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A Rare Case of Primary Ileal Lymphoma in A Situs Inversus Totalis

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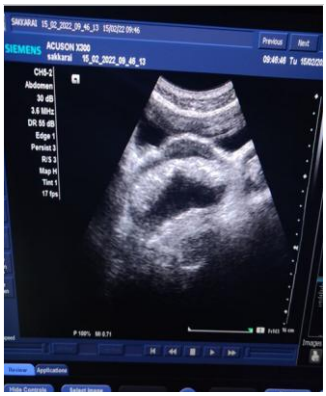
I. Introduction:

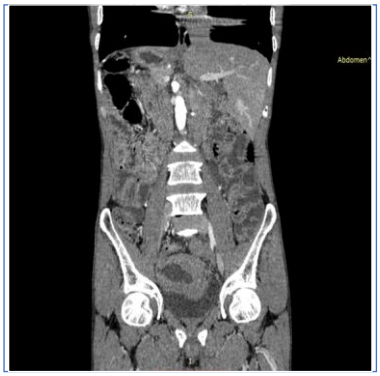
Gastrointestinal tract is the most common extranodal site involved by lymphoma with the majority being non-Hodgkin type. Although lymphoma can involve any part of the gastrointestinal tract, the most frequent sites in order of its occurrence are the stomach followed by small intestine and ileocecal region. Gastrointestinal tract lymphoma is usually secondary to the widespread nodal diseases and primary gastrointestinal tract lymphoma is relatively rare. Marginal zone B cell lymphoma is the most common pathological type of gastrointestinal lymphoma, other variants include diffuse large Bcell , burkitts, mantle cell lymphoma, T cell lymphoma. There has been a tremendous leap in the diagnosis, staging and management of gastrointestinal lymphoma in the last two decades attributed to a better insight into its etiology and molecular aspect as well as the knowledge about its critical signaling pathways helps introducing monoclonal antibodies.Certain risk factors have been implicated in the pathogenesis of gastrointestinal lymphoma including *Helicobacter pylori* (*H. pylori*) infection, human immunodeficiency virus (HIV), *Campylobacter jejuni* (*C. jejuni*), Epstein-Barr virus (EBV), hepatitis B virus (HBV), human T-cell lymphotropic virus-1 (HTLV-1), inflammatory bowel disease and immunosuppression. Primary intestinal lymphoma can be differentiated from secondary lymphoma by DAWSONS CRITERIA. Surgery can be done in isolated cases. Left untreated may cause obstruction, bleeding, perforation.

II. Case Report:

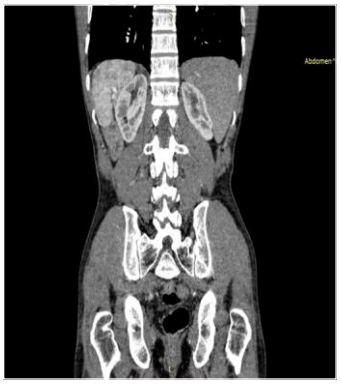
54years male presented to the hospital with chief complaints of abdominal pain for past 1 month lower abdomen, dull aching type, with other complaints and no other co morbidities.General physical examination was unremarkable.Abdomen examination shows a vague ill defined intraperitoneal mass palpable in left iliac fossa. Ultrasound abdomen shows Anuerysmal dilatation of distal ileum , multiple enlarged retroperitoneal nodes were noted alone with situs inversus(Fig1). Evaluated with Contrast enhanced CT abdomen which come as primary intestinal lymphoma involving terminal ileum(Fig2). We planned for elective surgery. Abdomen open in midline, thorough laparotomy done, right lobe of liver is found in left hypochondrium, spleen is on right hypochondrium, no liver surface nodules, pelvic nodules. 6x5cms mass lesion seen in mesenteric border of terminal ileum in left iliac fossa(Fig3). Ascending colon and caecum mobilized, resection of the tumour along with attached mesenteric nodes done, bowel continuity is maintained by ileoascending anastomosis(fig4). Abdomen closed in layers with a drain in left paracolic gutter. Following surgery post operative period was uneventful, orals started on 3rd post op day, drain removed on 5th post op day. Biopsy reviewed came as Maltoma with clear margins, stage IIE. Advised for IHC CD19,20,70a. IHC markers were found to be positive.



