



## Primary Small-Bowel Tumors; Our Clinical and Surgical Experience

### Primer İnce Barsak Tümörleri; Klinik ve Cerrahi Deneyimimiz

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#### ABSTRACT

**Objective:** Small bowel tumors are rare lesions and they constitute less than 5% of all gastrointestinal system (GIS) tumors. Diagnosis of small bowel tumors is very difficult due to nonspecific clinical findings. The aim of this study is to evaluate the data of patients with primary small bowel tumors who underwent surgical treatment in our clinic.

**Methods:** Thirty-five primary small bowel tumor cases operated in Dicle Hospital between January 2011 and December 2020 were retrospectively analyzed. The patients were evaluated in terms of age, gender, admission complaint, operative finding, surgical method, pathological findings, morbidity and mortality. The results of the patients were obtained from the file records and/or interviews with the patients.

**Results:** Twenty-two (62.9%) cases were female and 13 (37.1%) were male, with a mean age of 53.9 (21-79). Abdominal pain was present in all cases. Abdominal distension was seen in 15 (42.9%) cases, vomiting in 10 (28.6%) cases, invagination findings in 5 (14.3%) cases, GIS perforation in 4 (11.4%) cases, GIS bleeding in 2 (5.7%) cases. Abdominal computed tomography (CT) was used as the imaging method in all patients. The mean time between the onset of complaints and diagnosis was 1.6 (1-15) months. Laparoscopic surgery was used in six (17.1%) cases, and open surgery was used in others. Tumors were mostly located in the jejunum (68.6%). The most common type was adenocarcinoma (34.3%) detected in the postoperative histopathological examination. Mortality was observed in 3 (8.6%) cases in the 6-month postoperative follow-up.

**Conclusion:** Small intestinal tumor is a rare condition. Preoperative diagnosis is difficult due to the nonspecific nature of the symptoms. If there is high doubt in the diagnosis, surgical intervention should not be postponed.

**Keywords:** primary tumor, small intestine, surgical treatment

#### ÖZ

**Giriş:** İnce barsak tümörleri nadir görülen lezyonlar olup tüm gastrointestinal sistem (GİS) tümörlerinin %5' inden daha azını oluştururlar. Nonspesifik klinik bulgularına bağlı olarak ince barsak tümörlerinin tanısı oldukça güçtür. Bu çalışmanın amacı, kliniğimizde cerrahi tedavi uygulanan primer ince barsak tümörlü hastaların verilerinin değerlendirilmesidir.

**Yöntem:** Ocak 2011- Aralık 2020 yılları arasında Dicle Üniversitesi hastanesinde opere edilen 35 primer ince barsak tümör vakası geriye dönük olarak incelendi. Hastalarda yaş, cinsiyet, başvuru şikâyeti, operasyon bulgusu, uygulanan cerrahi yöntem, patolojik bulgular, morbidite ve mortalite açısından değerlendirildi. Hastaların sonuçları dosya kayıtlarından ve/veya hastalarla görüşülmesi sonucunda elde edildi.

**Bulgular:** Olguların 22'si (%62.9) kadın, 13'ü erkek (%37.1) olup, ortalama yaşları 53.9 (21- 79) idi. Tüm olgularda karın ağrısı mevcuttu. 15 (%42.9) olguda abdominal distansiyon, 10 (%28.6) olguda kusma, 5 (%14.3) olguda invaginasyon bulguları, 5 (%14.3) olguda GİS perforasyonu, 2 (%5.7) olguda GİS kanaması görüldü. Bütün hastalarda görüntüleme yöntemi olarak karın bilgisayarlı tomografisi (BT) kullanıldı. Şikayetlerin başlaması ile tanı konulması arasında geçen süre ortalama 1.6 (1 -15) aydı. Altı (%17.1) olguda laparoskopik cerrahi yöntem, diğerlerinde açık cerrahi yöntem uygulandı. Tümörler daha çok jejunum (%68.6) yerleşimli idi. Postoperatif histopatolojik incelemede en sık adenokarsinom (%34.3) tespit edildi. Postoperatif 6 aylık takipte 3 (%8.6) olguda mortalite gözlemlendi.

