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ANALYSIS OF PRIMARY SMALL BOWEL TUMORS : A SINGLE CENTRE EXPERIENCE AND REVIEW OF LITERATURE

General Surgery

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ABSTRACT

INTRODUCTION: Despite composing 75% of the length and 90% of the surface area of the gastrointestinal tract, the small bowel develops relatively few primary neoplasms and less than 2% of gastrointestinal malignant neoplasms. Variation in presenting symptoms may delay the treatment and therefore worsen the outcome. **MATERIALS AND METHODS:** 10 cases of small bowel tumors were operated in Government medical college hospital,Karur from June 2022 to September 2022. Their medical records were analysed retrospectively. **DISCUSSION:** Small bowel tumours are rare and in total account for less than 10% of gastrointestinal neoplasia. The majority of small bowel neoplasms are benign. They are frequently asymptomatic and identified incidentally. Four major histological types account for over 99% of small bowel malignancies that includes adenocarcinoma, carcinoid tumours, lymphomas and GIST. **CONCLUSION:** Small bowel tumors have a rare incidence and classically presents late, most often diagnosed after surgery for small bowel obstruction or perforation. Appropriate evaluation of presenting symptoms helps in early diagnosis and prompt treatment.

KEYWORDS

Small bowel tumors; Adenocarcinoma; GIST; Carcinoid; Extra skeletal Ewing sarcoma; Lymphoma

INTRODUCTION:

Despite composing 75% of the length and 90% of the surface area of the gastrointestinal tract, the small bowel develops relatively few primary neoplasms and less than 2% of gastrointestinal malignant neoplasms. The mean age at presentation is 62 years in the setting of benign tumors and approximately 57 years for malignant tumors[1]. The incidence of small bowel neoplasia varies considerably, with benign lesions identified more often at autopsy. In contrast, malignant neoplasms account for 75% of symptomatic lesions that lead to surgery. This reflects the fact that most benign neoplasms are asymptomatic and often identified as an incidental finding. Stromal tumors and adenomas are the most frequent of the benign tumors and appear to be more common in the distal small bowel but may be somewhat misleading because of the relatively short length of the duodenum. Adenocarcinoma is the most common malignant neoplasm, accounting for 30% to 50% of malignant neoplasms of the small intestine; neuroendocrine tumors (NETs) account for 25% to 30% of small intestine malignant neoplasms. Adenocarcinomas are more prevalent in the proximal small bowel, whereas the other malignant lesions are more common in the distal small bowel [2]. The most common presenting symptom is abdominal pain, typically intermittent and crampy. Weight loss, nausea and vomiting, gastrointestinal bleeding and intestinal obstruction are reported[3,4].

MATERIALS AND METHODS:

10 cases of small bowel tumors were operated in Government medical college hospital,Karur from June 2022 to September 2022. Their medical records were analysed retrospectively. Written informed consent was obtained from all 10 patients.

CASE 1:

A 20 year old female presented with severe lower abdominal pain and features of peritonitis. Pneumoperitoneum confirmed on X-ray erect abdomen with ultra sonogram suggesting loculated fluid collection with internal echoes. Intra operative findings revealed perforated intra luminal growth 50 cm proximal to ileocaecal junction with dense adhesions. Segmental resection of tumor with adjacent ileum and ileo-ileal anastomosis was done. Immunohistochemistry revealed CD 99 and FLI 1 positive suggesting extra skeletal Ewing sarcoma (Figure 1A and 1B). Follow up PET CT showed metabolically active peritoneal nodules in pelvis which are suspicious of metastasis. The patient was referred to medical oncologist for systemic chemotherapy. Patient started with VCD/ IE regimen (vincristine, cyclophosphamide and doxorubicin alternating with ifosfamide and etoposide) every 3 weeks.

