

• CLINICAL RESEARCH •

# Primary jejunoileal neoplasmas: a review of 60 cases

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## Abstract

**AIM:** Primary neoplasmas of the jejunum and ileum are infrequent and lack specific manifestations and inaccessibility of conventional endoscopy, so the diagnosis of these tumors are usually delayed. So far the data of primary jejunoileal neoplasmas is still scarce, especially in Chinese medical literature in English. There may be some differences among the Chinese and the westerners in jejunoileal neoplasmas.

**METHODS:** A retrospective analysis was made on clinical findings and pathological types.

**RESULTS:** Of the 60 patients with jejunal or ileal neoplasmas, the most frequent symptom was abdominal pain (57 %), followed by tarry stool (43 %) and hematochezia (10 %). Abdominal mass (40 %) was the most common finding on physical examination, followed by anemia and weight loss (35 %). 67 % of the jejunoileal neoplasms were located in the jejunum. Among the malignant neoplasmas (68 %), malignant stroma (47 %) was most common, while the benign stromoma (20 %) was the most common benign neoplasmas. Preoperatively, 40 patients (67 %) were diagnosed as small bowel neoplasmas, of which 34 were found by enteroclysis. Abdominal mass was shown by CT in 18 cases and by ultrasonography in 13. The mean duration of symptoms before diagnosis was 7 months. In 41 patients with malignant tumors, the duration of symptoms before diagnosis exceeded 12 months in 21 cases, lymphatic or distant metastases were found in 26 (63 %)cases during operation. An emergency laparotomy was performed in 4 patients (7 %) owing to intestinal obstruction or perforation.

**CONCLUSION:** Primary jejunoileal neoplasmas in Chinese present some difference from Westerners on clinical features and histopathologic types. Enteroclysis remains the major relevant diagnostic procedure in this study, the misdiagnostic rate is high preoperatively due to failure of detection by conventional imaging procedures such as CT and inaccessibility of routine endoscopy. For the suspected patients, combined application of aforementioned procedures may facilitate early diagnosis. The wireless capsule endoscopy may improve the diagnostic rate of jejunoileal neoplasmas in the future.

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## INTRODUCTION

Primary neoplasmas of jejunum and ileum, benign or malignant, are infrequent. Clinical manifestations of jejunoileal neoplasmas are nonspecific and the neoplasma-related symptoms occur late, moreover, such neoplasms are beyond the scope of conventional gastroscopy or colonoscopy. The rarity, nonspecific and inaccessibility make them liable to be overlooked, with delayed or erroneous diagnosis<sup>[1]</sup>, and are responsible for high mortality rates. So far the data on small bowel cancers are scarce<sup>[2]</sup>. To our knowledge, there may be some differences between the Chinese and the Westerner in jejunoileal neoplasmas. Herein we reported a series of consecutive cases of primary jejunal and ileal neoplasmas in our hospital over the past 8 years, and analyzed their clinical manifestations, pathologic types and diagnostic process.

## MATERIALS AND METHODS

Cases of primary jejunal and ileal neoplasmas from January 1993 to August 2001 were retrieved through the inpatient computer registry system of PLA General Hospital, Beijing. The medical records were reviewed by the authors. Cases of duodenal and metastatic small bowel tumors were excluded. A total of 60 cases of primary jejunal and ileal neoplasmas were collected. All cases underwent surgery and were confirmed by histopathology.

## RESULTS

### Demographic features

There were 42 males (70 %) and 18 females (30 %) with an average age of 48 years (16-77 years). The majority (51 %) of patients were over 50 years of age.

### Clinical manifestations

Symptoms, signs and complications were listed in the following Table 1. Abdominal pain and tarry stool were the most common symptoms in both benign and malignant jejunoileal tumors. Abdominal mass and anemia were common findings on physical examination in both benign and malignant neoplasmas, while the weight loss was frequent in malignant ones. Complications at presentation such as intestinal obstruction and perforation were more common in malignant tumors.

**Table 1** Clinical manifestations of 60 cases of jejunoileal tumors

	Benign (n)	Malignant (n)	Total
Symptoms			
Abdominal pain	8	26	34 (57%)
Tarry stool	9	17	26 (43%)
Hematochezia	2	4	6 (10%)
Signs			
Abdominal mass	9	15	24 (40%)
Anemia	6	15	21 (35%)
Weight loss	1	20	21 (35%)
Complications			
Intestinal obstruction	2	5	7 (12%)
Intestinal perforation	0	1	1

### Histopathology and sites of distribution

Histopathology and sites of distribution of jejunoileal neoplasmas were summarized in Table 2. Of the 60 cases, 40 (67 %) were located in the jejunum, and 20 (33 %) in the ileum. Malignant tumors ( $n=41$ , 68 %) was nearly twice that of the benign ones ( $n=19$ , 32 %). Benign and malignant stromoma were predominant histological entities.

