J Gastroenterol 2003; 38:186–189

Journal of Gastroenterology

Case report

Gastrointestinal stromal tumor of the rectal mesentery

Takamori Nakayama[1], Hayato Hirose[1], Kiyoshi Isobe[1], Kou Shiraishi[1], Takao Nishiumi[1], Syunji Mori[1], Yoshiaki Furuta[1], and Masao Kasahara[2]

[1] Department of Surgery, Shizuoka Red Cross Hospital, 8-2 Ohtemachi, Shizuoka 420-0853, Japan
[2] Department of Pathology, Shizuoka Red Cross Hospital, Shizuoka, Japan

We report a case of gastrointestinal stromal tumor (GIST) arising from the rectal mesentery. GIST of the large intestine is a rare tumor that accounts for only 0.1% of all colorectal cancers. The patient presented to our hospital with constipation and abdominal distension. Computed tomography (CT) revealed a huge mass in the pelvic cavity, and laparotomy disclosed diffuse peritoneal dissemination from the primary tumor. Radiochemotherapy was commenced, but the patient became too ill to complete it and died of the disease 2 months after surgery. A large high-grade tumor with diffuse dissemination was recognized as an indicator of poor survival in this patient.

Key words: gastrointestinal stromal tumor, rectum, mesenterium

Introduction

Sarcoma of the gastrointestinal tract arises from stromal cells of the intestinal wall. The vast majority of these tumors possess myogenic features, and historically they have been classified as leiomyosarcoma.[1] After the advent of immunostaining and electron microscopy, however, some gastrointestinal sarcomas were found to exhibit both myogenic and neurogenic features.[2,3] Accordingly, the term "gastrointestinal stromal tumor (GIST)" was adopted, in 1983.[4] It has recently been proposed that this tumor arises from the interstitial cells of Cajal. GIST is the designation for a major subset of gastrointestinal mesenchymal tumors that show histological, immunohistochemical, and genetic differences from typical leiomyoma, leiomyosarcoma, and schwannoma.[5]

Received: September 3, 2001 / Accepted: January 18, 2002
Reprint requests to: T. Nakayama

Colorectal GIST is a rare tumor that accounts for only 0.1% of all colorectal cancers, and its annual incidence has been estimated at 0.45 per million persons.[6] The prognosis of GIST is poor, with complete surgical excision being the only effective therapy.[7,8] The clinical course and treatment of GIST are similar regardless of its site of origin.[9]

We report a patient with GIST arising in the rectal mesentery who presented with a huge mass in the lower abdomen and underwent laparotomy and radiochemotherapy.

Case report

A 65-year-old man was admitted to our hospital on March 23, 2000. He had suffered from constipation and abdominal distension for 1 month. Examination revealed a mass in the lower abdomen. His inguinal lymph nodes were not palpable and there was no pretibial edema. Standard laboratory tests of serum and urine showed no abnormalities. Carcinoembryonic antigen (CEA) and carbohydrate antigen 19-9 (CA19-9) were within normal limits. He had no relevant past history or family history, apart from diabetes mellitus since the age of 50 years. Computed tomography (CT) and ultrasonography (US) revealed a mass, together with a small amount of ascites, in the pelvic cavity. The lesion was nonhomogeneous. Barium enema (Fig. 1A), magnetic resonance imaging (MRI; Fig. 1B), and angiography (Fig. 2) were also performed.

From the results of these examinations, rectal GIST was diagnosed. On July 3, 2000, laparotomy disclosed a mutilobular tumor was diffuse peritoneal seeding. Because of its poor resectability and curability, the tumor was only biopsied. Histological examination showed that the tumor was formed of spindle-shaped cells. The patient began receiving radiochemotherapy combined with hyperthermia from July 11, 2000. However, treat-

