

Primary cancers of the small bowel: About 20 cases

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ABSTRACT

Background: Cancers of the small bowel are rare. Diagnosis is late and difficult because of the lack of specific signs. Treatment is surgical. Prognosis is usually poor and depends on the histological type of tumor. **Aim of Study:** To specify the epidemiological, clinical and therapeutic characteristics of small bowel primary cancers in order to improve their prognosis. **Material and Methods:** This is a retrospective study about 20 cases of malignant tumors of the small bowel, collected in the department of general surgery of Habib Thameur Hospital in Tunis (Tunisia), from January 1994 through June 2011. **Results:** Our series involved 11 women and 9 men aged 62 on average (range: 44 - 80 years). In 45% of cases, the diagnosis was made in a patient rushed to hospital with clinical features of acute generalized peritonitis (66% of all surgical emergencies). Intestinal transit was performed in 5 patients only. Ultrasound abdominal examination was performed in 11 patients. Abdominal CT scan was performed in 7 patients, but the results were conclusive in 4 cases only (57%). Small bowel scanning was done in 5 patients only, but led to a positive diagnosis in all of them. All of our patients underwent surgery. Tumors of the small bowel were histologically divided as follows: carcinoid tumor (8 cases), leiomyosarcoma (7 cases), giant B-cell lymphoma (2 cases), malignant stromal tumor (2 cases) and malignant myxoid schwannoma (1 case). Malignant tumors of the small bowel most commonly arise in the ileum (60%) followed by the jejunum (35%). As for the long-term course, there was a recurrence at one year of a leiomyosarcoma and two recurrences of stromal tumors associated with liver metastases. **Conclusion:** Small bowel cancers are rare. Time to consultation is long and diagnosis is difficult and late due to the absence of typical

presentation. Treatment is surgical and progression depends essentially on histological findings.

KEYWORDS

Primary Cancers; Small Bowel; Epidemiology; Diagnosis

1. INTRODUCTION

Although the small bowel represents 75% of the length of the digestive tract and 90% of its absorptive mucosal surface area, primary cancers of the small bowel are uncommon accounting for less than 6% of all gastrointestinal tract tumors. They are characterized by a lack of specific clinical features which are generally vague. Diagnosis is therefore often made late following a complication. Primary treatment remains surgical consisting of resection of the small bowel and of the regional metastatic lymph nodes.

2. AIM OF STUDY

Our objective is to define the different epidemiological, clinical, therapeutic and prognostic characteristics of these cancers so that their management can be improved.

3. MATERIAL AND METHODS

This is a retrospective study about 20 cases of malignant tumors of the small bowel, collected in the department of general surgery of Habib Thameur Hospital in Tunis (Tunisia), from January 1994 through June 2011. All patients with primary malignant tumor of the small bowel, from the duodeno-jejunal angle until the last ileal loop, were included. We excluded from the study cancers of the duodenum and of Bauhin's valve for they constitute a different anatomico-pathological entity. We also excluded patients with small bowel metastases.

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4. RESULTS

Our series involved 11 women and 9 men aged 62 on average (range: 44 - 80 years). In 45% of cases, the diagnosis was made in a patient rushed to hospital with clinical features of acute generalized peritonitis (66% of all surgical emergencies). As for patients presenting to hospital for chronic symptoms, most of them primarily complained of abdominal pain (50%) and features of impaired general health (30%) (**Table 1**).

Intestinal transit was performed in 5 patients only; the results were conclusive in 3 cases. Thus, it had a positive predictive value of 60% and gave false negative results in 2 patients out of five (40%).

Ultrasound abdominal examination was performed in 11 patients. It revealed a small bowel tumor in 5 cases (43.5%), among which there were 3 cases of hepatic metastasis.

Ultrasound findings were normal in 4 patients among which there were two cases of leiomyosarcoma, a case of carcinoid tumor and a case of lymphoma.

Abdominal CT scan was performed in 7 patients, but the results were conclusive in 4 cases only (57%). Small bowel scanning was done in 5 patients only, but led to a positive diagnosis in all of them.

All of our patients underwent surgery. Midline laparotomy was adopted in 17 cases. Laparoscopy had to be converted into laparotomy in a patient with a lymphoma due to the difficulties encountered. Assisted laparoscopy was done in 2 patients. The site, size and extension of lesions are described in **Table 2**.

