

## Case Report


# Pediatric caecal lymphoma involving ovary in a 12 year old girl - A bad prognostic sign: A rarity

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## Abstract

Primary caecal lymphoma or the colonic lymphoma is a rare tumor of the gastrointestinal (GIT) tract and comprises only 0.2-1.2% of all colonic malignancies, both in adults and pediatric age group. The most common variety of colonic or caecal lymphoma is Non-Hodgkin's Lymphoma (NHL) which arises from the lymphoid elements of the intestine. GIT is the most frequently involved site, accounting for 50-60% of all extra nodal lymphomas, and most of them are NHL. In adults, the stomach is the most common location of GIT lymphomas, followed by the small intestine, but the most common GI site of NHL in children is the terminal ileum and the ileo-caecal region. Diagnosis is difficult since lymphoma presents with vague abdominal pain with loss of weight and appetite. It may present as lump abdomen with complications such as intestinal obstruction, bleeding, perforation and peritonitis and intussusception. Histologically it is B or T cell type with small or large cell variation, but frequently encountered is diffuse large B- cell lymphoma (DLBCL). DLBCL has low incidence but favorable outcome in young adults, lesions localized to one area or organ and children below 5 years of age, but has an aggressive course in children between 10-15 years of age and also in adults above 55 years of age. DLBCL or GIT Lymphomas in general have male preponderance. We present a rare case of caecal lymphoma (DLBCL) involving appendix and right ovary in a 12 year old girl who presented with vague symptoms of abdominal pain and abdominal mass. Clinically and radiologically, provisional diagnosis of Ileo-caecal tuberculosis with possibility of adhesions leading to a mass lesion was considered. Histopathological examination (HPE) revealed the diagnosis and prognosis of the case.

## Key words

Primary cecal lymphoma, DLBCL, B-Cell, CD-20, BCL-6, Ileo-cecal tuberculosis, Pediatric lymphoma, NHL.

## Introduction

Non Hodgkin's lymphoma mostly involves extra nodal lymphoid tissue. It can arise from organs like GIT, thyroid gland, ocular adnexae, waldeyer's ring, adenoids, tonsils, various sites of head and neck region, testis and ovary. GI tract is most common primary site of extra nodal non- Hodgkin's lymphoma (NHL). In adults, stomach is the most common site for NHL followed by small bowel 20-30% and very rarely colon [1]. In the colon, caecum is the most common site. In children between 5 to 15 years of age, terminal ileum and Ileo-caecal region are the most common sites. Gastrointestinal (GI) tract lymphomas comprises only 0.2-1.2% of all colonic malignancies [2, 3]. GI tract lymphoma is more common in males; male to female ratio is 2:1. Clinical diagnosis of GI tract lymphoma is very difficult since patients present with vague symptoms like abdominal pain, loss of weight and loss of appetite. Most of the cases reported till now are diagnosed at the time of surgery or on histopathological examination (HPE). Most of the extra nodal lymphomas belong to the group of Non-Hodgkin's Lymphomas with most common histological variant being Diffuse large B-cell lymphoma (DLBCL). Most of the DLBCLs in adults are encountered in patients with primary or secondary immunodeficiencies. In children they are seen in familial adenomatous polyposis syndromes. Surgical resection of the tumor or right hemicolectomy is the treatment of choice when the lesion is single or involving single area. In cases of higher stage of disease with involvement of surrounding structures, surgical resection followed by adjuvant chemoradiation is treatment of choice. However recurrence rate is high after chemoradiation in GI tract lymphomas [4].

## Case report

A 12 year old girl presented to the pediatric department with complaints of loss of appetite

