Original Research Article

Jejuno-Ileal tumors: a retrospective study

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ABSTRACT

Background: Primary tumors of the jejunum and ileum are rare and this segment of gastrointestinal tract is relatively inaccessible to conventional endoscopy, leading to a delay in diagnosis. Due to its rarity, data of primary jejunoileal tumors is still scarce, especially in India where diagnostic modalities like capsule endoscopy is not widely available. Herein we aim to discuss the clinico-radiologic findings, pathology and surgical management of such tumors.
Methods: Of the total 51 small bowel resections done in our institute during the period from year 2012 to 2015, 14 patients were identified who were diagnosed with jejunoileal benign/malignant jejunoileal tumors. Records were analysed with respect to patient demographic data, clinical features, radiologic findings, surgical management, histopathology and postoperative outcomes.
Results: Jejunoileal tumors are more common in males (77%) and first presentation was intestinal obstruction (50%) in majority of patients. A definite preoperative diagnosis based on radiologic/endoscopic means was possible in six patients (46%). Patients were operated and laparoscopic group showed superior outcomes in terms of postoperative complications.
Conclusions: Jejunoileal tumors are an infrequent finding in surgical practice. In our study malignant tumors were more common in proximal small bowel. Computed tomography (CT) enterography can detect small bowel lesions with low sensitivity but allows evaluation of lymphadenopathy/distant metastasis. Laparoscopic resection allows initial staging and has the advantage of early recovery, less wound infection rates and better cosmesis.

Keywords: Jejunoileal, Small bowel, Tumors

INTRODUCTION

Primary benign or malignant tumors in the small bowel are rare despite the fact that it contributes to a significant proportion of the length of gastrointestinal tract. Primary small bowel neoplasms constitute 1-5% of all tumors and less than 2% of the malignant lesions located at the digestive tube, with a population incidence of 1.6/100,000.[1,2]

This rarity has been well explained historically to be secondary to: fluid content; rapid intestinal transit; alkaline pH; lower bacterial population than in colon and difference in bacterial metabolism; high concentration of immunoglobulin A, particularly in the ileum; possible cytogenetic factors; rapid proliferation and replacement of mucosa; and stem-cells with more efficient apoptosis than in colon for removal of genetically modified cells.[3-6]

Clinical suspicion of primary jejuno-ileal tumours is uncommon in routine clinical practice as presenting signs and symptoms are not specific and often confused with other gastrointestinal (GI) manifestations, and therefore diagnosis is often delayed making future prognosis dismal. The more frequent clinical presentations of jejunoileal tumours are recurrent abdominal pain, palpable abdominal mass, intestinal obstruction or perforation, and GI hemorrhage.[7]

We recently witnessed a string of such patients who were operated in the department of surgical gastroentrology (B. L. Kapur Super Speciality Hospital, New Delhi) with varied histopathological findings. This gave us the necessary impetus to retrospectively analyze our data pertaining to primary jejuno-ileal tumors, their surgical management and postoperative outcome.

METHODS

We retrospectively reviewed the medical records in the hospital information system and case file sheets of all patients who underwent either laparoscopic or open small bowel resection (jejuno-ileal) in B. L. Kapur Super Speciality Hospital, New Delhi, India during the period from year 2012 to 2015. Institutional ethical approval was duly taken.

A total of 51 small bowel resections were done during this period for strictures (tubercular, Crohns or inflammatory), tumours (benign or malignant) and congenital lesions. Distribution of these lesions are given in Table 1.

Table 1: Broad distribution of jejunoileal lesions operated from 2012 to 2015 (n=15).

S. no.	Type of lesion	Subtype	N (%)
1	Strictures	Crohns disease	3 (6)
		Tuberculosis	10 (20)
		Inflammatory (non-specific)	16 (31)
2	Tumors	Benign/malignant	14 (27)
3	Congenital lesions	Meckels diverticulosis or duplications cyts	8 (16)

Inclusion criteria

All jejunoileal (i.e. ligament of Treitz till ileocaecal junction) solid benign or malignant tumors who underwent surgical resection.

Exclusion criteria

Jejunoileal inflamatory/infective lesions (i.e. tuberculosis, Crohns disease, non-specific ulcers, meckels diverticulosis). Duodenal and periampullary lesions were also not included in the study.

