

Case Report

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Atypical presentation of small bowel lymphoma as surgical emergency

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ABSTRACT

Gastrointestinal tract is the most frequent site of extranodal involvement by non-Hodgkin's lymphoma, though extranodal lymphoma may arise anywhere outside lymph nodes, stomach is the most commonly involved organ followed by the small intestine, pharynx, colon, and rarely, esophagus. 26 year old male with history of pain abdomen for 3 days, involving whole abdomen, associated with bilious vomiting, non passage of flatus and stools for 2 days. Patient also had multiple spikes of high grade fever associated with chills and rigors for 2 days. Abdominal examination revealed features of peritonitis. CXR revealed free gas under diaphragm. Patient planned for emergency exploratory laparotomy. Intraoperatively stricture perforation proximal to ileocaecal junction noted. Resection of stricture segment with ileo-ascending anastomosis performed. Biopsy revealed Diffuse large B cell lymphoma. 52 year male with history of central abdominal distension for 4 days with multiple episodes of bilious vomiting and non passage of flatus and stools for 2 days. Abdominal examination revealed marked abdominal distension with generalised tenderness and exaggerated bowel sounds. Abdomen X Ray revealed multiple air fluid levels. Intraoperatively growth of 8×5 cm present 10 cms proximal to ileocaecal junction. Resection of diseased segment with ileoascending anastomosis performed. Biopsy of resected segment revealed Diffuse large B cell lymphoma. Patient started on R-CHOP regimen, doing well. Here we present two cases of Diffuse large B cell lymphoma showing bimodal presentation with rare clinical presentation, one as a perforation and other as obstruction.

Keywords: Lymphoma, Acute abdomen, Emergency

INTRODUCTION

Gastrointestinal (GI) tract is the most frequent site of extranodal involvement by non-Hodgkin's lymphoma, though extranodal lymphoma may arise anywhere outside lymph nodes, stomach is the most commonly involved organ followed by the small intestine.^{1,2}

Primary lymphoma of the GI tract, as a single site, should be limited to the tract with no peripheral lymphadenopathy. Nodal involvement if present, is usually limited to the drainage area of the segment of the

GI tract which is involved while liver and spleen remain unaffected.³

Secondary involvement of the GI tract with lymphoma is common as mesenteric or retroperitoneal lymph nodes. They are common origins of lymphoma sharing their lymphatic drainage with lymphoid tissue in the GI tract.⁴

Symptoms usually depend on the site of involvement, but can include dysphagia, abdominal pain, nausea, vomiting, anorexia, weight loss, diarrhea, GI bleeding, malabsorption, and diarrhea, all being nonspecific. There

may be a palpable mass. Uncommonly, there may be small bowel obstruction. Bleeding, fever, and small bowel obstruction are poor prognosticators. Treatment for GI tract lymphoma include chemotherapy, radiation therapy, stem cell transplant, and antibiotic treatment for *H. pylori*.⁵

Here we report the varied presentation of Non Hodgkin Lymphoma involving the distal ileum with bimodal age presentation.

CASE REPORT

Case 1

A 26 year old gentleman presented to the surgical emergency with history of pain abdomen for 3 days, involving whole abdomen. Pain was associated with bilious vomiting with frequency of 5-7 episodes per day and non passage of flatus and stools for 2 days. Patient also had multiple spikes of high grade fever associated with chills and rigors for 2 days.



Figure 1: Case 1 intraoperative 3x3 cms perforation 10 cms from ICJ.

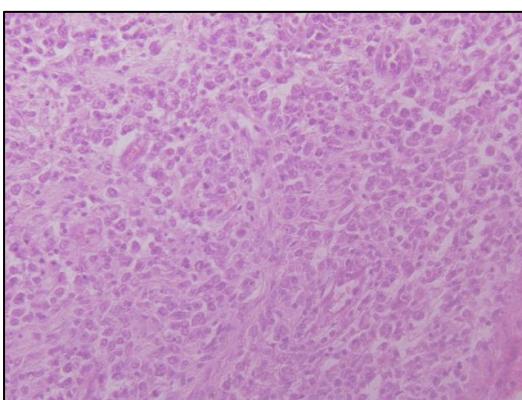


Figure 2: Case 1 histopathology.

On examination patient had Hippocratic facies. He was severely dehydrated, tachycardic and hypotensive and had decreased urine output. Abdominal examination

revealed features of peritonitis with guarding, rigidity, generalised tenderness and rebound tenderness with absent bowel sounds. On chest auscultation, coarse crepitations were heard in bilateral lower zones.

Hemoglobin was 10.7 and total leukocyte count 20,220. Liver function tests and kidney function tests were in normal range. Radiological investigation revealed free gas under diaphragm.

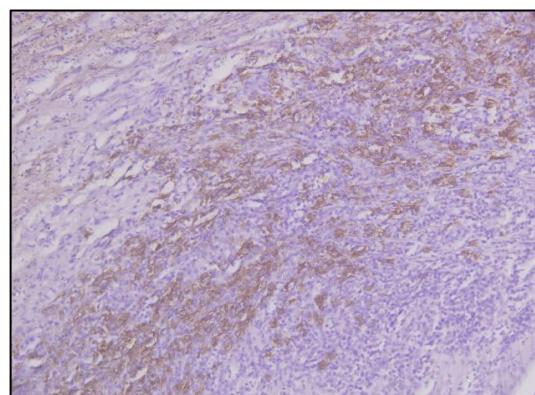


Figure 3: Case 1- immunohistochemistry.