and loss of weight since 2 months, abdomen swelling since one month, and pain abdomen since 15 days. There was also complaint of fever, diarrhoea and constipation, on and off. Bladder habits are normal. Patient was examined, in view of mass patient was referred to surgical department. On examination, patient was mild pallor, thin built. Local examination of the abdomen showed a mass occupying right iliac, right lumbar, right hypochondriac and umbilical regions. Ballotment of swelling is present, bimanual palpation of swelling is negative. Ultrasound abdomen and pelvis gave an opinion of ?Ileocaecal Tuberculosis which was well documented by Computerized Tomography (CT). Though the preoperative diagnosis was going in favor of inflammatory lesion, in view of large size of the mass, surgical risk was taken and explorative laparotomy was planned. On explorative laparotomy, caecum showed a large mass of size approximately 8.5 x 7 x 5 cm was noted which ruled out the possibility of preoperative diagnosis of tuberculosis. Finally right hemicolectomy and right side oophorectomy was done and the specimen sent for histopathological examination (HPE). Grossly, we received a right hemicolectomy specimen measuring approximately 43 cm in length (**Figure - 1**). A large mass noted in the caecum measuring 8x7x5.5 cm completely occupying the caecum. Wall of the ceacum appears to be involved grossly. Mass located 21 cm from the Ileal resected margin and 22 cm from the colonic resected margin. Also received appendix measuring 7 cm , right ovary measuring 3.5x3x1 cm (**Figure - 2**) , omentum measuring 17x12 cm. Dissected 4 lymph nodes from omentum and caecal fat. Microscopic examination showed histological features of Diffuse Large B-Cell Lymphoma (DLBCL) involving appendix and right ovary (**Figure - 3, 4, 5, 6, 7**). Immunohistochemistry (IHC) study

was done with markers CD-20 and BCL-6 (Figure – 8, 9).

**Figure - 1:** Gross picture of right hemicolectomy specimen along with mesentery. A large solid nodular tumor is seen in the caecum.



**Figure - 2:** Gross specimen of right ovary.

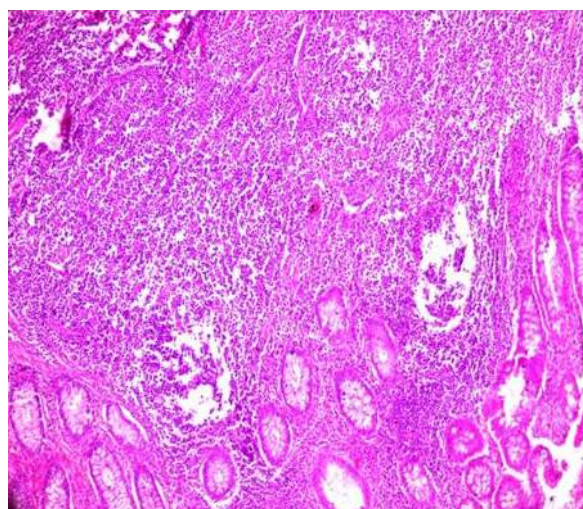


## Discussion

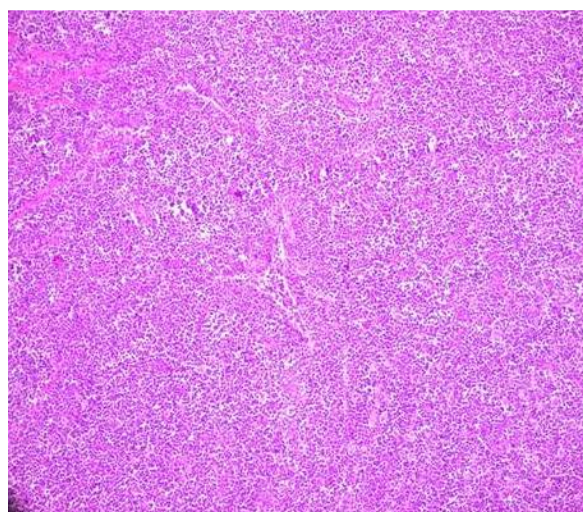
Lymphoma is a malignancy of the lymphatic cells of the immune system. Typically, lymphomas present as a solid tumor of lymphoid cells. Non-Hodgkin lymphoma is the third most common malignancy in childhood. According to Wong, et al. and Bairey, et al., Primary lymphoma of the colon is a rare tumor of the gastrointestinal (GI) tract that comprises only 0.2-1.2% of all colonic malignancies [3, 5].

According to the world literature, 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%) [3, 5, 6, 7]. Colon lymphomas are more frequent in men [3, 7] but are rarely seen in children [8]. When they occur in pediatric age group, they are mostly seen between age 5 and 15 years; commonly involving terminal ileum and ileo-caecal junction and present with vague symptoms which can lead to delayed diagnosis.

