

A case of primary jejunal cancer diagnosed by preoperative small intestinal endoscopy

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The patient was a 37-year-old female. She was brought to our hospital by ambulance with nausea and vomiting. Abdominal ultra sound and abdominal enhanced CT scan showed a tumor in left side of the abdominal aorta 6cm in size, and it showed an expanded stomach and duodenum. Upper gastrointestinal series revealed an apple core sign in upper jejunum near the Treitz' ligament. Small intestinal endoscopy (XSIF-240 endoscope, Olympus Inc.) revealed stenosis related to an epithelially protruding lesion with an irregular surface in the jejunum on the anal side of the horizontal duodenal peduncle. Biopsy suggested a well-differentiated adenocarcinoma. Scintigraphy showed hot spot in left middle abdomen. Under a diagnosis of primary jejunum cancer, Partial resection of the jejunum and partial resection of the transverse colon was performed. Histopathologically, the tumor was well differentiated adenocarcinoma exposed serosal surface. Postoperatively, the stage was evaluated as III (T3, N1, M0). Preoperative diagnosis to use small intestinal endoscopy was effectiveness. We report a patient with primary jejunum cancer in whom a definitive diagnosis was made before surgery.

Key words: Small intestinal cancer, primary jejunal cancer, Small intestinal endoscopy,

INTRODUCTION

Primary jejunum cancer is rare, and is difficult to diagnose before surgery. We report a patient in whom preoperative small intestinal endoscopy led to a definitive diagnosis of this cancer, and review the literature.

CASE REPORT

The patient was a 37-year-old female. On May, 2001, nausea and vomiting occurred. Two days after, vomiting became frequent, and she consulted a local clinic. However, it did not subside. Four days after, she was brought to our hospital by ambulance, and admitted. At 30 years of age she admitted by Pyelonephritis. Her father had cancer in which the primary focus was unclear.

Physical examination and laboratory data on admission: The abdomen was soft and flat. Palpation did not reveal any superficial lymph nodes. The leukocyte count was 12,900/ μ l. The C reactive protein (CRP), urea nitrogen (UN), and creatinine (Cr) levels were 0.6 mg/dl, 42 mg/dl, and 2.7 mg/dl, respectively (UA: 14.7 mg/dl, CPK: 550 U/l), suggesting an inflammatory response and dehydration. The levels of tumor markers were normal.

Abdominal ultrasound revealed dilatation of the stomach and duodenum. On the anal side, a small intestinal tumor was detected (Fig. 1).

Abdominal computed tomography (CT): Abdominal CT revealed a tumor measuring approximately 6 x 3 cm in diameter, in an area adjacent to the left iliopsoas

muscle. The stomach and duodenum on the orifice side of the tumor were markedly dilated, and rapidly tapered toward the tumor (Fig. 2).

Contrast-enhanced radiography of the upper digestive tract with Gastrografin revealed dilatation involving the horizontal duodenal peduncle. Circumferential apple-core stenosis was noted in an area adjacent to Treitz' ligament (Figs. 3).

Small intestinal endoscopy (SIF-240, Olympus Inc.): Endoscopy revealed stenosis related to an epithelially protruding lesion with an irregular surface in the jejunum on the anal side of the horizontal duodenal peduncle. It was difficult to insert an endoscope to the anal side of the tumor (Fig. 4). Biopsy at the same site suggested well-differentiated adenocarcinoma.

Abdominal angiography: Neither celiac nor superior mesenteric angiography revealed any abnormality.

Radiographic examination: Gallium scintigraphy identified a hot spot related to isotope uptake by the tumor, in the left middle abdomen (Fig. 5).

Based on these findings, surgery was performed under a diagnosis of primary jejunum cancer on June 5, 2001.

Intraoperative findings: The jejunal tumor was present in an area 2-cm distant from Treitz' ligament, and adhered to the transverse mesocolon and retroperitoneum, showing an indentation. Macroscopically, transverse mesocolon/retroperitoneal infiltration was suggested. Lymph node swelling was observed around the first vessel of the jejunal artery, suggesting metastasis. Neither liver nor peritoneal metastasis was de-

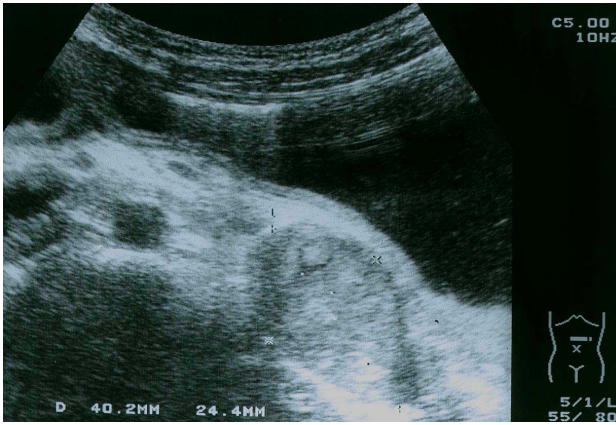


Fig. 1. Abdominal Ultra Sound showed gastroduodeno expansion, and anal side jejunal tumor was detected 4 x 2.4cm in size.

