jgc
J Gastric Cancer 2013;13(3):192-195 • http://dx.doi.org/10.5230/jgc.2013.13.3.192 Case Report

Gastrointestinal Stromal Tumor with Extensive Lymphatic Metastasis: A Case Report

Ki Young Kang, and Woong Lee[1]

Department of Anatomy, [1]Department of Surgery, Seonam University College of Medicine, Namwon, Korea

Gastrointestinal stromal tumor is a rare tumor which arises from the whole gastrointestinal tracts and most of it is detected in the stomach. It is uncommon with small intestine originated gastrointestinal stromal tumor and more uncommon with lymphatic metastasis. We experienced an unusual case of the small bowel gastrointestinal stromal tumor during experimental autopsy. Two primary tumors with central necrosis were detected in the ileum. The sizes of each tumor were 6.1×3.4×4.0 cm and 3.7×4.2×3.2 cm. There was extensive lymphatic metastasis on the greater omentum and mesenteric, iliac lymph nodes were also involved. With histologic findings, the eosinophilic spindle cells were densely distributed. Immunohistochemical findings were CD117 (-), CD34 (+), desmin (-), and S-100 protein (-). Therefore, we diagnosed the tumors as small bowel gastrointestinal stromal tumors with broad lymph node mestasis.

Key Words: Gastrointestinal stromal tumors; Lymph nodes; Neoplasm metastasis

Introduction

Gastrointestinal stromal tumor (GIST) is very rare tumor arisen from gastrointestinal tract that constitutes 0.1%~3.0% of gastrointestinal malignant tumor.[1-5] GISTs may be occurred at the esophagus, stomach, small intestine, large intestine, anal canal, mesentery, greater omentum, and retorperitoneum.[6] The small intestine constitutes 80% of the length, and 90% of the mucosal surface area of the digestive tract. However, the tumors originated from the small intestine are very rare because of various reasons, such as rare exposure to carcinogens due to rapid transit of food materials and rapid turnover of the epithelia, alkaline environment of bowel contents, richness of immunoglobulin A, and inconsiderable intestinal bacterial flora. The tumors arisen from the small intestine accounts for 5% of the tumors occurred in the digestive tract and the malignant tumors of the small intestine constitutes 1%~2% of the malignancies of the digestive tract. Fifty percents of the small intestine derived malignant tumors are the adenocarcinoma, and less than 20% are the malignant GISTs.[7]

In some cases, the researchers classified the leiomyoma and the leiomyosarcoma into the smooth muscle cell derived tumors or GISTs derived from the interstitial cell of Cajal.[8] However, in recent years, the pathologists clearly discriminate between the smooth muscle cell derived tumors and GISTs according to the immunohistochemical results.[9] GIST is occurred under the mucosa as a single mass. GIST may directly invade adjacent tissues or organs, and spread to other organs hematogenously, or metastasize to the liver or the omentum by peritoneal dissemination during the surgical operation. But lymphatic spread of GIST is very uncommon.[7] The authors experienced atypical case of GIST has two primary tumors in the ileum and extensive metastasis to the greater omentum and adjacent lymph nodes during cadaver dissection for medical students. Therefore, the authors report the case with review of the literatures.

Correspondence to: Woong Lee
Department of Surgery, Seonam University College of Medicine, 439 Chunhyang-ro, Namwon 590-711, Korea
Tel: +82-62-370-7750, Fax: +82-62-371-3092
E-mail: dkuma@naver.com
Received July 8, 2013
Revised July 23, 2013
Accepted August 2, 2013

Case Report

A 53-year-old male cadaver was donated to our laboratory. He was 175 cm tall and weighed 80 kg. The cause of death was acute myocardial infarction. There was no evidence of trauma or surgery. During an autopsy, the authors observed morphological features and distribution of the tumors and took photographs. The histological samples were obtained and analysed by hematoxylin-eosin stain and immunohistochemistry and then consulted with pathologist on the microscopic findings of the tumors.

