An Endoscopic Case Report of Jejunal Leiomyosarcoma and Review of the Japanese Literature

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ABSTRACT. Jejunoscopy for a small intestinal leiomyosarcoma was performed on a 73-year-old male. The patient was admitted because of a palpable abdominal mass. A barium meal study, abdominal ultrasonography, computed tomography and angiography suggested a leiomyogenic tumor with a central cavity arising from the jejunal loop extraluminally. Enteroscopy with a pediatric colonofiberscope (PCF) enabled us to observe a mucosal ulcer and a part of the cavity of the tumor. Histology of the resected tumor revealed it to be a leiomyosarcoma with a large central hollow connected to the mucosal ulcer. The authors discussed the endoscopic findings of 17 previously reported Japanese cases of jejunal leiomyosarcoma and the present case.

Key words: jejunoscopy — tumor of the small intestine — endoscopy

Tumors of the small intestine are generally rare compared with those of the stomach and the large intestine. Early detection and diagnosis of small intestinal tumors are also difficult and delayed. Recently, reports of jejunoileal leiomyosarcoma have increased as a result of the development of such diagnostic imaging methods as abdominal ultrasonography and computed tomography, but the number of cases in which enteroscopy has been performed has been small. We present a case of jejunal leiomyosarcoma in which we were able to observe a mucosal ulcer and a part of the cavity of the tumor endoscopically.

CASE REPORT

A 73-year-old man was admitted to our hospital on October 16, 1991 with a two-month history of an abdominal mass, low-grade fever, abdominal pain

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and blacky stools. On physical examination, the abdominal mass, which was about 10 cm in diameter and located on the left side of the navel, anemic conjunctiva and essential finger tremors were noted. No other abnormalities were found on the skin, or in the chest and abdomen. His laboratory data showed the immunological occult blood test of feces to be strongly positive. A hematological test revealed a microcytic hypochromic anemia (hemoglobin 8.4 g/dl, hematocrit 27.1%) and mild leukocytosis (white blood cell count 10,200/ μl). Serum C-reactive protein was 6.8 mg/dl and the erythrocyte sedimentation rate was 90 mm in an hour. Tumor markers such as carcinoembryonic antigen, alpha fetoprotein and carbohydrate antigenic determinant 19-9, were within normal limits.

Chest X ray films showed old tuberculous shadows in the right apical and middle lung fields, but sputum cultures for tubercle bacilli were negative.

A barium meal study revealed a so-called blank space with an irregular shaped barium-filled cavity in the upper portion of the jejunal loop (Fig. 1a, b).

Abdominal ultrasonography disclosed a pseudokidney sign that was a hypoechogenic shadow of the tumor with a central hyperechogenic cavity (Fig. 2a).

Abdominal computed tomography showed a low density mass which moved to the right side of the navel when tested, with an irregular Gastrografin-filled cavity connected to the jejunal loop (Fig. 2b).

