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CASE REPORT

NON-HODGKIN'S LYMPHOMA PRESENTING AS ACUTE INTESTINAL OBSTRUCTION: A CASE REPORT

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ABSTRACT:

Non-Hodgkin's lymphoma comprises about 7% of all paediatric cancers. The gastrointestinal tract is the most common site of primary extra-nodal lymphomas. In people under 20 years of age, lymphoma, the most common malignant neoplasm of GI tract, is almost universally non-Hodgkin's lymphoma (uncommon before age of 5 years). It is 2-3 times more common in male than female. Non-Hodgkin's lymphomas are classified as low, intermediate, or high grade on the basis of their pathological features. Low- and intermediate-grade tumours are commonly seen in adults, while high-grade non-Hodgkin's lymphoma are commonly (>90%) found in children. This occurs mainly due to the mutation at the time of cellular maturation and function of the immune system. High-grade non-Hodgkin's lymphoma comprises three histologic subtypes: small non-cleaved-cell, lymphoblastic, and large-cell lymphoma. Among children with small-noncleaved-cell tumours, both Burkitt's and non-Burkitt's lymphomas have been identified, with no clinical relevance so far[1].

Keywords: Non-Hodgkin's lymphoma; Burkitt's lymphoma; intussusception

INTRODUCTION:

Clinical presentations of non-Hodgkin's lymphoma in children depends upon the primary site of tumour, extent of the disease and its histological sub-type. The disease is mainly extra-nodal involving the abdomen, head and neck and the mediastinum. Sporadic cases of Burkitt's lymphoma typically involve the abdomen or head and neck. Abdominal tumors are often associated with pain, nausea, and vomiting resulting from intestinal obstruction caused by direct compression of the bowel lumen or by intussusception[1]. Due to the hematogenous dissemination early in the course of non-Hodgkin's lymphoma in children, systemic treatment is the mainstay of therapy[1]. Role of surgical intervention is very limited, mainly for diagnostic purposes and infrequently for cases in which complete resection of a gastrointestinal tract tumor is possible. The tumor in that case is down-staged, and the patient is then treated with less intensive chemotherapy

CASE PRESENTATION:

A 6 years old male child presented with complaints of abdominal pain since four months, distension, multiple episodes of vomiting and not passing stools since three days. On physical examination patient had tachycardia and pallor but was normotensive. There was no generalised lymphadenopathy. His abdomen was distended and tender on palpation. There was flank fullness, umbilicus was shifted upward and everted. Dilated cutaneous veins were visible all over the abdomen. There was no presence of free fluid. On visual analogue scale his score was 6/10. There were no bowel sounds heard on auscultation. Per rectal examination was normal. Nasogastric aspirate was bilious. Haematological investigations were within normal limits. Contrast enhanced computed tomography was suggestive of colo-colonic intussusception with conglomerated mesenteric lymphadenopathy. Considering the examination and imaging findings, possibility of abdominal Koch's or lymphoma was thought of. Patient was posted for exploratory laparotomy. Intraoperatively, small bowel appeared dilated with collapsed ascending colon. Ileocolic intussusception was found and was difficult to reduce. Old sealed off perforation was found in the part of terminal ileum which had telescoped into the caecum with feculent matter discharging from the caecal perforation site. Approximately 3 x 2 cm large paracolic lymph node was found which was excised **(Fig no 1,2).**

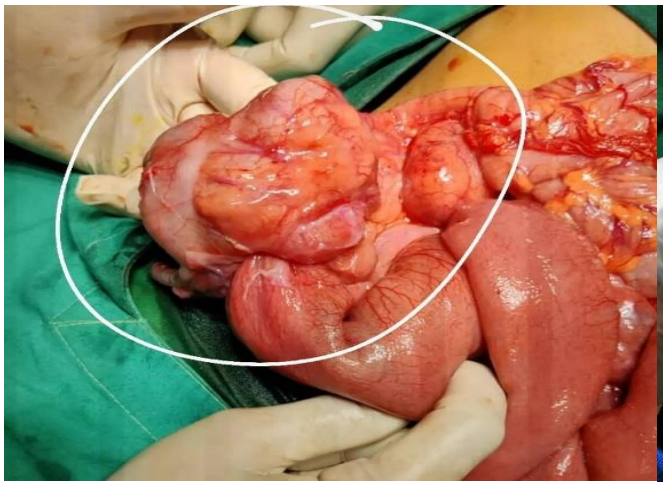

Fig no 1- ileo-colic intussusception

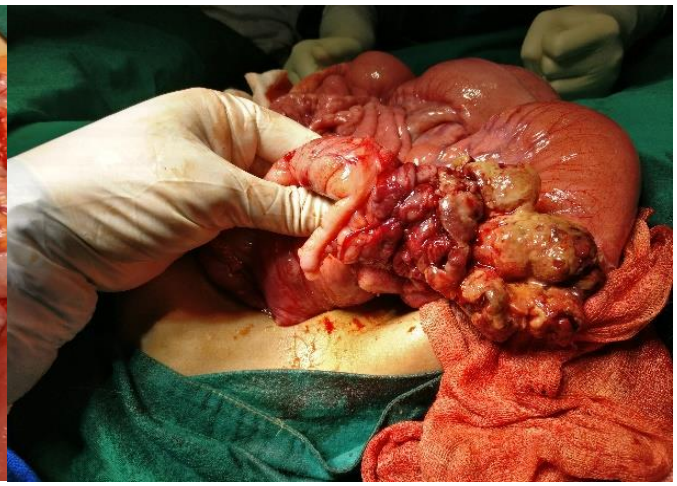

Fig no 2- old sealed perforation with feculent matter discharging

There was no evidence of retroperitoneal lymphadenopathy. Liver and spleen appeared normal. Right hemicolectomy was performed with ileo-transverse colic anastomosis **(Fig no 3).**

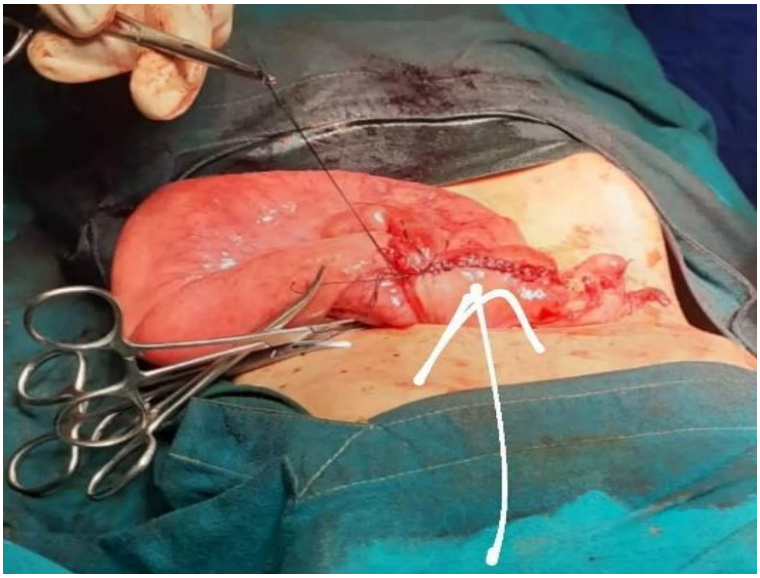

Fig no. 3- Ileo-transverse colic anastomosis

Resected bowel with the lymph node were sent for histopathology and immunohistochemistry (IHC). Histopathology of colon specimen and conglomerated mesenteric lymph node showed features of high- grade Non-Hodgkin's Lymphoma (Burkitt's lymphoma) with reactive lymphoid hyperplasia **(Fig. 4,5,6)**

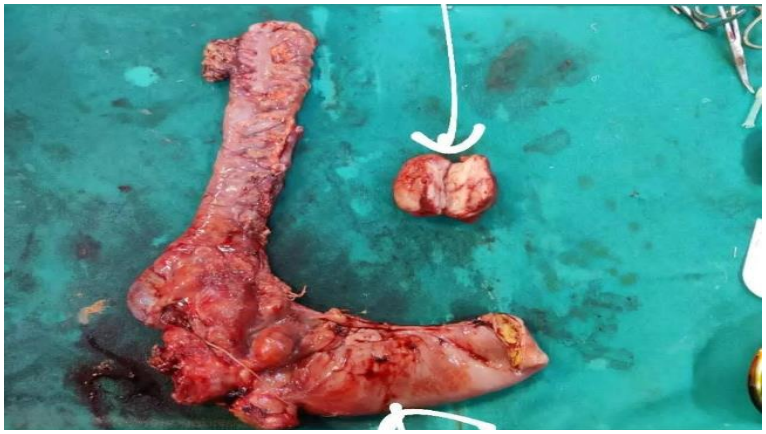

Fig no.4- Specimen of resected colon with mesenteric lymph node

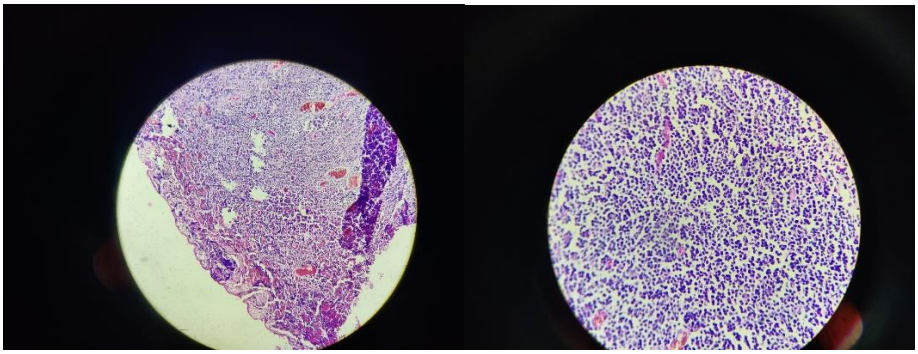

Fig no.5- features of lymphoma **Fig no. 6- 'starry-sky' appearance of Burkitt lymphoma**

On IHC, cells were positive for CD 20, Bcl 6, CD 10, EBV-LMP and showed focal reactivity for c-myc, Ki-67 index was 100%**.** CSF cytology showed total leucocyte count was 5 cells/cu.mm. No blasts were seen. Bone marrow aspiration was within normal limit**.** LDH was 432 U/L. PET-CT scan before starting adjuvant chemotherapy showed hypermetabolic heterogeneously enhancing aortocaval/mesenteric lymph nodal mass noted in the precaval region extending from right inferior pole of kidney into the bifurcation of aorta along the right common iliac vessels and extending into lower anterior abdomen closely abutting the adjacent small bowel loops. Focal increased metabolism along the right lobe of liver was noted (indeterminate in aetiology). Focal hypermetabolism involving midshaft of right femur – suspicious of lymphoproliferative malignant aetiology was noted. 2D echocardiography before starting adjuvant chemotherapy showed normal study with left ventricular ejection fraction of 66%. Risk stratification was done before starting chemotherapy and based on St. Jude's staging for lymphoma, he was classified as stage IV R3.

Adjuvant chemotherapy:

Patient was further managed as per BFM-NHL 95 protocol involving pre-phase, followed by intensive chemotherapy. Pre-phase chemotherapy was given with one cycle of cyclophosphamide, methotrexate, cytarabine and prednisone. This was followed by intensive chemotherapy over a period of 6 months and following agents were administered- vincristine, ifosfamide, etoposide, methotrexate, cytarabine, cyclophosphamide and doxorubicin. Immunotherapy with rituximab in each cycle of intensive phase chemotherapy was found to be effective. Serum potassium, phosphorus, calcium and uric acid repeated at regular intervals were within normal limits thus, ruling out tumour lysis syndrome. PET-CT after pre-phase chemotherapy showed some degree of remission. Complete remission was achieved post chemotherapy, wherein, there were no evidence of hypermetabolic lesion in the right lobe of liver and mid-shaft of right femur and negative aortocaval/precaval, external iliac nodal status.

DISCUSSION:

Non-Hodgkin's lymphoma comprises 7% of all paediatric tumours with gastro-intestinal tract as the most common site of extra-nodal lymphomas. Lymphomas are the most common malignant neoplasm of the GIT tract for children under 20 years of age with non-Hodgkin's lymphoma being the most common type. Paediatric gastrointestinal tumours are classified based on their tissue of origin into lymphoid, epithelial and mesenchymal. Lymphoid tumours are further classified as lympho-nodular hyperplasia and lymphoma. While ileum and colon are the most common sites of lympho-nodular hyperplasia, lymphoma commonly involves ileum, appendix and colon. Non-Hodgkin's lymphomas in general and abdominal undifferentiated tumour in particular have

rapid doubling times which can be as short as 12 hours[2]. Because tumour burden steadily increases with time it is imperative that chemotherapy be instituted promptly. Delay, because patients are recovering from surgical complications, will have significant negative effects on outcome. As is evident in this case, where in the patient was in relapse at the time of referral to oncology centre for adjuvant chemotherapy. Surgery plays an important role when tumour is completely resectable and there is lesser tumour burden. It also helps in the exact histo-pathological diagnosis and further staging of the tumour. Different protocols are used for treating NHL in children. LSA2-L2 protocol involves cyclophosphamide, vincristine, prednisone, daunomycin, methotrexate, cytosine arabinoside, thioguanine, L-asparaginase, BCNU and hydroxyurea in an induction, consolidation, and maintenance schedule[4]. COMP regimen consists of cyclophosphamide, vincristine, methotrexate, and prednisone[5]. The CCG-551 study, which compared these two regimens, showed that LSA2-L2 was more effective for disseminated lymphoblastic tumour and COMP was more effective in undifferentiated lesions. Our patient was however treated with Berlin–Frankfurt–Münster-95 (NHL-BFM-95) protocol[6]. Rituximab, when added to chemotherapy regimen prolongs overall survival for adult B-cell cancer. Based on the study conducted by Veronique Minard-colin et al, rituximab was added to the chemotherapy for this case[3]. Post-remission, 6 monthly follow up was taken which has been found to be event-free

CONCLUSION:

Emergency surgical intervention in children suffering from high-grade Non-Hodgkin's lymphoma (Burkitt's lymphoma), presenting with acute abdomen, favourably affects the outcome and overall survival. However, chemotherapy must be started as soon as the patient is fit for it post-operatively, owing to the high mitotic index of the disease. Rituximab was found to be a safe immunotherapy agent along with chemotherapy leading to an event-free survival.

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