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A rare case of giant gastrointestinal stromal tumor of the stomach involving the serosal surface



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ABSTRACT

INTRODUCTION: Although rare, gastrointestinal stromal tumors (GIST) are the most common mesenchymal tumors affecting the gastrointestinal tract.

PRESENTATION OF CASE: Here we report the case of a 43-year-old man complaining of abdominal pain along with a painless and palpable mass, which was confirmed on magnetic resonance and multislice computed tomography. Laparotomy revealed a nodular grayish-white firm noninfiltrative mass $(39 \times 27 \times 14 \, \text{cm}, 6109 \, \text{g})$ that was well localized within the extramuscular and peritoneal surface of the anterior wall of the stomach; complete tumor resection was performed. Histopathological examination revealed features typical of GIST, including increased cellularity, increased mitotic activity, and spindle shaped cells as well as positive immunoreactivity for KIT, CD34, and vimentin.

DISCUSSION: A review of literature revealed that GISTs of the size and weight similar to the present case has been rarely reported. GIST most frequently involves the stomach. Although the etiopathogenesis of this disease remains unclear, few well-documented familial cases have been associated with GIST syndromes.

CONCLUSION: The primary treatment preferred is complete surgical excision of the tumor.

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1. Introduction

Gastrointestinal stromal tumors (GISTs) are the most frequent mesenchymal neoplasms of the gastrointestinal tract [1]. They are rare, accounting for approximately 0.1-3% of all gastrointestinal tumors [2]. Most GISTs occur in patients aged >50 years at the time of diagnosis [3]. The etiological factors of GIST have not been determined so far [4], with the stomach beingthe most frequently (60%) affected site [5]. Clinically, most patients present with asymptomatic tumors are detected incidentally [6]. Generally, GISTs are thought to originate from interstitial cells of Cajal or their precursors [3]. These tumors are usually positive for KIT (CD117), and the combination of characteristic histopathological features and KIT (CD117) positivity is observed in 95% of cases [2,7]. GISTs have been associated with molecular alterations and mutations in platelet-derived growth factor receptor alpha (PDGFRA) [7]. The size of these tumors ranges from a few millimeters to several centimeters in diameter [6]. GISTs >10 cm in diameter are referred to as giant GISTs. To the best of our knowledge, giant GISTs of

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this size and weight have been rarely reported in the literature (Table 1).

2. Presentation of case

A 43-year-old male arrived at the emergency department of a state hospital with nonspecific systemic symptoms, abdominal pain and weakness, and local examination was detected to have a painless, palpable abdominal mass. Patient also gave a no history of trauma or mass in the abdomen. Abdominal ultrasonography, computed tomography (CT), and magnetic resonance (MR) imaging revealed the presence of a solid cystic mass in the intra-abdominal region, which was clinically considered as a tumor and was resected completely (Fig. 1).

The tumor measured $39 \times 27 \times 14\,\mathrm{cm}$ in diameter and weighed 6109 g. Serial sections of the surgical specimen did not reveal tumor infiltration to the any major organ compartment. Gross findings of the nonperitonealized perimuscular or mesentery tissues included the presence of a nodular, grayish-white, firm, well-localized and well-demarcated, smooth mass. The cut surface of the tumor revealed ill-defined solid areas that were diffuse gray, with focally hemorrhagic, myxoid, and cystic degeneration (Fig. 2). Macroscopically, tumor perforation and ulceration were not observed and the tumor margins were negative and focaly surgical margins close.

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Table 1Anatomic location, age, gender, size (cm) and weight (kg) distributions of gastrointestinal stromal tumor

References	Year	Anatomic location	Age (year)	Gender	Size (cm)	Weight (kg)
Kitabayashi et al. [25]	2001	Stomach	75	M	15 × 11 × 4.4	
Kimura et al. [26]	2004	Stomach**	84	F	20	
Mehta et al. [22]	2005	Stomach	75	M	13 × 10	
Dal Corso et al. [27]	2007	Stomach	88	F	$17\times13\times9$	1.630
Cruz Jr et al. [14]	2008	Stomach	37	M	$32\times25\times21$	3.750
Funahashi et al. [28]	2008	Stomach**	65	F	$25\times18\times11$	
Alder et al. [29]	2013	Rectum	70	M	10×8.5	
Cappellani et al. [30]	2013	Stomach	67	M	$37\times24\times13$	8.5
Colović et al. [31]	2013	Stomach	52	F	20.5×16	
Notani et al. [32]	2013	Stomach	58	M	22	
Skandalos et al. [2]	2013	Stomach ^a	79	F	10.77×9.67	
Misawa et al. [33]	2014	Jejunum	70	M	10 × 10	
Mu et al. [34]	2014	Esophagus	29	M	$13\times12\times5$	
Nakano et al. [35]	2014	Esophagus	65	F	18	
Schneider et al. [36]	2014	Stomach	71	M	$19\times18\times16$	2.6
In our case	This	Stomach ^b	43	M	$39\times27\times14$	6.109

kg: kilogram, cm: centimetre, M: male, F: female.

^b Extragastric growth.

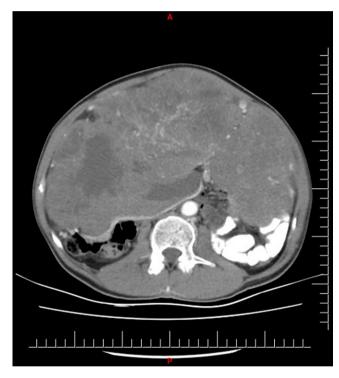


Fig. 1. Computed tomography (CT) scan of the abdominal showed tumors fulfilling.

Histologically, the tumor appeared to consist predominantly of spindle cells and multinucleated giant cells along with low-moderate levels of cytological pleomorphism. The cells were arranged in a prominent fascicular pattern with areas of collagenization and mitosis [mitotic rate; 30 per 50 high-power fields (HPF)]. In addition, focal necrosis and a myxoid stroma were also present. The histopathological features of the lesion in the present case were similar to those seen in high-risk tumors (Fig. 3). Therefore, it was classified as a high-grade (grade 2) tumor. Lymphovascular invasion and calcifications were not observed. However, the tumor cells were diffusely and strongly positive for KIT (CD117, cytoplasmic and membranous), CD34 (Figs. 4 and 5) and vimentin. The cells were focally positive for S-100 and neuron-specific enolase, but negative for desmin, smooth muscle actin, epithelial membrane antigen, synaptophysin, calretinin, and CK5/6.



Fig. 2. Gross morphology: the macroscopicappearance of gastrointestinal stromal tumors (GIST), multinodular features and gray-white firm, peritoneal surface intact.

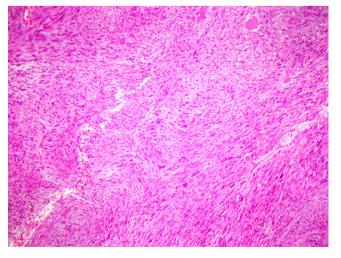


Fig. 3. GIST low power view demonstrating prominent fasicular pattern, predominantly spindle cell cytomorphology (hematoxylin–eosin, original magnification $\times 40$ objective).

^a Lesser curvature.

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