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A Rare Case of Ileal Mucosa associated Lymphoid Tissue-MALT Lymphoma

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Abstract: ***Background: Mucosa associated Lymphoid Tissue (MALT) Lymphoma is a subtype of Non Hodgkins Lymphoma, comprising 8%. The majority of the lymphomas within the gastrointestinal tract involve the stomach 60-75%. Primary ileal MALT lymphoma is rare, and has not been associated with a specific infectious disease. We report a case of Ileal MALT lymphoma in a Female which is presenting as sub acute Intestinal Obstruction. On colonoscopy, there is an Ulcerated non circumferential Exophytic Growth protruding from IC valve, blocking the lumen of ascending Colon. CECT revealed heterogeneously enhancing Irregular Circumferential Lesion involving terminal Ileum measuring 5.3cms with wall thickness of 1.5 cms, with external Iliac Lymph nodes. Biopsy demonstrated Low Grade B cell Lymphoma and was confirmed, as MALT Lymphoma on IHC. Chemotherapy started. Patient Partly responded to Chemotherapy at 2months and was on follow up.***

Keywords: GI tract, MALT Lymphoma, Small Intestine Ileum, Immune Histochemistry (IHC)

1. Introduction

The Gastrointestinal tract is involved by 30-40 % of all extra nodal non-Hodgkin lymphomas 1. Primary GI Lymphomas (PGIL) constitute 1-4% of all GI malignancies 2-3. . The incidence of PGIL varies from 0.58 and 1.31 per 100, 000 people and the usual age of diagnosis is between 50-70 years. The majority (60-75 %) of the extra nodal lymphomas within the gastrointestinal tract involve the stomach and are associated with Helicobacter pylori (H. Pylori) infection4 Involvement of other sites of the gastrointestinal tract by extra nodal lymphomas has been reported including colon, jejunum, ileum and rectum. Clinical presentation of lymphomas are abdominal pain, weight loss, loss of appetite, obscure GI bleed, low grade fever and intestinal obstruction, which is rare only seen in 10% of presentations. Here by we are reporting a maltoma arising from terminal ileum presenting as sub acute intestinal obstruction.

2. Case Report

A 34 yr old lady restaurant worker, presented with abdominal pain, vomiting since two months, increased in intensity two weeks before presentation to opd. Symptoms associated with low grade fever, reduced appetite, significant weight loss and altered bowel habits with diarrohea initially six months ago with constipation since three months. Physical examination revealed pallor, horizontal scar in hypogastrium from previous LSCS and hard ovoid mass palpable in the right iliac fossa. On imaging usg abdomen shows thickened terminal ileum with proximal dilated ileal loops, CECT revealed Heterogenously enhancing long segment thickening of terminal ileum noted for a length of 10 cms protruding into caecum and causing complete luminal obstruction. Extraluminal extension of the lesion noted in antimesentric border of terminal ileum, abutting the right psoas muscle Multiple mesenteric lymph nodes adjacent to the lesion largest measuring-27 X 13 mm s/o nodal deposits. (Figure 1)

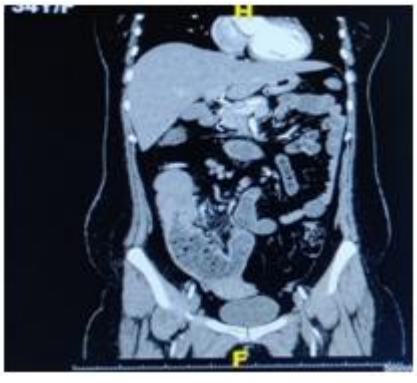

Figure 1 (a): CECT Abdomen:

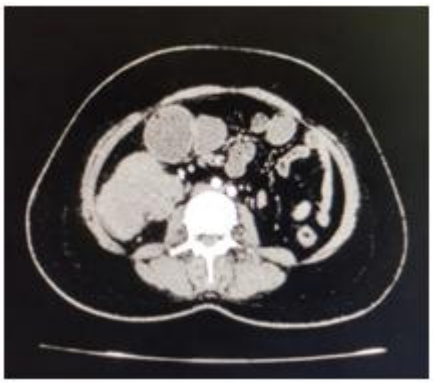

Figure 1 (b)

Colonoscopy: Large Exophytic growth noted with altered friable mucosa and ulceration at the tip protruding into caecum and causing luminal narrowing at IC valve. Scope could not be negotiated beyond (Figure: 2).