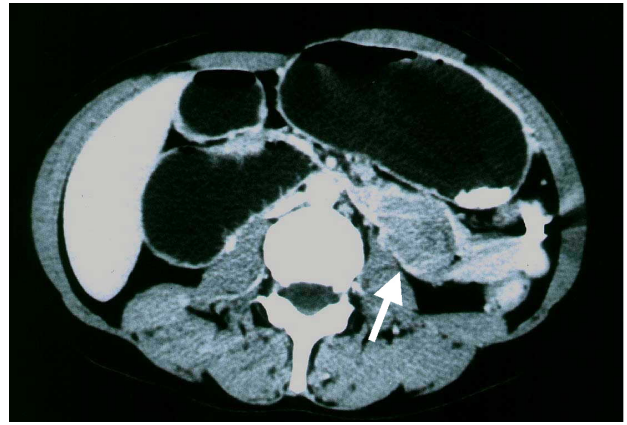


Fig. 2. Abdominal enhanced CT scan showed a tumor (white allow) in left side of the abdominal aorta 6cm in size, and it showed an expanded stomach and duodenum..

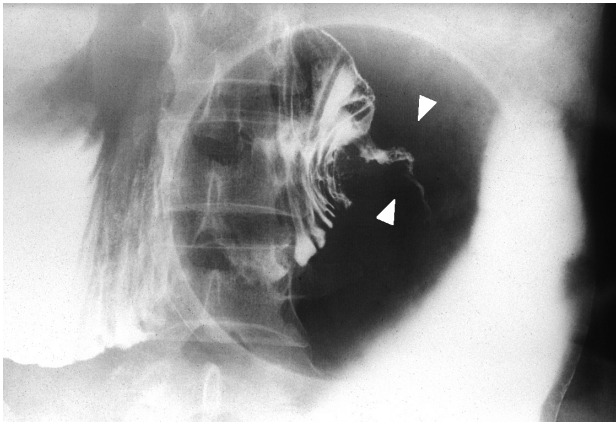


Fig. 3. Upper gastrointestinal series revealed an apple core sign in upper jejunum closed anal side of the Treitz' ligament (white triangle).

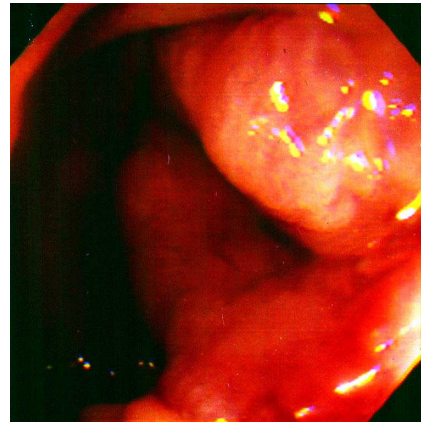


Fig. 4. Small intestinal endoscopy (XSIF-240: Olympus) revealed a jejunal tumor.

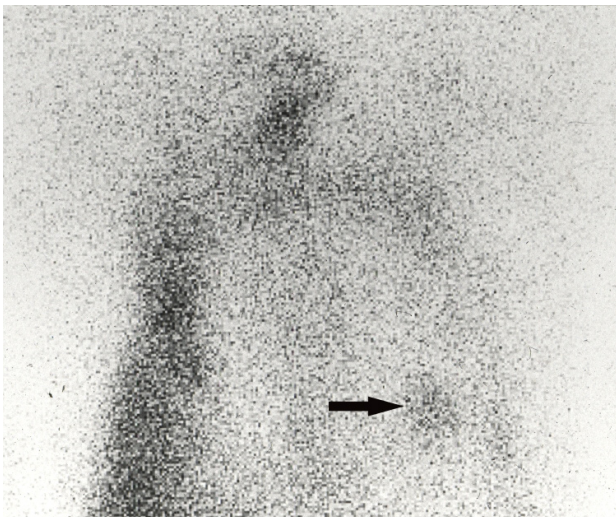


Fig. 5. Gallium scintigraphy showed hot spot in left middle abdomen (black allow)

tected. Based on these findings, we performed partial resection of the jejunum and partial resection of the transverse colon.

Macroscopic findings of the resected specimen: The tumor measured 8 x 6 cm, and was evaluated as a type II circumferential lesion (Fig. 6).