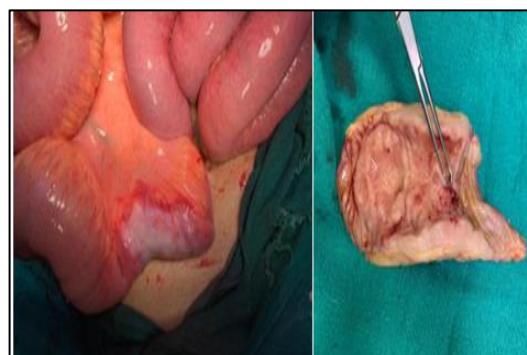


Figure 4: (A) Case 2 intraoperatively a growth of about 8x5 cms present 10 cm proximal to ICJ junction with a non negotiable stricture 4 cm proximal to ICJ and (B) cut section showing a growth.

Patient was resuscitated with IV fluids, IV antibiotics administered and was planned for emergency exploratory laparotomy.

Intraoperatively a stricture perforation of 3.0x3.0 cm found at about 10 cm from the ileocaecal junction (Figure 1). Resection of the stricture segment with ileo-ascending anastomosis was performed. Patient was shifted to ICU postoperatively in view of poor chest condition. On post operative day 5 patient had an anastomotic leak for which he was re explored but he succumbed to his illness. The biopsy of the resected segment revealed Diffuse large B cell lymphoma with resection margins being free of tumour with no isolated lymph nodes. Tumour cells were positive for CD20 and negative for CD3. (Figure 2, 3)

Case 2

A 52 year male presented to the surgical emergency with history of central abdominal distension for 4 days with multiple episodes of bilious vomiting. He also gave history of non passage of flatus and stools for 2 days. On examination patient's vitals were stable.

On abdominal examination, there was marked abdominal distension with generalised tenderness and bowel sounds were exaggerated. Per rectal examination revealed rectal ballooning. Other organ systems examination was normal.

Complete blood count, liver function test and kidney function tests were normal range. Abdomen X Ray revealed multiple air fluid levels. In view of these findings, patient was prepared for exploratory laparotomy.

Intraoperatively a growth of about 8×5 cm was present 10 cm proximal to ileocaecal junction with a non negotiable stricture about 4 cm proximal to ileocaecal junction (Figure 4a, 4b). Resection of diseased segment with ileoascending anastomosis was performed. Patient's post operative course was uneventful. Biopsy of the resected segment revealed Diffuse large B cell lymphoma with tumour cells positive for LCA and CD20.

Patient started on R-CHOP regimen consisting of Rituximab, Cyclophosphamide, Doxorubicin, Vincristine and Prednisolone. Patient is doing well.

DISCUSSION

Here we present two cases of Diffuse large B cell Lymphoma showing bimodal presentation with rare clinical presentation, one as a perforation and other as obstruction. A thorough literature review revealed some rare cases of lymphoma presenting with small-intestinal perforation.⁶⁻⁸

Analysis of the National Cancer Data Base and Surveillance Epidemiology and End Results database revealed a 17.3 % incidence of small bowel lymphoma. Primary gastrointestinal lymphoma, however, is very rare, constituting only about 1-4% of all gastrointestinal malignancies.

Diffuse large B-cell lymphoma is the most common type, representing about one third of all cases. Diffuse large B-cell lymphoma (DLBL) occurs in the gastrointestinal tract, both as de novo disease and following previous low-grade MALT lymphoma.

DLBCL was the most common lymphoma that perforated. The most common site of perforation was the small bowel (59%), followed by large bowel (22%) and gastric (16%). Despite latest imaging methods and effective chemoimmunotherapy, perforation of GI tract

lymphomas is an important clinical complication. In a large single-institution study, the rate of perforation in patients with biopsy-proven GI tract lymphomas was determined to be 9% with nearly half the perforation events representing the initial presentation of GI lymphoma.¹⁰ Patients presenting with obstruction present most commonly with a mass at the ileocaecal junction causing luminal narrowing. Ileocaecal junction being most common due to presence of Peyer's patches.

Thus all lymphoma patients with new abdominal pain should receive a prompt physical examination and evaluation for free air in the abdomen with appropriate imaging. PET-CT scan has been shown to have a greater sensitivity (97%) and specificity (100%) for staging of NHL and is superior to conventional CT scan in detecting recurrence and monitoring the response to chemotherapy.¹³ A delay in diagnosis and treatment results in increased morbidity and mortality.

Classically, small-bowel lymphoma most commonly arises in the ileum. Considering the standard treatments, localized small-intestinal lymphoma, should be treated with segmental resection of the involved intestine and adjacent mesentery. If the small intestine is diffusely affected by lymphoma, chemotherapy rather than surgical resection should be the primary treatment.^{9,10} Zinzani et al reported better outcome with surgical resection followed by chemotherapy in patients with limited disease.¹¹ Ibrahim et al. also noted improved event-free survival with resection followed by chemotherapy in 66 patients with intestinal DLBCL, although the effect on overall survival was not significant.¹²

CONCLUSION

Small Bowel Lymphoma among its various presentations can present as acute abdomen. Hence in patients with relevant clinical history and examination a differential diagnosis of the same must be considered for better patient management and outcomes.

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