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Unusual Presentation of Adult Ileo-Ileal Intususception- A Case Report of B-Cell NHL

KEYWORDS: Adult intussusception, NHL, small intestinal obstruction.

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ABSTRACT *The occurrence of adult intussusceptions due to small intestinal lymphoma is quite rare, hence pre-operative diagnosis often remains elusive. There are no explicit presenting symptoms except features of intestinal obstruction, hence can prove to be a surgical challenge. We present a case report of malignant lymphoma in a young 21 year adult male, causing ileo-ileal intussusception leading to intestinal obstruction. The patient underwent laparotomy with segmental ileal resection and anastamosis. Histopathology of the specimen suggested malignant lymphoma. Immunohistochemistry showed tumour cells staining positive for CD 20, BCl-2 and Ki-67 and negative for TdT and CD 10, concurrent with high grade B-cell NHL of intestine.*

Introduction:-
Intussusception is most often seen in infants and children and only 5% of all cases occur in adults, accounting for 1% of cases of adult small bowel obstruction. Adult intussusception is rare and about 40% are related to malignant lesion[1]. Primary malignant tumours of small intestine are very rare – less than 2% of all gastro intestinal malignancies, include adenocarcinoma, GISTS, lymphoma and carcinoid tumours[2].

The GIT is the most common site of primary extra nodal NHL- (20-40%) Of all extra nodal disease[3] the stomach is most frequently affected (50-60%) followed by small bowel (20-30%). Primary intestinal NHL is the 3rd most common intestinal neoplasm after adenocarcinoma and Carcinoid tumours. Intussusception is very rarely seen in intestinal NHL and the most common type of lymphoma causing intussusception is diffuse B cell NHL[4]. Presentation of intestinal lymphoma is of nonspecific abdominal pain, weight loss, fever, constipation and rarely presents as acute abdomen with obstruction/ perforation.

We present here a rare case of adult ileo-ileal intussusception in a young adult 21 years male, caused by B cell lymphoma of small bowel.

Case report:
A 21 year adult male presented to the casualty with a history of pain abdomen for 3 days, followed by vomiting and mild abdominal distention. He had history of passing a worm with his vomitus (after onset of pain abdomen). His vitals were stable except for mild dehydration and low grade fever. The patient was haemodynamically stable with only mild abdominal distention assosciated with features suggestive of sub-acute intestinal obstruction. Patient was started on IV fluids, antibiotics and kept nil orally with Ryle's tube aspiration. After 24 hours, his findings progressed to frank intestinal obstruction with dehydration, tachycardia, and foul Ryle's tube aspirate, increasing abdominal distention without any guarding / rigidity and increased peristaltic sounds. X-ray erect abdomen revealed multiple air fluid levels and USG abdomen suggested intestinal obstruction without pinpointing the possible cause. With a clinical suspicion of small intestinal obstruction, the patient was taken up for emergency exploratory laparotomy. On opening, proximal ileal loops were moderately distended and an ileo-ileal intussusception was detected about 50 cms from the ileo-caecal junction. The gut was viable but reduction was not successful due to the presence of a fleshy mass in the ileum. Segmental resection with end to end ileo-ileal anastamosis was done after decompression of the proximal ileal loops. No other anomaly or any lymphadenopathy was detected. Patient had an uneventful post operative period. The resected segment comprised of a fleshy mass which on histopathologicaly was reported as malignant small round cell tumour, possibly lymphoma. Immunohistochemistry showed tumour cells strongly positive for CD 20, BCl 2 and Ki67 and negative for CD10 and TdT consistent with high grade B cell NHL of intestine. Post operatively patient underwent whole body CT scan which suggested multiple mesenteric lymph node enlargement, largest measuring 10mm in short axis. Bone marrow aspiration revealed megaloblastic anaemia.

Discussion:
Intussusception occurs when a proximal segment of bowel (intussusceptum) telescopes into the lumen of an adjacent distal segment (intussuscepiens) anywhere along the gastrointestinal tract. The first report was made in 1674 by Barbette of Amsterdam[5] and in 1789 John Hunter gave a detailed report about intussusception or "introsusception" as it called then[6]. In 1871, Sir Jonathan Hutchison was the first to successfully operate on a child with intussusception.

The exact mechanism of intestinal intussusception is still unclear. However any lesion in the bowel wall or within the lumen that alters normal peristaltic activity, initiates an invagination. Ingested food and subsequent peristaltic activi-

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ty of the bowel produces an area of constriction above the stimulus and relaxation below, thus telescoping the lead point through the distal bowel lumen.

Adult intussusception can be classified according to etiological factors- primary/ idiopathic and secondary. Primary adult intussusception accounts of about 8% to 20% of cases- mostly occur in small intestine. Secondary intussusception more commonly seen in adults is associated with a pathological condition which is the causative lead point like benign polyp, sub mucosal lipoma, Meckel's diverticulum or malignant tumours like adenocarcinoma, GISTS, lymphoma or Carcinoid tumours.

