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Gastrointestinal Stromal Tumor of the Stomach with Narrow Stalk-Like Based, Uneven Protruding Appearance Presenting with Severe Acute Anemia despite Small Size

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Key Words
Gastrointestinal stromal tumor · Stomach · Stalk · Bleeding

Abstract
We report the case of a 56-year-old woman who had a gastrointestinal stromal tumor (GIST) of the stomach. She was admitted to our hospital for epigastric pain, nausea, and severe acute anemia (hemoglobin level 4.3 g/dl). Esophagogastroduodenoscopy revealed a narrow stalk-like based, hemorrhagic and uneven protruding lesion in the lesser curvature of the gastric upper corpus. Although the tumor was less than 2 cm in diameter and was probably a benign GIST according to histology, laparoscopy-assisted local resection was needed because the patient had continuous severe anemia and epigastric pain. Histological assessment showed that the elongated spindle-like tumor cells originated from the intrinsic muscle layer, and was shown with growth to the mucosal side, cropping out to the surface in most areas of the protruding lesion. Only a small part of the tumor was within nontumoral gastric mucosa. Most of the tumor cells demonstrated immunoreactivity for KIT and CD34 in the cytoplasm but not for αSMA,

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S100, and desmin. Mitotic activity (0/50 high power field) and the labeling index for MIB-1 (about 1%) were low. The GIST of the stomach described in this report was a rare case with a narrow stalk-like based, uneven protruding mass presenting with severe acute anemia despite small size.

Introduction

Gastrointestinal stromal tumors (GISTs), which arise primarily in the gut wall, are most commonly mesenchymal neoplasms closely related to the interstitial cells of Cajal. GISTs typically carry gain-of-function mutations in genes encoding the KIT receptor tyrosine kinase (CD117) or platelet-derived growth factor receptor a, both of which are involved in cell survival, development and proliferation [1]. Although GISTs can arise at any location in the gastrointestinal tract, they are found most often in the stomach (60–70%), followed by the small intestine (20–30%), colon and rectum (5%), and esophagus (<5%) [2–4]. In most cases in the stomach, GISTs show the appearance of a submucosal tumor, with a broad base and smooth surface within normal gastric mucosa, and sometimes have central 'delle' or depression [5, 6]. In larger cases, central ulceration or necrosis presenting with gastrointestinal bleeding are also observed [7, 8]. The GIST of the stomach described in this report was a rare case, with narrow stalk-like based, uneven protruding mass presenting with severe acute anemia despite small size.

Case Report

A 56-year-old woman was admitted to Fujita Health University with epigastric pain and nausea during the previous one month. Physical examination revealed pale conjunctiva and epigastric tenderness. Laboratory evaluation on admission showed severe anemia (red blood cell count 138×10^6/μl, hemoglobin level 4.3 g/dl, hematocrit 13.7%) (**table 1**). Esophagogastroduodenoscopy revealed a narrow stalk-like based, hemorrhagic and uneven protruding lesion in the lesser curvature of the gastric upper corpus (**fig. 1**). Endoscopic ultrasonography showed a low echoic mass, measuring 2.0 cm in diameter, within the fourth layer, suggesting that the tumor originated from the intrinsic muscle layer, and showed growth to the mucosal side (**fig. 2**). Biopsy specimens were taken from the protruding lesion during esophagogastroduodenoscopy. The histological finding showed spindle-like tumor cells positive for KIT and CD34. The labeling index (LI) for MIB-1, determined by counting positively stained nuclei among 1,000 tumor cells, was 1%. No lymph node or distant metastasis were detected by computed tomography and abdominal ultrasonography.

Although the histological findings suggested that the tumor was probably a benign GIST, laparoscopy-assisted local resection was performed after 2 weeks because the patient had continuous severe anemia and epigastric pain. The postoperative course of the patient was excellent. Anemia and epigastric pain dramatically improved after resection. She was discharged on the 8th postoperative day.

The resected tumor was 1.8 × 1.5 × 1.0 cm in size within the locally resected surgical speciemen. The tumor was elastic but hard in consistently, and its surface was uneven and irregular (**fig. 3**). The histological assessment was done cutting the specimen into 10 pieces (**fig. 4**a). The tumor cells originated from the intrinsic muscle layer and showed growth to the mucosal side, cropping out to the surface in most areas of the protruding lesion. Only a small part of the tumor was within nontumoral gastric mucosa (fig. 4a, b). Histological assessment revealed that the tumor was composed of elongated spindle-like cells, containing rounded or oval, relatively uniform nuclei without apparent atypia (**fig. 5**a). Mitotic activity was low (0/50 high power field [HPF]). The LI for MIB-1, determined by counting positively stained nuclei among 1,000 tumor cells, was about 1% (fig. 5b). Immunohistochemical staining of the tumor by the avidin-biotin peroxidase complex method showed that most of the tumor cells demonstrated immunoreactivity for KIT (fig. 5c) and CD34 (fig. 5d) in the cytoplasm, but not for αSMA (fig. 5e), S100 (fig. 5f) and desmin (fig. 5g).