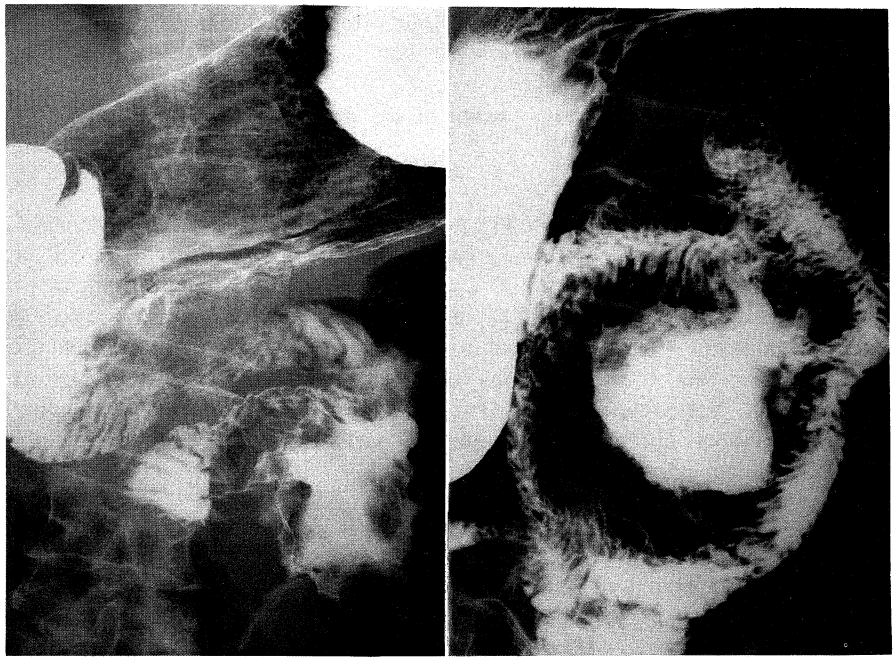

Fig. 1a,b. A barium meal study reveals extraductal irregular leakage of barium and a blank space in the upper jejunal loops.

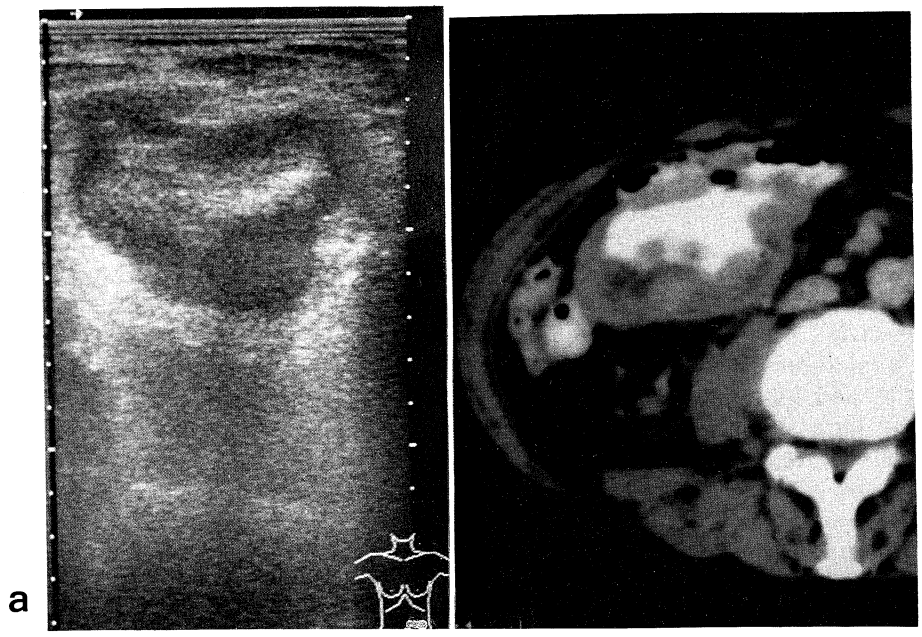

Fig. 2 a: An abdominal ultrasonogram shows a pseudokidney shadow.
b: Abdominal computed tomography shows a low density tumor with a cavity filled with contrast medium administered orally.

