



A CURIOUS CASE OF NON HODGKIN'S LYMPHOMA OF SMALL INTESTINE PRESENTING AS PERFORATION PERITONITIS

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KEYWORDS :

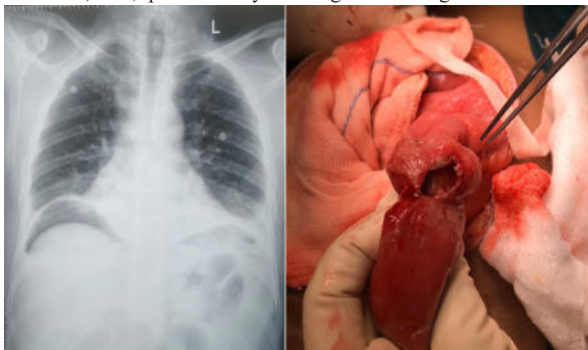
INTRODUCTION:

Malignant lymphomas involve small intestine either primarily or as a manifestation of systemic disease. It is rare for Non Hodgkin's lymphoma to affect small intestine and most western type lymphomas are Non Hodgkin's B cell lymphomas. Here we are reporting a rare case of 40 year old male presented to the emergency room with acute abdomen and subsequently found to have non Hodgkin's lymphoma of small intestine presenting as perforation peritonitis. Patient has recovered well from holistic treatment approach and presently receiving chemotherapy under medical oncology.

Case Summary

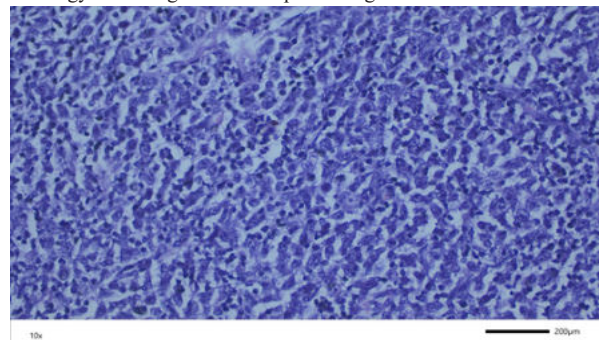
40 year old male patient presents to ER with complaints of pain abdomen for 8 days & it was colicky type gradually progressive in nature. Pain had started initially in the left iliac region and 2 days ago, it increased in intensity acutely and spread diffusely to entire abdomen. It was associated with non bilious vomiting. Patient had history of smoking for 10 years (half pack per day). There were no known comorbidities. At presentation, patient was in a state of hypovolemic shock with cold clammy hands.

Patient was immediately resuscitated with intravenous fluids and started on ionotropes. Abdominal examination revealed diffuse generalised distension with diffuse tenderness and guarding along with absent bowel sounds. X-ray erect abdomen showed free air under diaphragm indicating hollow viscus perforation. After adequate stabilisation, patient was taken up for emergency laparotomy. On exploration, 2.5cm x2.5cm perforation was found in the ileum along the anti mesenteric border, 70cm proximal to ileo-cecal junction. At the site of perforation, there was circumferential thickening of 3x4 cm with luminal narrowing suspicious of malignancy. However rest of the small intestine stomach, colon and rectum appeared normal. Also, there was no mesenteric lymphadenopathy, ascites or deposits in the omentum, liver, spleen and any other organ indicating metastasis.



Entire peritoneal cavity was contaminated with 2 liters of thin bilio-faecal fluid. Ileal segment with perforation site was resected with 10 cm margin on either side along with mesentery followed by end to end anastomosis. The resected specimen was sent for histopathological examination. Patient did not improve as expected after the surgery and was showing signs of prolonged paralytic ileus which increased on POD 7, with increased distension of abdomen indicating anastomotic leak/kink, for which he was taken for re-laparotomy. On re-exploration, the anastomotic site was found to be intact. However there were early post operative adhesions at the distal ileum with grossly

distended bowel loops. Adhesiolysis with loop ileostomy was done and bowel loops were decompressed. Post operatively, patient was on ventilator support because of poor respiratory efforts. Patient did not respond to weaning of ventilator because of ARDS changes till 3rd postoperative day. He was started on polymixin B based on ET tube culture and sensitivity report. Patient gradually weaned off from ventilator support and shifted out of ICU. Meantime, histopathology report came out as a surprise showing features of non Hodgkin's lymphoma. Medical oncologist's opinion was taken and was advised to send for IHC markers which was reported as large Bcell type of Non Hodgkin's Lymphoma with atypical lymphoid cells, positive for CD26, bcl-6, MUM-1 ; negative for BCL-2 and CD10. Patient developed surgical site infection on POD7. Pus was sent for culture and sensitivity and was treated with reg- ular bed side dressings and antibiotics according to sensitivity. Patient was also sup- plemented with total parenteral nutrition for severe weight loss. Patient improved clinically and started mobilising. He was then discharged and referred to medical on- cology for further management. Unfortunately patient developed stomal prolapse and was planned for ileostomy closure after 12 weeks. However, as patient had developed severe bone marrow suppression he was found to be unfit for anaesthesia and surgery was deferred. Presently patient is undergoing chemotherapy under medical oncology and on regular follow up with surgical team.