**Sonuç:** İnce bağırsak tümörü nadir görülen bir durumdur. Semptomların nonspesifik oluşu nedeniyle preoperatif tanı koymak zordur. Tanıda yüksek şüphesiz varsa cerrahi müdahale ertelenmemelidir.

**Anahtar Kelimeler:** primer tümör, ince barsak, cerrahi tedavi

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## INTRODUCTION

Although the small intestine contains 80% of the digestive tract mucosa, neoplasms, especially malignant tumors, are very rare. The reasons for this situation can be summarized as the rapid passage of the contents of the small intestine and the short contact time with the mucosa, the relatively low amount of carcinogens in the contents of the small intestine, and the presence of local protective mechanisms. Despite advanced diagnostic methods, small bowel tumors (SBTs) are difficult to diagnose and are usually found to be advanced at the time of diagnosis. These tumors can cause insidious abdominal pain and weight loss or emergencies that require surgery such as bleeding, obstruction and perforation (1). Computed Tomography (CT) is the most effective radiological method in diagnosis and preoperative staging. Unlike benign small bowel lesions, malignant lesions almost always cause symptoms. The most common symptoms are pain and weight loss (2). The most common malignant tumors of the small intestine are adenocarcinomas (50%). In order of frequency, malignant lesions of the small intestine are adenocarcinoma, carcinoid tumors, malignant gastrointestinal tumor (GIST), lymphomas, and sarcomas.

While the incidence of adenocarcinoma decreases from the duodenum to the ileum, lymphomas are mostly located in the distal small intestine. 20% of malignant tumors of the small intestine are located in the duodenum, 30% in the jejunum and 50% in the ileum. Apart from primary tumors of the small intestine, metastatic tumors may occur in the small intestines as a result of the spread of other organ cancers in the abdomen. The most common benign tumors are, in order of frequency, benign GISTs, adenomas, lipomas, hemangiomas, and fibromas (3). In this study, we aimed to retrospectively analyze the data and results of patients with primary SBTs who were operated in our clinic in the last 10 years.

## MATERIAL and METHODS

In this study, 35 primary small bowel tumor cases operated in Dicle University Faculty of Medicine General Surgery Clinic between January 2011 and December 2020 were retrospectively analyzed. The patients were evaluated in terms of age, gender, complaint, diagnostic methods, operative findings, tumor localization, surgical method, pathological findings, morbidity, mortality and survival. Long-term results were

obtained from file records and/or interviews with patients. Patients aged 18 years or older, who underwent emergency or elective surgery, and diagnosed with primary small bowel tumor (benign or malignant) were included in the study. Periampullary tumors, tumors localized in the duodenum adjacent to the ampulla vateri and metastatic tumors were excluded from the study. The study was approved by the local ethical committee of Dicle University, Turkey.

## RESULTS

Of the cases included in the study, 22 (62.9%) were female and 13 (37.1%) were male, with a mean age of 53.9 (21-79). All patients had abdominal pain. Abdominal distension (constipation) was seen in 15 (42.9%) patients, nausea in 8 (22.9%) patients, invagination findings in 5 (14.3%) patients, GIS perforation in 5 (14.3%) patients, and GIS bleeding in 2 (5.7%) patients. The mean time between the onset of complaints and diagnosis was 1.6 (1-15) months. Abdominal computed tomography (CT) was used as the imaging method in all patients. Five cases had previous abdominal surgery. Laparoscopic surgery was used in six (17.1%) cases, and open surgery was used in other cases. Small