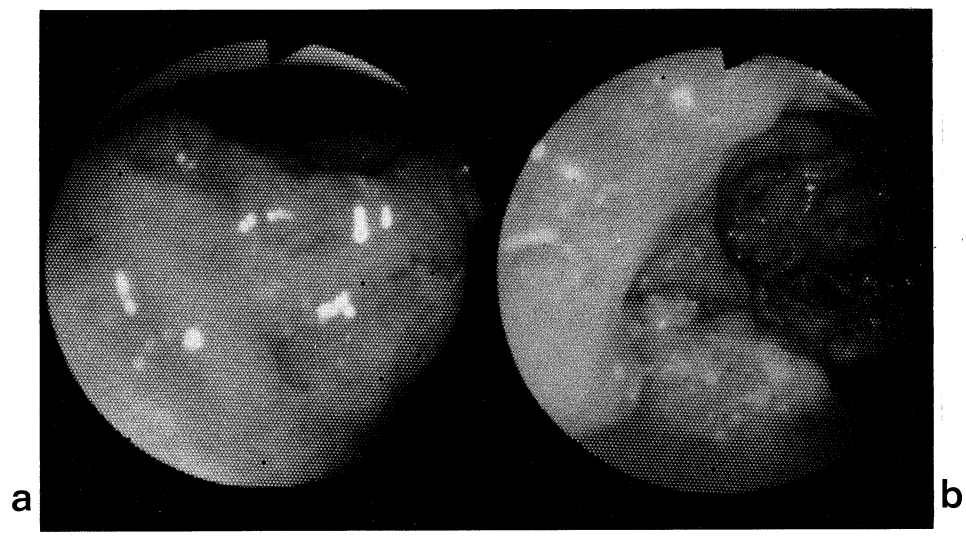

Fig. 3 a: Enteroscopy reveals a large jejunal ulcer opening into a cavity at the lower right. The upper lumen leads anally.
b: A part of the cavity shows a septal but relatively smooth inner surface and food contents.

Jejunoscopy with a pediatric colonofiberscope (PCF_{10}: Olympus Corp. Japan) disclosed a large ulcer of about 2 cm in diameter with a slightly elevated margin and a part of the cavity which had a ridged and smooth surface and stored jejunal food contents (Fig. 3a,b). We failed to take biopsy specimens from the rim of the ulcer and the surface of the cavity because of the difficulty of a forceps biopsy.

An abdominal superior mesenteric angiogram indicated that the mass was fed from the first and second jejunal branches, in which hypervascularities, tumor stains with large avascular areas and early venous returns were found (Fig. 4a,b).

The preoperative and presumptive diagnosis was a jejunal leiomyosarcoma in extraluminal development, with an mucosal ulcer connected to an autolyzed large cavity.

Laparotomy was performed on November 7, 1991. A large tumor with a whitish and lobular surface was found arising from the mesenteric side of the jejunal loop about 5 cm below the ligament of Treitz. An accessory nodule of 0.6 cm in size near the main tumor was separately attached on the anti-mesenteric side of the jejunal loop about 3 cm below the ligament of Treitz. A partial jejunectomy involving the main tumor and an extirpation of the nodule were performed.

The resected main tumor was $9 \times 7.8 \times 5$ cm in size. It had an irregular cavity about 5 cm in diameter on a cut surface, and there was a deep ulcer of 2.2×1.5 cm in size in the mucosal side of the tumor (Fig. 5a,b).

