CASE REPORT

Primary Diffuse Large B Cell Lymphoma of Colon Presenting as Chronic Diarrhea in a 3 Year Old Child - Case Report

Suraj Uppalapati, Prashant Kanni, Praveen Mathew, Manjunath Patil, Praveen Kumar

Department of Medical Gastroenterology, Vydehi Institute of Medical Sciences and Research Center, Bangalore, Karnataka.

Corresponding Author : Dr. Suraj Uppalapati (drsurajuppalapati@gmail.com)

Abstract

A 3 year old baby boy presented to hospital with a 2 months history of watery diarrhea (8-10 episodes per day) associated with abdominal pain, weight loss and swelling of face and limbs. Rehydration and electrolytic balance were restored with intravenous fluid therapy along with cover of antibiotics but diarrhea was worsening. Patient was planned for gastroscopy and ileo- colonoscopy with biopsies from duodenum and ileum. Gastroscopy was normal but colonoscopy showed ileal scalloping and transverse colon showed short segment stricture of approx. 5 cms with multiple ulcers. CT imaging showed a structuring mass involving the transverse colon and few adjacent jejunal loops with enlarged lymph nodes. Histopathological examination of the biopsies from transverse colon showed dense lymphoid aggregates. Hence Bone marrow aspiration and IHC panel of markers were advised. BMA was normal but IHC staining revealed Primary Diffuse large B cell Lymphoma of large intestine.

Keywords : GI Lymphoma, Chronic Diarrhea

Introduction:

Primary Non-Hodgkin's lymphomas (NHL) of the gastrointestinal tract are the most common extranodal lymphoma with increasing incidence in recent years, yet they are rare tumors.[1] Due to rarity and variable clinical presentation early detection is prevented while possibility of cure exists. Non-Hodgkin's lymphoma (NHL) remains the most common malignancy of the GI tract in children. They usually have different anatomic distribution and histologic appearance compared to common patterns in adult cases. Unlike adult patients in whom stomach is the most frequent site, small and large intestines are the most commonly involved sites in pediatric age group. Clinically, the patients present with varied symptoms ranging from abdominal mass to acute abdominal emergency caused by intussusception. Majority of the patients (81.4 %) present with abdominal pain as the presenting symptom, followed by abdominal swelling, vomiting, constipation, diarrhea, and intestinal obstruction. Nearly 50% of children with GI NHL have tumor infiltrates confined to GI tract with possible regional lymph node involvement.[2]

Case presentation:

A 3 year-old boy presented to hospital with a 2 month history of watery diarrhea. The child was reported to have had 8-10 episodes of stools per day which started with 5-6 episodes at presentation, watery in consistency, medium to large in quantity, associated with mucus occasionally, There was no blood, oil droplets or frothing. There was no fecal soiling of clothes ruling out anal incontinence or rectal pathology. It was associated with pain abdomen(child used to cry holding his hands around the umbilicus) prior to passage of stools and relieved after passage. No associated vomitings. Diarrhea was

not related to any specific food intake. Child also had lower limb swelling and facial swelling since 1 week along with weight loss of approximately 3 kgs(23.5% of total body weight).

His routine medical history revealed delivery via LSCS with normal development and good vaccination status. No history of recurrent chest or gastrointestinal infections. No history of previous hospitalisations.

On examination there was facial puffiness and bilateral pedal edema upto calf. There was no peripheral lymphadenopathy on examination. Respiratory, cardiac and neurological systems were normal on examination. His genital and rectal examination was normal. Abdominal examination revealed abdominal wall edema. No mass or organ palpable.

Laboratory tests showed normal blood picture with mild anemia (HB-11.7 g%). Liver function tests showed reduced protein (2.9) and albumin (1.48). His serum IgA levels were very low (<3 mg/100 ml). C-reactive protein and serum TTG (IgA)levels were normal (TABLE 1).