After the opening of the anterior abdominal wall and peritoneum, a large number of round and various-sized tumors (0.5~2.4 cm in diameter) were exposed on the greater omentum (Fig. 1). The superior part of the greater omentum was stuck in the right iliac fossa and adhered to adjacent peritoneal surfaces. The authors examined the intestinal loop following the removal of the greater omentum, and then the first primary mass was detected on the ileum at 90 cm proximal to the ileocecal junction. The mass was 6.1 ×3.4×4.0 cm sized. It was firm and adhered to adjacent parietal peritoneum and the greater omentum. A cutting plane of the mass was bright brown coloured and the cavity of the central necrosis was open to the lumen of the intestine (Fig. 2). The second primary mass was detected at the middle of the small intestinal loop and 3.7×4.2×3.2 cm sized (Fig. 3). It was adhered to adjacent small bowel and its cutting plane was bright brown coloured. The small foci of the central necrosis and a pattern of swirl were also observed on cutting planes. Moreover, a lot of mesenteric and iliac lymph nodes were enlarged.

The tumor mass lied within the tunica muscularis of the intestinal wall and there was no evidence of mucosal invasion. The tumor cells were spindle-shaped and densely packed in swirling pattern. The cytoplasm of the cells were eosinophilic and the nucleus did not show distinct mitotic figure. Lymphatic spread was confirmed by identification of the subcapsular lymphatic sinus within the pathologic samples obtained from the greater omentum, mesentery, and posterior abdominal wall (Fig. 4). The CD117 antigen was negative and the CD34 was partially positive in immunohistochemical assay (Fig. 5). The desmin and the S-100 protein were not detected. Consequently, the authors diagnosed this tumor as the GIST with lymph node metastasis on the basis of the diagnostic guide of Hirota and Isozaki.[10]

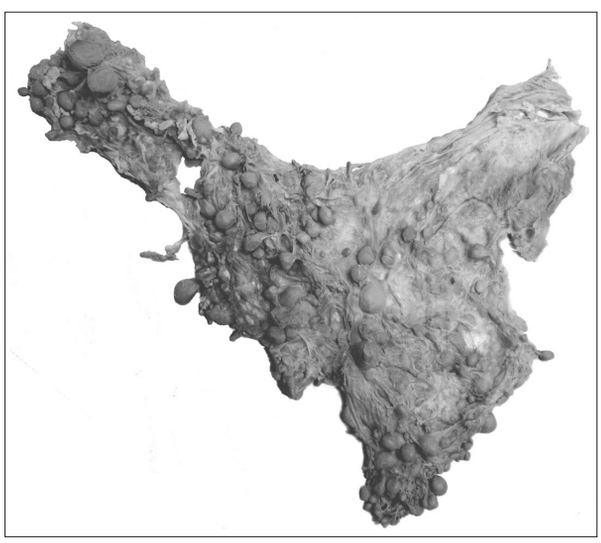

Fig. 1. Removed greater omentum shows numerous tumor masses.

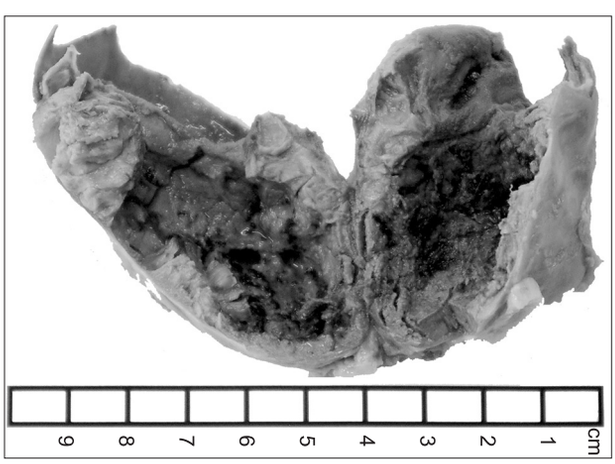

Fig. 2. Sectional planes of the first tumor. The cavity resulted from central necrosis is communicated with the intestinal lumen.

