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Original Article General Surgery

Primary extra gastrointestinal stromal tumors of the abdomen

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ABSTRACT

OBJECTIVE: In the present study, we aimed to evaluate the clinicopathological features and prognostic factors in extra-gastrointestinal stromal tumor (EGIST) cases, which are observed very rare, by examining the data the cases obtained in a single center.

METHODS: Data of 14 EGIST cases who were operated by a general surgeon between January 2007 and May 2020 were obtained and analyzed.

RESULTS: The median age was 47.5 (range: 34–87) years. A total of 135 patients were operated for GIST, and 14 (10.4%) of these patients were EGIST. The mean tumor diameter was 16.8±10.5 (range: 2.8–40) cm. The mitotic index was 5/50 high power field and below in seven (50%) cases. Twelve (85.7%) of the patients were in the high-risk group. The overall survival (OS) rate was 80%, and the 5-year survival rate was 88.9%. Mean OS was 78.5±50.7 months, 5-year OS and disease-free survival (DFS) were both 53.3±20.0 months, and overall DFS was 58.0±59.8 months. The mean OS and DFS durations were found to be significantly lower in women than men (p=0.006 for both comparisons). The mean OS was found to be significantly lower in patients over 60 years of age compared to those aged 60 and under (p=0.013).

CONCLUSION: In the present study, it has been determined that the rare EGISTs are large in size and that the mitotic index is often low. In addition, it has been observed that the prognosis may be similar to other GISTs, however, may be worse in elderly patients and in women.

Keywords: Extra-gastrointestinal stromal tumor; gastrointestinal stromal tumor; prognosis.

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Although gastrointestinal stromal tumors (GISTs) are among the most common mesenchymal tumors, they rarely develop outside the gastrointestinal tract and are called extra-GISTs (EGISTs) [1–3]. Although EGISTs mostly have similar features to GISTs, they show many differences in terms of both histopathological, clinical, and prognosis. It is important to know the characteristics of EGIST and to distinguish them with GISTs [3].

It has been reported that EGISTs are mostly aggressive compared to GISTs, their prognosis is poor, they are mostly detected in larger sizes, the mitotic index is generally low, and they more frequently affect women and older people [3, 4]. However, the fact that some different results were obtained in different studies raises questions about these generalizations. In addition, there are different opinions on the definitive diagnosis of EGISTs and their precise distinction from GISTs and other tumors [2, 5]. The fact that the studies on EGIST and the low number of cases in these studies, the fact that most of the published studies are case reports or case series, the limited information about the features of EGIST cases and the significant variability in the data indicate that new

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studies on EGIST are needed. In the present study, it was aimed to evaluate prognostic factors and indicators by examining 14 EGIST cases followed up in a single center.

MATERIALS AND METHODS

The present study was approved by the Kartal Dr. Lutfi Kirdar City Hospital Clinical Resources Ethics Committee (date: 11/11/2020; approval number: 514/189/5), and planned, retrospectively. The study was conducted in accordance with the principles of the Declaration of Helsinki.

Patients and Criteria

A total of 135 patients with GIST who were operated in the General Surgery Department of our hospital in the 13-year period between January 2007 and May 2020 were screened. A total of 17 of those tumors were out of the gastrointestinal tract. Three of those were not proved to be extra-intestinal GIST and were excluded from the study. Finally a total of 14 cases were included in the study. Demographic information, clinical, laboratory, pathology, and radiology findings of all patients were recorded. Patients were called by phone, and we learned whether the patients lived.

Since DOG-1 review has been studied in our hospital since November 2014, there is no DOG-1 data from before this date.

The risk classification developed according to the tumor diameters and mitotic indexes of patients were grouped based on the National Institutes of Health prognostic criteria. According to this classification, the high-risk category was defined as (i) tumor size in largest dimension >5 cm and mitotic count >5/5 high power field (HPF), (ii) tumor size >10 cm and mitotic count at any rate, or (iii) tumor with any size and mitotik count >10/50 HPF [6, 7].