CASE 2:

A 36 year old female on anti retro viral therapy presented to emergency department with features of intestinal obstruction. Radiological evidence showed obstructing mass in ileum.Patient was taken for emergency laparotomy. Intra operative finding showed intraluminal growth completely obstructing the lumen of ileum 20 cm proximal to ileocaecal junction . Resection of growth along with adjacent 5 cm bowel and double barrel ileostomy done in view of hemodynamic instability. Post operative HPE revealed high grade NHL. Patient subjected to R-CHOP chemotherapy regimen.

CASE 3:

A 26 year old male presented to emergency department with diffuse abdominal pain and features of peritonitis. X ray revealed pneumoperitoneum. Patient was subjected to emergency laparotomy and a perforating mid ileal growth of size 5*4 cm was found (Figure 2). Segmental resection with ileoileal anastomosis was done. Post operative HPE report revealed Burkitt lymphoma (aggressive Non-Hodgkin B-cell lymphoma) and patient was started with chemotherapy.

CASE 4:

A 66-year-old woman presented abdominal pain for 3 months. During investigation, CT scan evidenced a jejunal growth. Patient was submitted to exploratory laparotomy . During surgery a small bowel tumor was found in the jejunum and a 10 centimeters enterectomy was performed. The postoperative pathology showed a high-grade stromal tumor (cKit positive and more than 5 mitoses per 50 fields). The patient was started with Imatinib chemotherapy.

CASE 5:

A 72 year old male presented with abdominal pain and features of intestinal obstruction. The stromal tumor was found in ileum on laparotomy. Segemental resection and anastomosis was done.post operative period was uneventful. Pathology confirmed a stromal tumor (positive cKit, low grade) (Figure 3). The patients were subjected to post operative imatinib chemotherapy .

CASE 6:

A 70 year old male presented with bloody diarrhoea, abdominal cramp for the past 3 months. There was no history of fever or weight loss and physical examination was clear. Colonoscopy of no significance. Computed tomography (CT) and magnetic resonance image (MRI) showed a tumor lesion in the mid ileum. The patient was subjected to laparotomy. Segmental resection and anastomosis was done. Pathology defined a stage 1, low-grade stromal tumor (5

cm, cKit positive, 3 mitoses/50 field). Patient started with Imatinib therapy.

CASE 7:
A 63-year-old female patient presented with abdominal pain, abdominal distension, weight loss, anorexia and vomiting for 1 month. Comorbidities and family history were not present. Examination revealed abdominal distension, with no signs of peritonitis. CT scan showed an obstructive lesion in the terminal ileum with no significant distension. Exploratory laparotomy was performed . Terminal ileum resection with right hemicolectomy and ileotransverse anastomosis was done due to obstructing lesion (Figure 4A and 4B). Pathology showed a stage III adenocarcinoma. The patient was referred to the Oncology centre and underwent chemotherapy.

CASE 8:
A 57 year old male presented with vague abdominal pain and diarrhoea for 1 month.On evaluation CECT abdomen\pelvis showed a non-obstructing mass in the terminal ileum, with no evidence of metastasis. Laparotomy with right hemicolectomy was done . HPE confirmed the diagnosis of well-differentiated neuroendocrine tumor infiltrating muscularis propria with metastatic tumor in 2 out of 10 resected lymph nodes (T2N1M0). A follow-up laboratory workup showed 5-HIAA level of 2 mg\gCR and chromogranin A of 30 ng\ml.

CASE 9:
A 49 year old Male presented with intermittent abdominal pain and abdominal distension with features of intestinal obstruction. X ray showed multiple air fluid level and USG revealed target sign suggestive of intussusception. Patient was subjected to emergency laparotomy and ileoileal intussusception was found 30 cm proximal to ileocaecal junction (Figure 5A and 5B). On reducing the intussusception ,the lead point was found to be a 4*3 cm ileal growth. Segmental resection with ileoileal anastomosis was done. Post operative HPE report revealed Lipoma with no features of malignancy.

CASE 10:
A 53 year old female presented with vague lower abdominal pain for one month. Screening ultra-sonogram revealed small bowel pathology. CECT abdomen showed growth in jejunum with no evidence of metastasis. Exploratory laparotomy and enterectomy done. Post operative HPE report revealed lipoma with no features of malignancy.

