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EC GASTROENTEROLOGY AND DIGESTIVE SYSTEM

Case Report

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

Muthukumar R[1]*, Radha Krishna V[2], Nandha Kumar S[3], Pranav Prem[4]

[1]Professor, Department of General Surgery, Sri Venkateshwaraa Medical College Hospital and Research Centre, Puducherry, India

[2]Assistant Professor, Department of General Surgery, Sri Venkateshwaraa Medical College Hospital and Research Centre, Puducherry, India

[3]Senior Resident, Department of General Surgery, Sri Venkateshwaraa Medical College Hospital and Research Centre, Puducherry, India

[4]Postgraduate, Department of General Surgery, Sri Venkateshwaraa Medical College Hospital and Research Centre, Puducherry, India

***Corresponding Author:** Muthukumar R, Professor, Department of General Surgery, Sri Venkateshwaraa Medical College Hospital and Research Centre, Puducherry, India.

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Abstract

Introduction: Primary GI lymphoma is a rare disorder, most commonly seen in stomach followed by small intestine and ileocecal region majority being non-hodgkin type. Marginal zone B cell lymphoma is the most common pathological type. Risk factors are *H. pylori* infection, Human immunodeficiency virus (HIV), *Campylobacter jejuni*, Epstein bar virus, Hepatitis B virus, inflammatory bowel disease and immunosuppression. We discuss a case of primary Gi lymphoma in a patient with situs inversus totalis.

Case Report: 54-year-old patient with no co-morbidities and no prior hospitalization came to OPD with complaints of dull aching abdominal pain for past 1 month. USG abdomen and CECT abdomen was taken and showed focal aneurysmal dilatation of small bowel loops with multiple enlarged retroperitoneal lymph nodes suggestive of Primary GI lymphoma with Situs inversus totalis. Through midline laparotomy, resection of tumour with attached mesenteric nodes was done and bowel continuity was maintained by ileo-ascending anastomosis. Following surgery, postoperative period was uneventful. Biopsy reviewed came as Maltoma with clear margins - Stage IIe. IHC was done which showed Diffuse large B cell lymphoma not otherwise specified non germinal B cell subtype.

Conclusion: Primary GI lymphomas are a rare entity and are diagnosed late as they are mostly asymptomatic. USG/Ct abdomen helps in diagnosis. Surgery and chemotherapy remain main stay of treatment.

Keywords: *Primary GI Lymphoma; Maltoma; Situs Inversus Totalis; Diffuse Large B Cell Lymphoma*

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 B cell, burkitts, mantle cell lymphoma and 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.

Case Report

54 years male presented to the hospital with chief complaints of abdominal pain for the past 1 month, in the lower abdomen, dull aching type, with no other complaints and no other co morbidities. General physical examination was unremarkable. Abdomen examination showed a vague ill-defined intraperitoneal mass palpable in left iliac fossa. Ultrasound abdomen showed Aneurysmal dilatation of distal ileum, multiple enlarged retroperitoneal nodes were noted along with situs inversus (Figure 1). Evaluated with Contrast enhanced CT Abdomen which came as primary intestinal lymphoma involving terminal ileum (Figure 2). We planned for elective surgery. Abdomen was opened in midline, thorough laparotomy done, right lobe of liver was found in left hypochondrium, spleen on right hypochondrium, no liver surface nodules, pelvic nodules. 6 x 5 cms mass lesion seen in mesenteric border of terminal ileum in left iliac fossa (Figure 3). Ascending colon and caecum mobilized, resection of the tumour along with attached mesenteric nodes was done, bowel continuity was maintained by ileo-ascending anastomosis (Figure 4). Abdomen closed in layers with a drain in left paracolic gutter. Following surgery post operative period was uneventful, orals were 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.

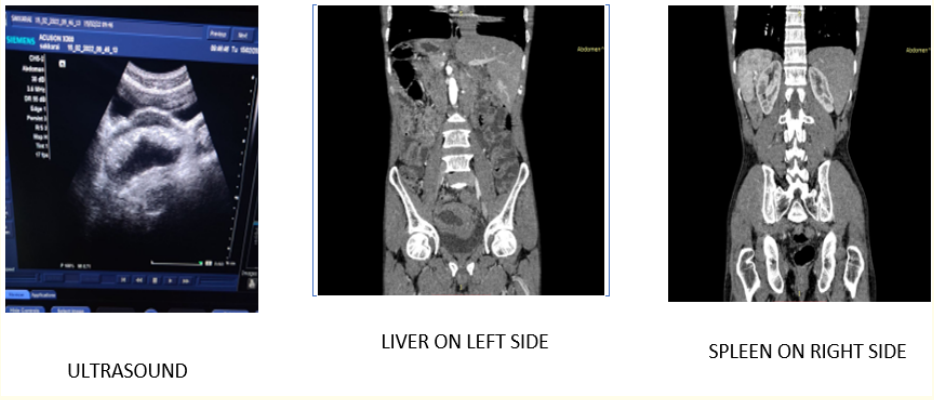
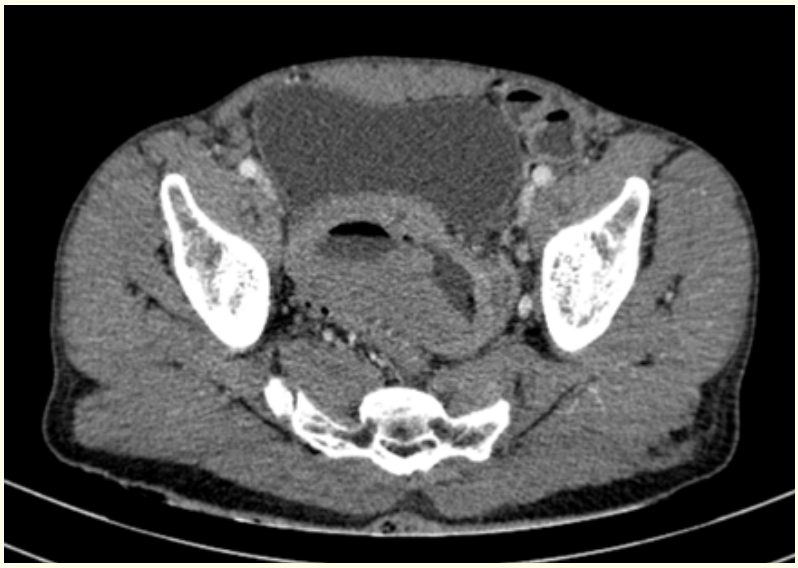


