CASE REPORT

Non-Hodgkin's lymphoma of caecum in children

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Abstract:
Lymphomas do not only arise in lymph nodes or other areas of the lymphatic system, but many arise (or present) in non-lymphoid sites such as the skin and gastrointestinal tract. The caecum is the one of rare extra-nodal site for lymphoma. It has low incidence but favorable outcome. Caecal involvement is very rare than other sites of gastrointestinal tract like stomach and spleen or MALT associated lymphomas. Here we report the successful surgical management of caecal Non-Hodgkin's lymphoma.

Keywords: Caecal Non-Hodgkin's Lymphoma, Extranodal, Lymphoma

Introduction:
Primary gastrointestinal (GI) non-Hodgkin lymphoma (NHL) is a heterogeneous group of B- and T-cell lymphoid malignancies. The clinical features, management, and prognosis of these lymphomas differ from lymphomas of lymph node origin. Here we report a successful surgical management of extra nodal caecal Non-Hodgkin's lymphoma in 7 years old child.

Case Report:
A 7 years old male child referred to us, with intermittent abdominal pain for couple of months and abdominal mass. On examination patient was looked pale but not anemic. Occasional vomiting.

On palpation abdomen was not distended, soft, rounded mass about 6X6 cm was felt just area between right hypochondrium and right lumber area, non tender, mobile, soft to firm.

Ultrasound was done which showed a complex mass at right lumber area just above the kidney, partly cystic and partly solid.

CT scan showed an space occupying lesion adjacent to caecum and ascending colon. There was no obstruction of the gut. No other organs were involved.

Exploratory laparotomy was done. Findings were intussusception of distal ileum and appendix in the caecum. Reduction was done. The caecal mass seen, which involving the posterior surface of caecum along with two large enlarged lymph nodes in the mesentery. Limited right hemicolectomy done with free margins and end to end anastomosis done as shown in figure 1-6. Post operative recovery was uneventful. Specimen was sent for histopathology and been reported as Non-Hodgkin's lymphoma of low grade with reactive changes in the lymph nodes as shown in figure 7 and 8.

Discussion:
Lymphoma is a malignancy of the lymphatic cells of the immune system. Typically, lymphomas present as a solid tumor of lymphoid cells. Treatment might involve chemotherapy and in some cases radiotherapy and/or bone marrow transplantation, and can be curable depending on the histology, type, and stage of the disease.

Extranodal presentation of non-Hodgkin's Lymphoma is rare; the gastrointestinal tract is the

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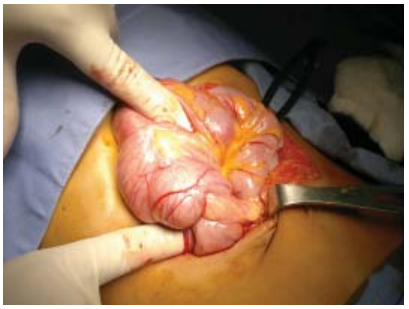
Figure 1: *Ileocolic intussusception*

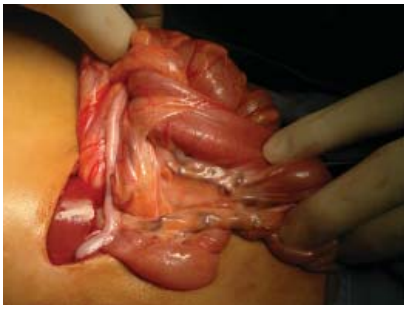
Figure 2: *Reduction of intussusception*

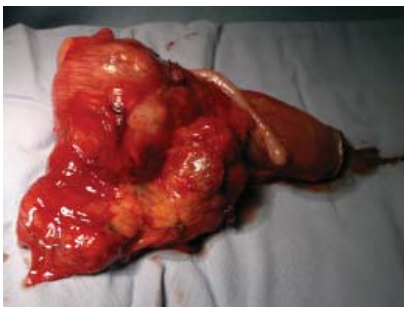
Figure 3: *Resection of caecal mass*

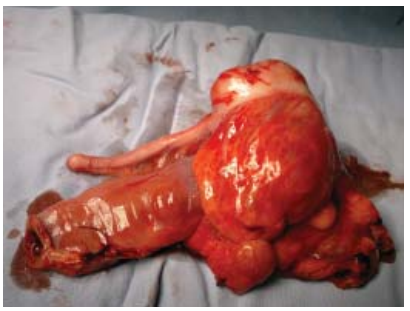
Figure 4: *Caecal mass with lymph nodes and appendix*

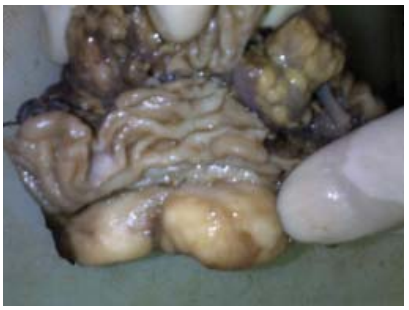
Figure 5: *After staining of caecal mass*

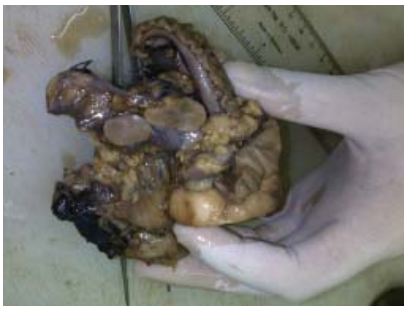
Figure 6: *Lymph node staining*

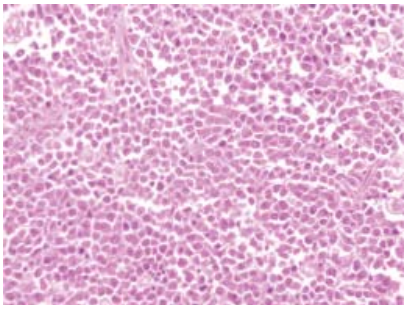
Figure 7: *Cellular proliferation*

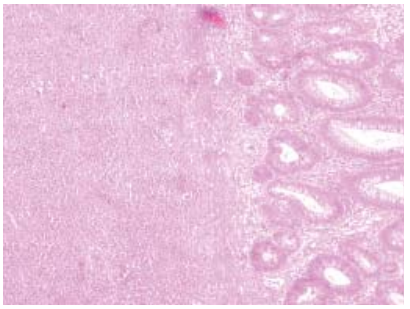
Figure 8: *Submucosal involvements of malignant cells*

most common site of extranodal involvement.

Lymphomas of the GI tract are the most common type of primary extranodal lymphomas, accounting for 5-10% of all non-Hodgkin's lymphomas. In particular, primary intestinal lymphomas represent about 15-20% of GI lymphomas. GI lymphomas are predominantly located in the stomach (50-60%), whereas intestinal lymphomas are more infrequent and appear in the small bowel (20-30%), the colon, and the rectum (10-20%)[1,2,3,4], but are rarely seen in children.[5]

The most common symptoms of colonic lymphoma are abdominal pain, nausea, vomiting, weight loss, abdominal mass, change in bowel habits, hematochezia, obstruction, intussusceptions,[6,7] and acute peritonitis due to intestinal perforation. The lack of specific complaints and the rarity of intestinal obstruction probably accounts for the delay in diagnosis. These bulky masses can usually be palpated by simple physical examination and viewed by ultrasonography.

Combined modality of approach that includes surgical debulking and systemic chemotherapy is the preferred treatment in more advanced cases.[8] Surgery alone can be considered as an adequate treatment for patients with low-grade NHL disease that has not infiltrated beyond the submucosa.[8]

However, it is still thought that the prognosis of intestinal lymphomas is related to surgery; therefore, it seems appropriate and cautious to resect intestinal lymphomas whenever possible.[9,10] Those with limited stage disease may enjoy prolonged survival when treated with aggressive chemotherapy. Radiotherapy is beneficial for incomplete resection or non resectable disease.

Conclusion:

Primary colonic lymphomas are rare; the cecum is the most common site of occurrence. Early diagnosis may prevent intestinal perforation; however, the diagnosis is often delayed in most cases. Surgical resection is the mainstay of treatment for localized primary lymphomas, followed by postoperative chemotherapy. Surgery alone can be considered as an adequate treatment for patients with low-grade Non-Hodgkin's lymphoma disease that does not infiltrate beyond the sub mucosa. Although resection plays an important role in the local control of the disease and in preventing bleeding and/or perforation, it rarely

eradicates the lymphoma by itself.

References:

1. Bairey O, Ruchlemer R, Shpilberg O. Non-Hodgkin's lymphomas of the colon. Isr Med Assoc J 2006; 8:832-5.
2. Zinzani PL, Magagnoli M, Pagliani G, Bendandi M, Gherlinzoni F, Merla E, et al. Primary intestinal lymphoma: Clinical and therapeutic features of 32 patients. Haematologica 1997; 82:305-8.
3. Doolabh N, Anthony T, Simmang C, Bieligk S, Lee E, Huber P, et al. Primary colonic lymphoma. J Surg Oncol 2000; 74:257-62.
4. Gonzalez QH, Heslin MJ, Davila-Cervantes A, Alvarez -Tostado J, de los Monteros AE, Shore G, et al. Primary colonic lymphoma. Am Surg 2008; 74:214-6.
5. Zhang KR, Jia HM. Primary non-Hodgkin's lymphoma of the sigmoid colon in a child. Am J Surg 2009; 197:e11-2.
6. Chiang JM, Lin YS. Tumor spectum of adult intussusception. J Surg Oncol 2008; 98:444-7.
7. Zubaidi A, Al-Saif F, Silverman R. Adult intussusception: A retrospective review. Dis Colon Rectum 2006; 49:1546-51.
8. Tondini C, Giardini R, Bozzetti F, Valagussa P, Santoro A, Bertulli R, et al. Combined modality treatment for primary gastrointestinal non- Hodgkin's lymphoma: The Milan cancer Institute experience. Ann Oncol 1993; 4:831-7.
9. Gobbi PG, Ghirardelli ML, Cavalli C, Baldini L, Broglia C, Clç V, et al. The role of surgery in the treatment of gastrointestinal lymphomas other than low grade MALT lymphomas. Haematologica 2000; 85:372-80.
10. Waisberg J, Bromberg SH, Franco MI, Matheus CO, Zanotto A, Petrolino LF, et al. Primary non-Hodgkin's lymphoma of the right colon: A retrospective clinical-pathological study. Int Surg 2001; 86:20-5.