GISTs that develop from the gastrointestinal tract and are associated with the serosa of the stomach, small, or large intestine and whose origin is not known exactly were excluded from the study. Three patients with retroperitoneal masses who were thought to be GIST but whose differential diagnosis could not be made from the other mesenchymal tumors by immunohistochemical methods and histopathological diagnosis could not be confirmed were excluded from the study. Tumors not originating from the GIS serosa or submucosa were included in the study.

Highlight key points

- Tumor size is mostly large in EGISTs.
- Although it is reported to be more common in women, in our study it is found to be frequent in men also.
- The prognosis may be worse in women and in elderly patients.

Statistical Analysis

All statistical analyzes in the study were performed using SPSS 25.0 software (IBM SPSS, Chicago, IL, USA). Descriptive data were given as numbers and percentages. Whether continuous variables were suitable for normal distribution was confirmed by Shapiro–Wilk Test. Differences between groups in terms of continuous variables were analyzed using Independent Samples' t test. The results were evaluated at 95% confidence interval, and $p<0.05$ values were considered significant. Bonferroni correction was made where appropriate.

RESULTS

The rate of EGISTs among GIST cases was 10.4% (14/135). Nine (64.3%) of the patients were male, and five (35.7%) were female. Eight (57.1%) patients had palpable mass, and five (35.7%) had abdominal pain. Nine (64.3%) of the tumors were in mixed histopathological type, four (28.6%) were in spindle cell type, and only one (7.1%) was in epithelioid structure. Colon adenocarcinoma was present in one (7.1%) of the cases, and EGIST was detected incidentally in that case (Table 1).

All patients were diagnosed by examining the resection material. Tumor was imaged with pre-operative computed tomography in all patients, and magnetic resonance imaging and/or endoscopy methods were also used in six patients. Three of the patients (21.3%) had comorbid diseases such as diabetes mellitus and/or hypertension (Table 1).

Metastasis was present in three (21.3%) patients at the time of diagnosis; in the peritoneum in two patients and in the liver in one patient. Lymph node metastasis was not detected in any of the patients. The most common location of the tumor was the abdominal cavity (7/14) (Table 1).

The majority of the tumors (10/14; 71.4%) had a diameter of 10 cm or more. Only one (7.1%) tumor was smaller than 5 cm. A total of 12 (85.7%) patients were in the high risk group, and two (14.3%) were in the me-

Table 1. Distribution and ratios of some variables

	n	%*
Gender		
Men	9	64.3
Women	5	35.7
Symptoms		
Palpabl mass	8	57.1
Pain	5	35.7
Asymptomatic	1	7.1
Histopathological cell type		
Mixed	9	64.3
Spindle	4	28.6
Epithelioid	1	7.1
Second primary tumor (colon adenocarcinoma)	1	7.1
Incidental	1	7.1
Diagnosis with resection material	14	100.0
Pre-op biopsy	1	7.1
Pre-op imaging		
CT	8	57.1
CT + MRI	1	7.1
CT + Endoscopy	3	21.4
MRI + Endoscopy	1	7.1
CT + MRI + Endoscopy	1	7.1
Concomitant disease	3	21.3
HT	1	7.1
DM + HT	1	7.1
Asthma	1	7.1
Lymph node metastasis	0	0
Metastasis (at the time of diagnosis)	3	21.4
Periton	2	14.3
Liver	1	7.1
Localization of the primary tumor		
Abdominal cavity	7	50.0
Retroperitoneum	2	14.3
Ampulla vater	1	7.1
Jejunum mesentery	1	7.1
Omentum	1	7.1
Pancreatic head	1	7.1
Sigmoid colon mesentery	1	7.1

*: Some of the total percentages may exceed (or may not reach) 100% due to the low number of the patients and rounding; CT: Computed tomography; MRI: Magnetic resonance imaging; HT: Hypertension; DM: Diabetes mellitus.

dium risk group. In immunohistochemistry, DOG1 and CD117 were positive in all patients, and CD34 was positive in 11 (78.6%) cases. Desmin and S100 were negative in all patients (Table 2).

Necrosis was detected in eight (57.1%) patients, and rupture in one (7.1%) case. Complete resection was achieved in nine (64.3%) patients. Synchronous GIST was not detected in any patients. One (7.1%) patient died within the post-operative 30 days. During the follow-up, three patients died due to non-EGIST related causes. The overall survival (OS) rate was 80%, and the 5-year survival rate was 88.9%. Imatinib treatment was given to 11 (78.6%) patients (Table 2).