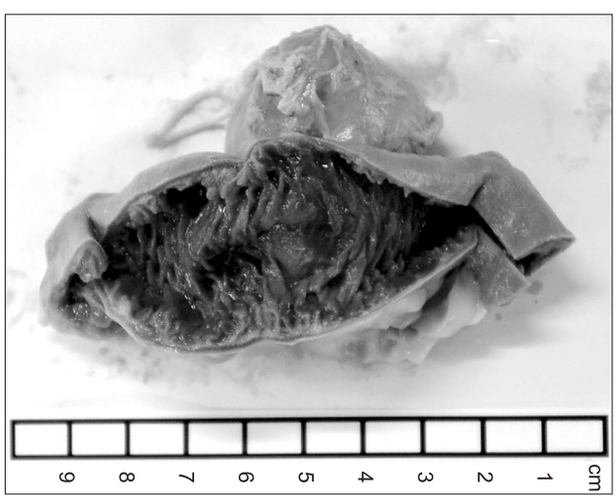

Fig. 3. Second tumor mass is located under the intact intestinal mucosa.

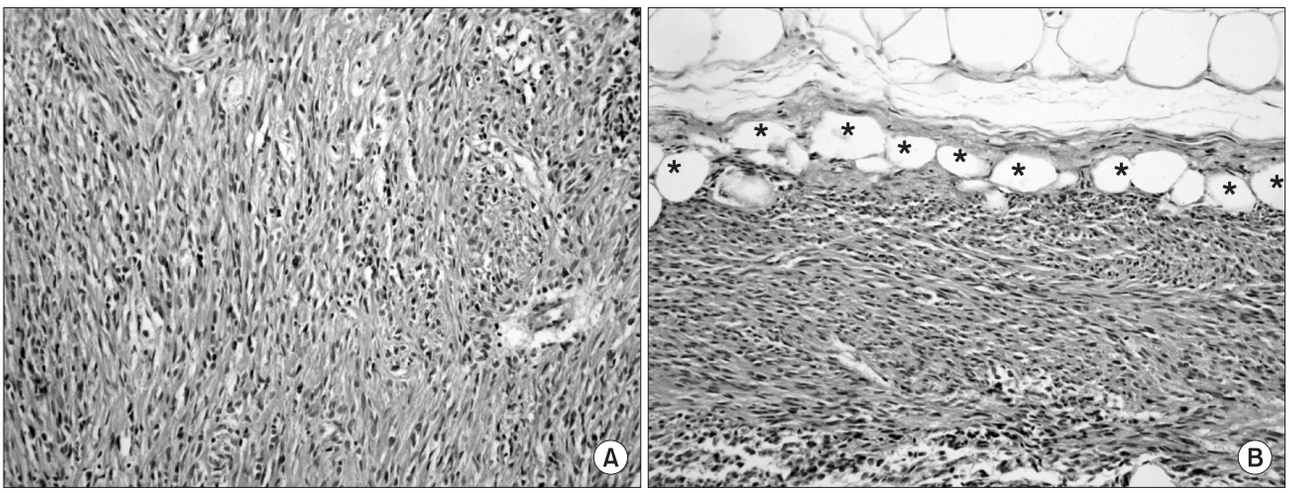

Fig. 4. H&E stain (×200). (A) Spindle tumor cells are arranged in interlacing fascicles. (B) Asterisks indicate the subcapsular sinusoids of the lymph node.