Curative resection was done in 15 patients (75% of cases). It consisted of surgical resection with end-to-end anastomosis in 9 cases and of resection with ileostomy in the cases of peritonitis.

Palliative resection was carried out in 5 patients (25% of cases) due to the extent of the local-regional involvement and to peritoneal carcinoses. Partial bladder removal was done in a patient with a leiomyosarcoma.

None of our patients received radiation therapy or hormone therapy.

Of the 20 patients of our series, 5 of them were given adjuvant chemotherapy: 3 cases of carcinoid tumor and 2 cases of lymphoma. Treatment by Gleevec was given to patients who were carriers of malignant stromal tumors.

In our series, tumors of the small bowel were histologically divided as follows:

- Carcinoid tumor: 8 cases
- Leiomyosarcoma: 7 cases
- Giant B-cell lymphoma: 2 cases
- Malignant stromal tumor: 2 cases
- Malignant myxoid schwannoma: 1 case

Malignant tumors of the small bowel most commonly arise in the ileum (60%) followed by the jejunum (35%) (**Table 3**).

Table 1. Circumstances of discovery of malignant tumors of the small bowel.

	n	%
Acute symptoms	9	45%
Peritonitis	6	30%
Acute occlusion of small bowel	1	5%
Massive lower gastrointestinal bleeding	2	10%
Chronic symptoms	11	55%
Abdominal pain	10	50%
Bowel habit disorder	3	15%
Palpable mass	2	10%
Distilling hemorrhage	3	15%
Impaired general health	6	30%

Table 2. Surgery findings.

		Malignant tumors
Site	Jejunum	7
	Ileum	12
	Bifocal	1
Size (cm)		Variable: 3 - 60 cm Average: 31.5 cm
Serosa involvement		7 cases
Metastatic lymph nodes		4 cases
Liver metastases		5 cases
Peritoneal carcinosis		5 cases
Bladder involvement		1 case

In our series there was only one death (5%). The victim had a malignant myxoid schwannoma which metastasized to the lungs. Death occurred three days after surgery, from septic shock following infectious lung disease that didn't yield to intensive care.

Immediate post operative course was uneventful for 17 patients (85% of cases). Apart from the above mentioned fatal complication, wound infection occurred in two patients with leiomyosarcomas who had undergone elective surgery. They responded well to local treatment.

As for the long term course, there was a recurrence at one year of a leiomyosarcoma and two recurrences of stromal tumors associated with liver metastases.

5. DISCUSSION

5.1. Epidemiology

Small bowel tumors are the least frequent among gastrointestinal tract tumors. Their epidemiological statistics, mainly based on hospital data, remain little known. They account for 6% of all GI tract tumors [1-5]. Incidence is 1.6 cases per 100.000 inhabitants per year [6].

Small bowel cancers are very rare. They represent 0.3 to 0.5% of all cancers and 1 to 3% of all malignant tumors of the GI tract [7-9]. According to NAEF and COLL, they are more frequent than benign tumors: 77%

Table 3. Distribution of small bowel malignant tumors according to sites.

	Carcinoid	Leiomyosarcoma	Lymphoma	Stromal	schwannoma
Jejunum	2	4	1	0	0
Ileum	5	3	1	2	1
Bifocal	1	0	0	0	0
Total	8	7	2	2	1

versus 23% respectively [4]. In our department, we collected over 16 years, for the purpose of the study, 25 cases of small bowel tumors, 20 (80%) of which were malignant. Average age of occurrence of malignant tumors is between 50 and 60 years, but it was 62 in our series. It is 56 in North's series [10].

According to the literature data, these cancers occur, in Africa, in patients aged between 34 and 42 and might therefore be correlated with living conditions and life expectancy of patients [11]. Nevertheless, the mean age depends on the histological type of tumor; it is lower for lymphomas and sarcomas [3,7]. Malignant tumors are more frequent in men than in women [12,13].

5.2. Etiology and Pathogenesis

Several hypotheses have been put forward to explain the relative infrequency of small bowel tumors in comparison with other tumors of the digestive tract.