**Table 2** Histopathology and distribution sites of 60 cases of jejunal and ileal tumors

Tumor types	Jejunum	Ileum	Total
Benign	14 (23.3%)	5 (8.3%)	19 (≈32%)
Stromoma	8	4	12 (20%)
Adenoma	2	0	2 (3.3%)
Angioleiomyoma	2	0	2 (3.3%)
Lipoma	0	1	1 (1.7%)
Lymphangioma	1	0	1 (1.7%)
Neurofibroma	1	0	1 (1.7%)
Malignant	26 (43%)	15 (25%)	41 (≈68%)
Malignant stromoma	17	11	28 (46.7%)
Adenocarcinoma	7	1	8 (13.3%)
Non-Hodgkin's lymphoma	0	2	2 (3.3%)
Carcinoid	0	1	1 (1.7%)
Neuroendocrine carcinoma	1		1 (1.7%)
Malignant histiocytosis	1	0	1 (1.7%)
Total	40(67%)	20(33%)	60 (100%)

### Diagnostic procedures

Before operation, small intestinal diseases were suspected in 73 % (44/60) on admission. There were 40/60 patients (67 %) correctly diagnosed preoperatively. Among the 40 patients, 34 jejunoileal neoplasmas were correctly diagnosed by enteroclysis. Other preoperative diagnoses were made by angiography, colonoscopy, CT and Ultrasonography. Angiography was performed in 5 cases, of which 4 (80 %) of them were discovered preoperatively. One case of lymphoma of terminal ileum was detected by colonoscopy, and one jejunal tumor was diagnosed by CT scan only.

Of the 60 patients, 56 underwent CT scan, abdominal masses or intestinal wall thickening were visualized in 18 (32 %) cases, but two of them were misinterpreted as ovarian cancer, one as renal lipoma and one as intestinal tuberculosis. Ultrasonography was carried out in all 60 patients, and solid abdominal masses were demonstrated in 13 (22 %).

20 patients remained obscure diagnosis preoperatively, final diagnoses were disclosed during laparotomy. The mean duration of symptoms before diagnosis was 7 months. In 41 patients with malignant neoplasms, the duration of symptoms exceeded 12 months in 21 (52 %) cases.

### Surgical findings

Of the 60 patients, three cases of intestinal obstruction and one case of intestinal perforation underwent emergency laparotomy (7 %). The rest underwent selective laparotomy (93%).

Of the 41 of malignant tumors, the masses within the intestinal walls were found in 15 without metastases; local lymph node metastasis in 12, distant and mesenteric lymph node metastasis in 9, liver metastases in 4 and urinary bladder metastasis in 1, the total rate of metastases was 63 % among the malignant tumors. lymph node metastasis was commonest in 51 % of malignant tumors.

## DISCUSSION

Primary neoplasmas of jejunum and ileum are only <2 % of

the gastrointestinal malignant tumors, the incidence was 1.4 per 100 000 compared to 35.7/100 000 for colorectal and 92.9/100 000 for breast cancer<sup>[2,3]</sup>. The jejunoileal neoplasmas are preponderant in male with a ratio of 2:1 between male and female<sup>[3,4]</sup>. Our result showed 70 % in male and 30 % in female, similar to that reported in the literature. The patients with jejunoileal tumors were usually old people with an average age of over 50 years<sup>[1-5]</sup>, in this article, 51 % were over 50 years.

The small bowel neoplasms located in the jejunum usually more frequent than that in ileal. In our results, the jejunal neoplasmas were twice as many as the ileal ones, higher than that reported in the literature<sup>[4]</sup>; but the ratio of malignant and benign in small bowel tumors was concordant with those in the literature<sup>[7]</sup>. As to the histopathologic types, the commonest pathologies of jejunoileal tumors were lymphoma, adenocarcinoma and carcinoids, followed by leiomyosarcomas, neuroendocrine tumors and other entities in Western countries<sup>[1-7]</sup>, which were significantly different from those in our study, in which the benign and malignant stromoma was predominant. In addition, some very rare tumors were seen in our investigation including angioleiomyoma, lymphangioma and malignant histiocytosis etc. The above results indicated that there were some differences between the Chinese and the Westerners in the pathologic types of jejunoileal tumors.

