Abstract:
Burkitt lymphoma is a highly malignant, aggressive and rapidly growing B-cell neoplasm. The abdomen is the most frequent onset site of non endemic Burkitt lymphoma. In children it usually presents as abdominal mass. Presented here is a case of Burkitt lymphoma presenting as intussusception.

Keyword: Burkitt lymphoma, B cell neoplasm, aggressive neoplasm, abdominal mass, ileum, Diffuse Large B Cell Lymphoma, immunohistochemistry

INTRODUCTION:
WHO classification has defined Burkitt lymphoma as a highly aggressive lymphoma (ICD-O 9687/3). Burkitt lymphoma represents 8-10% of all tumors in children less than 15 years of age. It was first described by Dennis Burkitt in 1958. These neoplasms were designated as Acute Lymphoblastic Leukemia, L3 type by the French-American-British group (ICD-O CODE 9826/3). The abdomen is the most frequent site of onset in non-endemic Burkitt Lymphoma. In children it usually presents as abdominal mass with intussusception.

CASE REPORT:
A three year old male child presented with complaints of abdominal pain, vomiting and refusal of feeds for two days duration. On examination a palpable mass was identified in right iliac fossa. Ultrasound abdomen showed alternating hypo and hyperechogenic bands and the impression was given as intussusception. The patient was posted for laparotomy. On table he was found to have ileocolic intussusception and was reduced manually. An ulceroproliferative growth was found 15cm proximal to ileo colic junction. Resection and anastomosis of ileum was done and specimen was sent for histopathological examination. The post operative period was uneventful. Retrospective clinical examination revealed no generalized lymphadenopathy or hepatosplenomegaly.
Complete haemogram was normal.

**GROSS FINDINGS:**
The external surface of ileal segment revealed focal congestion. On cut opening, the mucosa revealed a grey white polypoidal lesion measuring 2.5x2x1cm with small ulceration measuring 1x0.5cm in the luminal surface. No areas of necrosis or haemorrhage were noted. The lesion was 0.5cm from one cut end and 1.5cm from other cut end. The cut ends appeared normal. No abnormality was detected in the rest of the mucosa. No lymph nodes were made out in the mesentry.

**MICROSCOPIC FINDINGS:**

Histopathological examination revealed neoplastic proliferation of monotonous lymphoid cells arranged in diffuse infiltrative pattern involving mucosa as well as underlying wall up to muscular layer (fig1). Cells were medium sized with round nuclei, coarse chromatin and multiple basophilic nucleoli. 7 to 8 mitoses per HPF were noted. Many tingible body macrophages imparting a starry sky appearance were seen (fig2). No follicle formation or germinal centres were noted. Reed Sternberg giant cells were not seen. It was reported as high grade NonHodgkins Lymphoma- Burkitt lymphoma. The differential diagnosis was Diffuse Large B cell Lymphoma.

**Figure 1 monotonous lymphoid cells (10X)**

**Figure 2 starry sky pattern (40X)**

Immunohistochemistry was done to confirm the diagnosis.

**Figure 3 CD45 positivity (40X) Figure 4 CD20 positivity (40x)**
Immunohistochemical profiles were consistent with Burkitt lymphoma. An important differential diagnosis is DIFFUSE LARGE B CELL LYMPHOMA. It has to be distinguished because of the difference in treatment modality of these highly aggressive neoplasms.

Diffuse large B cell neoplasm most commonly affects adults. 40% being extra nodal occurring in gastro intestinal tract, skin. It is composed of heterogenous population of large cells with round nuclei having vesicular chromatin and multiple prominent nucleoli. Where as Burkitt lymphoma has monotonous population of medium sized cells. Starry sky pattern is usually absent in Diffuse Large B Cell Lymphoma. In our case the slide has predominantly medium sized cells with starry sky pattern because of the interspersed tingible body macrophages. Immunohistochemistry profile in Diffuse Large B Cell Lymphoma include positivity for CD19, CD 79a, bcl 6, bcl 2 and ki67 proliferation index around 40%. CD 10 is negative. Immunophenotypic features of Burkitt Lymphoma include positivity of tumor cells for CD19, CD20 (pan B cell marker) and CD10 (follicular centre cell), negativity for BCL2 and a proliferation fraction measured by Ki-67 index of nearly 100%.

Figure 5 CD 10 positivity (40x)  Figure 6 BCL2 negativity (40x)
CD 45 - strong positivity CD10 - strong membrane positivity CD20-strong membrane positivityKi 67 - >99% Bcl2 - Negative.