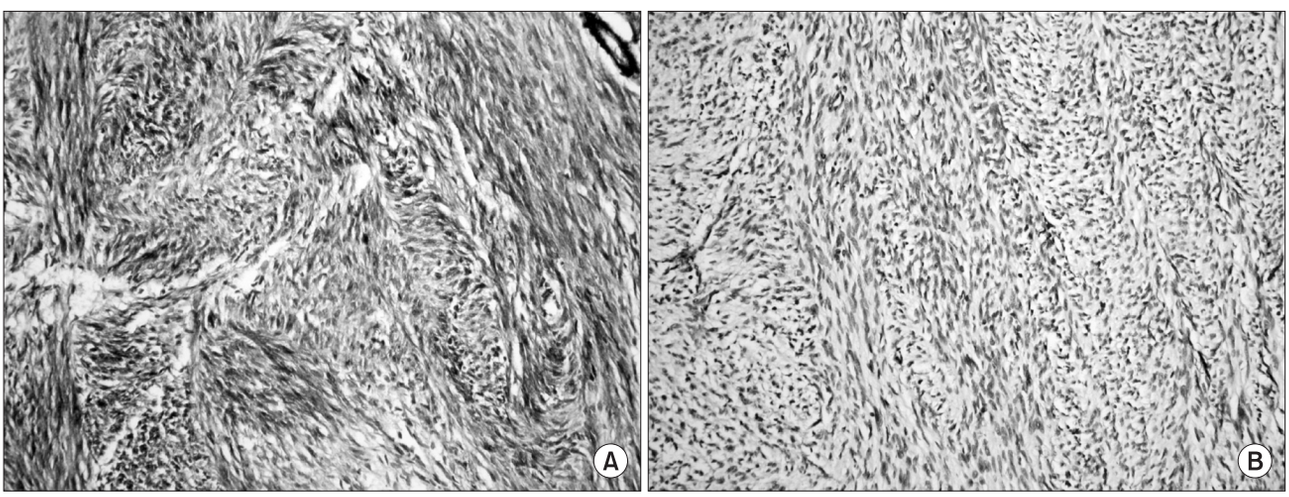

Fig. 5. Immunohistochemial stain (×200). (A) Smooth muscle actin is strongly expressed in whole cells of the tumor. (B) CD34 is expressed in the some of the tumor cells.

Discussion

The stromal tumor developed in the gastrointestinal tract is very uncommon and that occurred within the small intestine is extremely rare. The most common site of GISTs in the digestive tract is the stomach and it comprises more than 50%. The second common site is the small intestine and the esophagus is the most rare primary lesion.[8] Within the small intestine, the jejunum is the most common region of primary GISTs and the ileum is the most uncommon region. The jejunal GISTs are half as many as that occurred in the duodenum and the ileum.[8,11]

In the past, the stromal tumors developed in the digestive tract had been regarded as the smooth muscle derived tumors and classified into subtypes base on the histological characteristics until the cellular origin was confirmed. Since then, the researchers discovered that the cellular origin of GISTs was the interstitial cell of Cajal and this cell mostly expressed *c-kit* gene product CD117.[9] Nowadays, GISTs are diagnosed by the immunohistochemical analysis of several tumor markers, such as CD117, CD34, desmin, and smooth muscle actin (SMA), and detection of the CD117 strongly supports confirm diagnosis of the GIST.[8,9] The CD117 is detected at 95% of the GISTs. The CD34, SMA, S-100 protein, and desmin are positive at 60%~70%, 30%~40%, 5%, and 1%~2% of GISTs, respectively.[10] Hirota and Isozaki[10] classified the stromal tumors into GISTs, smooth muscle tumors, and Schwann cell tumors based on the above data. They categorized the stromal tumors

expressing the CD117 or the CD34 as GIST. The stromal tumors without the CD117 and the CD34 were classified into the smooth muscle tumors which were positive for the desmin or the Schwann cell tumors which were positive for the S-100 protein.

In this case, the authors diagnosed the tumors as GIST because it did not express the CD117, desmin, and S-100 protein, but expressed CD34 in abundance. It is well known that the SMA is detected around half of GISTs removed from the small intestine.[12] In our case, the tumors also expressed the SMA (Fig. 5).

