features of both carcinoid and adenocarcinoma. Although the existence of the disease had long been recognized, the term ‘adenocarcinoid’ was first coined by Warkel et al. (1) in 1978. It has been reported that the majority of adenocarcinoids occur in the appendix. Adenocarcinoid arising in the rectum is extremely rare and only six cases have been reported in the English literature (2–7). Here, we present a case of advanced rectal adenocarcinoid tumor with lateral lymph node metastasis treated with total pelvic exenteration and extended bilateral lymph node dissection resulting in long-term survival.

CASE REPORT

A 58-year-old man presented with pollakisuria at another hospital. Ultrasound-guided transrectal biopsy of the prostate revealed poorly differentiated adenocarcinoma; therefore, a radical prostatectomy was performed; however, no cancer lesion existed within the resected specimen. Reportedly, dissection between the rectum and prostate was easily performed, and no induration was palpable on the rectal wall during the operation. About a month after surgery, he presented with anal pain and a digital rectal exam revealed a hard mass on the anterior wall of the rectum. Ultrasound-guided biopsy of the mass showed a poorly differentiated adenocarcinoma and he was referred to our institution for further investigation and treatment.

Hematological examination was unremarkable except for mild anemia (hemoglobin 12.6 g/dl; hematocrit 35.8%). Tumor markers, including carcinoembryonic antigen (CEA), carbohydrate 19-9 and prostate-specific antigen (PSA), were within normal limits. Colonoscopy showed no tumorous lesion and the mucosa was intact. Abdominal MRI demonstrated a low-intensity mass, 4 × 6 cm, on the anterior wall of the lower rectum, involving the urethra (Fig. 1). Abdominal CT showed a low-density mass on the anterior wall of the rectum. Also, lymphadenopathy along the right internal iliac artery, suggesting metastasis, was pointed out (Fig. 2). At surgery, the rectal tumor had invaded the urethra, and lymph nodes along the internal iliac artery were
enlarged on both sides; therefore, total pelvic exenteration was performed. Internal iliac vessels and autonomic nerves were also resected for extended lateral lymph node dissection. The postoperative course was uneventful.

The surgical specimen contained a submucosal-tumor-like elevation on just the oral side of the dentate line; however, there was no mucosal lesion (Fig. 3).

Microscopically, the tumor was located mainly in the submucosal and muscular layers and had invaded the muscular layer of the urinary bladder. The tumor consisted of two types of tumor cells; a relatively small cell with a round nucleus and eosinophilic cytoplasm, and a signet-ring-cell-like cell. Neoplastic cells formed small nests in a solid, cord-like, or glandular pattern (Fig. 4).

Immunohistochemically, tumor cells were focally positive for synaptophysin and chromogranin A (Fig. 5). The Grimelius stain also showed argyrophilia, indicating a tendency to differentiate into neuroendocrine cells. The tumor contained the adenocarcinoma component, and the tumor was partly present in the lamina propria, suggesting that the tumor originated from the mucosa of the rectum. Immunohistochemically, the tumor was positive for CEA whereas negative for PSA, which also indicates the origin to be the rectum. Taking these findings into consideration, adenocarcinoid of the rectum was diagnosed.

Postoperatively, the patient received combination chemotherapy of fluorouracil (5-FU) and leucovorin (LV) as an
adjuvant therapy because an adenocarcinoma component had metastasized to regional lymph nodes. Three years after the initial surgery, abdomino-pelvic CT showed an enlarged left groin lymph node (Fig. 6). There was no distant metastasis in other organs; therefore, lymphadenectomy was performed. Pathologically, it was a poorly differentiated adenocarcinoma and signet ring cell carcinoma. Another 2 years later, left external iliac lymph node enlargement was detected again by abdomino-pelvic CT, and lymphadenectomy was performed. Pathologically, it was also adenocarcinoma. The patient is alive more than 5 years after the initial surgery.

DISCUSSION

Adenocarcinoid tumor arising in the rectum is extremely rare and only six cases have been reported in the English literature (2–7). More than 80% of reported adenocarcinoids have arisen in the appendix (2), and other sites, such as the stomach (2), duodenum (8,9), small intestine (2), biliary tract (10), and bronchus (11), have also been reported as origins of adenocarcinoid tumor.

Of the six cases of rectal adenocarcinoid tumor reported so far (2–7), three are male, two are female, and one is unknown. Grossly, two had an ulcerated lesion, two had a sessile polypoid lesion, and one was reported to have intact mucosa. Lymph node metastasis was confirmed in four cases and liver metastasis in three. Only one patient with lymph node metastasis is reported to be alive more than 3 years after surgery. Fortunately, aggressive surgery could be
indicated for primary and recurrent lesions without major complications in this case, and long-term survival was achieved.

One large population-based study demonstrated that the biological behavior of adenocarcinoid of the appendix is between malignant carcinoid and adenocarcinoma in terms of age at diagnosis and the proportion of lymph node involvement (12). Patients diagnosed with adenocarcinoid are less likely to have tumor spread beyond the colon; however, they have an unusual propensity for ovarian metastasis (13). As for prognosis, survival in adenocarcinoid tumor tends to be better than that in adenocarcinoma and worse than malignant carcinoid; however, the difference was not statistically significant (12).

There is no consensus on the surgical treatment for adenocarcinoid tumor and there is debate on how aggressive the operation should be even in a case of appendiceal adenocarcinoid tumor that constitutes the majority of the entity. In the present case, lateral lymph node metastasis was suspected on preoperative clinical imaging; therefore, extended lateral lymph node dissection with combined resection of internal iliac vessels was indicated. In addition, invasion to the urethra was suspected, and firm adhesion and a short distance between the rectum and the urethrovaginal anastomotic portion due to prostatectomy was expected; therefore, we chose total pelvic exenteration. It was a very aggressive surgical procedure; however, we consider that it was merited because there was invasion to the urinary bladder pathologically and there has been no pelvic regional recurrence. Three years and 5 years after the initial surgery, the patient developed recurrence confined to inguinal and external iliac lymph nodes and lymphadenectomy was performed each time. The patient has not experienced recurrence for 11 months since the last surgery. The 5-year survival of patients with lateral lymph node metastasis who have undergone rectal cancer surgery with lateral lymph node dissection is reported to be 37–45.8% (14,15). In our case, an aggressive surgical strategy might have resulted in long-term survival.

Large clinical trials have suggested that adjuvant chemotherapy with 5FU and LV provides a significant survival benefit in patients with Stage III colon cancer (16), and it was the standard adjuvant chemotherapy regimen in Japan at that time; however, there is no consensus on adjuvant chemotherapy for adenocarcinoid tumor. Mandai et al. (17) reported a patient with appendiceal adenocarcinoid with bilateral ovarian metastasis who survived for 2 years after receiving cisplatin-based adjuvant chemotherapy, which is standard for ovarian carcinoma. Hirschfield et al. (18) reported a similar patient with bilateral ovarian metastasis and peritoneal seeding who benefited from chemotherapy with 5-FU and streptozotocin. In our case, adjuvant chemotherapy with 5FU and LV was administered because metastatic lymph nodes involved only the adenocarcinoma component.

In conclusion, we experienced an extremely rare case of locally advanced rectal adenocarcinoid, successfully treated by TPE with bilateral lateral lymph node dissection, resulting in long-term survival. Moreover, groin lymph node recurrences were successfully removed twice, and it is possible that extended surgery is beneficial for patients with locally advanced rectal adenocarcinoid when resectable.

Conflict of interest statement
None declared.

References