Abstract:
AN INTERESTING CASE OF ILEAL MASS(NON-HODGKINS LYMPHOMA OF ILEUM)

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A 36 years old male patient presented with complaints of pain in the right lower abdomen for 3 days duration with history of vomiting and fever. On examination patient was afebrile, tachycardia was present. There was no generalized lymphadenopathy, per abdominal examination there was tenderness in the right iliac fossa, maximum tenderness in the Mc Burney's point with localised guarding and rebound tenderness, no mass palpable, Bowel sounds were sluggish.

Provisional clinical diagnosis was Acute Appendicitis. On Ultrasonography of the abdomen an irregular hypoechoic lesion measuring 6.3 x 4.3 x 5.4 cm with air pockets was seen with features suggestive of Appendicular Abscess. Emergency Exploratory Laparotomy was done, intra operatively an inflammatory mass measuring 8 x 6 cm was found around 10 cm proximal to the Ileo-caecal Junction in the distal Ileum with Omental adhesions. Hence Ileal mass Resection Anastomosis was done. Post operative period was uneventful. Histopathological examination proved to be high grade NON-HODGKINS LYMPHOMA.

Conclusion: Primary Gastrointestinal Non-Hodgkins Lymphoma is a rare condition. Incidence of Primary Ileal Lymphoma is 3 of all lymphomas. These patients can rarely present with intestinal perforation, bleeding and obstruction.
Keyword: PRIMARY NON-HODGKINS LYMPHOMA OF ILEUM

INTRODUCTION:
Non-Hodgkins Lymphoma is a proliferation of discrete masses with in Lymphnodes, Spleen or Extranodal tissues apart from Hodgkins Lymphoma which has its clinical and histological distinction. Gastrointestinal tract is the commonest extranodal site for Lymphoma. Ileum is 2nd commonest site for primary gastrointestinal tract Non-Hodgkins lymphoma next to stomach. Diagnostic criteria for primary gastrointestinal lymphoma includes absence of Superficial lymphadenopathy on physical examination, Absence of Mediastinal lymphadenopathy on chest imaging, Normal peripheral blood counts.

CASE REPORT:
A 36 year old male patient Mr. X presented with complains of pain in theright lower abdomen for past 3 days, there was history of vomiting for 2 days and fever for 1 day. There was no history of abdominal distension, constipation, malena, hematemesis, diarrhoea, burning micturition. No surgeries were done in the past, no history of comorbid illnesses like diabetes mellitus, tuberculosis, hypertension. Not a smoker or alcoholic, no history of altered bowel or bladder habits. No significant family history. On examination patient was conscious, oriented, afebrile, no pallor or icterus or generalised lymphadenopathy. Pulse rate was 108/minute, B.P was 110/80mm ofHg. Per abdominal examination: All quadrants move equally with respiration. No abdominal distension or dilated vessels or scars, umbilicus normal in position, hernia orifices free, no fullness in the flanks, no warmth, tenderness was present in the right iliac fossa region, maximum tenderness was in the Mc Burney’s point, localised guarding was present in the right iliac fossa region, no rigidity, no mass palpable or no organomegaly, no shifting dullness, bowel sounds were sluggish. Digital rectal examination was normal. Clinical diagnosis was ? ACUTE APPENDICITIS.

Investigations
Routine blood tests including Total and differential leucocyte count were within normal limits except raised ESR, urine routine examination was normal, HIV test negative, chest x-ray and x-ray abdomen erect were normal, Ultrasonogram of the abdomen: Irregular, hypoechoic lesion measuring 6.3×4.3×5.4 cms with air pockets and surrounding inflammatory changes seen in the right iliac region, features suggestive of ? Appendicular Abscess. Patient was planned for an emergency exploratory laparotomy.

Operative findings
eocaecal junction and appendix found to be normal, A firm inflammatory mass measuring 8×6 cms with haemorrhagic areas was found in distal ileum around 10 cms proximal to the ileocaecal junction with omental adhesions. Rest of the small and large bowel were normal, other solid viscera normal. No mesenteric lymphnodes or tubercles found. Intraoperative diagnosis was ILEAL MASS ? TUBERCULOUS IN ORIGIN.
2 cm margin and end to end anastomosis of ileum was done. Post operative period was uneventful. Histopathological examination of the specimen revealed HIGH GRADE NON-HODGKINS LYMPHOMA.

**HISTOPATHOLOGICAL REPORT**

**Figure 1:** Intra operative picture showing ileal mass measuring 8×6 cms with haemorrhagic areas.

**Figure 2:** Mass arising from distal ileum and is firm in consistency.

**Figure 3:** Resection of the mass with end to end anastomosis of ileum was done.

**Figure 4:** Cut section (gross appearance) of the resected ileal mass showing the lumen of the ileum and replacement of all layers of bowel with tumor infiltration. **Procedure done:** Emergency Exploratory Laparotomy through a rightparamedian incision with resection of ileal mass with a
Primary malignant neoplasms of small bowel include Adenocarcinoma, Carinoid, Gastrointestinal stromal tumour, Leiomyosarcoma, Lymphoma (7-25%). Rarely Liposarcoma, Myxoliposarcoma, Lymphangiosarcoma can occur. Metastatic tumors include Melanoma, Lymphoma can also be seen. Primary Gastrointestinal Lymphoma: The most common site is stomach (60%) followed by small bowel (30%), colon (10%).