The median age at diagnosis was 47.5 (inter-quartile range: 23; min-max: 34–87) years. The mean hospital stay was 10.6±6.4 days. The mean tumor diameter was 16.8±10.5 cm. The mean mitotic index was 16.8±22.0/50 HPF. The mean follow-up time was 72.2±48.0 months. The mean OS was 78.5±50.7 months, the 5-year OS and DFS were both 53.3±20.0 months, and the mean overall DFS was 58.0±59.8 months (Table 3).

The mean tumor diameter, mitotic index, duration of stay, 5-year OS, and DFS were all similar between genders, while mean OS and DFS durations were found to be significantly lower in women (p=0.006 for both comparisons). The mean OS was significantly lower in patients over 60 years of age compared to patients under 60 years (p=0.013) (Table 3).

The mean duration of hospital stay was significantly lower in patients with abdominal cavity compared to the tumors in other locations (p=0.026). The mean tumor diameter was also significantly higher in those with high mitotic index (p=0.021). The mean age at diagnosis, length of stay, OS, 5-year OS and DFS were all found to be similar in patients with complete resection and partial resection, in patients with or without necrosis, in patients with a tumor diameter greater than or smaller than 10 cm, in patients with a high or low mitotic index, in patients with lesions in the abdominal cavity and other localizations. Similarly, the mean age at diagnosis, tumor diameter, mitotic index, length of stay, and DFS were found to be similar in patients with mixed-type tumors and other histopathological types, and in the medium risk group and high risk group. It was found that mean OS was significantly lower in patients who received imatinib than those who did not receive treatment (p=0.013) (Table 4). The characteristics of the patients are summarized in Table 5. The median age of the patients whom were given imatinib was 43, and of those whom were not given was 68, three (21.4%) of the imatinib-given patients died within the follow-up period, three (21.4%) had metastasis at the time of diagnosis or within the follow-up period.

TABLE 2. Distribution and ratios of some variables

	n	%*		n	%*
Tumor diameter (cm)			Operation type		
2.01–5	1	7.1	Mass excision	4	28.6
5.01–9.99	3	21.4	Whipple + right colon resection	2	14.3
≥10	10	71.4	Anterior resection	1	7.1
≤5	1	7.1	Mass excision + right hemicolectomy	1	7.1
>5	13	92.9	Mass excision + segmental small bowel resection	1	7.1
<10	4	28.6	Mass excision + distal pancreatectomy + splenectomy	1	7.1
>10	10	71.4	Mass excision + segmental colon resection	1	7.1
Mitotic index (/50 HPF)			Mass excision + TAH BSO	1	7.1
≤5	7	50.0	Omentectomy	1	7.1
5.01–9.99	3	21.4	Segmental small intestine resection + partial bladder resection	1	7.1
≥10	4	28.6	Mortality (within the post-op 30 days)	1	7.1
≤5	7	50.0	Morbidity	4	28.6
>5	7	50.0	Mortality (during follow-up)	4	28.6
<10	10	71.4	5-year overall survival	8	80.0
>10	4	28.6	5-year disease-free survival	8	88.9
Markers			Adjuvant treatment (Imatinib)	11	78.6
CD34	11	78.6	Recurrence-Metastasis (during follow-up)	3	21.4
CD117	14	100	Liver	2	14.3
Desmin	0	0	Periton	1	7.1
DOG1	5	100**	Synchronous GIST	0	0
S100	0	0	Complete resection (R0)	9	64.3
Necrosis	8	57.1			
Rupture	1	7.1			
NIH risk category					
Intermediate	2	14.3			
High	12	85.7			

*: Some of the total percentages may exceed (or may not reach) 100% due to the low number of the patients and rounding; **: Among the tested patients; CT: Computed tomography; MRI: Magnetic resonance imaging; TAH BSO: Total abdominal hysterectomy + bilateral salpingooferectomy; GIST: Gastrointestinal stromal tumor.

DISCUSSION

EGISTs constitute about 5–7% of the GIST cases [8, 9]. Iqbal et al [9]. reported the rate of EGIST as 12%, and similar to this rate in the presented study, 10.4% of the GISTs were identified as EGIST (14/135).