bowel resection and anastomosis were the most common surgical techniques. Tumors were located in the jejunum in 24 (68.6%) cases and in the ileum in 11 (31.4%) cases. Postoperative histopathological examination revealed adenocarcinoma in 12 (34.3%) cases, lymphoma in 7 (20%) cases, gastrointestinal stromal tumor (GIST) in 6 cases (17.1%), neuroendocrine tumor (NET) in 2 (5.7%) cases, spindle cell mesenchymal tumor in 2 (5.7%) cases, inflammatory myofibroblastic tumor in 1 (2.8%) case, malignant melanoma in 1 (2.8%) case, polyp in 1 (2.8%) case, tubulovillous adenoma in 1 (2.8%) case, and spindle cell carcinoma in 1 (2.8%) case, and malignant mesenchymal tumor was detected in 1 (2.8%) case. Postoperative morbidity developed in 8 (22.8%) cases. These were wound infection in 3 cases with DM (evisceration was observed in 2 cases), ileus in 2 cases, intra-abdominal abscess in 1 case, pulmonary embolism in 1 case, and acute renal failure in 1 case. Mortality was observed in 2 (5.7%) patients in the 6-month postoperative follow-up. Clinical features of patients included in the study were seen in Table 1 and Table 2.

Case	Age/ Gender	Symptoms	Location	Pathology	Surgery
1	42, M	Abdominal pain, constipation	Terminal ileum	Adenocarcinoma	Resection + ileocolic anastomosis
2	41, F	Abdominal pain, nausea	10 cm from Treitz	Adenocarcinoma	Resection+anastomosis
3	58, M	Abdominal pain, nausea	20 cm from Treitz	Malignant melanoma	Resection+anastomosis
4	40, F	Abdominal pain, nausea	30 cm from Treitz	GIST	Resection+anastomosis
5	65, F	Acute abdomen (perforation)	10 cm proximal to ileocecal valve	GIST	Resection + ileocolic anastomosis
6	48, M	Abdominal pain	130 cm from Treitz	NET	Resection+anastomosis
7	37, F	Abdominal pain, constipation	20 cm proximal to ileocecal valve	Adenocarcinoma	Ileocection/anastomosis
8	79, F	Abdominal pain, constipation	200 cm from Treitz	Adenocarcinoma	Resection+anastomosis
9	72, M	Abdominal pain	150 cm from Treitz	GIST	Resection+anastomosis
10	51, F	Abdominal pain, invagination	40 cm from Treitz	Polyp	Resection+anastomosis
11	67, M	Abdominal pain, constipation	250 cm from Treitz	Adenocarcinoma	Resection+anastomosis
12	45, F	Abdominal pain, constipation	60 cm from Treitz	NET	Resection+anastomosis
13	55, F	Abdominal pain, constipation	40 cm from Treitz	Diffuse large B cell lymphoma	Ileocection/anastomosis
14	41, M	Abdominal pain, gastrointestinal bleeding, nausea	40 cm proximal to ileocecal valve	GIST	Ileocection/anastomosis
15	24, F	Abdominal pain, constipation	130 cm from Treitz	Inflammatory myofibroblastic tumor	Resection+anastomosis
16	74, F	Abdominal pain, gastrointestinal bleeding	40 cm from Treitz	GIST	Resection+anastomosis
17	53, F	Abdominal pain, invagination, nausea	30 cm from Treitz	Inflammatory pseudo tumor	Resection+anastomosis
18	21, M	Abdominal pain, constipation	50 cm from Treitz	Tubulovillous adenoma	Ileocection/anastomosis