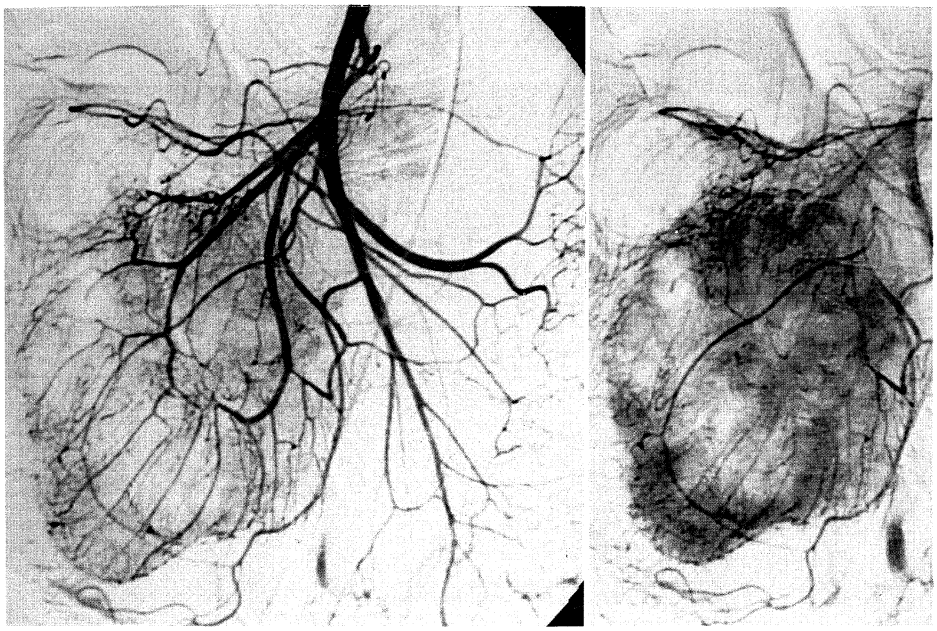

Fig. 4a,b. An abdominal angiogram reveals a hypervascullar tumor with outer stains and inner avascular areas supplied from the jejunal branches of the superior mesenteric artery.

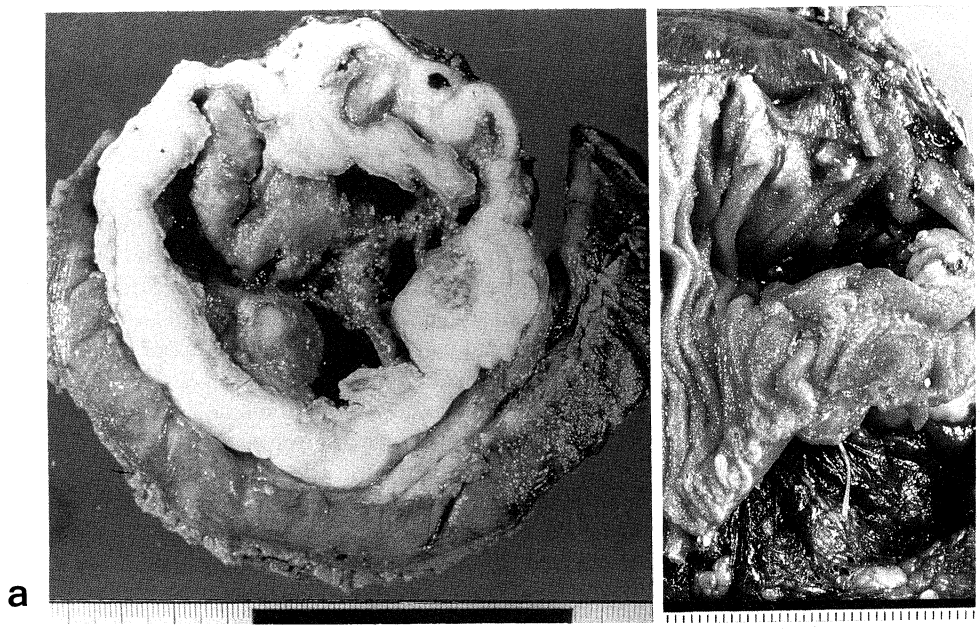

Fig. 5 a: The resected tumor on a cut surface shows a large cavity. (mm; scale)
b: A deep jejunal ulcer in the mucosal side of the tumor (mm; scale)

