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Mucinous Adenocarcinoma of Jejunum: A Rare Malignancy

Mahendra Lodha[1*], Ashok Kumar Puranik[1], Mahendra Singh[2], Banwari Bairwa[1]
[1]Department of surgery, AIIMS, Jodhpur, India
[2]Department of urology, Sir Ganga Ram Hospital, Delhi, India

***Corresponding author**
Mahendra Lodha

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Abstract: Small bowel malignancy is a rare entity. Adenocarcinoma is a most common histologic subtype. Primary jejunal carcinoma is rare and is difficult to diagnose clinically due to non-specific symptoms. Surgical resection is the treatment of choice. There is no standard protocol for unrespectable or metastatic disease. We report here a 52 year old female with mucinous adenocarcinoma of jejunum, lacking specific symptoms in early stage and later presented with occasional biliary vomiting with soft abdomen, leading to delay of 6 months in diagnosis. At laparotomy, growth was present in mid jejunum. The patient underwent segmental resection of intestine. The jejunal growth was diagnosed as mucinous adenocarcinoma on histopathology examination. The patient was given adjuvant chemotherapy after surgery and she is doing well after 6 months of follow up.

Keywords: Jejunum, Adenocarcinoma, Laparotomy.

INTRODUCTION

Small bowel cancer is a rare malignancy that comprises less than 5 % of all gastrointestinal malignancies, among them mucinous adenocarcinoma is scarcely known in literature. The estimated annual incidence is 0.3–2.0 cases per 100,000 persons, with a higher prevalence rates in the black population than the white, and has been recently increasing [1, 2].

Small bowel cancer classified into four common histological types: adenocarcinoma (30–40 %), carcinoid tumor (35–42 %), lymphoma (15–20 %), and sarcoma (10–15 %) [3,4]. Adenocarcinoma of the small bowel is most commonly located in the duodenum (57 %), while 29 % of cases are located in the jejunum and 10 % in the ileum [5]. The clinical presentation is nonspecific abdominal discomfort, such as abdominal pain, nausea, vomiting, gastrointestinal bleeding and intestinal obstruction, which leads to an average delay of 6–10 months at diagnosis [6]. Due to the rarity of this cancer and nonspecific presentation, there has been no good screening method developed for small bowel adenocarcinoma [6].

CASE PRESENTATION

A 52-years-old female presented in the surgical department with intermittent colicky pain involving whole abdomen for 6 months. Since last 15 -20 days, her abdominal pain has got aggravated and she also complained of occasional greenish vomiting. On general-physical examination, she was normal except mild pallor. Her abdomen was soft and non-tender without any palpable mass. The per rectal examination was normal. She was worked up in our institute by contrast enhanced computerized scan (CECT) of whole abdomen and pelvis, which revealed thickening of a small segment of small intestine probably suggestive of lymphoma or gastrointestinal stromal tumor (Figure 1). After routine work up, she was posted for diagnostic laparoscopy and proceeding. At laparoscopy, we found a small exophytic mass in mid jejunum which was measuring approximately 5x5 centimeters in size with ulceration and necrosis of the outer surface of the tumor, with free fluid and the presence of tumor bits in the pelvis (Figure 2). The rest of the intestine was normal with no gross metastasis to the liver and peritoneum. The decision was made for resection of segment of jejunum and end to end anastomosis after giving small midline incision at umbilical region. The resected segment of intestine and free tumor tissue present in pelvis were sent for histopathological examination. The patient was discharged on the fifth postoperative day and the biopsy came out to be adenocarcinoma jejunum with pathological stage T4N0M0 (Figure 3). After 21 days of surgery, patient was planned for adjuvant chemotherapy. We had a six month of follow-up for this patient and she is doing well.