A total of 14 patients who were diagnosed preoperatively or postoperatively with primary benign or malignant jejuno-ileal tumours were included in this study. Mean age was 48±10 years (30-62 years); 10 patients (77%) were male and 3 (23%) female. Mean length of stay was 8±7 days (4-30 days). Records were analysed with respect to patient demographic data, clinical features, radiologic findings, surgical management, histopathology and postoperative outcomes.

Statistics

Statistical package for the social sciences (SPSS) software was used for statistical analysis and data was presented as proportions, mean, standard deviation and range, and analysed with Chi square test.

RESULTS

The most common presenting symptoms was intestinal obstruction in six patients (50%) followed by colicy pain abdomen (22%), bleeding (14%) and perforation peritonitis (14%). Majority of patients who presented with obstruction were harbouring a malignant lesion, except one patient who was diagnosed with intussusception secondary to a jejunal lipoma. Bleeding and perforation was more common in gastrointestinal stromal tumors (GISTs) and lymphomas respectively. Eight patients (61%) had history of significant weight loss and two patients had chronic anaemia secondary to bleeding jejunal GISTs (Table 2).

On examination lump was palpable only in two patients, 1st was a known case of jejunal neuroendocrine tumor with hepatic metastasis who presented with symptoms of intestinal obstruction and underwent palliative laparoscopic jejunal resection and 2nd was case of jejunal amyloidoma.

Majority of patients were in normal body mass index (BMI) range (18.5-24.9 kg/m^2) except one patient a case of jejunal neuroendocrine tumors (NET) who was obese (BMI=31 kg/m^2) and another patient was undernourished (BMI=18 kg/m^2) who was diagnosed with an ileal lymphoma (Figure 1).

Majority of patients had no associated comorbidity (44%). Chronic anemia was found to be the most common comorbid condition in three patients (16%) who had history of malena, two of which were jejunal GISTs and one patient was a metastatic jejunal NET (Figure 2). Patients were preoperatively assessed with a contrast CT abdomen (Figure 3) and some also underwent PET CT whole abdomen (suspected neuroendocrine tumors) and enteroscopy/EUS (proximal jejunal lesions emnable for biopsy) (Figure 4).

A definite preoperative diagnosis based on radiologic/endoscopic means was possible in six patients (46%). Three patients were diagnosed only on basis of radiologic findings (2 GISTs and 1 NET) and in rest three, definite preoperative histopathologic diagnosis was made which correlated with postoperative biopsy findings as the lesions were located in the proximal 50 cm of jejunum, emnable to enteroscopy/EUS guided biopsy.

In this study primary small bowel tumors were more commonly encountered in the jejunum especially adenocarcinoma and NETs, however lymphomas were found in both ileum and jejunum.

Table 2: Distribution of symptoms/signs according to histologic type.

| S. no.	Symptom	AdenoCa	Lymphoma	GIST	Neuroendocrine	Amyloid	Lipoma	N	%
1	Vomiting*	2	3	0	1	0	1	7	50
2	Pain**	0	0	1	1	1	0	3	22
3	Bleeding	0	0	2	0	0	0	2	14
4	Perforation	0	2	0	0	0	0	2	14

*Associated with abdominal distention-suggestive of symptoms of intestinal obstruction, **colicky pain abdomen without intestinal obstruction

Table 3: Radiologic/endoscopic findings and correlation with histopathology.

S. no.	Age	CECT abdomen	Other imaging studies	Preop diagnosis	Postop diagnosis (histopathology)
1	37/m	Ileal perforation	-	Ileal perforation	Ileal lymphoma
2	54/m	Ileal stricture	-	Ileal stricture	Ileal lymphoma
3	40/m	DJ flexure mass	-	DJ flexure mass	Jejunal GIST
4	49/m	Ileal perforation with liver metastasis	-	Ileal perforation	Ileal lymphoma
5	57/m	Jejunal tumor	-	Jejunal tumor	Amyloidoma
6	62/m	Jejunal tumor	-	Jejunal GIST	GIST
7	51/f	Jejunal tumor with liver metastasis	PET CT: avid lesion in jejunum	Jejunal NET	Jejunal NET
8	57/m	Jejunal ?GIST	USG supraumbilical mass (6×3×3 cm)	Jejunal GIST	Jejunal GIST
9	36/f	Jejunal stricture	-	Jejunal stricture	Lymphoidfollicular hyperplasia
10	57/f	-	PET CT: avid lesion at root of mesentery 4×3 cm	EUS guided FNAC s/o Jejunal NET	Jejunal NET
11	44/m	Jejunal stricture	Enteroscopic biopsy s/o adenocarcinoma	Jejunal adenocarcinoma	Jejunal adenocarcinoma
12	30/m	CT- ileal stricture PET CT: ileal lesion	Enteroscopic biopsy s/o lymphoma	Ileal lymphoma	Ileal lymphoma
13	61/m	Jejunal intusuception	Enteroscopy: jejunal polyp	Jejunal polyp	Lipoma
14	61/m	Jejunal thickening	-	-	Adenocarcinoma

