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Rare Gastrointestinal Tumors: How Often do we Operate on Such Tumors? A Single-Center Study from a Regional Hospital

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Abstract

Background: Several types of rare tumors, benign and malignant, have infrequently been reported and dealing with them remains a challenge. Rare tumors can occur in every organ and in every age.

Methods: Forty-one rare tumors of the gastrointestinal tract were identified in forty patients, operated in a secondary, regional center, over a 15-year period. According to their biological behavior they were divided into three subgroups; benign, with uncertain behavior or low malignancy and malignant. The diagnosis was set by the pathology report.

Results: In most cases (61%) the tumor was the cause of surgery. Patient age ranged from 15 to 90 years old (median 61y; mean 58y), and male to female ratio was 1.22:1. Four tumors were benign (9.7%), eighteen were of uncertain behavior or low malignancy (43.9%) and nineteen were malignant (46.4%). Seventeen cases (41.4%) were located in the appendix, nine (22%) in colon, eight (19.5%) in small bowel, three in gallbladder (7.3%) and the rest occurred in stomach, omentum, Vater's ampulla and mesentery (1 case in each). Serious, life-threatening complications occurred in two patients, both with malignant tumors, but middle-aged (45 and 57 years old).

Conclusion: Even non-referral centers should reveal their rare cases, so that evidence-based medicine can be achieved in every patient. In most occasions, no guidelines were available and the decision was based on surgeon's knowledge and experience. Surgeons, even in non-referral centers, must be ready to deal with every kind of neoplasm, independent of its frequency and its site.

Keywords: Gastrointestinal tumors; Rare tumors; Gastric tumors; Small bowel tumors; Appendiceal tumors; Colorectal tumors

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Introduction

The majority of the tumors of the Gastrointestinal (GI) tract is adenocarcinomas. Other histotypes, either benign or malignant, might also occur, though much less frequently. Moreover, an infrequent location of a tumor makes it a rarity, even for an adenocarcinoma. Because of the rarity of these tumors, they present a diagnostic and therapeutic dilemma for the clinician when encountered in clinical practice. These histotypes include, but are not limited to, endocrine carcinoma of the small bowel, adenocarcinoid of the appendix, gastrointestinal stroma tumors, and gastrointestinal lymphomas. Current literature is limited to case reports or case series of such rare tumors. The purpose of our study was to estimate the prevalence in the workload of a General Surgery Department and evaluate the clinical management of rare gastrointestinal tumors at our institution.

Materials and Methods

We performed a single-center, observational, retrospective study of all gastrointestinal operations which were performed from January 2003 to December 2017 at the Department of General Surgery of General Hospital of Rethymno, Crete, Greece. Our institution is a secondary, regional center serving a prefecture with a population of more than 85,000 residents, which increases to over 140,000 during the summer months.

Inclusion criteria were:

(1) Resection of a part of the gastrointestinal tract (gastrectomy, enterectomy, colectomy, appendectomy, cholecystectomy, pancreatoduodenectomy or any combination of them in the context of a complex operation) (2) Elective or emergent/urgent operation (3) Tumor, either benign or malignant, in the resected segment of the gastrointestinal tract or omentum (4) Rarity of the tumor, incidence of tumor histotype <3% as referred to literature (5) Patient age greater than 15 years old. Operations, where only a palliative bypass or an ostomy was performed, were excluded from our analysis. To the best of our knowledge, there have never been reported so far, an analysis of the volume of rare gastrointestinal tumors, which have been managed in a regional hospital.

The study was organized according to ethical considerations, as described in the Declaration of Helsinki for human medical studies. Upon the approval of the study's protocol from the local Institutional Review Board and the Scientific and Ethics Committee, the patients were identified, and their charts were reviewed for data regarding patient's demographics, clinical presentation, indication for operation, operation performed, operative reports and histopathology report.

Tumors were classified according to the 8th edition of the TNM classification system of the American Joint Committee on Cancer, where applicable. Postoperative complications were classified according to the Clavien-Dindo classification system.

Results

During the studied period, 2806 resections in the gastrointestinal tract were performed (Table 1). The majority of these were cholecystectomies (45.74%) and appendectomies (35.02%), followed by colectomies (13.73%), small bowel resections (3.76%), gastric resections (1.52%) and pancreatic resections (0.21%). Of the operations performed, 42.37% were elective and the rest of them were emergent or urgent.