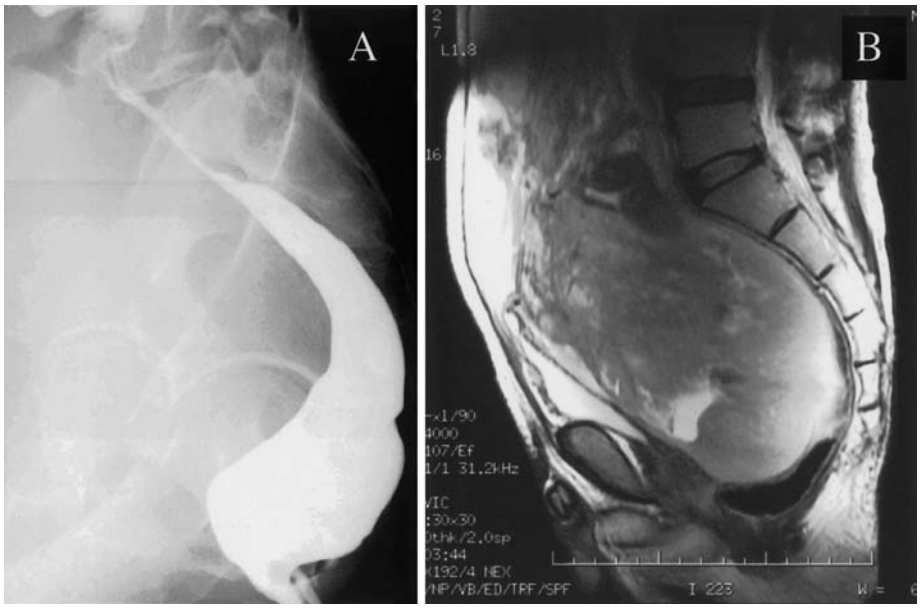

Fig. 1. A The barium-filled rectum gradually tapers to a narrow lumen. No irregular mucosal pattern and no barium-filled tumor cavities are recognized. **B** Magnetic resonance imaging (T2-weighted image) reveals a large tumor that shows a non-homogeneous structure and displaces the rectum

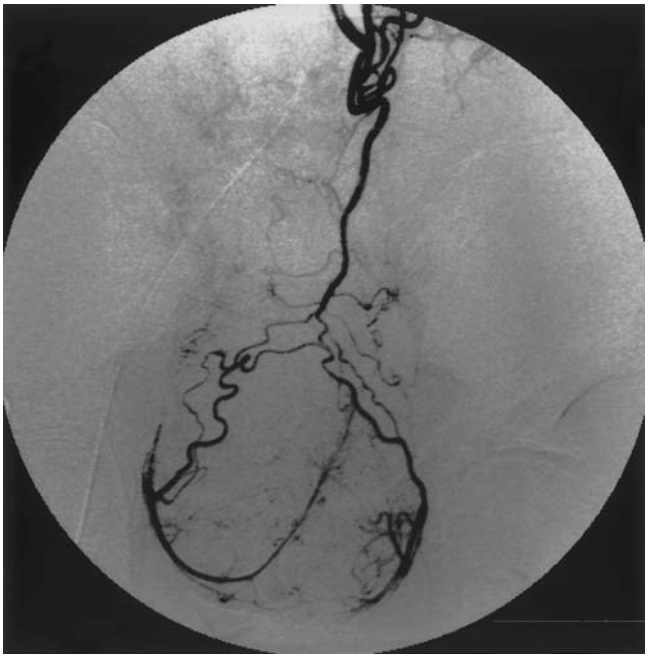

Fig. 2. Inferior mesenteric angiography (arterial phase) reveals that the superior rectal artery is irregularly narrowed and encased. There is also a faint tumor stain

ment was unable to be completed, because his general condition worsened. He developed pneumonia on August 26, 2000, and died of the disease on September 6, 2000.

At autopsy, the tumor measured 35 × 25 × 18 cm in size (Fig. 3A), and there was no involvement of lymph nodes or the liver. Microscopy disclosed hemorrhage, necrosis, hypercellularity, moderate nuclear atypia, and numerous mitotic figures (a mean of 2.8 per high-power field; ×400). Immunohistochemical studies showed that the tumor cells were positive for CD117 (Fig. 3B), CD34, and vimentin, but they were negative for smooth muscle actin (SMA) and neuron-specific enolase (NSE). The final diagnosis was high-grade, uncommitted GIST of the rectal mesentery.

Discussion

Primary malignant mesenchymal tumors can arise anywhere in the alimentary tract, with 5% occurring in the esophagus, 50% in the stomach, 30% in the small bowel, and 15% in the colon and rectum.[10] GISTs arising from the mesentery or the omentum are very uncommon, but such tumors show immunohistochemical features similar to those of gastrointestinal lesions. Tumor-related death occurs at a higher rate in patients with mesenteric GISTs than in those with omental GISTs,[11] but there is little information regarding the clinical behavior of GISTs arising in the rectal mesentery.

