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GASTRO-INTESTINAL LYMPHOMA PRESENTING AS INTUSSUSCEPTION

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ABSTRACT

Extranodal Lymphomas are most commonly located in gastrointestinal tract. The most common cause of small intestinal obstruction in children is intussusception. However GI lymphomas are a rare cause of intussusception. So we are reporting a case of extranodal lymphoma in a female child presenting as intussusception of small intestine. a 9 years old female child presented with features of subacute intestinal obstruction. On examination a lump was felt in the abdomen. On further evaluation, a computed tomography (CT) revealed a possibility of lymphoma. The bowel segment was excised and sent for histopathological examination.
On gross examination a growth was seen along with satellite lesion. 10 lymph nodes were also identified. Representative sections were taken. Microscopic examination showed the presence of medium size to large size lymphoid cells. After immunohistochemical staining a diagnosis of Non Hodgkins Lymphoma (Extranodal Marginal Zone) was made. 1 lymph node was positive for tumour invasion while other 9 were showing reactive hyperplasia. Since both lymphomas and intussesception are common in pediatric age group, lymphoma should always be kept as a differential diagnosis while dealing with a case of intussusception in pediatric age group.

KEYWORDS: Gastrointestinal lymphoma, Intussusception, Gastrointestinal obstruction.

INTRODUCTION

Gastrointestinal tract accounts for 5-20% of all cases of extranodal Lymphoma (1).

Primary lymphoma of gastrointestinal tract constitutes only about 1%-4% of all gastrointestinal malignancies (2).

Lymphoma of gastrointestinal tract most commonly occur secondary to widespread nodal diseases and the most commonly involved site is stomach followed by small intestine and ileocecal region (3).

Most commonly observed are two histologic subtypes -MALT and DLBC (4). As compared to gastric lymphoma, prognosis of intestinal lymphoma is poor (5).

NHL occurs in age group of 5-15 years in children (6).

Intussusception is second most common cause of obstruction in children and it is the most common cause of small bowel obstruction in children (7).

Lymphoma is very rare cause of intussusception. In between 2000 to 2011 only 36 cases have been reported in literature (8). Intussusception cause 5% of cases of obstruction in adults and 1% of all bowel obstruction and are idiopathic (9).

CASE REPORT

9 year old female child came to outpatient department with complaint of pain in whole abdomen since 2 months. Pain was intermittent, relieved on medication, associated nausea and 6 vomiting episodes

No significant family history.

No similar complaints in past.

General condition was poor condition with poor nutrition. Vitals were normal

Local examination showed distended abdomen, umbilicus centrally placed, no scar mark, no dilated vessel.

On palpation abdomen was soft and lump was felt 4x5 cm in size which was mobile and hard in consistency.

No fluid thrill, no shifting dullness was noticed.

CBC was within normal limits. Bilirubin was 1.1, SGOT 177, SGPT 40, ALP 222. All other investigations were also within normal limits.

Ultrasonography of whole abdomen revealed a well-defined, rounded hypoechoic lesion with internal echoes and debris in epigastrium adjacent to left lobe....?Abscess .Dilated bowel loop loaded with fluid, fecal matter giving doughnut appearance with

collapsed distal bowel. So intussusception was diagnosed on ultrasonography.

Abdominal and pelvic computed tomography with iv contrast of showed the following findings: Long segment of colocolic intussusception of transverse colon. No obstruction/bowel ischemia. Bulky non necrotic retroperitoneal, peripancreatic and mesentric lymph nodes with enhancing lesion in left lobe of liver and in left kidney. Hence a possibility of lymphoma was given.

GROSS: Specimen of intestine was received with attached mesentery, appendix, lymph node & intussusception part altogether measuring 13x11x3.5cm.

A growth measuring 6x5x2cm was seen close to one margin (1cm). Cut surface was grey-white with solid areas and few haemorrhagic points. A satellite lesion was present 8.5cm away from one margin measuring 4x2.5x2cm.

Appendix with attached mesoappendix was present 7 cms from one margin and measured 4cms in length. 10 lymph nodes were identified.

On microscopic examination of the growth, intense infiltration by atypical medium to large lymphoid cells was seen invading upto serosa.

Atypical cells were centrocyte-like having cleaved and non-cleaved nuclei with moderate to scant cytoplasm.

Adjacent tissue and resected margins were lined by tall columnar epithelium with basal nuclei and mild lymphoplasmacytic infiltrate.

