

**Case Report****Primary NHL (Non Hodgkin Lymphoma) Colon: A Rare Case Presentation**

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Abstract

The incidence of primary lymphoma of colon is only 0.2-1.2 % of all colonic malignancies. Non-Hodgkin's lymphoma (NHL) is the the most common among primary colonic lymphoma. The gastrointestinal (GI) tract accounting for 30-40% of all extra nodal lymphomas and approximately 4-20% of which are NHL. Most common site for GI lymphoma is stomach followed by the small intestine. The treatment depends upon the stage of the disease. Early stage disease can be managed by radical tumor resection (hemicolectomy) plus multi-agent chemotherapy (polychemotherapy). Advanced stage can be managed by biopsy plus multidrug chemotherapy. Surgery alone can be considered as an adequate treatment for patients with low-grade NHL disease that does not infiltrate beyond the sub-mucosa. Here, we have described a case, who presented with acute intestinal intussusception and vomiting in emergency, later on we found an intraluminal growth in ascending colon and on detailed histopathological examination (HPE) confirms that it was an NHL (diffuse large B-cell type).

Keywords: Non –Hodgkin lymphoma, chemotherapy, hemicolectomy.

Introduction

The third most common malignancy in the world is (CRC) colorectal carcinoma and accounting for 9.8% world-wide¹. Primary lymphoma comprises only 0.2-1.2% of all colonic malignancies^{2,3}. The most common variety of colonic lymphoma is NHL. Although GI tract is the most frequently

involved site, accounting for 30-40% of all extra nodal lymphomas, approximately 4-20% of which are NHL, colon and rectum are uncommonly involved^{3,4,5}. The stomach is the most common location of GI lymphomas; followed by the small intestine^{3,4}. The primary sites of origin in decreasing order of frequency include the stomach

(50– 70%), small bowel (20–35%), colon (especially the cecum) (5–10%) and the esophagus (<1%)^{6,7}. The most common site of involvement are the ileocecal region and caecum, probably because of more lymphoid tissue present normally in the ileocecal region than any other part of the colon.

Case Report

A 27-year-old female patient presented with moderate to severe pain abdomen since 4 days, which was insidious in onset, gradually progressive and colicky in nature. She gave history of occasional bilious, non-projectile vomiting, altered bowel habits, melena, intermittent fever, loss of appetite and loss of weight. General examination revealed pallor and patient was afebrile with blood pressure of 130/70 mm Hg, Pulse of 89/min, RR of 24/min. Abdominal examination revealed a single, mildly tender, firm, 9X 8 cm mass in the rt. lumbar region, extending into Rt. Hypochondrium & Rt. Iliac regions. The mass is moving downwards with respiration, has restricted intrinsic mobility, bimanually palpable, falling forwards in knee-elbow position and the abdomen is tender. There was no palpable Organomegaly. A dull note is heard on percussion with a band of resonance in front. P/R and other Systemic examination were normal. Haemogram revealed anaemia (Hb = 7.2gms%) and a normal peripheral smear study. Liver function tests, renal fuction tests and electrolyte panel were within normal limits. Chest radiograph showed no abnormalities. On Ultrasonography a diffuse lump was noted involving the ascending colon with an obstructed lumen. X-ray abdomen standing was suggestive of multiple air fluid levels. CECT abdomen revealed telescoping of caecum and ascending colon into distal colon along with fat and vessels, there is also diffuse and symmetrical wall thickening of colon with maximum thickness of 2cm and this thickening shows homogenous enhancement with C-T value of 80-85HU. This terminal ileum is dilated with a mass of diameter of 4.3 cm.

Multiple enlarged lymph-nodes were seen in right iliac fossa with largest one measuring 18x 11 mm. The base of appendix is also seen telescoping. There is evidence of stranding in surrounding fat in this region. The patient was immediately taken up for surgery. Patient was taken up for exploratory laparotomy and right hemicolectomy was done. There was intraluminal grey white growth of size 5x6 cm in ascending colon. The specimen was sent for detailed histopathological examination suggestive of all free margins. The lamina propria and muscularis is diffusely infiltrated by lymphoid cells. The neoplastic lymphoid cells were infiltrating into the sub-serosal adipose tissue. The overlying epithelium showed ulceration. The tumor cells were densely populated, with sheet like growth pattern, twice the size of mature lymphocyte, with scanty cytoplasm, irregular nucleus and dense chromatin. All the 18 dissected Lymph nodes shows sinus histiocytosis and follicular hyperplasia (non-specific reactive hyperplasia). All histopathological examination was suggestive of NHL (diffuse large B-cell type) as shown in (figure 1 and 2). The findings were further confirmed by immuno-histochemistry as shown in (figure 3). Post-operative period was uneventful. Chemotherapy based on R-CHOP Regimen (Rituximab, Cyclophosphamide, Doxorubicin, Vincristine, and Prednisolone) was started after 4 weeks (healing of incision). Patient completed 6 cycles of chemotherapy. Post treatment, positron emission tomography computed tomography (PETCT) was done, suggestive of complete response. Thereafter, patient was kept on regular follow up. Patient was on follow-up since last 5 years and doing well till last follow up.