According to its location adult intussusception can be classified into four categories- ileo-ileal, ileo-ceacal, ileo-colic or colo-colic. Enteric and colonic types are those that are confined to small gut and large gut respectively, while in ileocolic type the ileum prolapses through the ileocolic valve into the colon and constitutes 15% of all intussusceptions.[7]

It is rare in adults (5%) but it is the most common cause of obstruction in infants aged 6-18 months[1]. The characteristic triad of abdominal pain, palpable mass and blood in the stool is rarely seen in adults (less than 25% of the cases). Most patients present with sub acute (24.4%) or chronic (51.2%) symptoms of pain abdomen, nausea, vomiting and constipation, while 6-25% presents with complications of the disease like obstruction, haemorrhage or perforation[4]. Hence preoperative diagnosis is difficult. Barium enema, CT scan and Colonoscopy are helpful in diagnosis, with Barium enema being the most important diagnostic tool [8-10] . It shows the characteristic "coiled spring" or the "spiral sheath" appearance or crescent shaped appearance at the level of obstruction. Flexible sigmoidoscopy or colonoscopy is of paramount importance in evaluating the intussusception presenting with sub acute or chronic small bowel obstruction.

There is no universal approach to the treatment of adult intussusception, but the advocated treatment of choice is surgical resection of the intussusception without intraoperative reduction. The objections to reduction are because of (a) intraluminal seeding and venous embolization of malignant cells into the region of ulcerated mucosa. (b) Possible perforation during manipulation, (c) increased risk of anastomotic complications in the oedematous and inflammed bowel[11]. For small bowel intussusception, initial reduction of externally viable bowel is recommended. The incidence of malignancy in small intestinal intussusception ranges from 1-40%, and the vast majority are metastatic [11,12]. Thus the recommendation of initial reduction and then resection, allowing bowel preservation is prudent.

Four main sites of origin of gastrointestinal Non Hodgkin's Lymphoma (NHL) is distinguished at diagnosis. The most frequent location is the stomach (74%), followed by small bowel lymphoma (8.2%), ileo-caecal region (7.2%) and rarely only the duodenum or colon. Pain is the main diagnostic symptom (76%), followed by loss of appetite (46%), loss of weight (35%).Perforation and obstruction are rare initial manifestation. B symptom (fever, night sweat, loss of weight) occur in about 25% of cases[13].

The pathological diagnosis is made according to the Revised European – American Lymphoma (REAL) Classification or the WHO classification. In Ann Arbor classification ,the suffix 'E' should be restricted to describe invading growth , where as the suffix 'X' was proposed for extra nodal origin, followed by information of the actual stage (e9 X [stomach] II)[14].

Gastrointestinal NHL as such can be considered as a localized disease with the exception of multiple GI involvement.

Stage I- disease confined to intestine.

Stage II- disease extending to local (II-1) or distant nodes (II-2).

Stage II E- disease involving adjacent organs or tissues.

Stage III + IV- disseminated extra nodal involvement/ concomitant supradiaphragmatic lymph node involvement

Stage III & IV form about 16% of cases, while 70% cases are diagnosed in the very localized stages I E and II E. Stage II E was divided by Mushhoff et al into stages II 1E [involvement of regional lymph node only(gastric/mesenteric)] and stage II 2E [involvement of distant lymph nodes(para-aortic/paracaval)][15].

The ileo-caecal region is the most common site of involvement (40%) of primary sites and is mainly affected by B cell lymphomas (95.7%). T cell lymphomas are extremely rare in this region (4.3%) while involvement of the jejunum is more common (12.5%).

The optimal treatment strategy for intestinal lymphoma is still unclear. Primary surgical treatment has the most favourable influence on failure free survival in localised disease. And hence the resection may be appropriate as primary treatment[16]. The effectiveness of adjuvant therapy for localized NHL remains unclear because some cases can be cured only by surgical excision[17,18].

Combination of cyclophosphomide, doxorubicin, vencristin and prednisolone, given every 3 weeks (CHOP-21) is standard chemotherapy for aggressive lymphomas. CHOEP (CHOP with Etoposide) achieves better complete remission (87.6% versus 79.4%) and 5 year event free survival rates (69.2% versus 57.6%). CHOEP should be preferred regimen for young patients with good prognosis (normal LDH level) aggressive lymphomas[19]. A new anti CD-20 monoclonal antibody, Rituximab, is effective in the treatment of B cell lymphoma with slight adverse effects[20].

Conclusion: - We report a case of malignant lymphoma causing intussusception (ileo-ileal type) in a 21 year old adult male. High index of suspicion is required to diagnose adult intussusception in a case of intestinal obstruction, and adequate surgical treatment with resection anastamosis only (and not reduction) should be followed due to the high incidence of malignancy.

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