Seven patients (50%) underwent laparoscopic assisted bowel resection anastomosis and rest (50%) underwent open surgery. Open approach was preferred in patients who had abdominal distention on examination secondary to intestinal obstruction or perforation. There were no conversions from laparoscopic to open approach. A small midline 3 cm minilaparotomy incision was made to remove bulky specimen in an endobag. Lesions were more commonly found in the jejunum (69%) especially GISTs and NETs. Lymphomas had variable distribution with slight preponderance in ileum. One of the rare histopathological finding was that of a primary Jejunal Amyloidoma (Figure 6 and 7) in a 57 year old male patient who presented with colicky abdominal pain with CT findings (Figure 5) suggestive of a 5×5 cm jejunal growth invading the adjacent mesentery.

The overall mean duration of hospital stay was 7.9±6.8 days. Patients operated laparoscopically (5.5±2 days) were discharged earlier as compared to open approach (10±8.8 days).
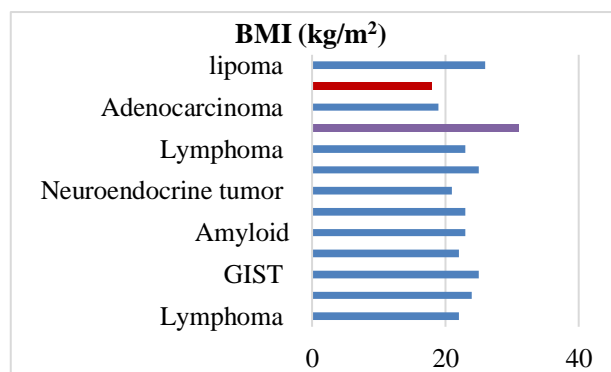

Figure 1: BMI distribution.

With regard to postoperative complications three patients (21%) developed superficial surgical site infection (SSI) identified on basis of obvious pus discharge noted from the wound by the consultant corroborated later by a positive microbiologic culture. *E. coli* was isolated in all three swab samples. Two patients (14%) developed pneumonitis. There were no other major immediate postoperative

complications. There was one mortality in a case of ileal lymphoma with immuno-compromised status who presented with perforation peritonitis expired following a prolonged intensive care unit (ICU) stay after surgery. All these patients were operated with open approach suggesting a higher morbidity and mortality rate in this group when compared to the laparoscopic group.
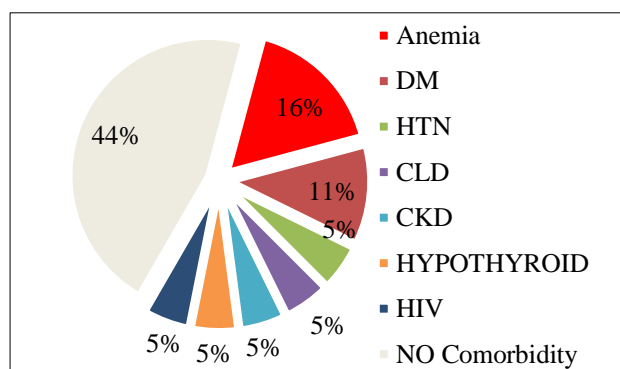
**Figure 2: Distribution of comorbidity.**
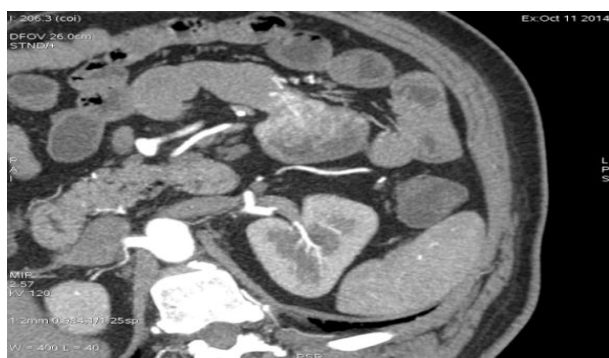
Figure 3: CT enterography (arterial phase-left and enteric phase-right) axial images of proximal jejunal loop showing well-defined eccentric intramural polypoidal soft tissue mass depicting heterogeneous enhancement on arterial phase.