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Discussion

The tumor described in this report showed immunoreactivity for KIT and CD34, but not for αSMA, S100 and desmin, implying that the tumor was pathologically a GIST [9] with a lack of differentiation toward smooth muscle, and neural elements. Most GISTs in the stomach generally show the appearance of submucosal tumors, with a broad base and smooth surface within normal gastric mucosa, and sometimes have small central 'delle' or depression [5, 6]. In larger cases, central ulceration or necrosis presenting with gastrointestinal bleeding are also observed [7, 8].

However, the GIST of the stomach described in this report showed narrow stalk-like based, uneven protruding mass presenting with severe acute anemia despite the small size of less than 2 cm, and local resection was needed due to continuous severe anemia and epigastric pain. Although the association between the patient's gastric symptoms and this small submucosal tumor is not fully understood, the considerable amount of blood in the stomach due to continuous bleeding may have led to the gastric symptoms such as epigastric pain and nausea. Histological assessment showed that the tumor cells showed growth to the mucosal side, cropping out to the surface in most areas of the protruding lesion, and only a small part of the tumor was within nontumoral gastric mucosa, despite the tumor cells originating from the intrinsic muscle layer. Thus the case of our report may be a rare case for its morphological appearance as well as clinical course.

Metastasis and/or recurrence after surgery may be observed for several GISTs, despite an initial diagnosis of a benign tumor. Therefore, it is recommended to apply risk classification based on tumor size and mitoic count under a HPF [10–12]. The MIB-1(Ki-67) LI or the presence or absence of tumor necrosis is also reported to reflect proliferation activities and malignant outcome [11, 13]. The GIST described in our report was less than 2 cm in diameter without necrosis, and had low mitotic activity (0/50 HPF) and LI for MIB-1 (about 1%). In this context, our GIST may have a very low malignant potential and thus metastasis and/or recurrence may not be observed in the future. Actually, the patient in this report has been disease-free 18 months after surgery.

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Table 1. Laboratory findings

WBC, /μl	10,500
RBC, $\times 10^6$/μl	138
Hb, g/dl	4.3
Ht, %	13.7
Plt, $\times 10^4$/μl	19.9
MCV, fl	99
MCH, pg	31.9
MCHC, %	32.1
TP, g/dl	5.4
Alb, g/dl	3.1
T-Bil, mg/dl	0.3
AST, IU/l	9
ALT, IU/l	8
ALP, IU/l	120
γ-GTP, IU/l	8
TG, mg/dl	106
BUN, mg/dl	18.3
CRE, mg/dl	0.46
Na, mEq/l	136
K, mEq/l	3.8
Cl, mEq/l	100
CRP, mg/dl	<0.3
FBS, mg/dl	168
CEA, ng/ml	2.1
CA19-9, U/ml	11.2

Fig. 1. Esophagogastroduodenoscopy finding. A narrow stalk-like based, hemorrhagic and uneven protruding lesion was found in the lesser curvature of the gastric upper corpus.

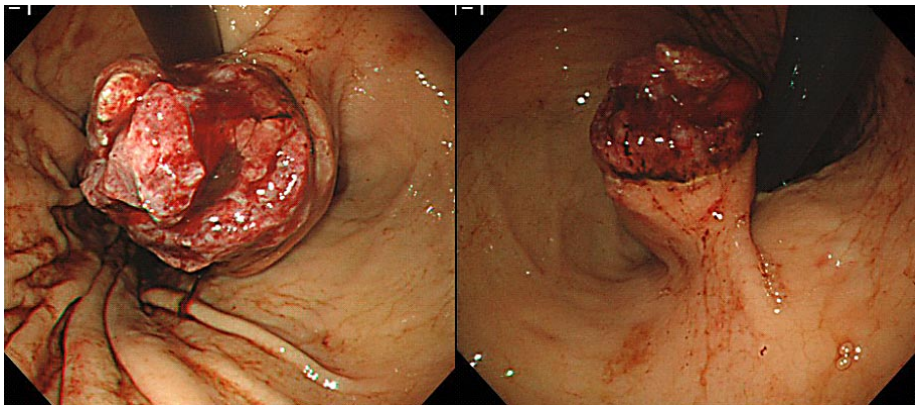

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Fig. 2. Endoscopic ultrasonography finding. A low echoic mass, measuring 2.5 cm in diameter, was found in the fourth layer, suggesting that the tumor originated from the intrinsic muscle layer, and showed growth to the mucosal side.