It has been reported that EGISTs are partially more common (60–77.8%) in women [9–14]. In the present study, it was found that most of the EGIST cases were male (64.3%). The lack of clear data on gender predominance in EGIST cases seems to be associated with the low number of cases.

The localizations where EGISTs are seen most frequently are presented differently in the reports. The most common localization was reported to be the omentum [15], mesentery [9], or the abdominal cavity [10, 11, 14]. In the present study, 50% of the primary tumors were found in the abdominal cavity and 14.3% in the retroperitoneum.

It has been reported that EGIST cases mostly involve elderly patients [9]. Studies reported the mean age between 45.8 and 59 years [9–13]. In the present study, the mean age at diagnosis was found to be 51.9±14.8 years (Min.–max.: 34–87 years), only three of the patients

Table 3. The mean values of some variables

	Mean	SD	Min.	Max.
Age at diagnosis (years)	51.9	14.8	34	87
Follow-up duration (months)	72.2	48.0	0	156
Duration of hospital stay (days)	10.6	6.4	1	22
Tumor diameter (cm)	16.8	10.5	2.8	40.0
Mitotic index (/50 HPF)	16.8	22.0	1	71
Overall survival (months)	78.5	50.7	0	156
5-year overall survival (months)	53.3	20.0	0	60
Disease-free survival (months)	58.0	59.8	0	156
5-year disease-free survival (months)	61.7	58.3	0	156

Min: Minimum; Max: Maximum; HPF: High power field; SD: Standard deviation.

were in the range of 60–70 years, and one was 87 years old. These findings show that EGISTs are not seen in elderly patients as expected. Accordingly, EGIST can occur in any time after the age of 30. Reith et al. [10] found no difference in terms of prognosis between the patients over 60 years old or younger. However, in the present study, the mean OS was found to be significantly lower in patients over 60 years old. These findings suggest that the prognosis may be worse in elderly patients.

Symptoms in GISTs vary according to tumor location and size. While GISTs are often diagnosed with bleeding and obstruction, EGISTs often present with pain and palpable mass [2–4]. Reith et al. [10] reported that the most common symptom for EGISTs is abdominal pain. In the present study, 57% of the patients were diagnosed due to a palpable mass, while the rate of patients diagnosed due to abdominal pain was found to be 35.7%. Since the lesions are not related to the gastrointestinal tract, no bleeding complaints were observed, although the size of the lesion was large, obstructive symptoms were not observed. In general, EGISTs are larger than GISTs at the time of diagnosis [2–4]. In the studies, the mean tumor diameter has been reported between 11 and 18 cm, and the tumor size can reach up to 40 cm [8–14]. In the present study, the mean tumor diameter is 16.8 cm, similar to the literature data, and 71.4% of the tumors are larger than 10 cm. The reason why EGISTs are diagnosed at a larger scale than GISTs is related to the tumor localization and the later recognition of the symptoms. It was observed in the present study that the diameter of the tumor also had no significant effect on prognosis and this was similar to the literature [10].

The majority of EGISTs have been reported histopathologically to be epithelioid type [10] or spindle type [9]. In the present study, the least observed type was epithelioid type, and most of the cases had mixed histopathology. While CD117, one of the immunohistochemical markers used in the identification of EGISTs, was found to be positive in almost all patients, CD34 was reported to be positive in more than half of the patients [9–15]. Among the other markers, desmin and S100 were reported to be low [11, 14], and DOG1 was reported to be highly positive [15]. Similar to the literature, CD117 and DOG1 were found to be positive in all patients, while CD34 was found to be highly positive (78.6%), and desmin and S100 were negative in all patients. Although lymph node metastasis can be observed, it has generally been reported with a low rate [9]. In the presented study, three patients had lymph node enlargement, while no metastasis was detected in their histopathological examination.

It has been reported that EGISTs can be more aggressive than GISTs [9], and the rate of metastasis development is observed between 6% and 31% [9–11, 14–17]. Recurrence rate during follow-up has been reported to be between 2% and 23% [9, 10, 14]. In the present study, the metastasis rate at the time of diagnosis was 21.3%, and the metastatic status continued in the follow-up. No recurrence or metastasis was observed in the 72-month follow-up of the other patients. Our result suggests that EGISTs are not as aggressive as expected.