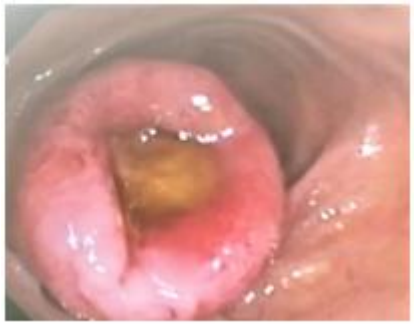

Figure 2: Colonoscopy

Large exophytic growth noted with altered friable mucosa and ulceration at the tip protruding into caecum and causing luminal narrowing at IC valve.

HISTOPATHOLOGY: Histologically, Lamina propria shows dense collection of monomorphic lymphocytes along with very few large atypical cells. **IHC: t**hese cells were CD20-positive B-lymphocytes that co-expressed BCL-2 and were negative for CD5, CD10, CD43, and cyclin D1 on immunohistochemical studies. The overall proliferation index was low with Ki-67 immunoreactivity in approximately 10 % of cells. No areas suspicious for large cell or high grade transformation were identified. The bone marrow biopsy showed no evidence of involvement by lymphoma (figure: 3 and 4).

Histopathology specimens:

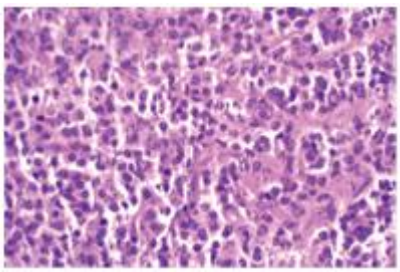

Figure 3: Ileal wall with transmural nodular unfilterate with monocytoid appearence

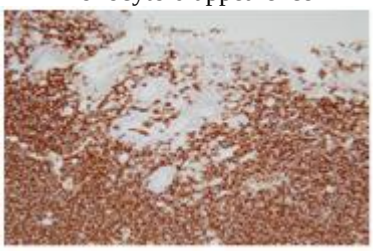

Figure 4: Immunohistochemistry staining-Lymphocytic infilterate predominantly containing CD20 positive B cells

Patient was advised surgical resection followed by chemotherapy. Patient refused surgery and opted for chemotherapy. . She has received 4 Cycles of CHOP regimen Chemotherapy started and on Follow Up lesion and symptoms responded Later pts developed Liver metastasis in 2months.

3. Discussion

MALT Lymphoma: also known as Extra nodal Marginal Zone Lymphoma comprises **8 %** of NHLs. One of the major series of MALT Lymphomas from south India Arora et al, Ileal Lymphoms are not descried, stomach constitutes 76.4%, small Intestine – 20.5% [12]In another series from North India, MALT Lymophoma are not described in Ileum and small Intestine [13].

Mucosa associated Lymphomas of GI tract are Unique as they involve the tissues that are Not Primary Organs of Immune system. First described in 1983, B-cell lymphoma of mucosa-associated lymphoid tissue (MALT) is a rare cause of primary gastrointestinal (GI) lymphoma. The very concept of MALT type of lymphoma was introduced by Isaacson and Wright. It is Defined by Jayabackthan et al,-- "MALT lymphoma is defined as an extra nodal marginal zone B-cell lymphoma of MALT type [5]as per the latest WHO classification.

Mucosa-associated lymphoid tissue can be found throughout the body, with the highest amount found in the GI tract. When this tissue undergoes abnormal cell proliferation, lymphoma can develop.