10	61, M	Acute abdomen (perforation)	150 cm from Treitz	Burkitt lymphoma	Resection+anastomosis
20	51, F	Abdominal pain, constipation	240 cm from Treitz	Diffuse large B-cell lymphoma	Resection+anastomosis
21	82, F	Abdominal pain, constipation	20 cm from Treitz	Adenocarcinoma	Resection+anastomosis
22	30, F	Acute abdomen (perforation)	50 cm from Treitz	Diffuse large B-cell lymphoma	Resection+anastomosis
26	77, M	Acute abdomen (perforation)	270 cm from Treitz	Adenocarcinoma	Resection+anastomosis
24	70, F	Abdominal pain, constipation	250 cm from Treitz	Adenocarcinoma	Laparoscopic resection+anastomosis
25	62, F	Abdominal pain, nausea	50 cm from Treitz	Spindle cell malignant tumor	Laparoscopic resection+anastomosis
26	54, M	Abdominal pain, invagination	110 cm from Treitz	Spindle cell mesenchymal tumor	Laparoscopic resection+anastomosis
27	74, F	Abdominal pain, constipation	200 cm from Treitz	T-cell lymphoma	Laparoscopic resection+anastomosis
28	30, F	Abdominal pain, invagination	150 cm from Treitz	Mesenchymal tumor	Laparoscopic resection+anastomosis
29	46, M	Abdominal pain, constipation	250 cm from Treitz	Diffuse large B-cell lymphoma	Resection+anastomosis
30	70, F	Abdominal pain, invagination	50 cm from Treitz	Adenocarcinoma	Resection+anastomosis
31	60, F	Abdominal pain	60 cm from Treitz	Adenocarcinoma	Resection+anastomosis
32	65, M	Acute abdomen (perforation)	100 cm from Treitz	B-cell lymphoma	Resection+anastomosis
33	58, M	Abdominal pain, constipation	30 cm from Treitz	GIST	Resection+anastomosis
34	56, F	Abdominal pain, nausea	110 cm from Treitz	Adenocarcinoma	Laparoscopic resection+anastomosis
35	62, F	Abdominal pain, constipation, nausea	240 cm from Treitz	Spindle cell mesenchymal tumor	Resection+anastomosis

(F: Female, M: Male, GIST: Gastrointestinal stromal tumor, BFL: B-cell lymphoma, TFL: T-cell lymphoma)

**Table 2. Distribution of the Patients**

	n	%
<b>Gender</b>		
Female	22	62.9
Male	13	37.1
<b>Tumor location</b>		
Jejunum	24	68.6
Ileum	11	31.4
<b>Histopathological type</b>		
Adenocarcinoma	12	34.3
Lymphoma	7	20
GIST	6	17.1
NET	2	5.7
Spindle cell mesenchymal tumor	2	5.7
Inflammatory myofibroblastic tumor	1	2.8
Malignant melanoma	1	2.8
Polyp	1	2.8
Tubulovillous adenoma	1	2.8
Spindle cell malignant tumor	1	2.8
Malignant mesenchymal tumor	1	2.8

(GIST: Gastrointestinal stromal tumor, NET: Neuroendocrine tumor)



**Figure 1.** Intraoperative view of the small bowel tumor localized in jejunum

## Statistical analysis

The statistical analyses were performed using the Statistical Package for the Social Sciences (SPSS version 21.0, SPSS Inc., Chicago, IL, USA) computer programme. Descriptive statistics were expressed as the mean and Standard deviation (SD) for numerical variables and as numbers and percentages for categorical variables.

## DISCUSSION

Primary SBTs are extremely rare. Chronic occult blood loss and associated iron deficiency anemia are the most common clinical findings in small bowel cancers. Other symptoms may vary according to the localization of the tumor. While epigastric pain, nausea, vomiting, and obstructive jaundice may be seen in tumors originating from the duodenum, recurrent abdominal pain, increased bowel sounds, abdominal distention, weight loss, and recurrent ileus attacks are more common findings in more distal tumors (4). The most common symptom in our cases was abdominal pain.