Histopathological findings: The histopathology suggested a well-differentiated adenocarcinoma. The grade of infiltration was evaluated as se, ly1, v0, n+(1/6: mesenteric lymph node), aw(-), ow(-), and ew(-)(Fig. 7). After surgery, the stage was evaluated as III (T3, N1, M0)(AJCC CANCER STAGING MANUAL¹⁾).

The postoperative course was good. We performed post operative adjuvant chemotherapy. UFT therapy has been performed at the outpatient clinic during the 3-year-and-3-month follow-up.

DISCUSSION

In Japan, the incidence of primary small intestinal cancer in malignant tumors of the digestive tract ranges from 0.1 to 0.3%²⁾. In Europe and the United States, it reportedly exceeds 1%³⁾. With respect to the type of malignant small intestinal tumors, Yao et al.⁴⁾ reported that the incidence of small intestinal cancer was highest (32.6%), followed by malignant lymphomas (30.4%)

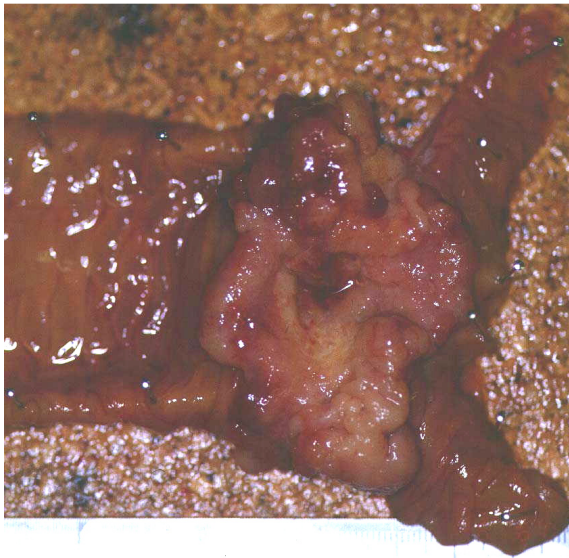


Fig. 6. The resected specimen showed a type 2 tumor 8 x 6cm in size.

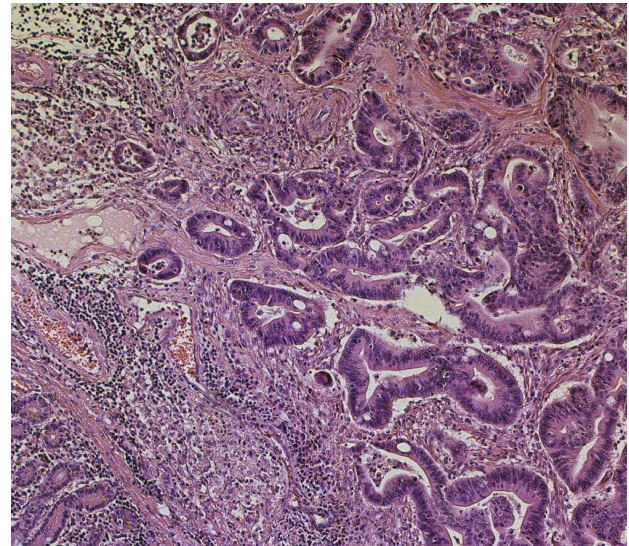


Fig. 7. Histological findings of resected specimen showed well differentiated tubular adenocarcinoma.

and leiomyosarcomas (29.1%). Kameoka *et al.*⁵⁾ indicated that leiomyosarcomas were most frequent (33.6%), followed by malignant lymphomas (30.9%) and small intestinal cancer (26.3%). In Japan, these 3 tumors account for more than 90%. In Europe and the United States, Forte *et al.*⁶⁾ analyzed a total of 1,086 patients presented in 20 publications, and reported that the incidence of carcinoids was highest among tumors of the small intestine excluding the duodenum, followed by adenocarcinomas and lymphomas.

Malignant small intestinal tumors frequently develop in persons aged 40 to 79 years. The male-to-female ratio is 1.4:1⁴⁾.

Concerning common sites, jejunum cancer develops in the jejunal region 50- to 60-cm distant from Treitz' ligament in more than 80% of patients. Ileal cancer is present in an area 50- to 60-cm distant from Bauhin's valve in most patients²⁾⁴⁾⁷⁾.

A consensus regarding macroscopic classification has not been reached. However, in many cases, macroscopic findings are classified into phyma, ulcer, and annular stenosis types. Annular stenosis-type lesions are most frequent⁸⁾. Furthermore, most patients have advanced cancer, frequently resulting in mesenteric lymph node metastasis, direct peripheral organ infiltration, hematogenous metastasis, and peritoneal dissemination. The pathogenesis is unclear. However, malignant small intestinal tumors are rare for the following reasons: there is no anatomically fixed flexion site in the small intestine; fluid intestinal contents pass in a short period, restricting carcinogen exposure; small intestinal contents are humoral/alkaline, minimizing the bacterial flora; the level of IgA is high, showing a protective activity against carcinogen-inducing viruses; and the level of benzyrene hydroxylase, which may detoxify carcinogens, is higher than that in the large intestine³⁾⁹⁾¹⁰⁾.