Pathological examination revealed 40 patients with 41 rare tumors, 4 of which were benign, 19 were malignant and 18 were of uncertain behavior or with low malignant potential (Tables 2-4). 1.42% of the resected specimens harbored a rare tumor of the gastrointestinal tract. Median age of patients with benign rare tumors was 73 years, while male/female and elective/emergent or urgent ratio was 1:1 for both. Regarding rare tumors with uncertain behavior or low malignancy potential, median age was 40 years, male/female and elective/emergent or urgent ratio was 1.125 and 0.214 respectively. Patients with rare malignant tumors had a median age of 71 years, male/female and elective/emergent or urgent ratio 1.375 and 1.11 respectively. Most patients had an uneventful postoperative period. Only one had a grade III complication after a total gastrectomy and another patient had a grade IV complication after he had been operated for an intraabdominal catastrophe. Both patients were middle aged (45 and 57 years old). No in-hospital death occurred. There are no data regarding follow up of the patients. Three of the patients were not from Hellenic origin, they were either north Europeans or immigrants.

From the oncological point of view, no patient received neo-adjuvant therapy. Adjuvant therapy was administered wherever indicated and according to the patient's will. All benign rare tumors were resected completely. All but one of the operations performed were R0 (patient 34 had a R1 operation). Moreover, in patient 8 the appendix was perforated. Right hemicolectomy was advised and performed in patients 35 and 36 after the pathology report of the initial specimen and the staging procedure. Patient 34 returned 2 months after the first operation and he underwent laparotomy with curative intent (right hemicolectomy) but peritoneal carcinomatosis was discovered. Patient 8 was referred to a tertiary center for possible HIPEC. Patient 13 with appendiceal NET was advised for a right hemicolectomy after the staging procedure.

Table 1: Type and number of gastrointestinal operations performed at our institution.

Operation	Number	Elective	Emergent/ Urgent
Gastric Resections	43	36	7
Total	23	22	1
Distal/Subtotal	20	14	6
Pancreatic Resections	6	5	1
Pancreaticoduodenectomies	5	5	-
Central Pancreatectomies	1	-	1
Cholecystectomies	1289	867	422
Small Bowel Resections	106	27	79
Appendectomies	987	-	987
Colectomies	387	259	128
Right Hemicolectomies	118	75	43
Extended Right Colectomies	23	15	8
Left Hemicolectomies	43	34	9
Sigmoidectomies	49	19	30
Anterior Resections	35	22	13
Low Anterior Resections	73	66	7
Abdominoperineal Resections	18	18	-
Subtotal Resections	28	10	18
Other abdominal operations (hernias, perforated peptic ulcers, splenectomies, hysterectomies, nephrectomies, adhesiolysis etc.)	2655	1491	1164
	5473	2685	2788

Discussion

All tumors mentioned above are rare, only few cases are reported in the literature and thus their treatment still remains a challenge. To the best of our knowledge, so far there has never been reported an analysis of the volume and type of rare gastrointestinal tumors managed in a single regional hospital, either referral or non-referral center. The majority were urgent/emergent cases (25/40), which highlights the need of awareness, knowledge and skills to cope with such findings and diseases intra- and post-operatively, even in a regional non-referral hospital. In 25 patients (62.5%) the tumor was itself the cause of surgery, whereas in 15 patients it was an incidental finding. Although only 50% of the benign tumors and 47% of the tumors with uncertain behavior/low malignancy potential were the cause of the operation, almost 79% of the malignant tumors were the cause of the operation. Most younger patients suffered from appendiceal tumors with uncertain behavior/low malignancy potential.

The diagnosis was set from the pathological examination of the surgical specimen and only in a few cases there was a suspicion of such tumor from the preoperative work up. Most rare tumors were

Table 2: Rare benign gastrointestinal tumors.

Patient	Histological Type	Location	Sex	Age	Cause of laparotomy/ Incidental finding	Indication of Operation	Elective/ Emergent or Urgent operation	Operation	Postoperative complications (Clavien-Dindo)
1	**Adenomyoma** d=0.8 cm	Vater	M	61	Cause	Obstructive jaundice, chronic pancreatitis, tumor of Vater	Elective	Whipple's procedure	II
2	**Leiomyomata**	Sigmoid colon	F	90	Incidental	Diverticulitis Hinchey IV	Emergent	Hartmann's procedure	II
3	**Lipoma** d=2.5 cm	Ascending colon	M	85	Incidental	Large bowel obstruction due to splenic flexure carcinoma, cecal perforation	Emergent	Extended right colectomy	-
4	**Lipoma** d=7 cm	Sigmoid colon	F	52	Cause	Large bowel partial obstruction	Elective	Sigmoidectomy	-

M: Male; F: Female

Table 3: Rare tumors with uncertain behavior/low malignancy potential.