Mitotic index in EGISTs was found to be 5/50 HPF and below in a significant number of patients [10, 14]. Similar to the literature, in the present study, 50% of the patients had a mitotic index of 5/50 HPF and below. It has been reported that mitotic index is important in terms of mortality, recurrence and metastasis as well as lesion size [10]; however, in the presented study, no association was found with mitotic index in terms of prognosis. Similarly, the presence of necrosis (18.8%) was reported in studies to negatively affect the prognosis [10], although more frequent necrosis was observed in the present study (57.1%), treatment and follow-up results were found to be similar in patients with and without necrosis. In the present study, the rate of patients with EGIST with high risk was determined at a high rate (85.7%), similar to the literature [9,14], and it was thought that the fact that the factors that could affect the prognosis could not be determined clearly could be due to the high rate and the absence of low risk patients.

Table 4. Comparisons between some variables in terms of the mean values

		Tumor diameter (cm)	Mitotic index (/50 HPF)	Duration of hospital stay (days)	OS (months)	5-year-OS (months)	DFS (months)
Gender	p	0.052	0.441	0.986	**0.006**	0.052	**0.006**
Men		12.9±6.2	20.3±26.2	10.7±6	106.3±35.3	60±0	93.7±53.8
Women		24±13.5	10.4±11.1	10.6±7.9	29.8±33.1	30±42.4	8±11.3
Age at diagnosis (years)	p	0.982	0.596	0.085	**0.013**	0.052	0.241
≤60		16.8±8.6	14.7±22.0	12.5±6.5	99.8±40.6	60±0	70.1±64.4
>60		17.0±16.0	22.0±24.4	6.0±3.4	21.7±20.6	30.0±42.4	21.7±20.6
Complete resection	p	0.587	0.773	0.729	0.794	0.516	0.219
Done		15.6±10.6	15.4±18.9	11.1±7.7	81.7±59.2	50±24.5	73.5±59.5
Undone		19±11.1	19.2±29.1	9.8±3.5	72.8±38.7	60±0	27±54
Necrosis	p	0.617	0.19	0.223	0.448	0.292	0.619
Present		18.1±9	23.6±26.2	12.5±6.2	92±51.6	60±0	48.8±70.6
Absent		15.1±12.9	7.7±11	8.2±6.3	67.2±51.7	45±30	67.2±51.7
Tumor diameter	p	–	0.179	0.635	0.91	0.749	0.926
<10 cm		–	4±3.6	12±7.3	74.5±47.4	60±	55±47.6
≥10 cm		–	21.9±24.4	10.1±6.3	79.3±54.1	52.5±21.2	59±65.9
Mitotic index		0.123	–	0.067	0.915	0.292	0.852
<10/50 HPF		14.1±9.1	–	12.6±6.4	77.4±43.8	60±0	60±53.5
≥10/50 HPF		23.8±11.8	–	5.8±3.3	81.3±78.2	45±30	52±90.1
Mitotic index		**0.021**	–	0.177	0.553	0.407	0.203
≤5/50 HPF		10.7±4.3	–	13±7.5	87.3±43.1	60±0	77.1±47.7
>5/50 HPF		23±11.5	–	8.3±4.3	67.8±61.9	48±26.8	31.2±69.8
Localization		0.33	0.424	**0.026**	0.791	0.407	0.694
Abdominal cavity		19.7±13.7	11.9±14.3	7±3.8	75.1±56.4	48±26.8	64.1±63.1
Others		14±5.7	21.7±28.1	14.3±6.6	84.3±46.2	60±0	49.4±60.8
Histopathological type		0.219	0.944	0.496	0.162	0.052	0.382
Epiteloid or spindle		21.6±12	16.2±15.1	9±7.8	49.5±71.7	30±42.4	39.2±66.1
Mixed		14.2±9.2	17.1±25.9	11.6±5.8	95±28.8	60±0	71.4±55.9
Risk category		0.113	0.341	0.378	–	–	0.471
Intermediate		5.9±4.4	2.5±2.1	14.5±10.6	–	–	28.5±17.7
High		18.7±10.1	19.2±23	10±5.9	–	–	63.9±64
Imatinib		0.757	0.668	0.106	**0.013**	–	0.241
Given		18.6±19.2	11.7±15.9	5.3±3.8	21.7±20.6	–	21.7±20.6
Not given		16.4±8.2	18.2±23.9	12.1±6.3	99.8±40.6	–	70.1±64.4

Independent Samples' t test was used. OS: Overall survival; DFS: Disease-free survival; HPF: High power field.