The symptoms and signs of GISTs are nonspecific, depending, to some extent, on their size, site, and pattern of growth.[12] Most patients with tumors of the upper gastrointestinal tract tend to complain of a mass, whereas patients who have tumors of the lower bowel usually complain of constipation. GISTs can grow to a considerable size, and the larger tumors tend to be lobulated. This tumor often undergoes central necrosis, which may result in hemorrhage or perforation. Intramural tumor growth occurs in 20% of cases, and usually

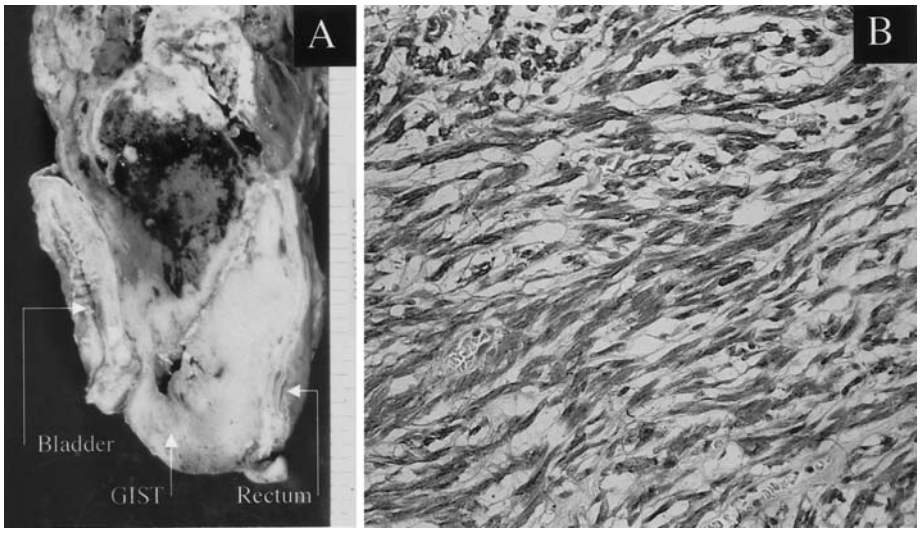

Fig. 3. A Macroscopic cross-section discloses a mesenteric tumor, revealing areas of hemorrhage and necrosis. *GIST*, Gastrointestinal stromal tumor. **B** Immunohistochemistry shows that the tumor cells are positive for CD117 stain. ×200

causes symptoms related to bleeding, even though the extent of involvement tends to be relatively small. In contrast, mesenteric and omental tumors can grow to a large size without causing symptoms.[13] For example, our patient came to our hospital with only the complaints of constipation and abdominal distension.

The preoperative histologic diagnosis is confirmed in fewer than half of all patients at the time of surgery. A large, dumbbell-shape and an irregular mucosal pattern in a tumor are signs suggestive of GIST on barium enema.[1] In our patient, the barium-filled rectum gradually tapered to a narrow lumen, and no tumor cavity was recognized. CT-guided biopsy is not always done because intra-abdominal tumor-cell seeding can compromise future resectability, and patients with a ruptured tumor have a prognosis similar to that in patients with noncurative resection. Percutaneous needle biopsy should only be performed when the tumor is clearly unresectable or when an alternative diagnosis such as lymphoma or germ-cell tumor is suspected. Tumor size, hemorrhage, necrosis, hypercellularity, nuclear atypia, and mitotic figures are the pathological indicators of malignancy. On microscopic examination, the tumor in our patient showed hemorrhage, necrosis, hypercellularity, moderate nuclear atypia, and numerous mitotic figures.

There is little agreement about the treatment and prognosis of lymph-node metastasis from GIST. Because such metastasis is considered an infrequent event in the natural history of these tumors,[14] local excision has a rational basis and elective lymphadenectomy is generally not indicated.

The overall survival rate for patients with a GIST is 28%–35% at 5 years, and the rate increases to 54%–65% after complete resection.[6,8] Patients with a low-grade rectal GIST have a significantly longer median survival than those with a high-grade tumor (median survival, 5–10 years versus 2–3 years).[1,9,12] The available data suggest that only 10%–20% of patients with rectal GISTs are cured by resection and that relapse remains a potential risk for up to 15 years.[1,9] Indolent growth and a slow clinical course have also been noted in gastric GIST.[15] The most common cause of death after the resection of rectal GIST is distant metastasis, rather than local recurrence,[9] and involvement of the liver is a frequent event. In contrast, it is extremely uncommon for limb sarcomas to spread in this way. Surgical resection of hepatic metastases from GIST has been tried, but no patient has survived for 5 years after hepatectomy.[16] The high incidence of liver metastasis, combined with a low incidence of lung metastasis, can be explained by tumor cells invading the portal circulation, with the pattern of hematogenous metastasis of rectal GIST resembling that of rectal carcinoma. The local recurrence rate of rectal GIST is quite high (67%), and primary tumors of the GI tract commonly metastasize to the mesentery.[12] However, autopsy demonstrated that the primary tumor in our patient was located outside the GI tract, in the rectal mesentery.

