

DOI: <https://doi.org/10.47391/JPMA.1214>

An atypical presentation of small bowel adenocarcinoma; a case report

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Abstract

Small bowel malignancy (SBM) is a rare malignancy in the gastrointestinal tract. Duodenum is the most commonly involved segment and the most common histological subtype is adenocarcinoma (40%). Due to a lack of screening tools and vague symptoms, its clinical detection is very challenging.

A 27-year-old man presented at the surgical emergency of Lahore General Hospital in February 2019 with non-specific gastrointestinal symptoms (vomiting, abdominal pain), for which he had previously visited the hospital multiple times. Later, on further workup, he had been diagnosed as a case of intussusception on CT scan. On exploration, he had an impassable stricture in the jejunum. Resection anastomosis of the jejunum was done, but later, on histopathology it turned out to be adenocarcinoma.

Adenocarcinoma of the small bowel is a rare entity, and, particularly in Pakistan, the available literature is limited. SBM should be included in the differentials of patients with vague abdominal symptoms. Future studies for the evaluation of new investigations and treatment modalities should be encouraged to improve the overall outcome of the patients.

28 **Keywords:** Small bowel malignancy, Intussusceptions, Intestinal obstruction.
 29 Intestinal stricture.

30

31 **Introduction**

32 Small bowel malignancy (SBM) is a relatively rare type of cancer when
 33 compared to other gastrointestinal diseases, with glaring similarities to the said
 34 malignancies. Despite it being rare, primarily of GI origin is 3-6% with only 1-2%
 35 being malignant¹. Mostly it is incidentally detected on histopathology. The most
 36 common histologic subtype is adenocarcinoma (40%), and duodenum is the
 37 most frequently involved segment, followed by jejunum and ileum². Clinically,
 38 SBM presents with vague abdominal symptoms, which, due to inaccessibility
 39 by endoscope and lack of proper screening tools, ordinarily takes 6-10 months
 40 for diagnosis³. There is limited knowledge about the clinical aspects, remedies,
 41 or prophecy of patients with SBM, especially in Asians⁴.

42 In this case report, is presented the history of a 27-year-old patient, who initially
 43 underwent surgery for a preliminary diagnosis of intussusception of the jejunum,
 44 which later turned out to be adenocarcinoma of the jejunum.

45

46 **Case presentation**

47 A 27-year-old man presented in the surgical emergency department of Lahore
 48 General Hospital, in February 2019, with a seven-month history of vomiting,
 49 loss of appetite, right lower abdominal pain and weight loss. These symptoms
 50 had worsened, and he had visited the hospital multiple times during the past
 51 three months. He was now admitted, with a preliminary diagnosis of intestinal
 52 obstruction. On examination, his vitals were within optimal range. He was pale,
 53 and his abdomen showed mild distention with sluggish bowel sounds. There
 54 was a visible sausage-shaped gut impression on the anterior abdominal wall in
 55 the right lower region. His laboratory investigations were well within the
 56 normal range. There were multiple air-fluid levels on abdominal X-ray.

57 Abdominal ultrasound showed dilated gut loops with increased peristaltic
58 activity and free fluid in the abdominopelvic cavity. A preliminary diagnosis of
59 intussusception was made based on CT scan findings. Thus, the patient
60 underwent exploratory laparotomy after informed consent. Per-operatively,
61 there was an unpassable stricture in the jejunum, one foot from DJ, with marked
62 dilation of proximal jejunum and stomach. (Figure1). There were also enlarged
63 mesenteric lymph nodes. Resection and primary anastomosis (figure 2) were
64 done along with a mesenteric lymph node biopsy. His recovery was uneventful,
65 and he was discharged on the sixth post-operative day.

66 The histopathology showed moderately differentiated adenocarcinoma of the
67 jejunum involving the serosa and two lymph nodes (LNs) showed reactive
68 lymphoid hyperplasia (Stage T3NxMx). His metastatic workup was negative,
69 and his case was discussed in a multidisciplinary team (MDT). He was advised
70 re-exploration for lymph node dissection followed by adjuvant chemotherapy.
71 Unfortunately, the patient refused further treatment. But he agreed to have a
72 regular follow up on OPD basis. The patient was advised to have clinical
73 examination and CEA levels every three to six months for the first two years
74 and then every six months for the next three years. CT scan of the chest,
75 abdomen and pelvis six monthly for the first two years and then annually for the
76 next three years were advised.

77

78 **Discussion**

79 Carcinoma of the small intestine is relatively rare as compared to other GI
80 carcinomas. Although it is a significant portion of the GI tract, covering 75% of
81 the length as well as 90% of the total mucosal surface, yet less than 2% of the
82 total GI carcinomas occur in the small intestine⁵. The incidence has been
83 recently increasing, with an annual frequency of 0.3–2.0 cases per 100,000
84 persons, with male predominance.⁶ It is common in the fifth to sixth decades of

85 life and prevalence increases after the fourth decade. But in this case, it occurred
86 in a 27-year-old young man.