The mean 5-year OS rate in EGIST patients has been reported to be between 38% and 79.1% [10, 11, 14, 16, 17]. The 5-year DFS rate was reported as 42.4% [11]. In addition, the 5-year OS rate was reported as 53.9% [9]. Yamamoto et al.[11] reported the 5-year DFS rate as 42.4%. Iqbal et al. [9] found the OS rate to be 53.9%. In the present study, the 5-year OS rate was 80%, and the 5-year DFS was 88.9%. One (7.1%) patient died within the post-operative 30 days. The cause of death of three patients lost during the follow-up was other than EGIST. In addition, the mean OS was 78.5 months, 5-year OS and DFS 53.3 months, overall DFS 58.0 months. In addition, in the present study, the rate of complete resection was higher than the study of Iqbal et al. [9] (23.1% vs. 64.3%). All these findings show that both OS rate and OS duration can be higher in EGIST cases in compar-

Table 5. Features of the cases

Age/ gender	Site	size (mm)	Morphology	Further findings	Risk	Operation
41/M	Abdominal cavity	15.0	Mixed	None	H	Mass excision
45/M	Retroperitoneal	11.0	Mixed	None	H	Mass excision + segmental small bowel resection
50/F	Ampulla vateri	9.0	Spindle	Necrosis	I	Whipple + right colon resection
55/M	Abdominal cavity	9.0	Mixed	None	H	Mass excision
50/M	Sigmoid colon mesentery	20.0	Mixed	Liver met. + necrosis	H	Anterior resection
41/F	Abdominal cavity	35.0	Mixed	Periton met. + necrosis	H	Mass excision + TAH BSO
68/F	Abdominal cavity	40.0	Spindle	None	H	Mass excision + distal pancreatectomy + splenectomy
87/M	Abdominal cavity	2.8	Mixed	None	I	Mass excision + right hemicolectomy
62/F	Abdominal cavity	13.0	Epithelioid	None	H	Mass excision
42/F	Retroperitoneal	23.0	Spindle	Periton met. + necrosis	H	Mass excision
43/M	Abdominal cavity	23.0	Spindle	Necrosis	H	Mass excision + segmental colon resection Omentectomy
69/M	Omentum	12.0	Mixed	Necrosis + rupture	H	Segmental small intestine resection +
34/M	Jejunum mesentery	15.0	Mixed	Necrosis	H	partial bladder resection
39/M	Pancreas head	8.0	Mixed	Necrosis	H	Whipple + right colon resection

M: Male, F: Female.

ison to the other GISTs, and this may be related to the application of aggressive surgical methods.

Imatinib was given to 11 (78.6%) of the patients. OS duration was lower in patients receiving imatinib treatment than others. In addition, the median age of the patients whom were given imatinib was 43, and of those whom were not given was 68, three (21.4%) of the imatinib-given patients died within the follow-up period, three (21.4%) had metastasis at the time of diagnosis or within the follow-up period. All these findings consider that the worse prognosis of the patients received imatinib was due to the fact that they had a more aggressive course.

The limitations of the study are that the diagnosis was not confirmed by PDGRF-alpha and C-KIT mutation analysis, and the study was retrospective. Although the number of patients for EGIST is high, it is not enough to allow an accurate evaluation of the prognostic factors.

Conclusion

Tumor size is mostly large in EGISTs. Although it is reported to be more common in women, in our study it is found to be frequent in men also. The prognosis may be worse in women and in elderly patients. Especially in patients with lymph node enlargement, lymph node metastasis should be confirmed.

Ethics Committee Approval: The Kartal Dr. Lutfi Kirdar City Hospital Clinical Resources Ethics Committee granted approval for this study (date: 11.11.2020, number: 514/189/5).

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