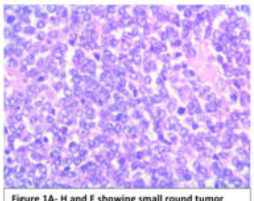
Figure 1A- H and E showing small round tumor cells with brisk mitotic activity

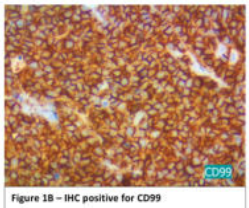

Figure 1B – IHC positive for CD99

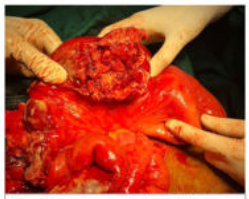
Figure 2- Lymphoma presenting as ileal perforation

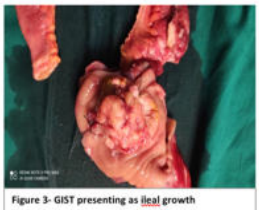
Figure 3- GIST presenting as ileal growth

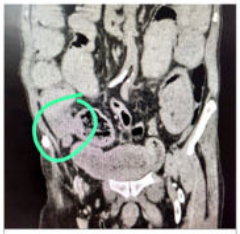
Figure 4A- CT showing mass in terminal ileum with intestinal obstruction

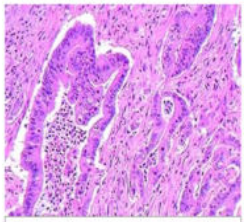
Figure 4B- Adenocarcinoma of small bowel in HPE

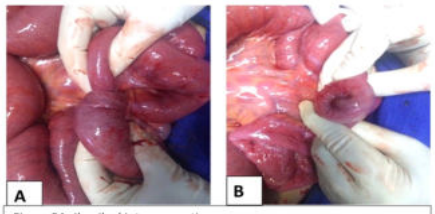

Figure 5A- Ileo ileal intussusception
Figure 5 B- Lipoma as lead point of intussusception.

RESULTS:
In this study 10 cases of both benign and malignant primary small bowel tumors were included. Analysis of small bowel tumors were done based on incidence (chart 1), sex, location and nature of the tumor (Table 1). In two patients (20%) the lesion was in the jejunum and eight (80%) was in the ileum. Diagnosis was done by surgical approach and histopathological evaluation. The procedures done included: segmental resection of bowel with lymphadenectomy in necessary cases. Two patients had right colectomy with ileo-transverse anastomosis .There were no anastomosis leak and the average hospitalisation time was 8 days. The pathological study of the resected specimens identified three GIST (30%), two lymphoma (20%), one adenocarcinoma (10%) , one carcinoid tumor (10%), one extra skeletal Ewing sarcoma (10%), two lipoma (20%). Seven cases underwent chemotherapy.

CHART 1: INCIDENCE OF SMALL BOWEL TUMORS

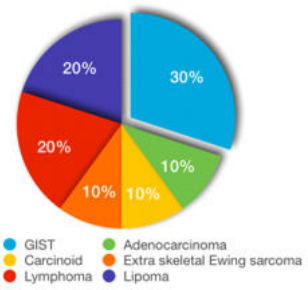

TABLE 1: ANALYSIS OF SMALL BOWEL TUMORS

Analysis of small Bowel tumors

SEX	Male-4	Female-6
LOCATION	Ileum-8	Jejunum-2
NATURE	Benign-2	Malignant-8

DISCUSSION:
The majority of small bowel neoplasms are benign, comprising adenomas, lipomas, haemangiomas and neurogenic tumours. They are frequently asymptomatic and identified incidentally, but can present with intussusception, small bowel obstruction and bleeding that may cause anaemia or may even be overt. Symptomatic lesions can be treated by small bowel resection and anastomosis [5].

Small bowel malignancy is rare. Four major histological types account for over 99% of small bowel malignancies that includes adenocarcinoma, carcinoid tumours, lymphomas and mesenchymal tumours (gastrointestinal stromal tumours [GIST])[6].