In our study, 22 patients were female and the number of female patients was 1.7 times higher. Unlike this, in a study conducted by Bilimoria et al. between 1985 and 2005 with



67.843 patients, small bowel cancer was found to be more frequent in men than in women (5). The time elapsed between the appearance of symptoms and diagnosis can vary between 3-16 months (6). In our study, the median time from onset of symptoms to diagnosis was 1.6 months. In the study of Minardi et al., the time from the onset of symptoms to surgery was reported as 54 days (7). Some patients may have no previous complaints and the diagnosis can be made after laparotomy for a sudden onset of ileus. The presence of weight loss along with cramp-like abdominal pain especially after meals should bring up the possibility of small bowel tumor. In a patient with such a clinic and a suspected small bowel tumor, the presence of annular narrowing in the small intestine, a vegetative mass and/or an irregularity in the mucosa in a local area in the double-contrast small bowel X-ray (Enteroclysis) supports the diagnosis. While reaching the lesions in the last parts of the duodenum and terminal ileum by gastroscopy and colonoscopy, tissue samples can be obtained, enteroscopy is required to reach the lesions in other parts of the small intestine. In cases where these examinations cannot be performed or results cannot be obtained, laparotomy should be performed

without delay. Computed tomography (CT) and/or magnetic resonance (MR) imaging are other examination methods that can help in diagnosis. We used CT for diagnosis in all our cases (8,9).

Adenocarcinomas are responsible for about half of small bowel tumors. They can also be seen in the jejunum and ileum, mostly in the duodenum (ampullary and periampullary region). In familial polyposis syndromes (Familial polyposis, Gardner syndrome, Peutz-Jeghers syndrome, etc.) and Crohn's disease of the small intestine, the incidence of adenocarcinoma in the small intestine has increased. There are ulcerations in the mucosa and narrowing of the intestinal lumen. Regional lymph nodes and liver are the most common sites of metastasis in small bowel adenocarcinomas. Treatment of small bowel adenocarcinomas is surgical removal of the tumor. The prognosis is better in ampullary and periampullary tumors. In jejunal and ileal cancers, wide resection is performed to include the mesentery and lymph nodes (10). In the study of Koc et al. the most common histologic subtype was reported as adenocancer (42.5%) in eighty patients (11). In our study 12 (34.3%) patients were reported as adenocarcinoma.



Carcinoid tumors are neuroendocrine tumors that originate from neuroendocrine cells, contain and secrete serotonin. It constitutes 1-2% of gastrointestinal tumors. Although they can be found in any part of the gastrointestinal tract, especially in the appendix, small intestine, stomach and rectum, they can also be located in extra-intestinal areas such as bronchi, biliary tract, ovary and testis. In the small intestine, it is mostly localized in the ileum. It is generally benign, but sometimes malignant phenotype may develop and liver metastases may occur. Carcinoid syndrome occurs in less than 10% of carcinoid tumors. In cases with liver metastases or when the tumor originates from the extraintestinal organs, carcinoid syndrome occurs because the active substances will be directly mixed into the systemic circulation. The main treatment is surgical resection. In the presence of multiple metastases, treatment is palliative after resection of the primary tumor (12).

Gastrointestinal stromal tumors (GIST) are mesenchymal/stromal neoplasms that appear as intramural masses in the upper gastrointestinal tract and originate from intestinal Cajal cells. They constitute less than 0.5% of all gastrointestinal tract

tumors and 80% of mesenchymal tumors originating from the gastrointestinal tract. GISTs constitute 2% of stomach tumors, 14% of small bowel tumors and 0.1% of colon tumors. Although GISTs can be found at any level of the gastrointestinal tract, from the esophagus to the anus, they occur mostly in the stomach (40-70%), small intestine (20-40%), and colon (5-15%). Most of the patients are asymptomatic. Tumors are usually asymptomatic until they reach large diameters and are often detected incidentally during examinations for other reasons. The symptoms that occur are related to the size and localization of the tumor. Advanced patients may present with a palpable mass, abdominal pain, or gastrointestinal bleeding. GISTs usually show exophytic growth. Approximately 1/3 of the patients have liver, peritoneal and retroperitoneal metastases at the time of first presentation. Lymphatic involvement and spread are rare in GISTs. All GISTs should be followed up as they may have malignant potential. Tumor size and mitotic activity in histopathological examination are two important factors affecting survival (13,14). In our study, GIST was detected in 6 cases.