Pt	Histological Type	Location	Sex	Age	Cause of laparotomy/ Incidental finding	Indication of Operation	Elective/ Emergent or Urgent operation	Operation	Postoperative complications (Clavien-Dindo)
5	**Hemangiopericytoma** d=1.6 cm, Ki67<1%, few mitoses	Small Bowel	M	77	Incidental	Small bowel ischemia due to obstruction (adhesions)	Emergent	Small bowel resection	-
6	**GIST** d=2 cm, T1N0, Ki67 5%, few mitoses	Small Bowel (terminal ileum)	F	88	Incidental	Cecal adenocarcinoma	Elective	Right hemicolectomy	I
7	**GIST** T1N0, d=2 cm, G1, Ki67 3%, mitoses <5/10HPF	Sigmoid Colon	M	84	Incidental	Adenocarcinoma of sigmoid colon	Elective	Low anterior resection	-
8*	**LAMN** d=6X3X2 cm at the base of the appendix	Appendix	M	72	Cause	Perforated appendicitis (fecal peritonitis)- appendiceal tumor	Emergent	Right hemicolectomy	-
9	**LAMN** d=2.4 cm	Appendix	M	38	Cause	Acute appendicitis	Urgent	Appendectomy	-
10	**LAMN** d=4 cm	Appendix	M	40	Cause	Acute appendicitis	Urgent	Appendectomy	II
11	**LAMN** d=0.9 cm	Appendix	F	22	Incidental	Acute appendicitis	Urgent	Appendectomy	-
12	**LAMN** d=2.2 cm **+** **NET** d=1.8 cm T1b	Appendix	M	31	Cause	Acute appendicitis	Urgent	Appendectomy	-
13*	**NET** T2, G1, well differentiated	Appendix	F	26	Cause	Acute appendicitis	Urgent	Appendectomy	-
14	**NET** d=0.4 cm-T1a, G1, Ki67<1%, well differentiated	Appendix	F	20	Cause	Acute appendicitis	Urgent	Appendectomy	-
15	**NET** d=1.4 cm T1, G1 well differentiated	Appendix	F	17	Incidental	Acute appendicitis	Urgent	Appendectomy	-
16	**NET** d=3 mm T1a, G1, well differentiated	Appendix	M	28	Incidental	Acute appendicitis	Urgent	Appendectomy	-
17	**NET** d=0.4 cm T1a, G1, well differentiated	Appendix	F	15	Incidental	Acute appendicitis	Urgent	Appendectomy	-
18	**Mesothelial inclusion cyst** d=2.3 cm	Appendix	M	40	Cause	Acute appendicitis	Urgent	Appendectomy	-
19	**Mesothelial inclusion cyst** d=0.5 cm	Appendix	F	48	Cause	Acute appendicitis	Urgent	Appendectomy	I
20	**GIST** d=2 cm, T1N0, Ki67 5%, few mitoses	Small Bowel (terminal ileum)	F	88	Incidental	Cecal adenocarcinoma	Elective	Right hemicolectomy	I
21	**Well-differentiated papillary mesothelioma** d=1.6 cm	Omentum	M	59	Incidental	Gastric ulcer perforation	Emergent	Suture of the perforation	I

M: Male; F: Female; LAMN: Low-grade Appendiceal Mucinous Neoplasm (mucinous cystadenoma); GIST: Gastrointestinal Stromal Tumor
*not from Hellenic origin

located in the appendix (17/41) (41.4%), which is expected since 35% of our surgeries were appendectomies. In nine cases (22%) the tumor was located in the large intestine and eight cases in the small intestine (19.5%). Small intestine is a rare tumor site by its nature and therefore such tumors are infrequently referred in the literature. One rare malignant tumor (2.4%) was located in stomach. Only 3 rare tumors derived from the gallbladder (7.3%), although 45% of our surgeries were cholecystectomies.

Table 4: Rare malignant gastrointestinal tumors.