Small bowel adenocarcinoma is more often found in the jejunum than the ileum and although the aetiology is unknown, it is more common in patients with CD, coeliac disease, familial adenomatous polyposis, hereditary non-polyposis colon cancer and Peutz–Jeghers syndrome. . Prognosis is poor, particularly in patients with CD, in whom these tumours often present late, because the symptoms are commonly mistaken for those of CD and treated conservatively. Adenocarcinoma of ileum presents with pain and weight loss. Intestinal obstruction and bleeding are seen but perforation is uncommon[7,8].When suspected,

the advised surgical treatment is a resection of 5 cm of non-involved bowel either side of the lesion and the affected mesentery. A right hemicolectomy is likely to be required for tumours of the distal ileum[9,10].

Small bowel lymphoma may be primary or, more commonly, secondary to systemic lymphoma. The incidence of small bowel lymphoma is increased in patients with CD and immunodeficiency syndromes. Hodgkin's lymphoma of the small bowel is rare and most are non-Hodgkin's B-cell lymphomas. It presents as pain, weight loss, vomiting and changes in bowel habits. Perforation may occur in up to 25% of patients.The mainstay of treatment for these conditions is chemotherapy; however, surgery may be required for obstruction, perforation or bleeding[11,12].

Gastrointestinal stromal tumour is a mesenchymal neoplasm most commonly occurring in stomach followed by small intestine. Most common in 5th to 6th decade with slightly Male preponderance. It causes bleeding, anaemia and pain but never obstruction. It grows outside the lumen and large tumors are prone for necrosis and haemorrhage. It rarely shows local lymph node metastasis. Increased size and high levels of c-kit (CD117) staining are associated with malignant potential. Surgery is the most effective way of treating GISTs, as the tumour is radioresistant and is not sensitive to conventional chemotherapy. Imatinib is a tyrosine kinase inhibitor that has been shown to be effective in advanced cases and may also have a role in adjuvant treatment[13,14].

The Ewing sarcoma family of tumors [ESFT] are a high grade small round cell tumors. Most common genetic event in Ewing's sarcoma is a reciprocal translocation between chromosomes 11 and 22, t(11,22), which fuses the Ewing Sarcoma Breakpoint Region 1 (EWSR1) gene of chromosome 22 to the Friend Leukaemia Virus Integration 1 (FLI 1) gene of Chromosome 11 as diagnosed by Fluorescence in situ hybridisation [15]. Mhawech-Fauceglia et al suggested combination of CD99 and FLI1 for identifying EWS/PNET[16].

Carcinoid tumors are neuroendocrine tumours that occur throughout the gastrointestinal tract, most commonly in the appendix, ileum and rectum in decreasing order of frequency. The tumour arises from Kulchitsky cells at the base of intestinal crypts. The primary is usually small, although significant lymph node metastases can occur. They may produce dense fibrosis in the surrounding tissues, resulting in distortion and scarring of the bowel and associated mesentery. Carcinoid tumours can produce vasoactive peptides, most commonly 5-hydroxytryptamine (serotonin).When they metastasise to the liver, the 'carcinoid syndrome' can become evident, because the vasoactive substances escape the filtering actions of the liver. The clinical syndrome itself consists of reddish-blue cyanosis, flushing attacks, diarrhoea, asthmatic attacks and, eventually, pulmonary and tricuspid stenosis[17]. Hepatic resection can be carried out in patients with metastatic disease. The treatment includes the use of octreotide which reduces both flushing and diarrhoea, and octreotide cover is used in patients with a carcinoid syndrome who have surgery to prevent a carcinoid crisis resulting from liberation of vasoactive substances following handling of the tumour[18].

CONCLUSION:

Small bowel tumors have a rare incidence and classically present late, most often diagnosed after surgery for small bowel obstruction. Appropriate evaluation of presenting symptoms helps in early diagnosis and prompt treatment.

CONSENT: Written informed consent was obtained from all 10 patients.

CONFLICT OF INTEREST: The authors declare no conflict of interest.

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