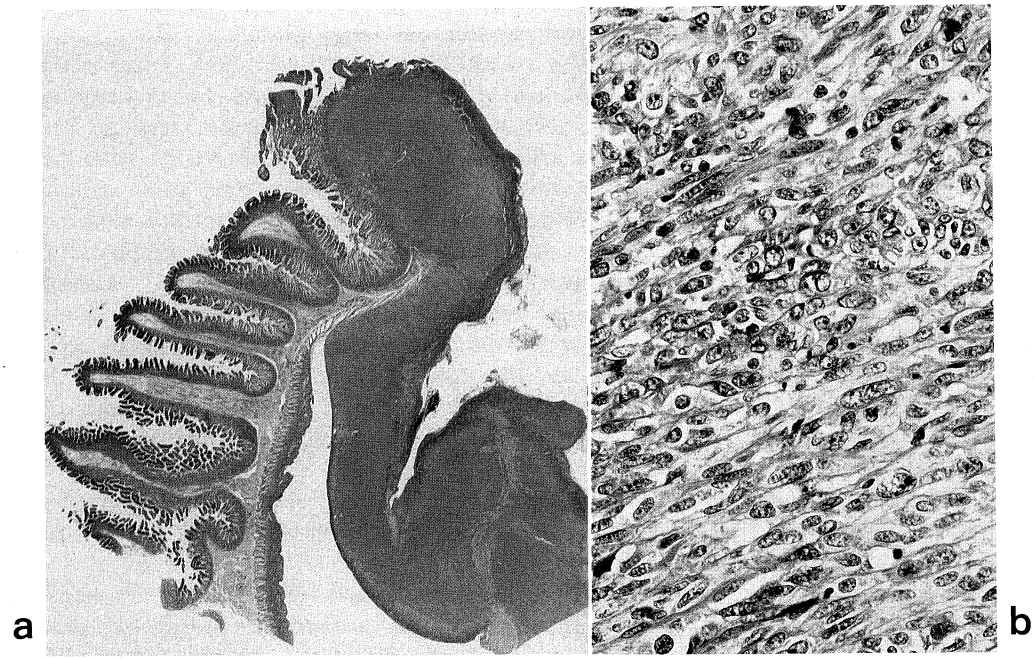

Fig. 6 a: A part of the tumor shows a margin of the ulcer leading into the cavity at the upper right. (×3.6, H.E.)
b: Proliferations of epitheloid and spindle cells reveal a leiomyosarcoma. (×290, H.E.)

Histology revealed a leiomyosarcoma originating in the proper muscle layer of the jejunal wall, with more dominant proliferation of epithelioid cells than spindle cells and areas of moderate pleomorphism with mitosis (Fig. 6a, b). Desmin immunohistostaining was slightly positive. The other tumor was a disseminated leiomyosarcomatous nodule in fibrous tissues.

DISCUSSION

In the American literature,[1,2] malignant tumors of the small intestine involving the duodenum make up 0.2-2 per cent of all malignant tumors of the gastrointestinal tract. Carcinoids, adenocarcinomas, malignant lymphomas and leiomyosarcomas are the most common malignant tumors in order of incidence.[2] In Japan, tumors of duodenal origin are excluded from those of the small intestine.[3] Among jejunoileal malignant tumors, malignant lymphomas, adenocarcinomas and leiomyosarcomas are most common, but carcinoid tumors are rare.[3] The incidence of leiomyosarcomas of jejunal origin is about three times greater than those of ileal origin, and about 80 per cent of jejunal leiomyosarcomas arise from the loop within 60 cm below the ligament of Treitz. The clinical aspects of leiomyosarcoma in Japan include common findings of male predominance with a male to female ratio of approximately 2 to 1, a high incidence in the fifth and sixth decades of age, an average size of about 10 cm in diameter and extraluminal development of about 90 per cent of the tumor.[4,5] Pain, a palpable mass and recurrent gastrointestinal bleeding are a common clinical triad. Other manifestations include anemia, perforative peritonitis[6] and intussusception.[7]

Preoperative diagnostic procedures are generally a barium meal study with small intestine follow-through and abdominal angiography. Roentgenographically, a larger extraluminal mass shows a so-called blank space with separated and distorted loops. V-shaped deformity of the loop due to hanging of the tumor, a crater with a defect and extraluminal barium leakage are definite findings that the tumor has arisen from or is closely connected with the small intestine. A smaller tumor, however, is difficult to detect even with a selective small bowel study involving selective infusion of contrast medium through a long tube advanced past the ligament of Treitz. Angiographically, leiomyogenic tumors are hypervascular during the arterial phase. Tumor staining with or without an avascular area due to inner necrosis of the tumor and early venous returns due to arteriovenous shunting are also characteristic. Although determination of whether the tumor is benign or malignant is difficult, arteriography is very useful and valuable for the diagnosis of leiomyogenic tumors, for identifying their feeding artery, and for making a decision about resectability.[8]

