

Linitis Plastica of Terminal Ileum

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ABSTRACT

Linitis plastica generally involves stomach. Most common cause is diffuse signet ring adenocarcinoma, metastatic disease from breast and lungs can also lead to Linitis plastica and also caustic burns. In Linitis plastica the stomach becomes rigid and pipe like (leather bottle). We are presenting a rare case of Linitis plastica of terminal ileum. Ours is a 55-year female patient presenting in casualty with abdominal pain, distension, vomiting and bilious vomiting. Her terminal ileum had stricturous rigid tube-like lesion with multiple mesenteric lymphadenitis. We did curative resection (ileoascending colon) and anastomosis. She had uneventful recovery. Though small bowel malignancies are rare, one should have high index of suspicion when one encounters rigid pipe like strictures lesion of ileum, as these can be malignant lesion and one should perform curative resection. Surgery improves survival rate along with adjuvant chemotherapy as required.

Key Words: Linitis plastica, small bowel malignancy, Linitis plastica ileum, Linitis plastica stomach.

INTRODUCTION

Signet ring cell carcinoma (SRCC) is a rare highly aggressive malignant adenocarcinoma that generally involves the stomach, ileal involvement is very rare. We report a case of Linitis plastica of ileum (SRCC) in an elderly female, very few cases are reported in literature about Linitis plastica of ileum. Signet-ring cell carcinoma is defined according to the WHO's classification as a poorly cohesive carcinoma composed predominantly of tumour cells with prominent cytoplasmic mucin and a crescent-shaped nucleus eccentrically placed.⁽¹⁾

CASE REPORT

A 55 yrs. female patient came to casualty with complaints of abdomen pain for 2-3 months, which was more severe for the last 8-10 days. It was associated with abdomen distension, constipation & bilious vomiting (postprandial). There was no history suggestive of chronic inflammatory

bowel disease or Crohn's disease. No history suggesting per rectal bleeding. No past surgical history. Patient was known diabetic, hypertensive and asthmatic on regular medication.

On general examination, patient was having pallor, tachycardia with low volume pulse. On per abdomen examination, abdomen was distended, tense with generalized tenderness, no lump was palpable. Hyperperistalsis was observed. Rectum was empty.

X-ray erect abdomen revealed air fluid levels with dilated small bowel loops. USG abdomen revealed small bowel obstruction (ileal).

On emergency exploration, distal 8cms of ileum was stricturous with rigid pipe like till ileocecal junction with multiple mesenteric lymphadenopathies. (figure 1) Ileo-ascending colon resection and anastomosis was done. Histopathology revealed signet ring cell carcinoma of ileum

PT3pN0M0 (stage II A). (Figure 2.) Patient

had uneventful post-operative period.



FIGURE 1

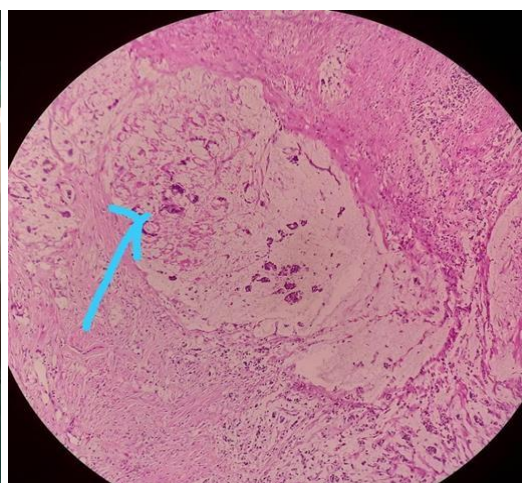


FIGURE 2

DISCUSSION

Malignant neoplasms of the small bowel are among the rarest types of cancer, accounting for 1-2% of all GI cancers despite composing 75% of bowel length and 90% of surface area of mucosa⁽²⁾. Rates of small bowel cancer are increasing an average of approximately 2% each year during last 10 years. Around 98% of small-bowel tumors are made up of adenocarcinomas, carcinoid tumors, lymphomas, sarcoma (most commonly leiomyosarcoma and more rarely angiosarcoma or liposarcoma) and gastrointestinal stromal tumors (GISTs)⁽³⁾.