DIFFERENTIALS

MARGINAL ZONE LYMPHOMA	Monotypic sIg+, cIg+/-, (IgM>IgG or IgA), CD20+, Cd5-, CD 10-, bcl6-, bcl2+,CD43-/+,cyclin D1-
DLBCL	Monotypic s Ig+, CD 20+, Bcl 6-, CD10-/+, CD43+/-
Mantle cell lymphoma	Monotypic sIgMD +, Cd20+, CD5+, CD10-, CD43+, cyclin D1+
Follicular lymphoma	Monotypic sIg+,CD 20+, Cd10+, bcl 6+,bcl 2+, CD5-, CD43-, cyclinD1-
Burkitt Lymphoma	Monotypic sIgM+, Cd20+, CD10+, bcl 6+, bcl 2-, Ki67=100%

A final diagnosius of Non-Hodgkins lymphoma ?? Marginal zone lymphoma of ileo-caecal region was made. Appendix was unremarkable. One lymph node was positive for malignant cells whereas nine were showing reactive lymphadenitis. Resected margins were free from malignant cells.

DISCUSSION

Intussusception is a very common pediatric surgical emergency. Intussusception is defined as the telescoping of a proximal segment of the intestine within the lumen of the adjacent segment and was first reported in1674. (10)

Worldwide incidence is approximately one to four in 2000 infants and male: female ratio 2:1 or 3:2 ratio.75% cases occur within 2 years of life and 90% within 3 years of age. 1.5–12.0% of cases show underlying pathological causes of intussusception. (11). Meckel's diverticulum, polyps, duplications, mesentery cysts, intestinal hematoma and lymphoma are the frequent causes (12).Lymphoma, although uncommon, causes the most concern due to its malignant nature, and represents 6.5% of pathologic lead points of intussusception in children (11).

In Children GI lymphoma typically presents with diffuse extranodal disease while adults show primary nodal disease commonly. Primary malignancy of gastrointestinal tract is rare in children, with limited information from Asian population.(13)

The male to female ratio of childhood GI NHL is reported to be from 7:1 to 1.8:1(14). Primary lymphoma of GI tract accounts for 30-40% of extranodal lymphomas and comprises 10-15% of all non-hodgkins lymphomas (15) .

Almost half of the children with GI NHL have tumor infiltrates confined to GI tract with possible regional lymph node involvement(16)

A higher prevalence of DLBCL amongst primary pediatric GI lymphomas (17) .

Marginal zone lymphomas are indolent lymphomas that arise at the post-germinal center stage of differentiation. They are found to be morphologically and clinically heterogeneous. According to the 2008 WHO classification there are 3 major categories: nodal marginal zone lymphoma, extranodal marginal zone lymphoma of mucosa-associated lymphoid tissue,and splenic marginal zone lymphoma(18).

Extranodal marginal zone lymphomas (MALT lymphoma) occur more frequently than nodal marginal zone lymphomas among adults, and extranodal marginal zone lymphomas are associated with specific genetic aberrations not present in nodal or splenic marginal zone lymphomas (19,20) Marginal zone lymphomas lack a specific immunophenotype unlike other B cell lymphoma.

Thus, the recognition of specific genetic aberrations may help in greater diagnostic accuracy, or provide additional prognostic data. One example is t(11;18)(q21;q21) in gastric MALT lymphomas (19). Its presence is associated with a lack of regression of the lymphoma upon eradication of H. pylori (21,22).

Symptoms may range from dyspepsia, epigastric pain, abdominal pain, nausea, vomiting to diarrhea, weight loss, malabsorption, obstruction, anemia and to a lesser extent ulceration, perforation and intussusceptions (23-25)

B-cells are most commonly derived from the lymphoid tissue of the lamina propria and submucosa of the ileum, which has greatest concentration of gut associated lymphoid tissue. (26,27).

Volumetric doubling of this malignancy leads to an acute abdomen presentation which may mimic other diseases while some presentations may mimic acute appendicitis (28,29).

Immunohistochemistry: CD19 and CD20, CD43 was coexpressed in 70% of atypical cells, and negative for CD5, CD10, CD23, and CD3. 48% cases show light chain restriction by immunohistochemistry, while 84% show monoclonal IgH gene rearrangement. CD10 and BCL6 were negative in tumor cells however residual non-malignant germinal centers were seen. BCL2 was positive in 42% and p53 was negative (30).

The ultimate treatment approach in GI lymphoma remains debatable. Recent studies have shown use of chemotherapy alone as an effective treatment option in primary GI lymphoma in all stages (31).Surgery is reserved for complications such as obstruction, perforation, and bleeding. Disease stage at presentation and extent of involvement remain the most important criteria determining survival (32,33). Study showed that advanced Stage (III and IV) at presentation and diffuse disease at presentation carry increased mortality.

The overall 5- and 10-year survival rates of pediatric GI lymphoma cases treated with surgery and combined chemotherapy and radiation therapy is 52% and 44%, respectively. Study showed 61.9% patients are free of disease and on follow-up.

Differential diagnosis includes enteritis, inflammatory bowel disease, leiomyoma, leiomyosarcoma, or gastrointestinal stromal tumor.

CONCLUSION

A diagnosis of lymphoma should always be kept in mind while dealing with case of intussusception in children.

DECLARATIONS

Ethics approval: Ethical approval obtained from institutional ethics committee of Eras Lucknow Medical College and Hospital.

Consent for publication: Consent was given the parents/guardians of the patient.

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