ULTRASOUND



Liver on left side



Spleen on right side

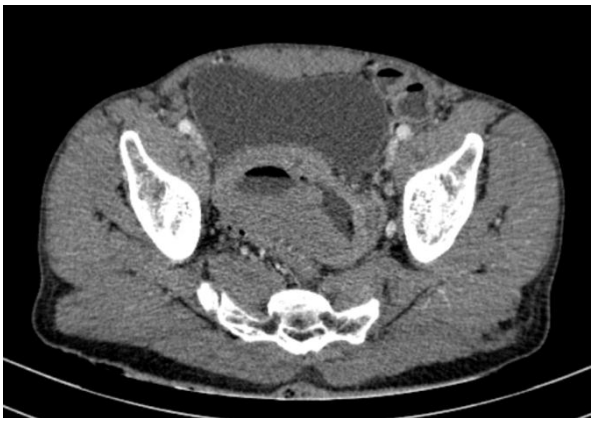


Fig 2 -FOCAL ANEURYSMAL

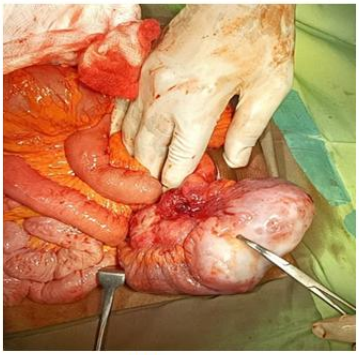


Fig 3

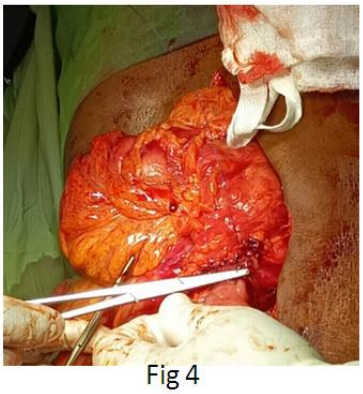
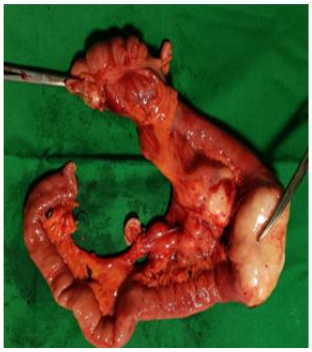


Fig 4



specimen

III. Discussion:

Primary gastro-intestinal lymphoma accounts for 1%-4% of all gastrointestinal malignancies.Majority of these arise in the stomach(65%) followed by the small bowel (20- 30%) with rest arising in the colon and rectum. Most common site involving in lymphoma of small intestine is ileum followed by jejunum, duodenum. The ileocaecal region is the most common site. They arise from the lymphoid tissue present in the mucosa of the bowel wall, they are marginal zone type of lymphomas due to continuous exposure to the high flow of external antigens.

Small bowel lymphomas tend to have non specific symptoms, high level of suspicion is needed to diagnose. Primary lesions do not have lymphadenopathy, DAWSONS CRITERIA is used to label primary gastrointestinal lymphoma, that include (1) absence of peripheral lymphadenopathy at the time of presentation; (2) lack of enlarged mediastinal lymph nodes; (3) normal total and differential white blood cell count; (4)

predominance of bowel lesion at the time of laparotomy with only lymph nodes obviously affected in the immediate vicinity; and (5) no lymphomatous involvement of liver and spleen.

Elevated total counts warrantsperipheral smear and bone marrow aspiration.Radiological investigations like ultrasound and CT helps in picking up mass lesions.PET CT role in primary GI lymphomas is for follow up and recurrent cases, along with LDH andB2 macroglobulin. With advent of capsule endoscopy and push and pull enteroscopy biopsy is made possible, they appear as mass, polyp or ulcers.

ANN ARBOR staging modified by Musshoff is used for staging the disease.

StageI-tumour limited to digestive tract either single or multiple locations

stageII- tumour with intra abdominal location,II1- local, gastric, or intestinal lymph nodes, II2- distinct nodes (aortic, mesenteric, vena cava, pelvic, inguinal)

stage IIIE- perforation of serosa, affected by contiguity to adjacent organs

stage IV-diffuse intestinal compromise, outside the diaphragm and spine.

With exception of T cell lymphoma, small bowel lymphoma are chemosensitive, and tend to be poorer prognosis than gastric lymphomas. Regardless the type, resection is needed in symptomatic subsets. Chemotherapy regimen consists of R-CHOP Ritiximab, cyclophosphamide, Doxorubicin (hydroxydaunomycin), Vincristine (oncovin) Prednisolone.

Immunohistochemistry must be done,a panel of markers areavailable (no single marker is specific) which includes leukocyte common antigen (LCA), B-cell markers (CD20 and CD79a), T-cell markers (CD3 and CD5) and other markers like CD23, bcl-2, CD10, cyclinD1, CD15, CD30, ALK-1, CD138 (based on cytoarchitectural pattern). Theyhelp in subtyping, prognostication and targeted therapy, Rituximab in B-cell lymphomas, CD22 – IgG 1 antibody (Epratuzumab) in relapsed and refractory, indolent and aggressive NHL, CD30 – Anti CD30 (SGN 30) in Hodgkin's lymphoma and CD30 positive T-cell lymphomas.

IV. Conclusion:

Primary GI lymphomas are rare, high level of suspicion must be there, vague symptoms will make it difficult to diagnose, often made by ultrasound /CT/ endoscopy. Surgical exploration and chemotherapy remains the mainstay of treatment. Our case is a rare occurrence in a sistus inversus patient.

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