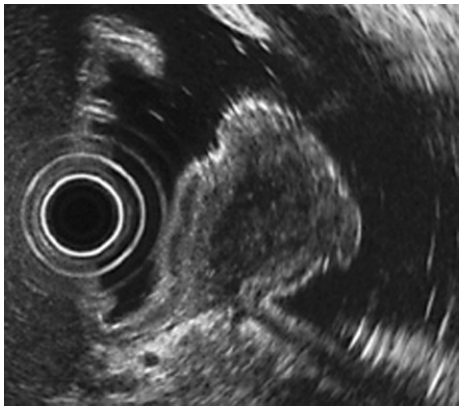

Fig. 3. The surgical specimen. The resected tumor was 1.8 × 1.5 × 1.0 cm in size, elastic but hard in consistently, and its surface was uneven and irregular.

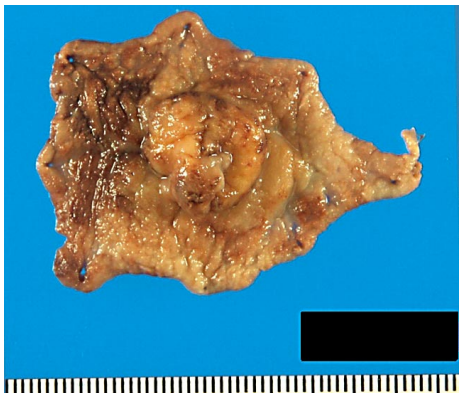

Fig. 4. Schema of the tumor extension, and low-power histological view. The histological assessment was done cutting the specimen into 10 pieces (**a**; white lines). The tumor cells originated from the intrinsic muscle layer and showed growth to the mucosal side, cropping out to the surface in most areas of the protruding lesion (**a**, red lines, and **b**). Only a small part of the tumor was with in nontumoral gastric mucosa (**a**; yellow lines).