Recently the use of abdominal ultrasonography and computed tomography with enhancement by contrast medium is also increasing and being applied to the diagnosis of these tumors.[2]

We studied and summed up the clinical aspects of 18 cases of small intestinal leiomyosarcoma endoscopically reported in Japan including our case (Table 1). The male to female ratio was 1.6 to 1 and the average age was 57 years old. Abdominal pain, a palpable mass and recurrent melena were equally chief complaints. In many cases, these findings were almost the same as those

TABLE 1. Summary of 18 cases of jejunal leiomyosarcoma observed endoscopically in Japan

	Name	Year	Age Sex	Complaint	Site	Type	SMT*	Biopsy 1/2	Type of Endoscope	Size of Tumor(cm)	Literature (in Japanese)	
1	Saito	1977	38M	P T	5cm	IIIa	+	+/+[3]	SIF-B	7.5×6×5	Gastroenterol Endosc	19 : 563-70
2	Nagase	1977	76 F	T B	5cm	IIIa		+/+	JF-B_2	14×18×9	Progr Digest Endosc	11 : 185-8
3	Nishimura	1980	43 F	B	?cm	III	+	?	?	?	Jpn J Gastroenterol	77 : 842-3
4	Uchida	1980	31M	P	7cm	IIb	+	+/−	JF-B_3	8	Progr Digest Endosc	17 : 245-8
5	Asakura	1980	55M	P	7cm	IIIb	+	+/+[4]	SIF-B	3×4	Gastroenterol Endosc	22 : 1761-9
6	Nakagami	1981	54M	T	just	I a	+	+/−	SIF?	14×10×8	J Jpn Soc Clin Surg	42 : 800-5
7	Masuda	1982	49M	T B	60cm	III(b)		+/+	SIF?	10×10×10	Gastroenterol Endosc	24 : 997
8	Shimada	1982	35M	P T	anal	III		+/+	SIF-B	11×18×19	Progr Digest Endosc	21 : 265-8
9	Miyachi	1982	51 F	P B	20cm	IIIa	+	+/−	JF-B_4	1.5×2×5	Progr Digest Endosc	21 : 269-72
10	Enoki	1982	70 F	T	7cm	III		+/−	FIS-V_a	17×9×9	Progr Digest Endosc	21 : 273-5
11	Miyashita	1983	61M	P T	?cm	?		+/−	?	?	Jpn J Gastroenterol	81 : 1724
12	Sakakibara	1984	54M	B	just	IIIa		+/+	SIF-B	5×6	Progr Digest Endosc	24 : 311-4
13	Kimura	1984	64 F	T	70cm	IIIa	+	−	SIF-B**	7×9×12	Shimane Chu-Byo-I-Shi	11 : 170-4
14	Oikawa	1986	69M	P B	5cm	IIIa	+	−	JF	4.5×4.2×4	Progr Digest Endosc	29 : 359-61
15	Tsuno	1987	63 F	B	just	IIIa	+	−	SIF-P_3	10×8×7.5	Nagasaki Igk Z	62 : 68-72
16	Hondo	1988	68M	B	11cm	IIa	+	+/−	?	14×12×11	J Hiroshima Med Ass	41 : 977-81
17	Harada	1990	63 F	B	20cm	?		+/?	JF?	5	Gastroenterol Endosc	32 : 2768
18	Authors' case	1993	73M	P T B	5cm	IIIa		−	PCF_{10}	9×7.8×5		