- Reduced transit time of food in the small intestine which shortens the exposure time of the mucosal lining to carcinogens, in addition to fluid circulation of alkaline intestinal chyme [14,15].
- Small amount of bacteria susceptible to produce carcinogens [7].
- Rapid turnover of the small bowel mucosa which inhibits the growth of cancer cells.
- High concentration of microsomal hydrolases likely to inactivate some carcinogens.
- High level of Ig A, that is evidence of an important anti-virus activity.

The corollary of all these hypotheses is that these tumors are more frequent in patients with congenital or acquired immuno-deficiency [7,16,17].

In the same way as we look for esophageal, gastric and colorectal cancers, precancerous lesions should be identified for they pave the way for small bowel cancers [18]. Several authors have shown a link between adenoma and adenocarcinoma of the small intestine. In Perzin's series [19], of the 51 patients involved in the study, 33 of them (65%) had concomitant adenomas and adenocarcinomas in the same lesions. Other precancerous lesions have been mentioned: Familial polyposis [20-22], Crohn's disease [23-28], leiomyomas [29] and coeliac disease [30].

5.3. Clinical Presentation

Given the nonspecific and latent clinical manifestations

of small bowel cancers, diagnosis is made late with several month's lag (5 to 11 months), and it is very difficult to suspect the site of a lesion and to foresee its nature. Moreover, clinical features vary according to anatomic characteristics of the tumor (size, site, shape) [31].

Abdominal pain which was reported by half of our patients, is the primary complaint made by more than 50% of patients [32]. It is due to obstruction of the intestinal lumen or to inflammatory manifestations secondary to tumor ulceration or necrosis [9].

Bowel habit disorders may reveal tumor of the small bowel in 6% to 30% of cases [33-35] in the form of alternating episodes of diarrhea and constipation especially in the presence of Koenig's syndrome. According to some authors, diarrhea is frequently encountered with lymphomas and carcinoid tumors [21,36]. In our Series, the three patients who had bowel habit disorders had carcinoid tumors. Two of our patients complained of diarrhea; a third patient had a sub-obstructive syndrome. Lower G I bleeding is also noted but in patients with leiomyomas and leiomyosarcomas [21,27]. Thus, in our series, there were only 3 cases of minor lower gastrointestinal bleeding: 2 patients with leiomyosarcoma and a third patient with a stromal tumor.

According to several authors, small bowel tumors are frequently revealed by a complication such as an obstructive syndrome or massive G I bleeding or peritonitis [20,37,38]. Nine of our patients actually had to undergo emergency surgery.

Acute intestinal obstruction reveals tumor of the small bowel in 30% of cases [16,39]. This mechanical obstruction results from obstruction of the lumen or from strangulation. Among the 9 patients who underwent emergency surgery, only one had acute intestinal obstruction.

According to Gore and Johnson, cases of peritonitis from tumoral perforation are rare [27]. On the contrary, such cases are frequent in Desa's series [39]; they represent 31% of all cases and they are essentially associated with lymphomas and leiomyosarcomas [40,41]. In our series, 6 patients (30%) had peritonitis following tumoral perforation, involving a lymphoma, a leiomyosarcoma, two carcinoid tumors and a schwannosarcoma.

Recurrent episodes of massive gastro-intestinal bleeding associated with a state of shock represent a revealing clinical feature especially in case of schwannoma [41-43]. Incidence ranges from 8% to 24% in cases of malignancy

[33,44].

In our series, emergency surgery was required for massive lower G I bleeding in two patients, a carrier of leiomyosarcoma and a carrier of stromal tumor.

Physical examination usually provides little information in case of small bowel tumor. Palpation reveals an abdominal mass in only 30% to 50% of cases [45]. In our series, palpation disclosed an abdominal mass in 2 patients only (10%): a carrier of leiomyoma and a carrier of malignant stromal tumor. Liver enlargement is rarely observed (7% of cases) [33]. This finding was encountered in two of our patients who were both carriers of carcinoid tumors.

5.4. Special Investigations

Intestinal transit remains the best diagnostic procedure for small bowel tumors [9]. It is best performed by enteroclysis employing double contrast method [46]. In addition, it is the most reliable method for detecting small lesions in an early stage [47]. Small bowel scanning is a very efficient technique as it combines the advantages of the two other techniques enteroclysis and multibarett scanning [48]. It detects and defines small bowel tumors without yielding false negatives like other investigation procedures (sensitivity 100%, specificity 90%) [49].