87 Several risk factors predispose to small bowel cancer, including genetic,
88 environmental, and medical conditions. Genetic risks include hereditary
89 nonpolyposis colorectal cancer, familial adenomatous polyposis (FAP), and
90 Peutz-Jegher syndrome.⁵ Whether environmental elements cause, small bowel
91 cancer is unknown. There is an increased risk of SBA in alcoholics and
92 smokers.⁴ It is also common amongst the people who use sugar, carbohydrates,
93 red meat and smoked food in large quantities. However, a reduced risk has been
94 observed with higher intakes of coffee, fish, fruit, and vegetables. Diseases like
95 Crohn's disease and Coeliac disease may represent a subset of a pre-existing
96 condition for small bowel cancer. However, in our patient, no such risk factor
97 was present.

98 The clinical features and diagnosis of SBM are usually late. The initial
99 symptoms are typically vague and include abdominal pain and discomfort.⁶ The
100 common presentations include abdominal pain, abdominal distension, bleeding,
101 and jaundice. Bowel obstruction is a common phenomenon in cases of the
102 jejunal and ileal tumour but is less common in proximal duodenal tumours (47%
103 vs 34%; $p = 0.06$). SBM has usually presented as emergencies such as occlusion
104 (40%), bleeding (24%), perforation, and intussusception,⁶ which was also the
105 case in our patient, who had emergency surgery on the suspicion of
106 intussusception.

107 Previously, vague clinical presentation, along with limited sensitivity of contrast
108 studies and abdominal X-rays for small bowel neoplasms, had been the reason
109 for delayed diagnosis. Different tools are widely available now for the diagnosis
110 of SBM, which include Small bowel follow through (sensitivity is 50%), CT
111 scan (accuracy of 47%), capsule endoscopy (sensitivity is between 88.9% and
112 95%) and CT Enterocolysis.⁶ Studies have shown that CT Enterocolysis using

113 spiral and multi-detector CT with an oral contrast agent has matched the
114 radiographic test of choice for SBM, with the sensitivity of 100%.⁷

115 According to the French guidelines, a CT scan of thoraco-abdominal and pelvic
116 cavities to evaluate distant metastases, along with EGD and colonoscopy to
117 exclude synchronous growths, are advised.⁸ CEA and CA 19-9 assay have a
118 prognostic value, especially in advanced disease. Testing for coeliac disease,
119 Crohn's disease, and Lynch syndrome are also recommended in the presence of
120 genetic predisposition.^{7,8} In this case, the patient underwent CT scan of the
121 abdomen preoperatively, which helped in the diagnosis of intussusception. Later,
122 he had a CT scan of the chest to rule out metastasis, which was normal.

123 Sadly, there had been no significant enhancements in consequences and survival
124 in SBM in the last two decades, and the treating modalities have mainly
125 continued unchanged.² Although surgical resection is the treatment of choice for
126 SBM, curative resection of the tumour is possible in only 40-65% of the cases
127 as by the time it is diagnosed; it is too late for cure.² For localised SBM,
128 complete en bloc removal of growth along with locoregional lymph node
129 resection is associated with better outcomes. It is often required even in
130 metastatic disease due to the high probability of obstruction or severe
131 haemorrhage.^{5,9} For unresectable SBM, by-pass surgery as the palliative
132 procedure is a suitable option. Recurrence, even after surgery, is high (40-60%),
133 and it is mainly systemic. Also, until now, no effective adjuvant or palliative
134 regimen has been recommended for SBM.^{9,10} Our patient had resection and
135 anastomosis of the jejunum along with lymph node biopsy. His histopathology
136 confirmed moderately differentiated adenocarcinoma with clear resection
137 margins and reactive lymphoid hyperplasia without any signs of malignancy in
138 the lymph node. Although our patient is doing fine at a six-month follow-up, the
139 prognosis of SBM is poor and correlated to the tumour stage, with a five-year
140 overall survival rate ranging from 14% - 33%.¹⁰

Conclusion

The global prevalence of small intestine adenocarcinoma is rare, and, particularly in Pakistan, the literature available is scarce. This report highlights the need for physicians to broaden their diagnostic vision for the suspicion index for these adenomas in patients with vague abdominal symptoms.

Informed Consent: Patient has agreed for publication of his case report.

Disclaimer: None to declare

Conflict of Interest: None to declare.

Funding Disclosure: None to declare.

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Fig 1: Stricture obstruction.



Fig 2: Jejunum-jejunum anastomosis.