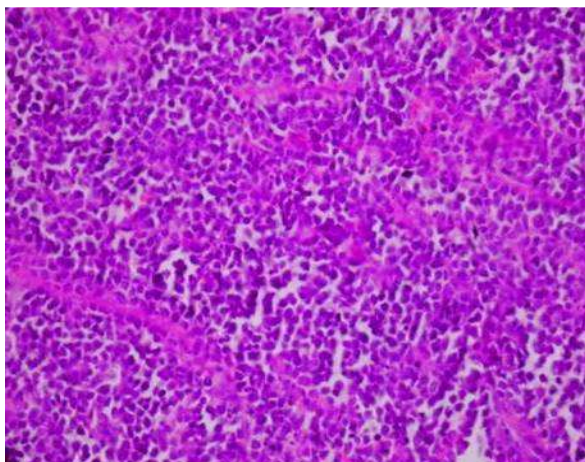
**Figure - 3:** Photomicrograph showing colonic mucosa with submucosal tumor tissue.



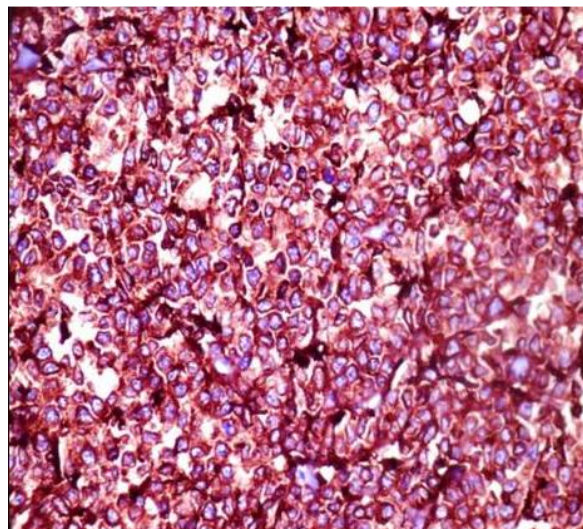
**Figure - 4:** (10X view) Photomicrograph showing tumor tissue arranged in sheets.



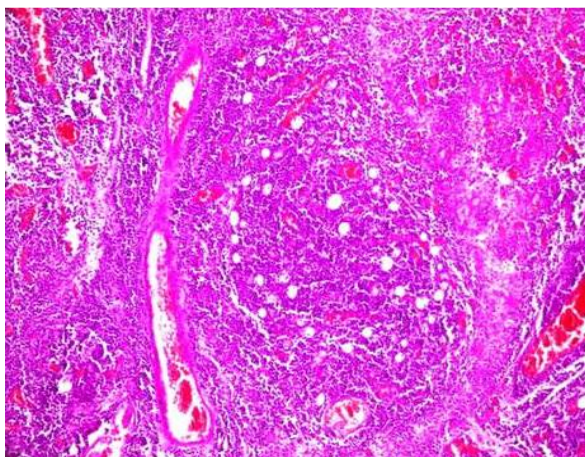
**Figure - 5:** (40X view) Photomicrograph showing tumor tissue with large cells having scant cytoplasm.



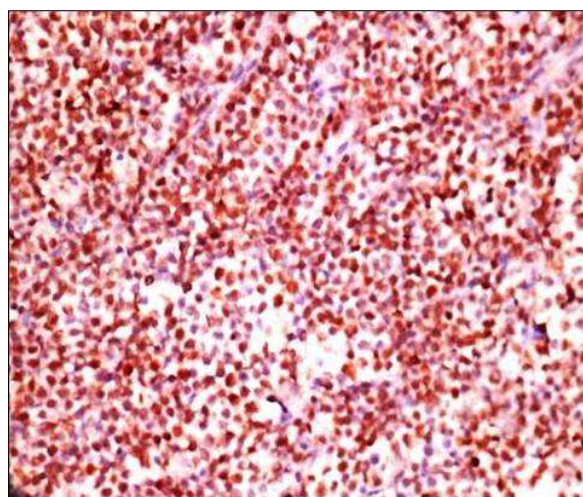
**Figure - 8:** CD-20 showing cytoplasmic positivity (Brown color).



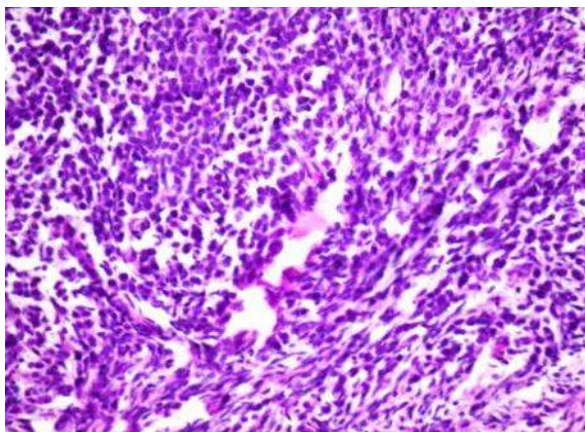
**Figure - 6:** Photomicrograph showing appendix involved by tumor.