The clinical manifestations of jejunoileal neoplasmas were nonspecific and symptoms usually occurred late. The abdominal pain, gastrointestinal bleeding and weight loss were most frequent complaints in our patients and as well as in the other reports<sup>[4-6]</sup>, followed by nausea and/or vomiting<sup>[4,6]</sup>. Abdominal mass and anemia were the most common physical findings in our study and related literatures<sup>[4,6]</sup>, followed by abdominal distension, muscle guarding and rigidity<sup>[4]</sup>. The diagnosis of small bowel neoplasms was often made when symptoms presented several months late, the median duration of symptoms before diagnosis was 7 months in our study, 3.6 and 6 months in others<sup>[4,6]</sup>. The metastasises of jejunoileal neoplasms were common as they were usually identified late, the metastatic rate of leiomyosarcoma ranged from 24 % to 50 %, with the liver being most frequently involved<sup>[5]</sup>, distant metastasis occurred in 27 %<sup>[2]</sup>. In our study, metastasis was found in 63 % of the malignant tumors, lymph node metastasis was the commonest (51 %) similar to that reported in the literature<sup>[4]</sup>, followed by liver metastases in 10 % of cases.

The preoperative diagnosis of primary jejunoileal neoplasmas remains difficult for clinicians owing to the nonspecific symptomatic presentation. The enteroclysis, enteroscopy and imaging methods have been used generally for small bowel diseases. Radiographic study of small bowel malignant tumors showed abnormality in 87 %<sup>[4]</sup>. In our study, 34 cases of jejunoileal neoplasmas were diagnosed by enteroclysis. Both ultrasonography (US) and CT are also useful for detection of small bowel tumours. CT was found to detect leiomyoma and leiomyosarcoma most successfully and had the additional advantage of locating metastatic lesions<sup>[5]</sup>. Moreover, CT offered the possibility of a preoperative staging by evaluating tumour extension through the bowel wall, involvement of lymph node and possible metastases<sup>[8]</sup>. Other preoperative diagnoses were made through angiography, colonoscopy, CT and Ultrasonography. Angiography was performed in 5 cases, of which 4 (80 %) of small intestinal neoplasmas were discovered preoperatively. A case of lymphoma of terminal ileum was detected by colonoscopy, and one case of jejunum tumor was diagnosed by CT scan only.

Of the 60 patients, 56 underwent CT scan, abdominal masses or intestinal wall thickening were visualized in 18 (32 %), but two cases were misinterpreted as ovarian cancer, one as renal lipoma and one as intestinal tuberculosis. Ultrasonography was

carried out in all 60 patients, and the solid abdominal masses were demonstrated in 13 cases (22 %).

Final diagnoses were disclosed during laparotomy for those 20 patients whose diagnoses remained obscure preoperatively. The mean duration of symptoms before diagnosis was 7 months. In 41 patients with malignant neoplasms, the duration of symptoms exceeded 12 months in 21 cases (52 %).

Jejunum and ileum are difficult to visualize directly. Colonoscopy with retrograde ileoscopy is useful in the diagnosis of neoplasm of the terminal ileum<sup>[4]</sup>. Enteroscopy is capable of observing most of the small intestine<sup>[5]</sup>, but it is hard for the patients to endure. Small bowel enteroclysis is still a good choice for the detection of jejunoileal neoplasms<sup>[6]</sup>, albeit a third of cases of this series may be missed in the examination. Angiography is carried out in only a small proportion of patients, though it is relatively sensitive, especially for those lesions with abundant vascularity, the expense and invasive property limit its application<sup>[7]</sup>. Spiral CT is thought to be comparatively accurate in detecting small bowel tumors<sup>[8]</sup>, however its diagnostic accuracy is poor in this series with possible misinterpretations, probably due to lack of experience of some radiologists in this field. Despite the interference by intestinal gas, ultrasonography proves to be helpful in a fifth of cases to demonstrate thickened intestinal wall or unsuspected mass. Wireless capsule endoscopy is optimal for those patients without intestinal obstruction when it is available. Laparotomy is justified in those suspected cases as a last resort.

Thanks to a variety of investigations, the correct diagnosis is reached in the majority of patients before operation, though the delay between the onset of symptoms and the final diagnosis

is often significant. As a result, more than 60 % of patients with malignant tumors has advanced beyond the early stage of neoplasm. Recognition of this entity, with high index of suspicion, rational application of aforementioned investigation procedures, the advent of capsule endoscopy and justified early laparotomy, may facilitate the diagnosis and improve the outcome.

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