Pt	Histological Type	Location	Sex	Age	Cause of laparotomy/ Incidental finding	Indication of Operation	Elective/ Emergent or Urgent operation	Operation	Postoperative complications (Clavien-Dindo)
22	**Non-Hodgkin Lymphoma (large B cells)** d=10.6 × 9 cm stage IIE MUSSHOF	Stomach	M	57	Cause	Gastric tumor-Hemorrhage	Elective	Total Gastrectomy D2	III
23	**Non-Hodgkin Lymphoma (diffuse large B cells)** d=4 × 3 × 2 cm, stage IIE1 MUSSHOFF	Terminal ileum	F	71	Cause	Small bowel obstruction	Urgent	Right hemicolectomy	-
24*	**Non-Hodgkin Lymphoma (diffuse large B cells)** d=11 cm	Ascending colon	M	79	Cause	Tumor perforation-Fecal peritonitis	Emergent	Right hemicolectomy	-
25	**Neuroendocrine Carcinoma** d=2 × 1.3 × 1.8 cm, G3, T3N1, poorly differentiated G3, infiltration of lymphatics and veins	Sigmoid	M	81	Cause	Sigmoid colon tumor	Elective	Low anterior resection	II
26	**Adenosquamous carcinoma** d=5.8 × 4 cm, T4N1b	Cecum	M	75	Cause	Cecal tumor	Elective	Right hemicolectomy	-
27	**Leiomyosarcoma** d=4.5 × 4 × 2 cm, no lymph node metastasis	Cecum	M	63	Cause	Cecal tumor	Elective	Right hemicolectomy	-
28	**Medullary carcinoma** T3N0	Ascending colon	M	57	Cause	Ascending colon cancer	Elective	Right hemicolectomy	I
29	**Adenocarcinoma** d=5.6 × 4.5 × 1.9 cm, T2N1	Small Bowel	F	61	Cause	Small bowel obstruction	Urgent	Small bowel resection	-
30	**Neuroendocrine Carcinoma** d=1.5εκ, T2N1 infiltration of lymphatics and veins	Small Bowel (ileum)	F	35	Cause	Small bowel tumor-GI hemorrhage	Elective	Small bowel resection	I
31	**GIST** d=5.2 cm, T3N0, mitoses 8/10HPF	Small Bowel (jejunum)	F	82	Incidental	Strangulated umbilical hernia	Urgent	Small bowel resection	–
32	**GIST** d=8.5 cm, T3N0, mitoses <5/50HPF	Small Bowel (terminal ileum)	F	69	Cause	Small bowel tumor (hemorrhage)	Elective	Right colectomy	-
33	**GCC** T2N0	Appendix	M	41	Cause	Acute appendicitis	Urgent	1.Appendectomy	-
34	**Adenocarcinoma** d=1.5 cm, T3 moderate differentiation	Appendix	M	79	Cause	Ruptured appendicitis-retroperitoneal abscess	Urgent	1.Appendectomy	I
35	**Adenocarcinoma (mucinous)** T3	Appendix	F	71	Cause	Acute appendicitis	Urgent	1.Appendectomy	–
36	**Adenocarcinoma (mucinous)** T4a, on the ground of a LAMN	Appendix	M	39	Cause	Acute appendicitis	Urgent	1.Appendectomy	–
37	**Adenocarcinoma** T3	Gallbladder	F	81	Incidental	Symptomatic cholelithiasis	Elective	Laparoscopic Cholecystectomy	–
38	**Adenocarcinoma** T3N0	Gallbladder	M	74	Incidental	Chronic Cholecystitis	Elective	Radical cholecystectomy	-
39	**Adenocarcinoma** T2, well differentiated	Gallbladder	F	81	Incidental	Symptomatic cholelithiasis	Elective	Cholecystectomy	-
40	**Desmoid tumor** d=10.5 × 9.5 × 7 cm	Mesentery	M	45	Cause	Small Bowel Obstruction-Strangulated incisional hernia. Necrotizing infection of the abdominal wall	Urgent	Small bowel resection. Debridement of the abdominal wall	IV

M: Male; F: Female; LAMN: Low-grade Appendiceal Mucinous Neoplasm (mucinous cystadenoma); GCC: Goblet Cell Carcinoid; GIST: Gastrointestinal Stromal Tumor; GI: Gastrointestinal

