Abstract:
Several tumors can arise within the small bowel, both malignant (adenocarcinoma, carcinoid, lymphoma, and GIST) and benign (adenoma, leiomyoma, lipoma). Although these malignancies may be found throughout the different regions of the intestine, certain subtypes have a predilection for specific regions. Primary lymphomas of the GI tract are rare accounting for only 1 to 4 percent of malignancies arising in the stomach, small intestine, or colon, while secondary GI involvement is relatively common. We report a case of primary small bowel lymphoma of the jejunum and a review of the literature.

Keyword: Small bowel, Lymphoma
CASE REPORT:
CECT showed multiple focal soft tissue lesions in the small bowel and a short segment concentric nonobstructive thickening of the bowel wall with multiple enlarged mesenteric lymph nodes. Patient was taken up for laparotomy on 9th July 2013 after multiple preoperative transfusions. Peroperative findings revealed an 8 X 6 cm mass in jejunum with three loops of small bowel entangled in it. There was another intraluminal lesion in between the second and the third entangled loops, which had caused an irreducible intussusception of the bowel. There were multiple, soft, discrete mesenteric lymphadenopathies. Since there was a peroperative suspicion of lymphoma, in which case, chemotherapy forms the primary modality of treatment, intraoperative diagnosis by frozen section was sought. Repeated frozen sections of the nodes were reported as reactive adenitis and a core needle biopsy from the primary was also inconclusive. Hence proceeded
with bowel resection. The involved portions of small bowel was removed en bloc with adjoining mesentery and intestinal continuity re established by a stapled anastomosis. Patient had an uneventful postoperative recovery. Cut section of the specimen showed 3 independent lesions, all arising from the jejunum, the second lesion was the lead point of an intussusception, the two proximal lesions were polypoidal and the third lesion was concentric, involving almost the entire circumference of the bowel. On microscopy, the lesions showed ulceration of mucosa with numerous lymphoepithelial lesions and submucous sheets of monotonous population of small lymphocytes , involving full thickness of bowel wall and breaching the serosa to involve the adjacent mesentery. Margins were free. Mesenteric nodes, 8/16 showed involvement by tumor. IHC showed positivity for CD 45, CD 3 AND Bcl 2 and negativity to Bcl 6, Cyclin D1, CK 20 and CD 20, consistent with Diffuse Large B cell Lymphoma. Subsequently patient was staged with a CT chest and bone marrow biopsy, was confirmed to be Stage IIE Non Hodgkins lymphoma and is on chemotherapy.

DISCUSSION
Primary lymphomas account for only 1 to 4 percent of malignancies arising in the gastrointestinal tract [1], while secondary GI involvement is common and occurs in 10 percent of limited stage NHL and up to 60 percent of advanced NHL [2,3]. In the GIT, Stomach is most commonly involved, accounting for 68 to 75 percent, small bowel (including duodenum) 9 percent, ileo-cecal region 7 percent and rectum, 2 percent. Most of them are Non Hodgkins lymphomas. The criteria for diagnosis of a primary GI lymphoma are [4]:

No peripheral or mediastinal lymphadenopathy

A normal WBC count and differential count on the peripheral blood smear

Tumor involvement must be predominantly in the GI tract

No evidence of liver or spleen involvement These lymphomas are categorized into three main groups:

Immunoproliferative small intestinal disease (IPSID or alpha heavy chain disease or Mediterranean lymphoma or Seligmann disease) This lymphoma is characterized by secretion of alpha heavy chains.

Enteropathy-associated T cell lymphoma (EATL), also called intestinal T cell lymphoma This type is associated with gluten-sensitive enteropathy Other western-type non-IPSID lymphomas (diffuse large B cell lymphoma, mantle cell lymphoma, Burkitt lymphoma, follicular lymphoma [5]). Clinical features Patients with IPSID typically present with abdominal pain, chronic diarrhea and malabsorption whereas those with with enteropathy-associated T cell lymphoma (EATL) often present with acute bleeding, obstruction, or perforation [6,7] or as deterioration in a known case of celiac disease. Patients with other non-IPSID lymphomas present with non specific complaints of GI bleed, abdominal pain, intestinal obstruction or perforation and/or a palpable abdominal mass [8,9]

Evaluation : A suspected lymphoma of the small intestine should be evaluated with a contrast-enhanced
computed tomography (CT), conventional endoscopy, and capsule endoscopy if available.

Diagnostic laparotomy with resection of the involved bowel is indicated if there is obstruction, perforation, or major bleeding at presentation. Computed tomography — The following findings may be present on CT [10]:

- Multiple, large tumors
- Bowel segments with lumen that are enlarged, narrowed or both
- Mesenteric nodal masses resulting in nodal masses that surround the mesenteric vessels, called the sandwich or hamburger sign
- Homogenous thickening greater than 2 cm with a normal or enlarged bowel lumen.