Concerning the grade of histopathological infiltration, Moriyama *et al.*¹¹⁾ indicated that tumor infiltration in the serous membrane or deeper was observed in 60.5% of their patients. Toochika *et al.*⁸⁾ reported that

22 (71%) of 31 patients had se or deeper lesions. With regard to the histological type, most patients (87.5%) had adenocarcinomas, and 1 to 6.3% showed undifferentiated carcinomas¹¹⁾.

Clinical symptoms mainly consist of ileus, hemorrhage of the digestive tract, and phymas on palpation. In addition, anemia is observed. Occlusive symptoms are frequent in the presence of cancer²⁾. The present case was also typical because cancer-related occlusion led to vomiting.

For diagnosis, contrast-enhanced radiography of the small intestine, small intestinal endoscopy, abdominal angiography, abdominal echography, and abdominal CT are employed. As characteristic findings on contrast-enhanced radiography of the small intestine, Good³⁾ indicated that circumferential, small shadow defects with a clear border, the disappearance of normal mucosa and ulcer formation, irregular narrowing of the lumen around the lesion, and intestinal dilatation on the orifice side of the cancer lesion were important. The present case also showed typical findings: tumor-related narrowing of the lesion site and intestinal dilatation on the orifice side.

The mean interval from the initial onset of symptoms until diagnosis is 5 months. It is relatively rare to make a definitive diagnosis before surgery, suggesting difficulty in diagnosis. According to Moriyama *et al.*¹¹⁾, a definitive diagnosis was made via endoscopic biopsy in 28.3% of jejunal and 5.7% of ileal cancer patients among small intestinal cancer patients in whom the site was detected before surgery using a conventional small intestinal endoscope; preoperative diagnosis was rare. However, recent advances in medical instruments, such as double-balloon small intestinal/capsule endoscopes, have facilitated a breakthrough in diagnosing small intestinal disorders¹²⁾¹³⁾. Capsule endoscopes have limitations: delayed capsule passage, stenosis-related retention, and no allowance for biopsy. In our patient, occlusion of the upper small intestine was observed; therefore, we did not consider that a capsule endoscope should be indicated. The upper small intestine can be

observed and biopsied using a conventional small intestinal endoscope, as demonstrated in our patient. In the middle to lower small intestine, a double-balloon endoscope may facilitate the reaching of the lesion site and biopsy-based diagnosis. In the future, advances in/the widespread use of new medical instruments such as capsule/double-balloon endoscopes for small intestinal lesions will shorten the interval until diagnosis and increase the diagnostic rate.

As a rule, resection of the small intestine with lymph node dissection is performed. However, thorough dissection involving the superior mesenteric artery requires extensive small intestinal resection; therefore, it is difficult¹⁴. In our patient, colonic ligamentum infiltration was noted during laparotomy. Therefore, concurrent colectomy was added. In the future, radical surgery should be performed in a larger number of patients. Concerning adjuvant chemotherapy, several studies have reported combination therapy with 5-FU/adriamycin⁹/Taxol and cisplatin¹⁵. However, no therapy has been established. In our patient, UFT was orally administered for postoperative adjuvant chemotherapy.

According to some studies, the 5-year survival rates of primary small intestinal cancer patients range from 9.1 to 38.5%^{11,16}. James *et al.*¹⁷ investigated 144 patients with small intestinal tumors, and reported that the 5-year survival rate for small intestinal cancer was 59%. In particular, the 5-year survival rate in patients undergoing total resection was 81%. However, the prognosis of small intestinal cancer is poor, possibly because cancer is advanced at the time of detection in many patients due to a delayed diagnosis. Our patient also had advanced cancer. However, there has been no relapse during the 3-year-and-3-month postoperative follow-up. In the future, the further development of medical instrument-based techniques may facilitate the early diagnosis of small intestinal cancer in an increasing number of patients, improving treatment results (we searched the literature from the Central Medical Journals issued between 1983 and 2006, using “small intestinal cancer”, “small intestinal endoscope”, and “primary jejunum cancer” as key words. Among the publications, we employed reference/quoted ones as references).

CONCLUSION

We have reported a patient with small intestinal cancer in whom small intestinal endoscopy facilitated a preoperative diagnosis, and reviewed the literature. An abstract of this study was presented at the 88th meeting held by the Japanese Society of Gastroenterology (Asahikawa, April 2002).

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