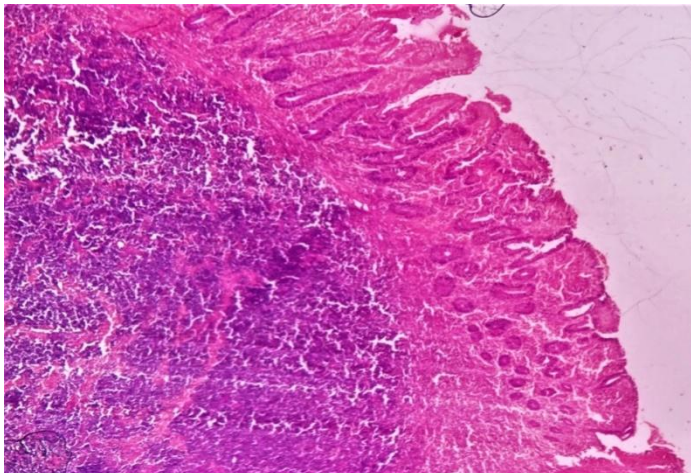


Figure 1: (Haematoxylin &Eosin stained, 10x) Normal Intestinal lining epithelium. Submucosa shows diffuse infiltration by atypical lymphoid cells.

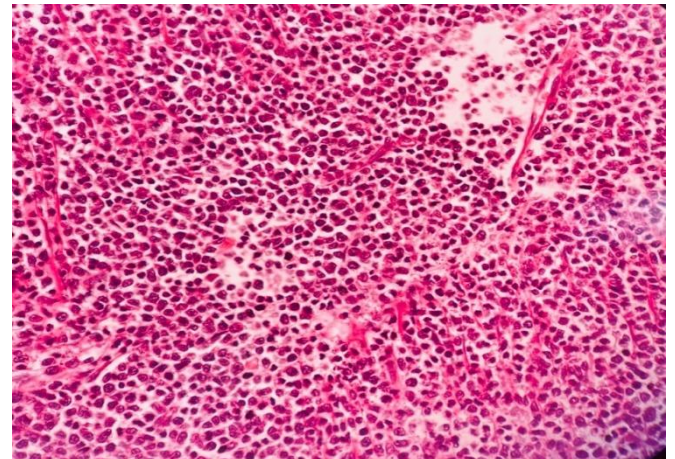


Figure 2: (Haematoxylin &Eosin stained, 40x) showing sheets of lymphoma cells.