Lymphomas constitute 1% of gastrointestinal system tumors. 50-60% of gastrointestinal system primary lymphomas are seen in the stomach, 20-30% in the small intestine, and 10-20% in the colon and rectum. The most common localization of small intestinal lymphoma is the ileum, which parallels the localization of the mucosa-associated lymphoid tissue. While B-cell lymphomas are frequently observed in the ileum, those seen in the jejunum are mostly T-cell lymphomas. Patients are usually over 50 years old. It is more common in women. Patients often complain of abdominal pain, weight loss, diarrhea, vomiting, and bleeding. Lymphomas in the histological classification of small bowel tumors published by the World Health Organization (WHO) in 2000;

- 1) Immunoproliferative small bowel disease
- 2) Western type B-cell MALT lymphoma
- 3) Mantle cell lymphoma
- 4) Diffuse large B-cell lymphoma
- 5) Burkitt lymphoma
- 6) Atypical Burkitt lymphoma
- 7) T-cell lymphoma (15,16).

Of our lymphoma cases, 4 were diagnosed with diffuse large B-cell lymphoma, 2 with T-cell lymphoma, and 1 with Burkitt lymphoma.

Inflammatory pseudotumor of the small intestine is a very rare condition. It is a

localized predominantly submucosal mass characterized histologically by inflamed granulation tissue with variable numbers of eosinophils. They are clinically interpreted to be neoplasm and treated by segmental resection. Although these lesions generally pursue a benign clinical course, some intraabdominal and retroperitoneal lesions of this type have typically shown local recurrence and even distant metastases. The etiology of these tumors is not known but they may represent an unusual inflammatory response to precipitating factors such as surgery, trauma and localized infection (17).

Metastatic tumors of the small intestine are more common than primary neoplasms. The most common metastases to the small intestine originate from intra-abdominal organs, including the uterus-cervix, ovaries, kidneys, stomach, colon, and pancreas. Due to its anatomy and localization, the radiological diagnosis of small bowel lesions is difficult. The efficacy of direct radiographs, contrast radiographs, ultrasonography, CT and MR imaging is limited in some lesions. Even endoscopic methods may be insufficient. In some cases, the diagnosis can be made by laparotomy (18,19). The primary treatment of small bowel malignant tumors is surgical,

which consists of segmental resection including wide excision of the lymph node bearing mesentery. Radical resection is often chosen for relatively early stage patients, whereas the palliative operation is applied to very late stage patients. The prognosis for malignant small bowel tumors depends on the extension of tumor cells through the bowel wall and the metastatic spread to lymph nodes and distant organs (20).

In differential diagnosis Magnetic resonance (MR) imaging is rapidly increasing clinical acceptance to evaluate the small bowel and can be the initial imaging method to investigate small bowel diseases. MR examinations may provide the first opportunity to detect and characterize tumours of the small bowel. Intra- and extraluminal MR findings, combined with contrast enhancement and functional information, help to make an accurate diagnosis and consequently characterize SBTs. MR enteroclysis should be recommended for the initial investigation in patients suspected of having SBTs. In some cases, conventional ultrasound and elastography can be used in differential diagnosis. Multidetector CT (MDCT) have been introduced as feasible and accurate diagnostic techniques for the identification and staging of small bowel

neoplasms. Videocapsule endoscopy (VCE) and double-balloon enteroscopy (DBE) have revolutionized the diagnosis and management of patients with small bowel diseases, including SBTs. By using all these imaging methods, the differential diagnosis of these diseases from colon tumors and intestinal malrotations can be made (21-23).

### **Conclusion**

Small bowel tumors are not common. The late onset and nonspecificity of the symptoms, the low growth rate, and the inadequacy of definitive diagnostic methods create difficulties in diagnosis. Since the delay in treatment is the most important factor affecting the prognosis in primary small bowel tumors, surgery should not be avoided if there is a high degree of doubt in the diagnosis.

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