*not from Hellenic origin

GISTs are found throughout the gastrointestinal tract, most commonly in stomach (30-45%) and small bowel (20-30%) and the rest in esophagus and colon [1,2]. In our hospital four GISTs were found in small bowel, three of which in terminal ileum, treated with right hemicolectomy, one in jejunum treated with small bowel resection and one in sigmoid colon, treated with low anterior resection. Interestingly, in only one case GIST was the cause of a laparotomy, whereas in three other cases a synchronous tumor was the cause and all patients were above 80 years old. Adjuvant imatinib for at least 3 years should be administered in high malignancy potential GISTs [1]. GISTs can cause bleeding, as was the indication of surgery in one of our cases [3,4]. Furthermore, 20% of GISTs coexist with other tumors but only 4.3% of them can be diagnosed preoperatively [5]. It still remains uncertain whether this is a coincidence or not [6,7].

Three patients were diagnosed with Non-Hodgkin Lymphoma (NHL), all DLBC type. In all of our patients the tumors presented with complications. One was located in stomach, one in terminal ileum and one in right colon. Only 4% to 20% of NHLs are located in the GI tract. Stomach is the most common site (60-75%), but NHL accounts for 3% of all gastric, 2% of small and 0.2% of large intestine neoplasms [8]. In intestinal lymphomas surgery followed by chemotherapy (R-CHOP) is usually performed [9,10]. In contrary, recent studies suggest primary treatment with R-CHOP for gastric NHLs, while surgery should be preserved for elective cases, usually with complications [8-11].

Gallbladder adenocarcinomas represent 76% to 95% of all gallbladder malignancies. Median age is 70 years old and female to male ratio is 4:1 [12-14]. In our study, three cases were documented, aged between 74 to 81 years old, two of which were females. All three cancers were incidental findings.Two of them were treated with cholecystectomy alone due to advanced age and one with radical surgery, with the decision been made intraoperatively. Adjuvant chemotherapy still remains a question waiting clinical trials to be answered.

Adenomyoma in the ampulla of Vater is a rare benign situation with only 58 cases in English literature until 2018 [15]. We treated our patient with Whipple's procedure due to symptoms and importantly due to suspected malignancy. Most cases are identified postoperatively. Biopsies *via*EUS and ERCP should be a first step approach, however false negative rates range from 16% to 60% [15]. Since Ki-67 tends to be <1%, a more conservative strategy with endoscopic papillectomy should be first-line approach, if symptoms appear and preoperative diagnosis is made [16].

Two colon lipomas were found, one in sigmoid colon (7cm) which led to colonic partial obstruction and one asymptomatic in ascending colon (2.5cm), which is the most common site (90%), and were treated with colectomy [17,18]. In general, colon lipomas bigger than 2 cm tend to be symptomatic and are unsafe to be excised endoscopically. They are extremely rare, consisting 0.3% of all colorectal tumors, show no sex predominance and have almost 100% 10-year survival [17,19].

Leiomyomas are rarely found in the colon (3%) [20]. They can be removed entirely endoscopically. In our case it was an incidental finding in a Hinchey IV diverticulitis and was resected with the specimen of a Hartmann's procedure. Colonic leiomyosarcomas are aggressive with high mitotic index. Aggarwal et al. described 11 cases, where nine patients died of this tumor within an average of 20 months [20,21].

Other rare colonic tumors included an Adenosquamous Carcinoma (ASC) in cecum (0.06% of all colon carcinomas [22]), a NET in sigmoid colon and a medullary carcinoma in right colon. Two reviews concluded different parts of colon as ASC's primary site [23,24]. Masoomi et al. reported that ASCs had worse overall survival, higher rate of poor differentiation and distant metastasis than colon adenocarcinomas [25]. Colonic NETs account for 7.8% of GI's NETs, 13% of whom are located in sigmoid colon and are usually larger than 2 cm [26]. Colonic medullary carcinoma is a very rare, poorly differentiated adenocarcinoma, discovered the last two decades, consisting 0.05% to 0.08% of all colon carcinomas [27].