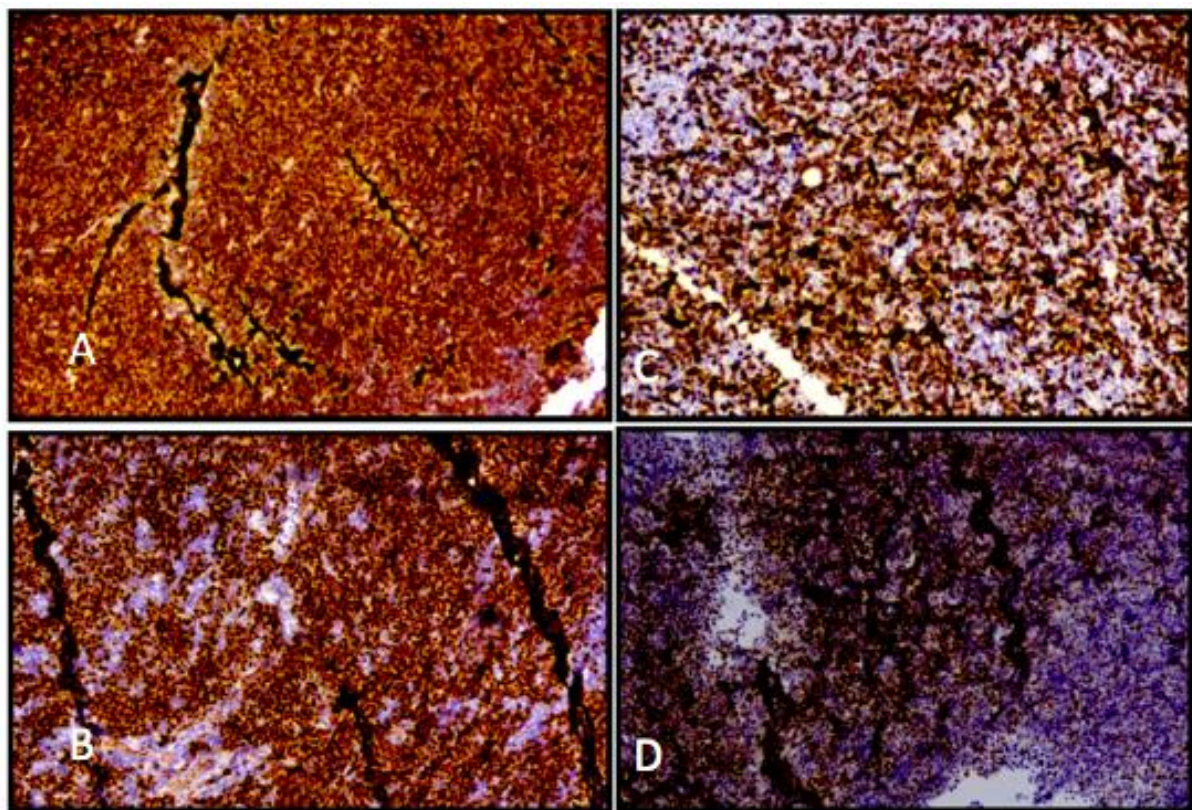


Figure 3: Immunohistochemistry showing positivity for A: BCL2, B: MUM1, C:CD20, D: BCL6

Discussion

GI tract lymphomas are the most common type of primary extranodal lymphomas, constituting 5-10% of all non-Hodgkin’s lymphomas. In aspect of pathology, clinical features, treatment and prognosis intestinal lymphomas behave differently than gastric lymphoma^{4,8}. The majority of the colon lymphomas are single (86%)⁴ but can be

multiple or diffuse in nature. Primary lymphomas arise from the B- cells of the gut, “Mucous associated lymphoid tissue” (MALT). Abdominal pain is the most common symptom followed by anaemia and weight loss. There can be an abdominal mass, found during physical examination^{3,4}. Other symptoms can be nausea, vomiting, change in bowel habits, haematochezia,

obstruction, intussusceptions and acute peritonitis due to intestinal perforation⁴. There is more predilection of incidence in males as compared to females⁹. Most of the time there is no specific association with any pre-existing pathology/disease¹⁰. It is found that Helicobacter-pylori infection is involved in the pathogenesis of MALT gastric B-cell lymphomas. Hence it has been proposed that lymphomas of MALT arise in the setting of mucosal lymphoid activities that may result from helicobacter associated chronic gastritis. Colorectal cancer and inflammatory bowel disease can be a reason for lymphomas¹¹. The Immuno-histochemical (IHC) studies on tissue blocks helps in distinguishing the immunological classification of lymphoma into either B or T- cell lineage. CD20 is the most widely used pan B-cell marker and CD3 is pan T-cell marker. CD5, CD10 and BCL-2 markers help in further sub classification of B-Cell phenotypes. The best treatment for GI lymphomas of any location remains uncertain. Most of the literature suggests laparotomy and tumor resection. Combined modality of approach that includes surgical debulking and systemic chemotherapy is the most preferred treatment¹². Polychemotherapy includes CHOP (cyclophosphamide, doxorubicin, vincristine, and prednisolone) or CHOP-like combination chemotherapy or MACOP-B-like regimens. Surgery alone can be an option for patients with low-grade NHL disease that does not infiltrate beyond the sub mucosa¹². Radiotherapy is beneficial for incomplete resection or non-resectable disease.

Conclusion

Gastrointestinal lymphomas are a heterogeneous group of lymphomas. These are rare entities and many questions regarding their management remained unanswered. Though GIT is the most common site of NHL, primary colonic lymphomas of the large intestine are rare, with caecum being the most common site of occurrence. Early diagnosis and treatment may prevent intestinal perforation and associated complications.

However, the diagnosis is often delayed in most cases. The major presenting clinical symptom is only abdominal pain and diagnosis is usually based on incidental findings. Surgical resection is the mainstay of treatment for localized primary lymphomas, followed by postoperative chemotherapy. Those with limited stage disease may have prolonged survival when treated with aggressive chemotherapy.

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