DISCUSSION

Despite the fact that the small bowel represents 75% of the total length and over 90% of the mucosal surface of the intestinal tract, malignant tumours of the small intestine account for less than 1% of all the malignancies. Primary gastrointestinal lymphoma(GIL) as compared to all GI malignancies is very rare, constituting only about 1%-4% of all gastrointestinal malignancies. Small intestine is unique in being highly resistant to GIL , because of the most obvious reasons such as bacteriological factors and the rapid passage of its liquid contents[1]. The most common pathological type of gastrointestinal lymphoma is Diffuse Large B-cell Lymphoma, even though other types have been shown to increase recently.

Clinical presentation of intestinal lymphomas may vary from nonspecific abdominal complaints like fatigue, malaise, weight loss and abdominal pain. Over a period of time, patients may develop complications like malabsorption, obstruction or palpable mass.

Perforation and peritonitis are known and dreaded complications of GIL that can occur either at diagnosis or during the course of treatment. [2]

A study Group on intestinal NHL reported a lesser incidence of perforation in in- testinal B-cell lymphomas compared with T cell lymphomas[3].

In small bowel lymphomas CT scan may show findings like mass, bowel wall thick- ening, displacement of adjacent organs or luminal obstruction. Dawson's criteria are used for labelling primary gastrointestinal lymphoma(GIL), that include (1) absence of peripheral lymphadenopathy at the time of presentation; (2) lack of enlarged medi- astinal lymph nodes; (3) normal total and differential white blood cell count; (4) pre- dominance of bowel lesion at the time of laparotomy with only lymph nodes obvious- ly affected in the immediate vicinity; and (5) no lymphomatous involvement of liver and spleen [4]. Ann Arbor staging is commonly employed to stage gastrointestinal lymphoma and the international prognostic index has been used to define the prog- nostic subgroups. Accurate staging and diagnosis of gastrointestinal lymphoma are important for planning the treatment in this heterogeneous group of lymphoma [5]. [Table. 1.0]: Modified Ann Arbor Classification of Primary Non-Hodgkin's Gastroin- testinal Lymphoma

^I E	Tumor confined to small intestine without Nodal Involvement
^{II} E	Regional Lymph node involvement
^{III} E	Involvement of non resectable lymph nodes beyond re- gional basin
^{IV} E	Spread to other nonlymphatic organs

[Table. 2.0]: Staging of gastrointestinal non-Hodgkin's lymphoma (GI-NHL) according to the In- ternational Workshop

I	Tumor confined to the gastrointestinal (GI) tract (Single pri- mary site or multiple non-contiguous lesions)
II	Tumor extending in abdomen from primary GI siteIII - Local (paragastric or paraintestinal)II2 - Distant (mesenteric, para- aortic, paracaval, pelvic, inguinal)
III	Penetration of serosa to involve adjacent organs or tissues
IV	Disseminated extranodal involvement or a GI tract lesion with supradiaphragmatic nodal involvement

Morbidity and mortality of intestinal lymphoma presenting with perforation is found to be high, as the perforation may go unrecognised until shock follows peritonitis [6]. The time interval from the onset of symptoms caused by the perforation to the time of operation can affect the outcome. B cell lymphomas are more chemo sensitive than T cell lymphomas and have high remission rates with or without surgeries.

Treatment of localised (stage I_E and stage II_E) lymphoma involves resection of the in- volved segment and adjacent mesentery. Stage III_E and stage IV_E is treated with chemotherapy. The role of adjuvant therapy after curative resection for stage I_E and stage II_E lymphomas is controversial. The 5-year survival in patients with resectable lymphoma approaches 80%. Surgery plays a limited role in diffuse cases although it may be required for accurate diagnosis. Lymphoma primarily located in the small in- testine usually warrants laparotomy with the affected segment removed both for its diagnosis and for its treatment. Low-grade B-cell lymphoma of the small intestine (stage IE) only requires surgical resection[7].

Sakakura et al reported that perforation is the most impending complications in pa- tients with Non Hodgkin's lymphoma undergoing systemic chemotherapy [8]. One study done by Ara, C., Coban, S., Kayaalp, C. et al. states that early diagnoses and treatment are necessary to improve the prognosis in non Hodgkin's lymphoma and that possibility of intestinal lymphoma should be kept in all cases of intestinal perfo- rations[9].

Dughayli, M.S et al. states that the prognosis of synchronous primary lymphoma in the small and large bowel correlates better with the depth of invasion, tumor size, and lymphadenopathy. A higher percentage of perforations occurred in the small bowel (59%), compared with the stomach (16%) or large bowel (22%) [10].

Ara et al. reported similar findings in their case series of eight patients with bowel perforations from lymphoma, where six of the eight perforations occurred in the small bowel and only two involved the large bowel because the larger lumen of the colon, which delays obstruction leading to lower tendency (4%) of large intestinal lymphomas to produce abdominal emergencies due to perforation and bleeding [9].

CONCLUSION:

One of the uncommon complications of GIL that can occur either at diagnosis or dur- ing the course of treatment is perforation and peritonitis. The incidence of perfora- tions is life threatening and can lead to morbidity from wound infection, increase in duration of hospital stay, SIRS, MODS undue delays in chemotherapy and ultimately mortality. Many studies have reported cases of perforation in case of Non hodgkins lymphoma while receiving chemotherapy but here this case study reports the unusual incidence of the Non Hodgkin's lymphoma presenting as perforation where in-time surgical resection and appropriate systemic chemotherapy afterwards saved the pa- tient from significant morbidity and mortality.

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