FIGURE 7 KI67 Labelling
Our patient’s immunohistochemical profile was CD10 positive, CD20 positive, CD19 positive, bcl2 negative, CD 79a negative and ki67 index more than 99%. With such clinical features, gross findings, histopathology and immunohistochemistry picture a diagnosis of BURKITT LYMPHOMA was arrived.

**DISCUSSION:**

Non-Hodgkin’s Lymphoma (NHL) represents the third most frequent cancer in childhood and usually is subdivided into 3 histological subtypes (1) B-cell NHL including both Burkitt lymphoma and diffuse large Bcell lymphoma (65%); (2) Lymphoblastic lymphoma (20%) and (3) Anaplastic large cell lymphoma (30%). Burkitts lymphoma represents 40%-50% of all NHL cases in childhood, and is more common in males than females. It is one of the rapidly growing tumors. It has a doubling time of 24-48 hours. This type of lymphoma has three clinical forms. Endemic form occurring in Africa affecting jaw bones. (2)Non endemic form occurring throughout the world and presenting as abdominal mass (3) Immunodeficiency associated seen in patients with immunodeficiency states and presenting as lymphadenopathy.

Our patient has a nonendemic form of Burkitt Lymphoma occurring in South East Asian country like India. The age of presentation is usually under 15years. In our case it is being 3 years. Male to female ratio is 2to3:1. Our patient is a male patient.

With reference to ‘clinical implications and surgical management of intussusception in paediatric patients with Burkitt lymphoma’ Gupta H et al presented 33 patients (17.5%) out of 189 patients with primary abdominal Burkitt lymphoma presenting as intussusception. Our case also presents with intussusception.

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<th><strong>BURKITT LYMPHOMA</strong></th>
<th><strong>DIFFUSE LARGE B CELL LYMPHOMA</strong></th>
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<tr>
<td><strong>AGE</strong></td>
<td>CHILDREN (15 years)</td>
<td>ADULTS &gt; 70 YEARS (COMMON)</td>
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<td><strong>MONOTONOUS POPULATION OF MEDIUM SIZED CELLS</strong></td>
<td><strong>HETEROGONEOUS POPULATION OF LARGE CELLS</strong></td>
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<td><strong>PROMINT STARRY SKY PATTERN</strong></td>
<td><strong>STARRY SKY PATTERN IS INFREQUENT</strong></td>
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<tr>
<td><strong>CD19</strong></td>
<td>POSITIVE</td>
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<td><strong>BCL2</strong></td>
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<td><strong>CD 79A</strong></td>
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<td><strong>KI67 INDEX</strong></td>
<td>&gt;99%</td>
<td>USUALLY 40%</td>
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With reference to ‘American Burkitt Lymphoma- a clinicopathological study of 30 cases’ Arseneau et al found that abdomen was the most frequent site of involvement of Burkitt lymphoma. Typically our patient had the site as small intestine. With the above mentioned histopathological and immunohistochemical picture the patient was diagnosed as Burkitt Lymphoma. Further studies used to confirm the diagnosis are cytogenetics which shows the translocation involving the MYC oncogene and immunoglobulin heavy or light chain genes as follows. t(8;14)(q24;q32), t(2;8)(p12;q24), t(8;22)(q24;q11). Cyto genetic studies were not done in this case.

CONCLUSION:
Burkitt lymphoma is a rapidly growing highly aggressive neoplasm requiring rapid diagnosis for initiation of treatment at an early stage. Rightly in our patient the mitotic figures were seven per high power field. The ki67 index was about 99% reflecting the high grade nature of the tumor. However they respond well to surgical resection of the affected portion of intestine and chemotherapy with high dose cyclophosphamide resulting in cure rate of 70-80%. Prompt diagnosis aided in targeted therapy and the patient improved.

BIBLIOGRAPHY:
2. Balanescu et al, Ileocolic intussusception due to Burkitt lymphoma a case report-Journal of medicine and life. volume 6; issue 1; January to March; 2013 61-64.
4. Juan Rosai: Rosai and Ackerman’s surgical pathology 10th edition; volume 2; Elsevier; 1842-1844.

12. Naoya et al; Distinction between Burkitt lymphoma and Diffuse Large B cell Lymphoma with cmyc rearrangement. The US and Canadian academy of pathology. volume 5; no7 ; 771-776.

13. Dan L.Longo, Dennis L.Kasper: Harrison’s principles of internal medicine. 18\textsuperscript{th} edition; volume 1; mc Graw Hill; 1931.