PERFORATION OF SMALL BOWEL LYMPHOMA - A CASE REPORT

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Abstract:
Malignant tumors of small bowel contribute only 1.1 to 2.4 of all gastro intestinal tract malignancies. Among these, lymphomas contribute 10 -15. Of all lymphomas primary Gastrointestinal Lymphomas account for 5, of which one third occurs from small bowel. Its manifestation as perforation is uncommon. We report a case of small bowel lymphoma presenting as perforation.

Keyword : Gastro, intestinal, lymphoma, small, bowel, perforation

INTRODUCTION: Primary malignant tumours of the small bowel are rare and they correspond to 2% of all the tumours of the alimentary canal. The gastrointestinal tract is the most common location of extra nodal lymphoma. The stomach is affected in 75% of cases and small intestine only in 9% 4. They are classified into primary (predominance of intestinal involvement with regional node) or secondary (superficial lymphadenopathy or mediastinal with liver or splenic involvement). This case is presented for its rarity in frequency.

CASE REPORT: A 75 years old male patient was admitted with complaints of abdominal pain, constant in nature, all over the abdomen more in the right lower abdomen for the past 4 days which was aggravated by movements and relieved by lying still. This was associated with vomiting and abdominal distension. The patient has not passed stools and flatus for the past two days. There was history of breathlessness and high grade fever for the past two days. He denied history of diarrhoea, hematemesis, and melena, loss of appetite or weight. He was operated for left inguinal hernia 2 years ago. There were no other comorbid diseases. He was a smoker, not an alcoholic. His physical examination revealed that he was febrile, pale, dehydrated, dyspnoic, not icteric, no generalised lymphadenopathy and no pedal oedema. His vitals were PR-108/min, BP-100/60mmhg, and RR-22/min. On inspection the abdomen was mildly distended, restriction of movements with respiration was present. There was no other positive finding on inspection other than scar of left inguinal hernioplasty. On palpation the abdomen was
warm and tenderness was present all over the abdomen. Guarding and rigidity were present. On percussion, liver dullness was obliterated. Bowel sounds were sluggish on auscultation. P/R- was normal except for tenderness in anterior rectal wall. X ray abdomen erect was taken which showed air under diaphragm.

Liver and spleen were normal. We proceeded with the resection of the involved segment of ileum and mesentry with local proximal and distal clearance of about 10 cm. There was no residual tumor. As there was heavy peritoneal contamination, bowel oedema and sepsis, primary anastomosis was deferred. End ileostomy and mucous fistula was done. Postoperatively patient developed wound infection and burst abdomen. Histopathological examination showed features of Non-Hodgkin’s lymphoma (diffuse large cell variant) the resected margins were free of tumor cells. Immunohistochemistry showed CK negative, CD 45 positive. Postoperative peripheral smear and bone marrow biopsy found to be normal. There was no other lymph node involvement in the postoperative CT chest and abdomen. We planned for adjuvant chemotherapy, after the improvement of general condition of the patient. The patient is currently on chemotherapy - CHOP regimen and follow up.

**fig 1:** Gross- small bowel perforation showing wall thickening.
**fig 2:** Gross- resected segment.
With the above findings we came to a diagnosis of hollow viscus perforation and planned for exploratory laparotomy after adequate resuscitation. There was 1 litre of faecal peritoneal contamination which was drained. We found asymmetric circumferential wall thickening of ileum for about 30-35 cm with about 3x3cm perforation in the antimesenteric border, which was 40 cm proximal to ileo-caecal junction. No mesenteric lymphadenopathy was present. No other lesions were noted.
fig 4: IHC Neoplastic cells – Positive for CD45

fig 5: H&E Small and large lymphoid cells 40X.