**Figure - 9:** BCL-6 showing nuclear positivity (Brown color).



**Figure - 7:** Photomicrograph showing ovary with infiltration by tumor cells (Top field showing large B-cells).



Etiopathogenesis of small bowel lymphoma is not clearly known but research suggests that small bowel lymphoma originates from local antigen-responsive B cells. Because of this reason 70% of the neoplasms are DLBCL. Incidentally our case was also reported as Diffuse Large B-cell Lymphoma (DLBCL). However known risk factors like celiac disease and various inflammatory bowel diseases, immune suppression after organ transplant, retroviral infection (HIV), infections with Epstein Barr virus (EBV) and Human T-Cell Leukemia Virus 1 (HTLV-1) have been

identified [9-12]. In this case, we could not find the exact cause of the disease. Dysregulation of BCL-6 gene is the most common cytogenetic abnormality seen in cases of DLBCL. BCL-6 positivity was strong in this case indicating the presence of dysregulation of BCL-6 gene and protein. According to the literature and author's Chiang, et al. and Zubaidi, et al., the most

common symptoms of colonic lymphoma is abdominal pain, nausea, vomiting, weight loss, abdominal mass, change in bowel habits, hematochezia, obstruction, intussusceptions [13, 14], 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.

### **Paris staging of the tumor**

	<b>Extent of tumor</b>
<b>T x</b>	Lymphoma extent not specified
<b>T 1</b>	Lymphoma confined to the Mucosa/ Submucosa
	T1m- Lymphoma confined to Mucosa T1sm-Lymphoma confined to Submucosa
<b>T 2</b>	Lymphoma infiltrates Muscularis propria/ Subserosa
<b>T 3</b>	Lymphoma penetrates Serosa without invasion of adjacent structures
<b>T 4</b>	Lymphoma invades adjacent structures or organs
	<b>Nodal involvement</b>
<b>N 0</b>	No evidence of lymph node involvement
<b>N x</b>	Involvement of lymph nodes not assessed
<b>N 1</b>	Involvement of regional lymph nodes
<b>N 2</b>	Involvement of intra-abdominal lymph nodes beyond the regional area
<b>N 3</b>	Spread to extra-abdominal lymph nodes
	<b>Metastasis</b>
<b>M 0</b>	No evidence of extra nodal dissemination
<b>M x</b>	Dissemination of lymphoma not assessed
<b>M 1</b>	Non-continuous involvement of separate site in gastrointestinal tract (e.g. stomach and rectum)
<b>M 2</b>	Non-continuous involvement of other tissues (e.g. Peritoneum, Pleura) or organs (e.g. Lung, Liver, Bone marrow etc.)

According to the Paris Staging system, our case belongs to **T4 N1 M2 Staging**.

### **Treatment**

Surgery is an important treatment modality for primary, low-grade and localized intestinal NHL, because surgery not only removes the tumor but also reduces tumor burden and prevent hemorrhage, perforation, and secondary infection caused by chemotherapy-induced tumor necrosis [15]. Combined modality of approach that includes surgical debulking and systemic chemotherapy is the preferred treatment in more advanced cases [16].

### **Conclusion**

Primary alimentary tract malignancies, including primary gastrointestinal Non-Hodgkin lymphoma (NHL), are rare in children. The most common site of non-Hodgkin lymphoma in children is terminal ileum and the ileocaecal region. DLBCL or Non-Hodgkin's Lymphomas (NHL) usually present with vague symptoms which may lead to delay in diagnosis. Histopathological examination forms a valuable postoperative test to diagnose and to know the extent of involvement. Immunohistochemistry (IHC) can

be used in support to the diagnosis, to know the cell of origin and also to know the prognosis of the case. In our case, CD-20 was done to confirm the cell of origin and BCL-6 was done to know the prognosis of this case. Prognosis in this case was bad as the tumor involved peritoneal organs/structures and the BCL-6 IHC was strongly positive. Patient expired 50 days after the surgery.

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