Most of the malignancies of small bowel will be diagnosed very late as they don't show any earlier symptoms and patients present in advanced stage⁽⁴⁾. Only 31% patients are diagnosed with local disease. 5-year survival for localized small bowel cancer is about 83%. Patient with regional disease has 71% and distant disease has 40% survival⁽²⁾. Mean age for malignant small bowel cancer is 57 years⁽²⁾. Incidence of small bowel cancer is very low in India.

Approximately 40% of small bowel malignancies are adenocarcinomas and it is the most common histologic type⁽²⁾. Adenocarcinomas are most commonly seen in duodenum, jejunum. Ileum being the least common site for adenocarcinoma. Neuro

endocrine tumors account for 25 to 30% of small bowel malignancies⁽²⁾.

Linitis plastica also called leather bottle is a morphological variant of diffuse infiltrating carcinoma most commonly seen in stomach, there are very few Linitis plastica reported in ileum. Linitis plastica as a type of adenocarcinoma in stomach accounts for 3 to 19% of gastric cancer⁽⁵⁾, rarely cases may be due to metastatic infiltration from breast and lung carcinoma⁽³⁾. Non-malignant conditions like caustic injury to stomach also known to cause Linitis plastica. In our case it is the diffuse infiltrating adenocarcinoma, which has caused Linitis plastica of ileum.

Pre disposing medical conditions for small bowel malignancies are Crohn's disease, celiac disease, familial adenomatous polyposis, hereditary nonpolyposis colorectal cancer, Peutz-Jeghers syndrome and lynch syndrome.^(7, 8) Mutation of the KRAS gene is commonly found in small bowel neoplasm. Allelic losses, particularly involving tumor suppresser genes at chromosome location 5q, 17q, 18q, DCC, DPC4 have been noted in small bowel cancer.

Recent finding demonstrated that approximately 15% of adenocarcinoma has inactivation of DNA mismatch repair genes and display a high level of microsatellite

instability. MSI-H is associated with celiac disease and carcinogenesis.

Ileal tumors are more likely to develop intestinal obstruction than jejunal tumors⁽⁹⁾. Due to insidious nature of small bowel malignancies (symptoms may include dyspepsia, anorexia, malaise, dull abdominal pain and colicky pain) high index of clinical suspicion must be present for early diagnosis of these neoplasms.

Workup procedures such as upper gastrointestinal follow through studies, CT and MRI enteroclysis, push enteroscopy, colonoscopy with retrograde ileoscopy, video capsule endoscopy & double balloon enteroscopy can be used pre-operatively for appropriate diagnosis⁽¹⁰⁾.

The treatment of choice is wide resection of the small bowel segment harbouring the carcinoma as well as resection of the corresponding mesentery and lymph nodes⁽³⁾. Surgery improves the survival rate.

Long term monitoring includes follow-up visit in the outpatient setting every 3 months to assess for symptoms or signs suggestive of recurrent disease. Periodic CBC and LFT to check for anaemia & hepatic Metastasis. Abdomen CT, PET CT and colonoscopy screening should be done every 6 monthlies to identify subclinical recurrent disease early.

Recommended post-op chemotherapy in present case is with Capecitabine, 5-fluorouracil (5FU), Oxaliplatin, Irinotecan.

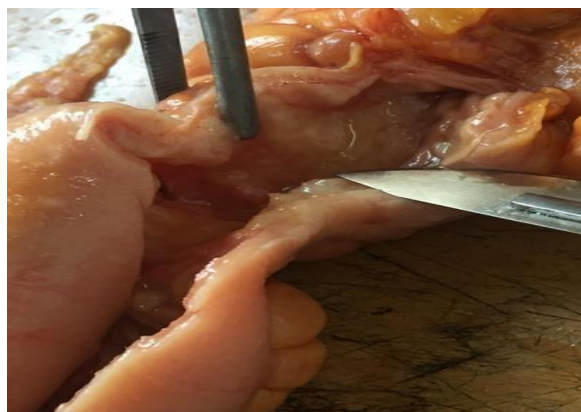


Figure 3

CONCLUSION

In cases of small bowel obstruction due to rigid pipe like strictures, one should have high index of suspicion of malignancies like signet ring adenocarcinoma & do curative resection, to give better prognosis. (Figure 3.)

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