DISCUSSION:
Primary gastrointestinal lymphomas constitute 5% of all lymphomas of which 1/3rd occurs in small intestine. It represents 10-15% of all small bowel malignancies. Small bowel is second most frequent site of gastrointestinal tract involvement by lymphoma next to the stomach. Ileum is the most common site of occurrence as ileum has most lymphoid tissue followed by colon and others. Duodenal is least frequently involved. Most cases of small bowel lymphoma are due to Non-Hodgkin’s lymphoma. It includes 4 (1) Diffuse large B cell lymphoma (DLBCL) (2) mucosa associated lymphoid tissue lymphoma, (3) Burkitt’s lymphoma (4) T cell lymphoma. Diffuse large B cell type is the most common variety. It may be primary or secondary. It has bimodal age distribution. Usually occurs in 5th or 6th decade of life with slight male preponderance. In children less than 10 years these are the most common intestinal neoplasm. Criteria for primary lymphomas include 4 (1) confinement of disease to a small bowel segment as confirmed by diagnostic imaging, endoscopy or laparotomy (2) only regionallymphadenopathy (3) no hepatic or splenic involvement except by direct tumor extension (4) palpable or mediastinal lymphadenopathy (5) normal peripheral blood smear and bone marrow biopsy. Mesenteric involvement by lymphoma may occur by direct extension from bowel, indirectly by displacement due to mass effect. Risk factors include (a) immunocompromised or immunosuppressed state (b) long-term celiac sprue (c) chronic lymphocytic leukemia (d) mucosa-associated lymphoid tissue – lymphoma. Grossly small bowel lymphomas are large, with most larger than 5 cm may extend beneath the mucosa with infiltration of muscular layer with destruction of the myenteric plexus leading to aneurysmal dilatation, often at the antimesenteric segment leading to widening of the lumen rather than narrowing. Polypoidal lesions are sometimes reported to cause intussusception. Microscopically diffuse infiltration of intestinal wall may be seen. Definitive diagnosis is based on histopathological examination from biopsy of the lesion.

CLINICAL FEATURES
Patients may present with pain abdomen, weight loss, nausea, vomiting, change in bowel habits. Perforation occurs in 10-25% of patients. Fever is uncommon and if present suggests systemic involvement. The possible mechanism for perforation includes substitution of mucus membrane by tumor cells followed by necrosis and ischemia of tumor segment, it may also follow chemotherapy. Systemic steroids can induce tumor necrosis and intestinal perforation.

INVESTIGATIONS:
Investigations for gastrointestinal lymphoma include complete blood count with differential count, peripheral smear, serum lactate dehydrogenase.
level, bone marrow biopsy, computerised tomography (CT) of the chest and abdomen, small bowel barium series and lymph node biopsy. Small bowel barium series can show luminal narrowing of the involved segment with loss of mucosal pattern and thickening of the plica circularis with intraluminal filling defects possibly with dilatation of the involved segment. Ultrasound may demonstrate a hypoechoic lesion of the affected bowel and presence of abdominal lymphadenopathy. CT scan may show a sausage shaped loop of bowel of relatively homogenous tissue density. Also asymmetric wall thickening of usually greater than 2 cm, aneurysmal dilatation, polyiodal mass and abdominal lymphadenopathy can be seen. 

**STAGING SYSTEM:** Based on the classic Ann Arbor staging system modified systems of staging for gastrointestinal lymphoma have been proposed which include (1) Blackledge staging system (2) Mussoff and Schmid-volmer modification. According to the Mussoff’s system stage 1E refers to small bowel tumor without nodal involvement and 2E refers to small bowel lymphoma with nodal disease below the diaphragm. Stage 3E denotes small bowel lymphoma with nodal disease above and below the diaphragm and 4E refers to widespread disseminated disease. For accurate staging endoscopy, ultrasonography, chest radiography, bone marrow biopsy and CT abdomen has to be done.

**TREATMENT:** Resection followed by adjuvant chemotherapy or radiotherapy. Indications of surgery include (1) establishing the diagnosis (2) to stage disease (3) to relieve obstruction (4) to prevent perforation (5) to prevent tumor bulk (6) to treat peritonitis from perforation. Surgical options are varied ranging from simple biopsy to resection and anastomosis. Surgical resection should be done with 10 cm proximal and distal clearance. The standard protocol for chemotherapy is CHOP (Cyclophosphamide, Doxorubicin, Vincristine, and Prednisolone) - 6 cycles 3 weekly is the standard treatment. For diffuse large B cell lymphomas standard CHOP can be used and rituximab can be added with this for mantle cell/follicular variant. For Burkitt’s lymphoma prednisone in CHOP is replaced by methotrexate. Poor prognosis is associated with (a) stage greater than 2E (nodal disease above and below the diaphragm) (b) tumor size greater than 10 cm (c) T cell type (d) immunoblastic histology (e) presence of aneuploidy (f) presentation as an acute abdomen. 5 year survival rate is 50-70% with combined modality of treatment. Patients recovering from surgery have mean survival of eight months. The overall 5 year survival rate for aggressive small intestinal lymphomas are 25%-30%. Among these intestinal B cell lymphomas are associated with better prognosis than T cell lymphoma (75% and 25% respectively). Radiotherapy is useful in bulky residual disease; cases underwent partial resection or debulking.

The dose is limited to 30 Gy due to intrinsic radio sensitivity of tumor and adjacent structures. Dose per fraction should be 1.5-1.8Gy. Whole abdominal irradiation is associated with greater toxicity.

**References:**