Chemotherapy does not have a substantial impact on GIST,[6] which shows less response to conventional chemotherapy than carcinoma. There have been conflicting reports concerning the value of adjuvant chemotherapy for rectal GIST, although some studies of limb sarcomas have shown a better response to adjuvant chemotherapy than have GISTs.[9] In view of the rarity of rectal GIST, a prospective study to investigate the role of adjuvant chemotherapy would be difficult.

Adjuvant radiotherapy has been shown to decrease recurrence and to improve survival after curative resection of rectal GIST.[10] Radiotherapy was not completed in our the patient, because he became too ill to continue

it. Brachytherapy, in particular, has been used to minimize visceral and neurological toxicity while delivering a sufficient radiation dose to the tumor bed.[17] There seems to be a strong rationale for the performance of adjuvant radiotherapy in patients with rectal GIST. Local excision and radiation therapy could possibly be an alternative to radical surgery.

In conclusion, we treated a patient who had GIST arising in the rectal mesentery. Exploratory laparotomy and radiochemotherapy were performed, but the patient died of this disease 2 months after surgery. Although our efforts seemed ineffective, we doubt that more aggressive treatment would have saved this patient. A large, high-grade tumor primary tumor associated with peritoneal dissemination is strongly suggestive of poor survival.

References

1. Akwari OE, Dozois RR, Weiland LH, Beahrs OH. Leiomyosarcoma of the small and large bowel. Cancer 1978;42:1375–84.
2. Lauwers GY, Erlandson RA, Casper ES, Brennan MF, Woodruff JM. Gastrointestinal autonomic nerve tumors. A clinicopathological, immunohistochemical, and ultrastructural study of 12 cases. Am J Surg Pathol 1993;17:887–97.
3. Erlandson RA, Klimstra DS, Woodruff JM. Subclassification of gastrointestinal stromal tumors based on evaluation by electron microscopy and immunohistochemistry. Ultrastruct Pathol 1996;20:373–93.
4. Mazur MT, Clark HB. Gastric stromal tumors. Reappraisal of histogenesis. Am J Surg Pathol 1983;7:507–19.
5. Hirota S, Isozaki K, Moriyama Y, Hashimoto K, Nishida T, Ishiguro S, et al. Gain-of-function mutations of c-kit in human gastrointestinal stromal tumors. Science 1998;279:577–80.
6. Meijer S, Peretz T, Gaynor JJ, Tan C, Hajdu SI, Brennan MF. Primary colorectal sarcoma. A retrospective review and prognostic factor study of 50 consecutive patients. Arch Surg 1990;125:1163–8.
7. Conlon KC, Casper ES, Brennan MF. Primary gastrointestinal sarcomas: analysis of prognostic variables. Ann Surg Oncol 1995;2:26–31.
8. DeMatteo RP, Lewis JJ, Leung D, Mudan SS, Woodruff JM, Brennan MF. Two hundred gastrointestinal tumors. Recurrence patterns and prognostic factors for survival. Ann Surg 2000;231:51–8.
9. Lewis JJ, Leung D, Woodruff JM, Brennan MF. Retroperitoneal soft-tissue sarcoma. Ann Surg 1998;228:355–65.
10. Minsky BD, Cohen AM, Hajdu SI. Conservative management of anal leiomyosarcoma. Cancer 1991;68:1640–3.
11. Miettinen M, Monihan JM, Sarlomo-Rikala M, Kovatich AJ, Carr NJ, Emory TS, et al. Gastrointestinal stromal tumors/smooth muscle tumors (GISTs) primary in the omentum and mesentery: clinicopathologic and immunohistochemical study of 26 cases. Am J Surg Pathol 1999;23:1109–18.
12. Khalifa AA, Bong WL, Rao VK, Williams MJ. Leiomyosarcoma of the rectum. Dis Colon Rectum 1986;29:427–32.
13. Shiu MH, Farr GH, Egeli RA, Quan SH, Hajdu SI. Myosarcomas of the small and large intestine. J Surg Oncol 1983;24:67–72.
14. Fong Y, Coit DG, Woodruff JM, Brennan MF. Lymph node metastasis from soft tissue sarcoma in adults. Ann Surg 1993;217:72–7.
15. Shiu MH, Farr GH, Papachristoou DN, Hajdu SI. Myosarcomas of the stomach. Cancer 1982;48:177–87.
16. Japues DP, Coit DG, Casper ES, Brennan MF. Hepatic metastases from soft-tissue sarcoma. Ann Surg 1995;221:392–7.
17. Pisters PW, Harrison LB, Leung DH, Woodruff JM, Casper ES, Brennan MF. Long-term results of a prospective randomized trial of adjuvant brachytherapy in soft tissue sarcoma. J Clin Oncol 1996;14:859–68.