Endoscopy — Although technically difficult, endoscopic evaluation of the small bowel with biopsy of lesions can be diagnostic. Proximal small bowel lesions can be detected by "push" enteroscopy, while lesions in the distal small bowel may be assessed with intubation of the terminal ileum during colonoscopy. Endoscopic findings:

- EATL of the jejunum typically demonstrate large circumferential ulcers with biopsy showing evidence of celiac disease in the normal mucosa
- Mantle cell lymphoma may demonstrate polypoid tumors (2 mm to more than 2 cm in size), referred to as "lymphomatous polyposis": Primary intestinal follicular lymphoma most commonly present with multiple small (1 to 5 mm) polypoid lesions. Capsule endoscopy is another useful technique for evaluating the small bowel, but with limited availability. Capsule endoscopy, however, does not permit tissue sampling [11].

Laparotomy — Exploratory laparotomy must be performed when the lesion is not accessible via endoscopy or when endoscopic biopsies are unavailable or non-diagnostic. Laboratory studies — With the exception of alpha heavy chain paraproteinemia in immunoproliferative small intestinal disease (IPSID)-associated disease, the laboratory studies are usually normal in these patients. [12]. STAGING — The Ann Arbor staging system is considered to be inadequate for the staging of GI lymphoma since it does not incorporate information on the depth of tumor invasion, which is known to affect prognosis. The most widely accepted staging system is the Lugano staging system. In this system Early stage [I / II] : Single primary lesion / Multiple noncontiguous lesions confined to the GI tract that may have nodal involvement. There is no stage III. Advanced stage [IV] : Disseminated extranodal involvement or concomitant supra-diaphragmatic nodal involvement

Management Immunoproliferative small intestinal disease Surgery is rarely indicated since intestinal involvement is generally diffuse. Antibiotics against H. pylori or campylobacter jejuni, a regimen of ampicillin and metronidazole, may be tried in early lesions which may lead to regression. [13,14]. Most patients ultimately relapse and present with an aggressive high-grade histology. Combination chemotherapy with or without radiotherapy are the mainstays of treatment for such cases. Five-year survival rates are as high as 70 percent.

Diffuse large B cell Lymphoma Patients with this type of lymphoma have an aggressive course and poor prognosis. Lower rates of complete remission (68 vs 92%).
and five year survival (46 vs 75%) have been reported when comparing with low grade MALToma. [15]. Treatment options that have been evaluated in GI DLBCL include surgery, radiation therapy (RT), chemoimmunotherapy, H pylori eradication therapy, and combinations of the above. A prospective trial of 589 patients with early stage primary gastric DLBCL randomly assigned therapy with surgery, surgery plus RT, surgery plus CHOP (cyclophosphamide, doxorubicin, vincristine, and prednisone) chemotherapy, or CHOP chemotherapy alone [16]. Ten-year event free survival rates were 28, 23, 82, and 92 percent, respectively. Late toxicity was more frequent and severe in patients undergoing surgery. For most patients with GI DLBCL, combination chemotherapy plus rituximab with or without involved-field RT is recommended. ENTEROPATHY-ASSOCIATED T CELL INTESTINAL LYMPHOMA — Enteropathy-associated T cell intestinal lymphoma (EATL) is almost always of high-grade histology; the prognosis is poor and is worse than that of other intestinal lymphomas. Treatment consists of combination chemotherapy [17]. The five-year overall survival rate with anthracycline-based chemotherapy alone is approximately 10 to 20 percent [18]. MANTLE CELL LYMPHOMA & BURKITT'S LYMPHOMA Surgery has a relatively small role in the management, but may be of value in patients presenting with bowel obstruction or intractable bleeding. Systemic chemotherapy is the treatment of choice for advanced disease PRIMARY INTESTINAL FOLLICULAR LYMPHOMA No consistent treatment recommendation is available. The treatment options would include observation, chemotherapy with or without radiotherapy, radiotherapy (30 to 43 gy) and rituximab monotherapy.

CONCLUSION Lymphomas of the small intestine are rare disorders and the clinical presentation varies depending upon whether the tumor is associated with immunoproliferative small intestinal disease (IPSID), celiac disease (enteropathy-associated T cell lymphoma, EATL), or others. The diagnosis may be suggested on computed tomography (CT) and/or contrast radiography, but requires a biopsy for confirmation. Exploratory laparotomy is indicated when preoperative diagnosis is uncertain or when the patient presents with an acute complication like obstruction or bleeding. Management is by combination of chemotherapy or chemoimmunotherapy with or without radiotherapy. Surgery is indicated in specific situations. Antibiotic therapy may be indicated in early stage IPSID.

References:


