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Case Report

A rare case of diffuse large B cell lymphoma presenting as jejunal perforation

Althaf Ahmed*, Philip Umman

Department of General Surgery, Believers Church Medical College, Tiruvalla, Kerala, India

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***Correspondence:**
Dr. Althaf Ahmed,
E-mail: althafalways9@gmail.com

ABSTRACT

Diffuse large B-cell lymphoma can present as either primary lymph nodal disease or with involvement of extra-nodal sites. In gastrointestinal involvement, the clinical presentation is often insidious, such as abdominal pain, nausea, vomiting, anemia, weight loss, fever, and occasionally small bowel obstruction or a palpable mass. Small-bowel perforation as the first manifestation of non-Hodgkin's lymphoma is extremely rare. The case of a 73-year-old woman is reported who presented to the emergency department complaining of a generalized abdominal pain and vomiting. Contrast enhanced computer tomogram (CECT) of the abdomen showed multiple air pockets in left side of abdomen, right hemidiaphragm, edematous small bowel loops with fat stranding with multiple tiny air pockets. Laparotomy was performed that revealed a focally perforated segment of the jejunum. Histological evaluation reported diffuse large B-cell non-Hodgkin's lymphoma. This case illustrates a rare complication of large B-cell non-Hodgkin's lymphoma of the jejunum that was responsible for small-bowel perforation and peritonitis. This clinical pattern may appear as a leading presentation of small intestinal neoplastic disease in which surgical resection may have to be considered.

Keywords: Gastrointestinal tract, Non-Hodgkin's lymphoma, Peritonitis, Small-bowel perforation

INTRODUCTION

The gastrointestinal (GI) tract is the most common extra-nodal location for non-Hodgkin's lymphoma (NHL) and accounts for up to 20% of cases. Involvement may be seen from the pharynx to the rectum.[1] A world standardized incidence of 1 case in 100,000 individuals has been estimated. The incidence is increasing, mainly because of various environmental and exogenous factors, especially the increasing incidence of HIV infection.[2]

Primary GI tract lymphoma is defined as a tumour that predominantly involves the GI tract with lymph node involvement confined to the drainage area of the primary tumour site. There is no liver or spleen involvement and palpable lymph nodes are not present. Chest radiography and peripheral white blood cells are normal.[2] One analysis of the National Cancer Data Base and Surveillance Epidemiology and End Results database found the following percentage distribution of histologic subtypes of small bowel cancer: carcinoid (37.4%), adenocarcinoma (36.9%), lymphoma (17.3%) and gastrointestinal stromal tumor (8.4%).[3] Primary gastrointestinal lymphoma, however, is very rare, constituting only about 1%-4% of all gastrointestinal malignancies.[4]

Differentiation between primary GI lymphoma and systemic lymphoma with GI tract involvement has important implications because the prognosis is better in the primary form when diagnosed early on, with five-year survival rates as high as 62 to 90 percentage.[2] In the western world, GI tract lymphoma, whether primary or systemic, most often involves the stomach (50%),

followed by the small intestine (33%), colon (10-16%), and esophagus (1%). NHL of the GI tract is more common in men than in women (3:2).[2] Although it is predominantly a disease of middle-aged persons, there is a double peak, with the first peak in patients less than 10 years of age and the second at a mean age of 53.[2]

Table 1: Features of small-intestinal malignancies.

Tumor type	Cell of origin	Frequency	Predominant site
Adenocarcinoma	Epithelial cell	35-50%	Duodenum
Carcinoid	Enterochromaffin cell	20-40%	Ileum
Lymphoma	Lymphocyte	10-15%	Ileum
GIST	Interstitial cell of Cajal	10-15%	-

Diffuse large B-cell lymphoma is the most common type of non-Hodgkin's lymphoma, representing approximately one third of all cases.[5] This lymphoma makes up the majority of cases in previous clinical trials of "aggressive" or "intermediate- grade" lymphoma.[5] Diffuse large B-cell lymphoma (DLBL) occurs in the gastrointestinal tract, both as denovo disease and following previous low-grade MALT lymphoma.

Symptoms will depend on the site of involvement, but may include dysphagia, abdominal pain, nausea, vomiting, anorexia, weight loss, diarrhea, GI bleeding, malabsorption, and diarrhea, all of which are nonspecific.[2] There may be a palpable mass.[2] Uncommonly, there may be small-bowel obstruction. Small bowel involvement usually presents as an acute emergency and is rarely diagnosed electively. Bleeding, fever, and small bowel obstruction are poor prognosticators.[2] Treatment modalities for GI tract lymphomas include chemotherapy, radiation therapy, stem cell transplant, and antibiotic treatment for H. pylori.[2] Recently, PET CT has been increasingly used for both staging and treatment follow-up in patients with lymphoma.

CASE REPORT

A 73-year-old woman presented to the emergency department with a one-day history of sudden onset abdominal pain in the left upper quadrant. She is a diabetic on oral hypoglycemics. She was hemodynamically stable. On abdominal examination, generalized tenderness and guarding was present. Chest x-ray was non-contributory. Ultrasonogram of the abdomen revealed moderate free fluid. A CECT abdomen showed multiple air pockets in left side of abdomen, right hemidiaphragm, edematous small bowel loops with fat stranding with multiple tiny air pockets were revealed.

On laparotomy, a pin point perforation of size around 0.2cms, nearly 20 cms from the duodenojejunal flexure with surrounding induration of the jejunum with extension of the induration into the mesentery for around 2cms was noted. Another intramural swelling was palpable about 1 feet distal to the perforation, on the mesenteric border. A resection of the perforated segment of jejunum encompassing the perforation proximally and the intramural swelling was done.

Hematoxylin and eosin staining of the histological examination showed extensive infiltration of large cells with pleomorphic vesicular nuclei, conspicuous nucleoli, and pale eosinophilic cytoplasm deep in the mesenteric adipose tissue, which was compatible with non-Hodgkin's lymphoma of the small bowel. Immunohistochemical analysis revealed atypical cells positive for CD20 and Bcl-6, Ki 67, MUM and negative for CD3, CD30, ALK, and cytokeratin. Therefore the diagnosis of diffuse large B-cell non-Hodgkin's lymphoma was made.

DISCUSSION

Study report the case of a woman who presented with small-bowel lymphoma resulting in perforation of a jejunal segment and subsequent peritonitis. A thorough literature review revealed some rare cases of lymphoma presenting with small-intestinal perforation.[6-8] In the presented case an intramural mass about 1 feet distal to the perforation was palpable with erosion of the swelling onto the mucosal surface of the jejunum resulting in perforation of the adjacent thickened jejunum was found.

Classically, small-bowel lymphoma most commonly arises in the ileum. Considering the standard treatments, localized small-intestinal lymphoma, as in our case, 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]

However, the value of adjuvant chemotherapy after resection of localized lymphoma is controversial.[9,10] A pin point perforation 20cms from the duodenojejunal flexure and surrounding induration of the jejunum with another intramural swelling 1 feet distal to the perforation, on the mesenteric border.

Figure 1: Pin point jejunal perforation with surrounding induration; note the intramural swelling distal to the perforation site.

In our patient, based on her history and physical examination that confirmed the diagnosis of acute surgical abdomen (generalized peritonitis), a decision was made to proceed to an explorative laparotomy. Our patient was discharged in good general condition and was healthy on regular postoperative visits.

CONCLUSION

Large B-cell non-Hodgkin's lymphoma of the jejunum may present as small bowel perforation. In the emergent setting the treatment is directed to control of sepsis and resection of the involved segment. Adjuvant chemotherapy should be considered after recovery.

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