Figure 4: Enteroscopic image showing a submucosal nodular lesion in jejunum.

All patients diagnosed with malignant small bowel tumors (i.e. adenocarcinoma, NET and lymphoma) received adjuvant/palliative chemotherapy and GISTs were started
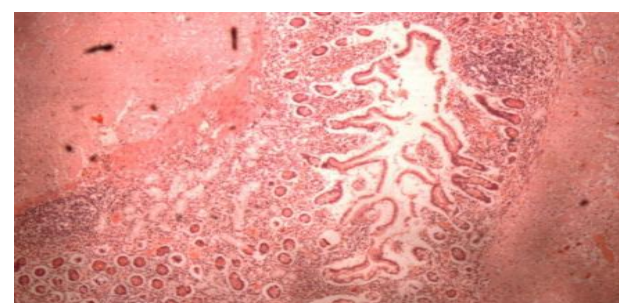
on imitinib mesylate. There were no recurrences noted as per the follow up records of these patients till date.
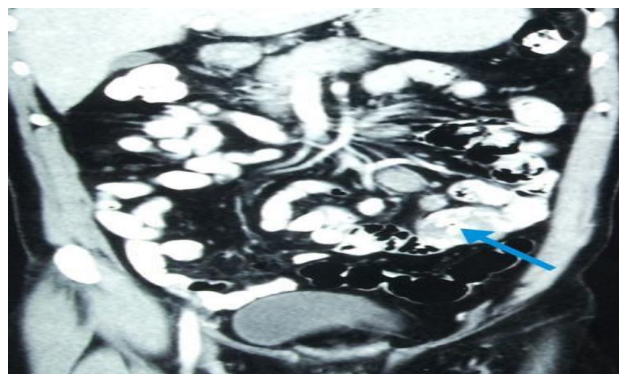
Figure 5: CECT abdomen showing focal jejunal growth involving mesentery.
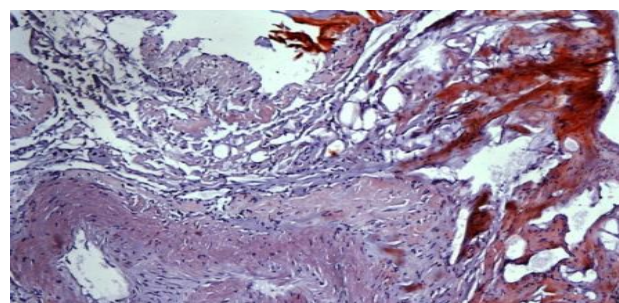
Figure 6: Classical congo red stain (acellular hylinization of tumor s/o amyloidoma).

Figure 7: H and E stain 10x view.

Table 4: Postoperative complications.

S no.	Type of complications	N	%
1	Wound infection (superficial SSI)	3	21
2	Pneumonitis	2	14
3	Anastomotic leak	0	0
4	Urinary tract infection	0	0
5	Deep vein thrombosis	0	0
6	Pulmonary embolism	0	0

DISCUSSION

Primary jejuno-ileal tumors are rare and unlikely to found in routine surgical practice. Its rarity as mentioned before

is said to be secondary to multiple factors which renders this segment of gastrointestinal tract comparatively immune to malignant transformation. Due to absence of specific symptoms/signs and relative inaccessibilty of this region to routine endoscopy and low sensitivity of imaging modalities, diagnosis is usually delayed. Malignant tumors often produce more exuberant clinical manifestations than benign tumors. However, because of the unspecific signs and symptoms, the mean time required for diagnosis was close to 6 months, a fact that is corroborated by the literature.[8,9] It likely that a case of intestinal obstruction may reveal a jejunal tumor during exploration as a surprise to the surgeon.