Radical resection is recommended for the treatment of GISTs. But localized resection and chemotherapy are performed in some cases. Recently, clinicians have been using the tyrosine kinase inhibitor STI-671 (imatinib mesylate, Gleevec; Novartis pharmaceuticals, Basel, Switzerland) to cure GISTs and it brings good results. It is remarkable consensus that resection of lymph nodes are unnecessary because lymphatic spread of GISTs is very uncommon. Morinaga et al.[13] proposed that complete resection of GISTs, even if they are small in size, is compulsory, but routine lymph node dissection is unnecessory. Pross et al.[14] also reported that lymph node dissection was useless in treatment of GISTs of the stomach. According to the National Institutes of Health consensus classification system,[9] our case falls under the intermediate grade, but it shows unusual feature that the tumor cells extensively metastasize to the lymph nodes and the greater omentum.

In our case, two primary GIST masses were found at the ileum and they showed clinically malignant features, such as size of the tumors, central necroses, lymphatic spread of the tumor cells, but they peculiarly did not show local invasion of the tumor cells and mitotic figures on high power field of microscopy. This specific case has not been reported in published literatures. Therefore, the authors report this unusual case of GIST with brief review of the literatures and we hope that this report will be helpful to the researchers engaged in the basic and clinical medical sciences.

References

1. Sircar K, Hewlett BR, Huizinga JD, Chorneyko K, Berezin I, Riddell RH. Interstitial cells of Cajal as precursors of gastrointestinal stromal tumors. Am J Surg Pathol 1999;23:377-389.
2. Rubin BP, Singer S, Tsao C, Duensing A, Lux ML, Ruiz R, et al. KIT activation is a ubiquitous feature of gastrointestinal stromal tumors. Cancer Res 2001;61:8118-8121.
3. Kim CJ, Day S, Yeh KA. Gastrointestinal stromal tumors: analysis of clinical and pathologic factors. Am Surg 2001;67:135-137.
4. Kim EK, Lee HC, Lee MR, Lim SB, Kang SB, Park KJ, et al. Gastrointestinal stromal tumors (GISTs) of the colon and rectum: clinicopathological analysis. J Korean Soc Coloproctol 2003;19:282-289.
5. Eisenberg BL, Judson I. Surgery and imatinib in the management of GIST: emerging approaches to adjuvant and neoadjuvant therapy. Review Ann Surg Oncol 2004;11:465-475.
6. Miettinen M, Sarlomo-rikala M, Lasota J. Gastrointestinal stromal tumors: recent advances in understanding of their biology. Hum Pathol 1999;30:1213-1220.
7. Townsend CM Jr, Beauchamp RD, Evers BM, Mattox KL. Sabiston Textbook of Surgery: The Biological Basis of Modern Surgical Practice. 18th ed. Philadelphia: Saunders Elsevier, 2008:1271-1272.
8. Pidhorecky I, Cheney RT, Kraybill WG, Gibbs JF. Gastrointestinal stromal tumors: current diagnosis, biologic behavior, and management. Ann Surg Oncol 2000;7:705-712.
9. Fletcher CD, Berman JJ, Corless C, Gorstein F, Lasota J, Longley BJ, et al. Diagnosis of gastrointestinal stromal tumors: a consensus approach. Hum Pathol 2002;33:459-465.
10. Hirota S, Isozaki K. Pathology of gastrointestinal stromal tumors. Pathol Int 2006;56:1-9.
11. Howe JR, Karnell LH, Scott-conner C. Small bowel sarcoma: analysis of survival from the National Cancer Data Base. Ann Surg Oncol 2001;8:496-508.
12. Heo GW, Shin DW, Paik SY, Kim ID, Kim KH, Suh BS, et al. Comparative study of the clinicopathologic characteristics between gastrointestinal stromal tumors arising from the stomach and small bowel. J Korean Gastric Cancer Assoc 2007;7:254-260.
13. Morinaga N, Sano A, Katayama K, Suzuki K, Kamisaka K, Asao T, et al. Laparoscopic transgastric tumor-everting resection of the gastric submucosal tumor located near the esophagogastric junction. Surg Laparosc Endosc Percutan Tech 2004;14:344-348.
14. Pross M, Manger T, Schulz HU, Lippert H, Roessner A, Günther T. Gastrointestinal stromal tumors--problems in diagnosis and therapy. Chirurg 1999;70:807-812.