In our study, five patients suffered from Low-grade Appendiceal Mucinous Neoplasm (LAMN), one of whom interestingly had a synchronous Appendiceal Neuroendocrine Neoplasm (ANEN). Four were treated with appendectomy and one with right hemicolectomy due to the size and site of the lesion and suspected malignancy.Smeenk et al. reported that 20% of such patients developed pseudomyxoma peritonei [28], while 17% of them had a synchronous tumor in colon [29]. Lesions larger than 6 cm had increased chance for LAMN and perforation [30]. In six of our appendectomies (0.6%) ANEN was the cause, correlating with the global average of 0.16% to 0.52%. They appear mostly during the second and third decade of life as did in our patients (age 15-28 years old) [31,32]. ANENs in our study ranged from 0.3 cm to 1.8 cm. If the lesion is less than 1 cm, appendectomy alone is safe, whereas for ANENs 1 cm to 2 cm, right colectomy should be discussed, as it was advised to patient 12 and 13 in our series [31-33].

In three appendiceal specimens (0.3%), adenocarcinomas were found, two of which were mucinous. Adenocarcinoma of the appendix, as a primary tumor, is a rare malignancy that constitutes less than 0.5% of all gastrointestinal neoplasms. One patient was 39 years old, much younger than the median age of 59 years old of tumor occurrence [34]. Two patients were treated with right hemicolectomy, after the diagnosis was set initially with appendectomy, which is the mainstay of management. One patient had peritoneal carcinomatosis by the time he decided to underwent curative right hemicolectomy. However right hemicolectomy is challenged lately, especially in the presence of peritoneal or nodal metastasis [35]. Survival between mucinous and non-mucinous adenocarcinomas does not differ [36]. One case of a young patient with appendiceal goblet cell carcinoma was presented. He was treated with right colectomy, as advised due to the aggressiveness of such tumors [37].

With only 130 cases referred in the literature until 2016, we present two mesothelial inclusion cysts as the cause of acute appendicitis. Although benign with excellent prognosis, recurrence is frequent and thus follow-up is necessary [38]. In one study it was noticed that 84% of such tumors appear in women of reproductive age [38].

Our series also includes an adenocarcinoma (SBA) and a NET of small bowel. NETs are the most common type of small bowel neoplasms (2% of all GI's neoplasms). They are treated surgically whenever possible, as was performed in our case, and follow-up is recommended [39]. One third of small bowel's tumors are SBA. Mostly discovered in stage III such tumors have low 5-year survival (14-33%) and are treated surgically, whereas adjuvant chemotherapy is an option in advanced stages [40,41]. A rare case of hemangiopericytoma of small bowel, which represents <1% of vascular tumors, was an

incidental finding in a case of small bowel ischemia. The lesion was excised radically. In the literature, chemotherapy or radiation are recommended for larger lesions or metastasis. Recurrence rates are high, often in distant sites, but 5-year survival is satisfactory, even in metastatic diseases [42,43].

A Well-Differentiated Papillary Mesothelioma (WDPM) was found incidentally in the omentum during a laparotomy in a 59-year patient and was treated with omentectomy. Such tumors are infrequently found in women of reproductive age and rarely cause symptoms. Since WPDM is a tumor of uncertain malignancy, close follow-up is essential [44].

The frequency of many of these tumors is not well described and in fact is not known in the literature. Although our results are limited to the population of an area in southern Greece, we should mention the marked increase of the population during summer period in addition to the number of the immigrants. Three of the patients (7.5%) were not from Hellenic origin.

The drawback of our study is the lack of follow up of the patients. Moreover, there is no data about few of the patients, to whom further management, either adjuvant chemotherapy or radical operation, was suggested after the diagnosis was set in the initial operation in our hospital and chose another hospital for their definitive oncological management. More multi-center studies should be performed in order to confirm our results and reveal the exact incidence of rare GI tumors in the daily clinical practice.

Conclusion

In summary, our study demonstrates the importance of knowledge and skills needed to cope with rare situations in a surgical unit, independent of the level of healthcare a hospital belongs. To the best of our knowledge this is the first single-center study which retrospectively reported the volume of rare GI tumors operated in a Department of General Surgery of a regional hospital. Moreover, this is the first study examining these tumors regarding their nature and behavior related to the different histological types, the mode of presentation, indication for the operation and demographic characteristics of the patients. In most occasions, pre-operative diagnosis was not available so the surgeon must be alert and ready for intraoperative decisions. In addition, most cases underwent emergent/urgent laparotomy. Since in most such tumors no official guidelines are available, more reports must come to light in order actions to be taken. Knowledge of epidemiology of rare tumors may help, but is not panacea, since not every case correlate with them.

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