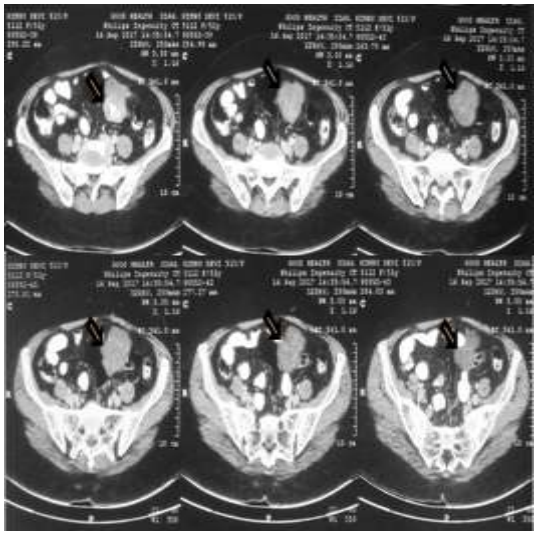

Fig-1: CECT-findings

Fig-2: Resected growth

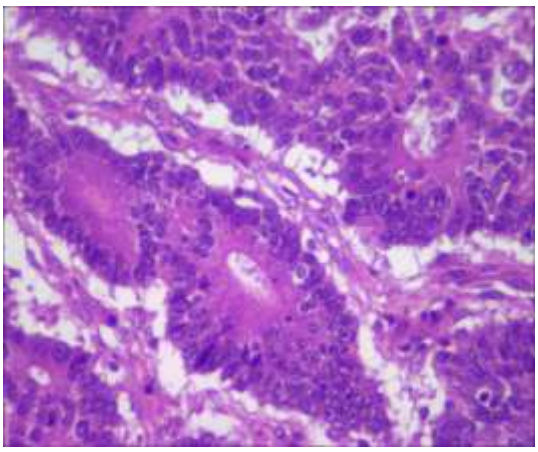

Fig-3: Histopathology suggestive of adenocarcinoma

DISCUSSION

Small bowel comprises 75% of the length of the gastrointestinal tract and 90% of mucosal surface area, but less than 2% of GI malignancies arise from the small intestine [1]. The possible explanation includes the low bacterial colonization, large amount of alkaline fluid secretion, rapid transit time and high concentration of the enzyme e.g. benzopyrene, hydroxylase [7]. The

high Immunoglobulin-A levels cause dilution and detoxification of potential carcinogens, and also prevent prolonged contact of such carcinogens with the mucosa. Small Bowel also has a very limited number of bacteria as compared to the colon that are capable of transforming potential pro-carcinogens in their active breakdown products [7]Small intestine cancers have a high prevalence in western countries as compared to the Asian. Males have a high predilection for these malignancies. Increasing age is associated with higher incidence of small intestinal cancers [2, 8].

The symptoms are initially nonspecific abdominal discomfort; so diagnosis is delayed. The diagnosis of small bowel adenocarcinoma is mainly obtained at advanced stages; ~40 % of patients have lymph node metastasis (stage III), and 35 to 40 % have distant metastasis (stage IV) [5, 9]. In late stages, it may present as obstruction (40 %) or bleeding (24 %) [9]. our patient was a female and the mass remained undetectable until she developed small bowel obstruction.

The diagnosis of small bowel carcinoma may be elusive. Upper GI series with small bowel follow through shows abnormalities in 53-83% of patients with small bowel cancers. An abdominal CECT scan will reveal the exact site and extent of local disease as well as the presence of liver metastasis. The accuracy of abdominal CT in detecting primary small bowel tumors is poor, reported as 57% [8]. Upper GI endoscopy with small bowel enteroscopy may allow identification and biopsy of lesions in the duodenum and jejunum. Capsule endoscopy has become more sensitive and specific diagnostic method for small bowel disease. It has increased the diagnostic rate of small bowel tumors, most of them are detected as cases of obscure gastrointestinal bleeding, with 50–60% of these being malignant. The capsule endoscopy has overcome the low sensitivity of barium studies (30–50%), and the discomfort and complexity of enteroscopy [9].

The PET/CT scan technique is being used to differentiate small intestinal malignant tumors from benign. The uptake of 18F-FDG is related to tumor size, infiltration and lymph node metastasis; the higher the uptake of 18F-FDG, the higher the tumor invasiveness [10] a baseline plasmatic assay of CEA and CA 19-9 is necessary, especially in patients with advanced disease because CEA and CA 19-9 levels are of prognostic value [11].

The treatment of choice is the surgical resection with clear margins and regional lymph node resection in localized small bowel adenocarcinoma; indeed, it is often required even in metastatic tumor due to the high probability of obstruction or severe hemorrhage [12]. Still, there has been no standard chemotherapy regimen against small bowel adenocarcinoma. Several studies have explored the role of palliative chemotherapy in advanced small bowel tumors. Hong et al. have been shown in stage IV patients who received palliative chemotherapy that overall survival (OS) increased significantly compared to those who did not receive chemotherapy (8 vs. 3 months, $p = 0.025$) [13]. In limited clinical reports, a combination of fluoropyrimidine with a platinum compound (FOLFOX or CAPOX) reported as first line palliative chemotherapy in metastatic malignancy of small bowel treatment [13, 14]. Zaanan *et al*. have been shown that median overall survival in advanced small bowel malignancy patients treated with FOLFOX was 17.8 months, the longest survival among different chemotherapy regimens [14].

CONCLUSIONS

We report a rare case of mucinous adenocarcinoma of jejunum in elderly women. Due to the non-specific clinical presentation, inaccessibility to endoscopic instruments, often diagnosis is delayed and has a poorer prognosis. Surgery remains the mainstay of treatment. The role of chemotherapy is in palliative treatment and not as a primary treatment. Incidence of small bowel malignancy is very low; there is a need for further evaluation of diagnostic and therapeutic strategies.

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