Figure 2: *Focal aneurysmal dilatation.*



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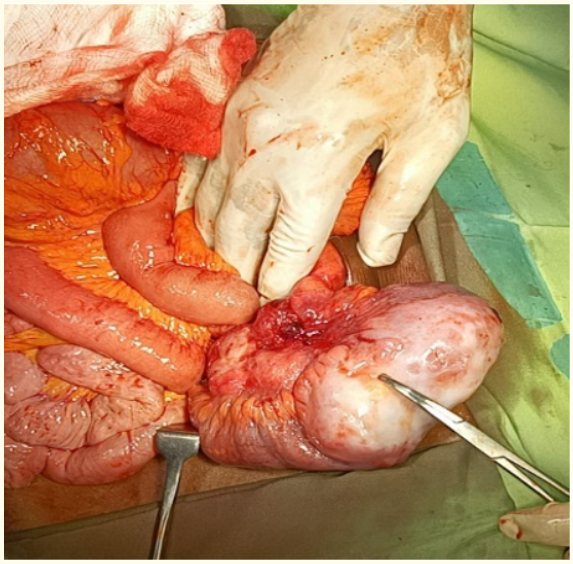

Figure 3

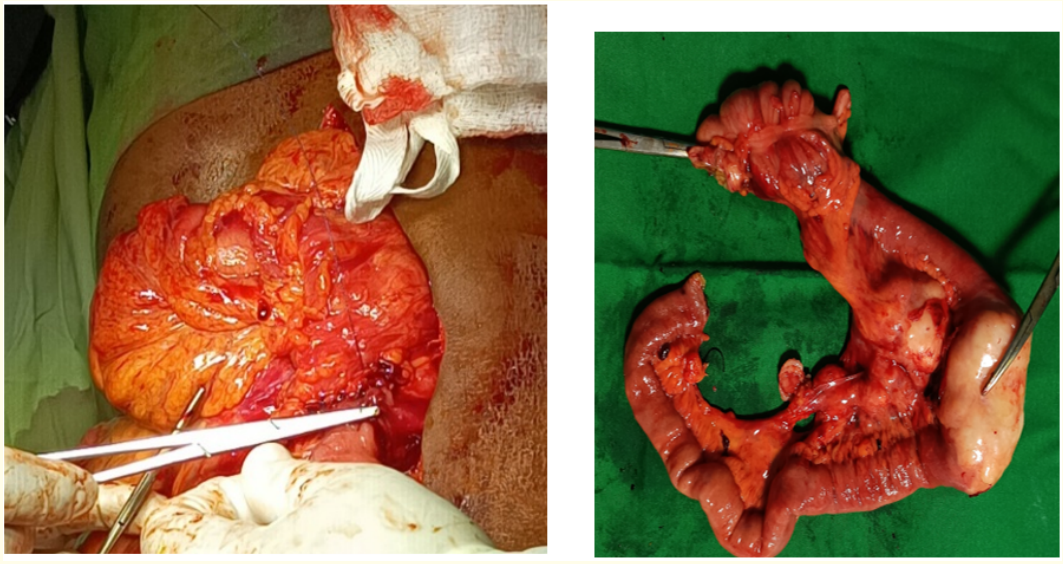

Figure 4*: Specimen.*

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 involved in lymphoma of small intestine is ileum followed by jejunum and 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 warrant peripheral 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 and B2 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:

- Stage I-tumour limited to digestive tract either single or multiple locations.

- Stage II- tumour with intra-abdominal location, II1- local, gastric, or intestinal lymph nodes, II2- distinct nodes (aortic, mesenteric, vena cava, pelvic, inguinal).
- Stage III E- 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 is 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 Rituximab, cyclophosphamide, Doxorubicin (hydroxy daunomycin), Vincristine (oncovin) Prednisolone.

Immunohistochemistry must be done, a panel of markers are available (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). They help 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 [1-4].

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 situs inversus patient.

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