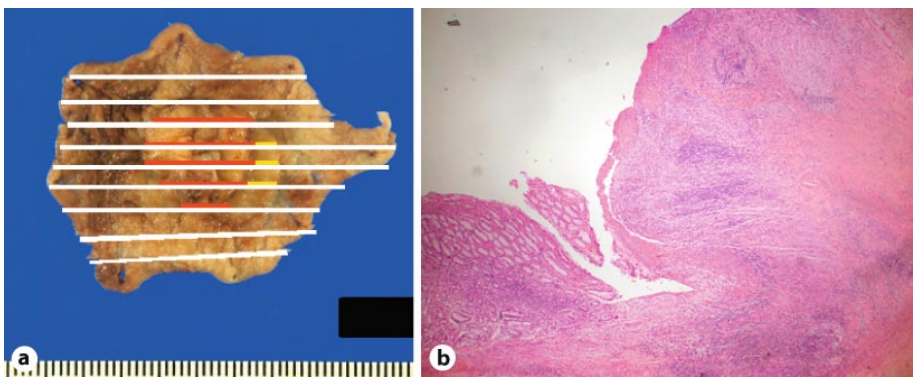

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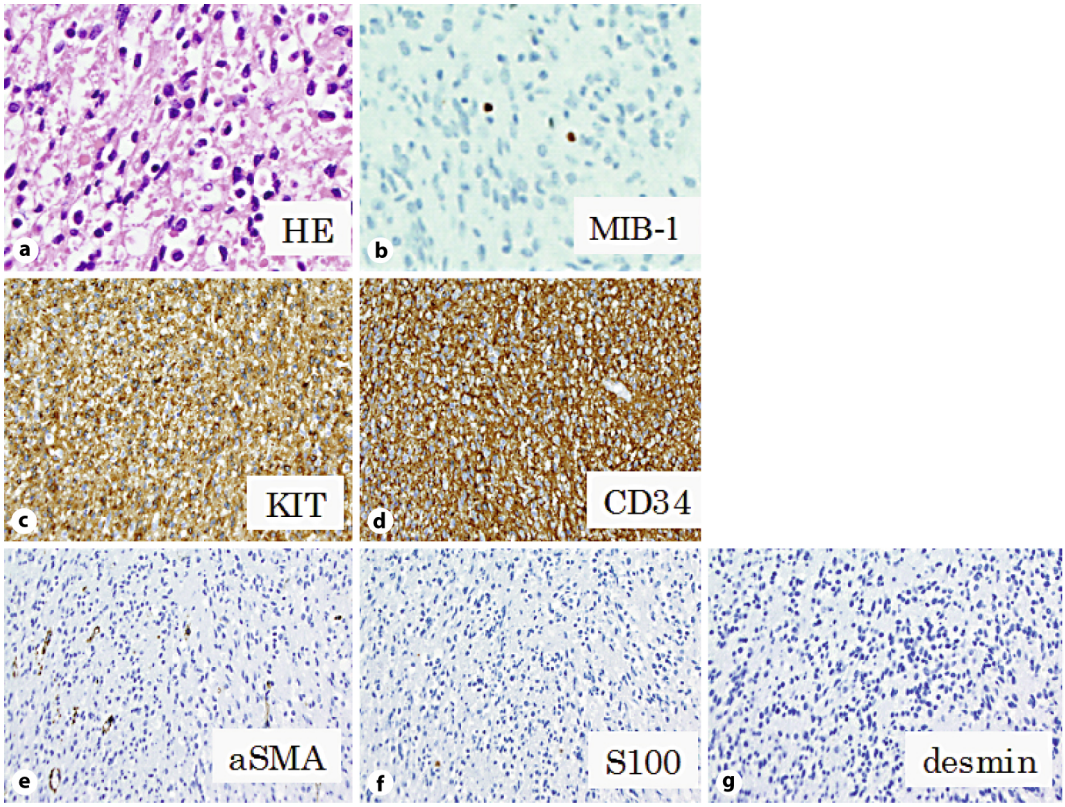
Fig. 5. Histological view. The tumor was composed of elongated spindle-like cells. The spindle-like tumor cells contained rounded or oval, relatively uniform nuclei without apparent atypia. Mitotic activity was low (0/50 HPF) (**a**). The LI for MIB-1, determined by counting positively stained nuclei among 1,000 tumor cells, was about 1% (**b**). Immnohistochemical staining of the tumor by the avidin-biotin peroxidase complex method showed that most of the tumor cells demonstrated immunoreactivity for KIT (**c**) and CD34 (**d**) in the cytoplasm but not for αSMA (**e**), S100 (**f**), and desmin (**g**).



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References

1 Hirota S, Isozaki K, Moriyama Y, et al: Gain-of-function mutations of c-kit in human gastrointestinal stromal tumors. Science 1998;279:577–580.

2 Judson I: Gastrointestinal stromal tumours (GIST): biology and treatment. Ann Oncol 2002;13(suppl 4):287–289.

3 Miettinen M, Majidi M, Lasota J: Pathology and diagnostic criteria of gastrointestinal stromal tumors (GISTs): a review. Eur J Cancer 2002;38(suppl 5):S39–S51.

4 Pidhorecky I, Cheney RT, Kraybill WG, Gibbs JF: Gastro-intestinal stromal tumors: current diagnosis, biologic behavior, and management. Ann Surg Oncol 2000;7:705–712.

5 Kamimura H, Mochiki E, Kamiyama Y, Aihara R, Kimura H, Kuwano H: Gastrointestinal stromal tumor of the stomach: report of a case. Hepatogastroenterology 2005;52:1297–1300.

6 Shirai H, Takeuchi T, Naka T, et al: Gastrointestinal stromal tumor of the stomach: report of a case. Surg Today 2001;31:346–349.

7 Djukić V, Karamarković A, Mijatović S, et al: Gastrointestinal gastric tumor (GIST) as a cause of massive hemorrhage from the upper digestive tract. Acta Chir Iugosl 2007;54:169–171.

8 Kimura H, Yoshida T, Kinoshita S, Takahashi I: Pedunculated giant gastrointestinal stromal tumor of the stomach showing extragastric growth: report of a case. Surg Today 2004;34:159–162.

9 Fletcher CD, Berman JJ, Corless C, et al: Diagnosis of gastrointestinal stromal tumors: A consensus approach. Hum Pathol 2002;33:459–465.

10 Wong NA, Young R, Malcomson RD, et al: Prognostic indicators for gastrointestinal stromal tumours: a clinicopathological and immunohistochemical study of 108 resected cases of the stomach. Histopathology 2003;43:118–126.

11 Singer S, Rubin BP, Lux ML, et al: Prognostic value of KIT mutation type, mitotic activity, and histologic subtype in gastrointestinal stromal tumors. J Clin Oncol 2002;20:3898–3905.

12 Fujimoto Y, Nakanishi Y, Yoshimura K, Shimoda T: Clinicopathologic study of primary malignant gastrointestinal stromal tumor of the stomach, with special reference to prognostic factors: analysis of results in 140 surgically resected patients. Gastric Cancer 2003;6:39–48.

13 Hasegawa T, Matsuno Y, Shimoda T, Hirohashi S: Gastrointestinal stromal tumor: consistent CD117 immunostaining for diagnosis, and prognostic classification based on tumor size and MIB-1 grade. Hum Pathol 2002;33:669–676.