In accordance to other authors, abdominal pain, weight loss, anemia and nausea/vomiting represented the signs and symptoms commonly observed in our patients with small bowel tumors.[10,11] Majority of these patients presented with intestinal obstruction (50%). Anemia (16%) was the most common comorbidity in our patients secondary to occult gastrointestinal bleed. Lesions that lead to digestive hemorrhage occur more frequently in the proximal small bowel segment, what is likely to be explained by the larger diameter of jejunum, allowing for tumoral growth with posterior ulceration and hemorrhage. Distal lesions obstruct the ileal lumen before the occurrence of such alterations.

Association of malignancies with obesity is well established in one of the largest meta-analysis conducted till date however this was not corroborated as majority of patients in our study had normal BMI and overall sample size was small.[12]

With regard to imaging modalities magnetic resonance imaging (MRI) has proved to be more sensitive than computed tomography CT for detecting mucosal lesions of the small bowel and it appears to facilitate superior detection of segments with only superficial abnormalities. These findings may be due to the better soft-tissue contrast afforded by MRI, which is required for tissue characterization and the detection of subtle areas of abnormality.[13]

In our study a definitive preoperative radiological diagnosis of small bowel tumor was made in 46% patients on basis of contrast enhanced CT (CECT)/PET scan abdomen. Moreover, CT offered the possibility of a preoperative staging by evaluating tumour extension through the bowel wall, involvement of lymph node and possible metastases. PET-CT is especially helpful in cases of well differentiated small bowel neuroendocrine tumors. In proximal jejunal/termial ileal lesions preop endoscopic biopsy was possible in three cases as mentioned previously. Rest of the small bowel being inaccessible to routine endoscopy lead to the introduction of capsule endoscopy .Capsule endoscopy has rapidly gained acceptance as a standard method for small bowel evaluation for occult gi bleeed even though it is contraindicated in patients with suspected or documented

intestinal obstruction and does not allow therapeutic intervention.[14,15] In our study capsule endoscopy was not done .

In our study adenocarcinoma/neuroendocrine tumors were more common in jejunum and lymphomas were found in ileum due to its high lymphoid concentration. Studies by Zollinger, Cunningham et al, and Ojha et al found that the site of the primary lesion did not influence survival.[16-18]

Laparoscopy has emerged as valuable tool for diagnostic as well as therapeutic interventions in jejunoileal tumors. It allows initial staging of neoplastic lesions and segmental resection. Our study showed a shorter duration of hospital stay and lower incidence of post-operative complications in the laparoscopy group. Only contraindication being patients presenting with intestinal obstruction/perforation .We readily acknowledge the limitations of such a small study size. Given the relative rarity of these tumours, our numbers did not demonstrate statistically significant differences in length of stay or post-operative complicationsPrimary jejuno-ileal tumors are rare and unlikely to found in routine surgical practice. Its rarity as mentioned before is said to be secondary to multiple factors which renders this segment of gastrointestinal tract comparatively immune to malignant transformation. Due to absence of specific symptoms/signs and relative inaccessibilty of this region to routine endoscopy and low sensitivity of imaging modalities, diagnosis is usually delayed. Malignant tumors often produce more exuberant clinical manifestations than benign tumors. However, because of the unspecific signs and symptoms, the mean time required for diagnosis was close to 6 months, a fact that is corroborated by the literature.[8,9] It likely that a case of intestinal obstruction may reveal a jejunal tumor during exploration as a surprise to the surgeon.

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CONCLUSION

Jejunoileal tumors are an infrequent finding in surgical practice and is usually a diagnosis of exclusion or intraoperative surprise. However, when evaluating any patient with nonspecific abdominal symptoms, particularly occult GI bleed with normal endoscopy findings, such tumors should be high on the differential diagnosis list. In our study malignant tumors were more common in proximal small bowel. CT enterography can detect small bowel lesions with low sensitivity but allows evaluation of lymphadenopathy/distant metastasis. Surgical resection of small bowel tumors is the treatment of choice. Laparoscopy has its advantages as it allows initial staging

and associated with early recovery, less wound infection rates and better cosmesis.

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