By immunohistochemistry, the neoplastic cells express the pan-B-cell markers CD19, 20, 22, and 79a. Plasma cell differentiation can be demonstrated by CD 138 expression and concurrent light chain restriction[6]. CD5, CD10, and cyclin D1 are classically negative, aiding in differentiation of MALT from other low-grade B-cell lymphomas such as chronic lymphocytic lymphoma (typically CD5 positive), mantle cell lymphoma (typically CD5 positive), and follicular lymphoma (typically CD10 positive). Rarely, the malignant B cells in marginal zone lymphoma may express CD5, posing a diagnostic challenge [7]. CD43 is positive in about half of the cases, and can help to differentiate a malignant proliferation (CD43 positive B-cells) from reactive MALT Lymphoma.

Table 1: Immune Histochemistry of GI Lymphomas -

Type	CD5	CD 10	CD19	CD20	CD22	CD23	CD43	CD79a	CD3	CD30	CD45RO
Diffuse B cell Lymphoma	+/ -	+/ -	+	+	+	-	-	+	-	-	+/ -
Malt Lymphoma	_	-	+	+	+	-	+/ -	+	-	-	+
Follicular Lymphoma mantle cell Lymphoma	-	+/ -	+	+	+	+/ -	-	+	-	-	-
Extanodal Peripheral T cell Lymphoma	+/ -	- /+	-	-	-	-	+/ -	- /+	- /+	- /+	+/ -

Pathogenesis: The pathogenesis can be in a setting of chronic Inflammation it develops. Most commonly in stomach. About 72-98% are associated with H. Pylori infection[4]. Extra nodal marginal zone lymphoma typically arises in areas where mucosa-associated lymphoid tissue (MALT) has been acquired, but not necessarily where it is found under normal circumstances, such as the Peyer's patches. In these acquired areas, lymphoma is believed to arise as a result of some chronic antigenic or inflammatory stimulus such as H. pylori gastritis or autoimmune process such as celiac disease, Sjogren syndrome, or Hashimoto's thyroiditis, Gastric cases associated with Helicobacter pylori.

The clinical presentation depends on the site of involvement. Pain, loss of appetite, weight loss, and gross gastrointestinal Bleeding are the most common symptoms. Perforation and hemorrhage is reported as one of the complications. MALtoma commonly presents as Abdominal Pain. The symptoms of MALT Lymphoma are Non specific which makes the diagnosis difficult.

When lymphomas arise in the small bowel, they are most often diffuse large B-cell lymphomas; however, MALT lymphomas represent 1/3rd of the small bowel lymphomas [8, 9]. MALT lymphomas of Ileum are extremely rare and only few reports published [10]

Small Intestine MALT Lymphoma: H. Pylori infection is not commonly documented in small intestinal MALT lymphoma cases. MALT lymphoma of small intestine is seen more in elderly age group. Risk factors for small bowel lymphomas has been found to include immunodeficiency, inflammatory bowel disease and mal absorption syndromes.[14]Among the available Treatment options there is No role for anti-H Pylori antibiotic treatment in non-gastric MALT lymphomas. Surgery is indicated in cases of obstruction and perforation. Chemotherapy and immunotherapy are main treatment modalities.

Prognosis
Prognosis depend on-3 parameters-age ≥70, stage III or IV disease, sr. LDH level s.5 year event free survival in low, intermediate and high risk groups – 70%, 56%, 29% respectively.5 year survival in small intestinal MALT Lymphoma is approximately 75% [15]

4. Conclusion

This case represents a rare case of MALT Lymphoma, involving ileocaecal region, in a young, female patient, H. Pylori Negative without any immunodeficiency, inflammatory bowel disease and malabsorption syndrome. Appropriate identification of these cases is important for early intervention and proper management.

Conflict of interest
The authors declare that they have no conflicts of interest.

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Ethical statement
Informed consent taken from patient

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