Primary Gastrointestinal Lymphoma comprises approximately 20% of all Lymphomas. Small bowel lymphomas account for 5% of all Lymphomas. Primary ileal lymphoma accounts for 3% of all lymphomas and 0.1% of all malignancies. Primary ileal lymphoma accounts for 1-3% of all Gastrointestinal malignancies. World wide Incidence of primary ileal lymphoma is 1-5/100000/year. Incidence in India is 0.17/100000, incidence in Chennai is 0.23/100000. An excess in male incidence was found in primary ileal lymphoma, male:female ratio is 1.5:1. Most common site for lymphoma of small intestine is ileum as it contains highest concentration of Gut associated lymphoid tissue (GALT) in the intestine. Primary Ileal Non-Hodgkins lymphoma originates in the lymphoid follicle of the submucosa.

GROSS APPEARANCE OF ILEAL LYMPHOMA CAUSING LUMINAL OBSTRUCTION

GROSS APPEARANCE OF A PERFORATED PRIMARY ILEAL NON-HODGKINS LYMPHOMA

There is increased increased risk of developing Lymphoma for patients with celiac disease, Helicobacter pylori, Inflammatory bowel disease, Immunosupression after solid viscera transplantation and immunodeficiency states(e.g: AIDS).

Characteristics of Non-Hodgkins Lymphoma
Ø More frequent involvement of multiple peripheral lymph nodes.Ø Noncontiguous spread.Ø Mesenteric
nodes and Waldeyers ring commonly involved.Ø Bone marrow and Extranodal involvement common.Ø Common in elderly.Ø Responds well to chemotherapy.

**THE WHO(UPDATED REAL) CLASSIFICATION OF THE LYMPHOID NEOPLASMS**

**I. Precursor B-Cell Neoplasms**
- Precursor-B lymphoblastic leukemia/lymphoma

**II. Peripheral B-Cell Neoplasms**
- Chronic lymphocytic leukemia/small lymphocytic lymphoma
- B-cell prolymphocytic leukemia
- Lymphoplasmacytic lymphoma
- Splenic and nodal marginal zone lymphomas
- Extranodal marginal zone lymphoma
- Mantle cell lymphoma
- Follicular lymphoma
- Marginal zone lymphoma
- Hairy cell leukemia
- Plasmacytoma/plasma cell myeloma
- Diffuse large B-cell lymphoma
- Burkitt lymphoma

**V. Hodgkin Lymphoma**
- Classical subtypes
- Nodular sclerosis
- Mixed cellularity
- Lymphocyte-rich
- Lymphocyte depletion
- Lymphocyte predominance

**Clinical features**
- Patients with primary ileal Lymphoma presents with nonspecific abdominal complaints like malabsorption, weight loss, abdominal pain, nausea, vomiting, anorexia, acute or chronic blood loss.
- 10% of patients may present with intestinal obstruction (Acute or Subacute), GI bleeding, Bowel perforation, Intussusception, Fistulization.

**DIAGNOSIS DIAGNOSTIC CRITERIA FOR PRIMARY GI LYMPHOMA**
(DAWSON ET.AL s criteria)
- 1.Absence of superficial Lymphadenopathy on physical examination.
- 3.Normal peripheral blood counts.
- 4.There is no involvement of Liver and spleen.
- 5.At Laparotomy, the Alimentary lesion is predominantly involved, with lymph node involvement (if any) confined to the drainage area of the involved segment gut.

These cases are rarely diagnosed preoperatively, high index of suspicion is required for early diagnosis. Primary Ileal Lymphomas can grow to large size before the onset of symptoms. USG Abdomen: Not sensitive. CT Abdomen: Lymphoma may appear as a Mass or Bowel wall thickening or Displacement of adjacent organs or Luminal Obstruction.

**Tissue Diagnosis:** Can be achieved by Endoscopic or CT guided Biopsy.

**STAGING AND PROGNOSIS**
Most of the patients present with stage 3 or 4 disease and have poor prognosis. Less than 30% of patients: Surgically resectable.

**TREATMENT**
**MEDICAL TREATMENT**
CHOP Regimen is the most commonly used combination chemotherapy which includes Cyclophosphamide (750mg/m2), Doxorubicin (50mg/m2), Vincristine (1.4mg/m2 with maximum of 2 mg), Prednisolone (100mg for 5 days every 21 days). Late stage disease which is not amenable to surgery should be given chemotherapy. Risk of perforation in patients received chemotherapy approaches to 5%. Early stage (1E, 2E) Chemotherapy is given along with Radiotherapy.

RADIOTherAPY
Radiotherapy has a limited usefulness for large tumors (>6 cms). Radiotherapy is given in combination with chemotherapy (Chemoradiation). Dosage: 30-35 Gy delivered over 4-5 weeks is the usual standard. Late complications of radiotherapy includes stricture Enteritis, secondary tumor formation. Treatment with radiotherapy should be individualized.

SURGICAL TREATMENT
Wide resection including regional Lymph nodes should be done for cases presenting with Bleeding, Perforation, Obstruction. Often surgical resection for cure is not possible, palliative resection should be performed in such cases to prevent complications. If palliative surgery not possible, bypass of the involved segment may be done to relieve symptoms like obstruction. If discovered unexpectedly at operation: Confirmed by Frozen section. Additionally fresh tissue should be sent to Fluorescence-Activated cell sorting, Immunohistochemistry and Genetic Analysis. If isolated Stage 1E or 2E Lymphoma is encountered. Surgical removal of all gross disease is ideal. Disseminated Lymphoma: Surgical cure not possible, operation done for obtaining tissue for diagnosis.

and repair of perforations

CONCLUSION
Primary Ileal (Non-Hodgkins) Lymphoma is a rare condition which can cause complications like intestinal obstruction, gastrointestinal bleeding, perforation, Intussception (Ileocaecal), Fistulization.

High index of suspicion is required for early diagnosis and treatment.

Primary Ileal Lymphoma should be considered for differential diagnosis of lower Gastrointestinal bleeding.

Can mimic other causes of acute abdomen especially when it develops complications.

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