In our series, small bowel scanning was done in 5 patients and led to the diagnosis of small bowel tumor in all of them.

Video capsule endoscopy represents a major advance in the field of medical diagnostic investigation. This non-invasive technique makes it possible to visualize all the small bowel mucosal lining, even the zones that can't be reached by other diagnostic methods [50].

5.5. Pathology

More than 2/3 of small bowel tumors are malignant. Most of them are adenocarcinomas [1,2,7,51] though they do not represent more than 1% of adenocarcinomas of the digestive tract. Like in other series, there were no adenocarcinomas in our own series. This finding might be explained by regional differences.

Carcinoid tumors rank second. They arise from the enterochromaffin cells of the neural crest. They represent 20% to 70% of tumors of the small bowel [44] and 13% to 34% of all endocrine tumors [52,53]. Most of them occur in the ileum. In our series, 5 patients out of 8 (62.5%) had ileal carcinoid tumors.

Malignant non-Hodgkin's lymphomas come in third place. They represent 20% to 30% of all primary gastrointestinal lymphomas and 12% to 31% of malignant small bowel tumors. They are located in the ileum in 53% of cases, in the jejunum in 35% of cases and in the duodenum in 12% of cases [10,54]. Anatomic sites of gastro-

intestinal lymphomas vary according to geographic zones. Gastric lymphomas are thus three times more frequent than small bowel lymphomas in the West; it is the other way round that is observed in the East [55].

B-cell lymphomas are present in 65% of cases [25,56]. T-cell lymphomas are less common and represent less than 5% of non-Hodgkin's lymphomas of the gastrointestinal track [56]. Nearly all of them are located in the small bowel and mainly in the jejunum [56].

Leiomyosarcomas account for 10 to 20% of cancers of the small bowel [27].

They are very often located in the ileum and mainly in Meckel's diverticulum [56]. Schwannosarcomas are very rare as they represent no more than 4.9% of sarcomas [42].

5.6. Treatment

Primary treatment of small bowel tumors consists of wide segmental surgical resection (a 5 cm margin on either side of tumor) of the small bowel tumor and of its regional metastatic lymph nodes [57,58].

Apart from lymphomas, chemotherapy is indicated for the treatment of primary small bowel tumors only if the disease is beyond all other means of treatment [18]. In our series, adjuvant chemotherapy was administered to 5 patients, two of whom were carriers of lymphomas. The other 3 patients had carcinoid tumors and liver metastases in addition to peritoneal carcinosis in one of them.

Radiation therapy as adjuvant treatment for small bowel tumors remains a matter for debate [59,60].

5.7. Prognosis

In Miles's series, surgery-related mortality in cases of tumors of the small bowel is 10% [61,62]. The author doesn't mention any specific cause. Death occurred following general causes such as pulmonary embolism or myocardial infarction or after unspecified causes.

In our series, we had one surgery-related death (5%). The patient had a widespread malignant myxoid schwannoma with pulmonary metastases. Death occurred from septic shock following a lung disease.

Prognosis is poor [27]. It depends on several factors: age and general condition of patient, age of symptoms, time of diagnosis, site of tumor, histological type of tumor, extent of invasion of surrounding tissues, presence of lymph nodes or distant metastases and type of surgery performed, palliative or curative.

Five-year survivals are lower than in colorectal cancers, but higher than in other cancers of the digestive tract. Data analysis according to histological type shows an important prognostic variation [18]. Thus, adenocarcinomas are associated with the poorest prognosis [16, 59]. Overall survival rate at 5 years is 30.5%; median

survival is 19.7 months [63].

Five-year survival is lower for leiomyosarcomas than for other gastrointestinal tract cancers [64]; it ranges from 2% to 50% [54]. This survival rate is 25% after palliative surgery against 50% after curative operation [65].

Carcinoid tumors have the best prognosis [16]. Five-year survival ranges between 55% and 75% [51,52]. When they are localized in the small bowel, carcinoid cancers have a bleak prognosis in comparison with other localization in the digestive tract.

6. CONCLUSION

Small bowel cancers are rare. Time to consultation is long and diagnosis is difficult and late due to the absence of typical presentation. Ultrasound scanning, CT scanning, enteroclysis and double contrast examination may reveal the tumor but positive diagnosis of cancer is determined by histology. Treatment is surgical and progression depends essentially on histological findings.

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