P : abdominal pain T : palpable tumor B : gastrointestinal bleeding Site : distance below the ligament of Treitz
Type : endoscopic classification of small intestinal tumors I : sessile tumor II : (semi-) pedunculated tumor
III : large tumor with central depression a ; partially encircling b ; completely encircling * : like a submucosal tumor endoscopically
[1] : + ; biopsy performed [2] : + ; diagnosed as leiomyosarcoma − ; not performed/diagnosed
[3] : diagnosed as leiomyogenic tumor [4] : diagnosed as sarcoma ** : by a ropeway method
SIF-B, FIS-V_a, SIF-P_3 : enteroscope JF : often used for ERCP PCF : pediatric colonofiberscope

in the American[1,9] and Japanese[4] literature in general for small intestinal leiomyosarcomas. In the endoscopic classification of small intestinal tumors by Kawai, *et al*,[10] type III (a large tumor with a central depression) was observed in 12/15 cases. Among these cases, there were more of the partially encircling type (IIIa) than of the completely encircling type (IIIb). A submucosal tumor-like appearance was also detectable in half of the cases. In 72 per cent of cases (13/18) an endoscopic biopsy was successful. In nearly half (6/13) of those cases the biopsy was valuable for preoperative diagnosis. A push type enteroscope of a side-view JF scope, which is usually employed for endoscopic retrograde cholangiopancreatography (ERCP) was used. Although a PCF has not been used for enteroscopy in Japan, jejunoscopy with a PCF was useful beyond the ligament of Treitz in our case and those of Maglinte, *et al*.[11] With the exception of one case, jejunal endoscopy within 60 cm below the ligament of Treitz was performed by a push method.

Survival from primary malignancies of the small intestine has not improved.[11] The major delay in diagnosis occurred after the patient received medical help and not from the onset of symptoms to the first medical consultation. Physicians must order the appropriately selected imaging studies and the studies must not be misread by radiologists. Only by considering the small bowel as a potential source of unexplained abdominal symptoms can the prompt application of selective methods of examination be made with the hope of improving the patient's prognosis.[11]

Enteroscopy is rarely selected as the first method of investigation for small bowel diseases in Japan, but in one case, emergency endoscopy with an ordinary gastroduodenoscope for acute intestinal bleeding revealed a submucosal leiomyoma in the stenotic jejunal loop.[12] Lewis, *et al* reported that as further examination for chronic gastrointestinal bleeding of obscure origin, sonde-type enteroscopy disclosed a small intestinal leiomyosarcoma.[13]

In addition, fine needle aspiration cytology guided by ultrasonography for a large abdominal mass has revealed a leiomyosarcoma arising from the jejunum at operation,[14,15] although aspiration might affect the incidence of peritoneal dissemination. There have been other reports of initial scintigraphic detection and localization of jejunal leiomyosarcoma utilizing Tc-99m labeled sulfur colloid[16] or red blood cells.[17] This technique may be also used for obscure gastrointestinal bleeding.

As treatment,[4] complete resection of the tumor, the adjacent small intestinal loop and regional lymph nodes is necessary in the same manner as surgery for an adenocarcinoma. Local recurrence, peritoneal dissemination and hematogenous spread to the liver are common, but lymphatic spread is not ordinary. Recent technical developments in hepatic resectioning have made the treatment of hepatic metastases possible.[8]

Histologically, the tumor shows spindle cell proliferations with elongated nuclei and an arrangement of interdigitating or pallisading fascicles. This makes it possible to recognize its derivation from smooth muscle in some areas of the tumor on careful observation by Masson's trichrome staining.[9] It also exhibits nuclear mitosis and pleomorphism ; *e.g.*, sparse and rough chromatin of tumor cells. Dominant proliferations of oval cells with round nuclei showing clear cytoplasm are sometimes visible in leiomyosarcomas, which are also called

epithelioid leiomyosarcomas or malignant leiomyoblastomas.[18] However, there seems to be no apparent clinical significance in classifying small intestinal leiomyosarcomas into spindle cell type and epithelioid cell type because a mixture of both cell types in the same tumor is detectable in varying degrees. Therefore, a marker of the mitotic rate is more useful and significant.[2] Even if a leiomyogenic tumor is diagnosed as a benign tumor because of no histological metastasis and mitosis, there must be a postoperative follow up of a patient with a mass larger than 5-6 cm in diameter, extraluminal growth and central necrosis and/or mucosal ulceration.

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