Wbc/pmn (*10^9/L)	7900
Hb (g/dL)/Ht (%)	11.7
Platelets (*10^9/L)	4,07,000
CRP (mg/dL)	0.501(Normal range - <0.6)
Total bilirubin (mg/dl)	0.3
AST / ALT (U/L)	26/ 38
Total protein(g/dl)	2.9
Serum albumin (g/dl)	1.48
Sodium / potassium (mEq/l)	138.2/2.92
Calcium (mg/Dl)	7.8
Stool examination	Normal
Stool for occult blood	negative
Serum IgA	< 3mg / 100 ml
TTG (IgA)	1.24 U/ml

TABLE 1 : Lab Parameter

He was rehydrated with IV fluids and started on broad spectrum I.V antibiotics along with enteral nutrition. Upper gastrointestinal endoscopy and Ileo- colonoscopy were advised along with duodenal and ileal biopsies. Gastroscopy and duodenal histopathological examination revealed no abnormality. Ileo-colonoscopy showed ileal scalloping (FIG 1) along with transverse colon showing short segment stricturous lesion extending for approximately 5 cm with multiple ulcerations and friable mucosa (FIG 2). Ileal microscopic examination revealed focal blunting and broadening of villi associated with focal cryptitis. Transverse colon lesion on microscopic examination showed large intestinal mucosa with ulcer replaced by fibrin, inflammatory exudate and bacterial colonies. Crypt atrophy was seen along with mild infiltrates of plamsa cells and eosinophils (40/hpf). A dense lymphoid aggregate composed of fairly monotonous cells having scanty cytoplasm and round to oval nuclei displaying 2-4 nucleoli is seen. No granulomas or parasites were seen. AFB staining was negative (FIG 3 & 4).

Contrast enhanced CT imaging was done which showed irregular structuring mass involving the transverse colon near the splenic flexure causing partial luminal obstruction with involvement of few adjacent jejunal loops in left upper abdomen with mild focal strictures.

Diffuse mesenteric edema is seen with multiple enlarged lymph nodes(FIGURE 5 AND 6).

Meanwhile patient was shifted to Intensive care in view of increased frequency of stools to 15-20 episodes per day and started on Total parenteral nutrition. In view of dense lymphoid aggregate with multiple nucleoli in the large intestinal lamina propria and a stricturing mass noted in CT imaging, IHC panel of markers and bone marrow aspiration was advised to r/o Lymphoid neoplasm. Bone marrow aspiration was normal and IHC was consistent with diffuse large b cell

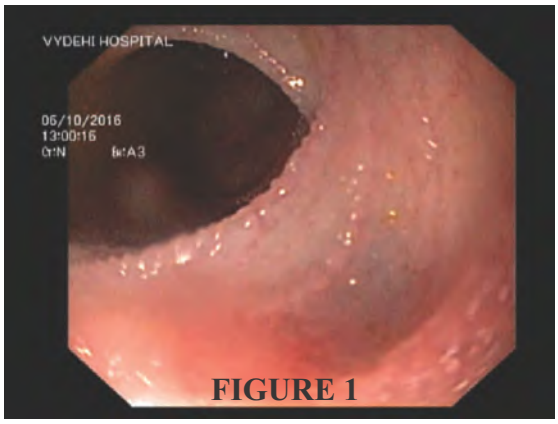

FIGURE 1

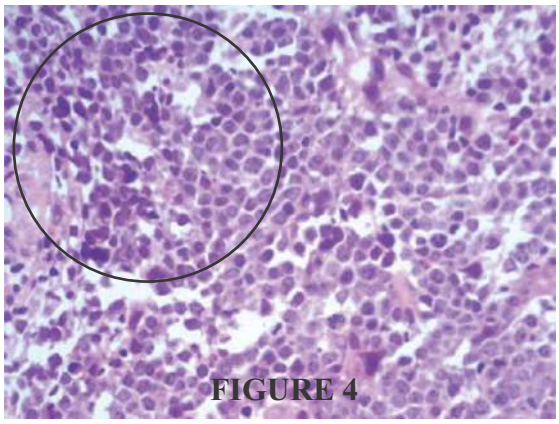

FIGURE 4

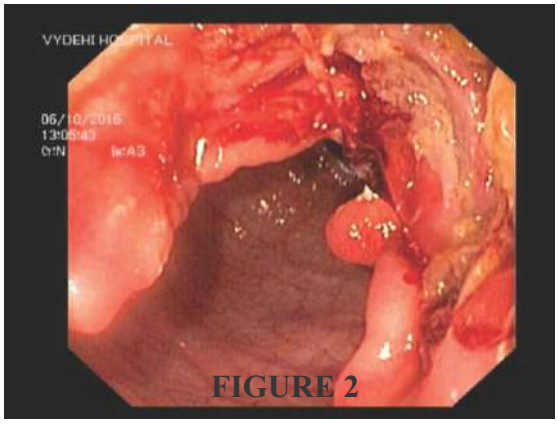

FIGURE 2

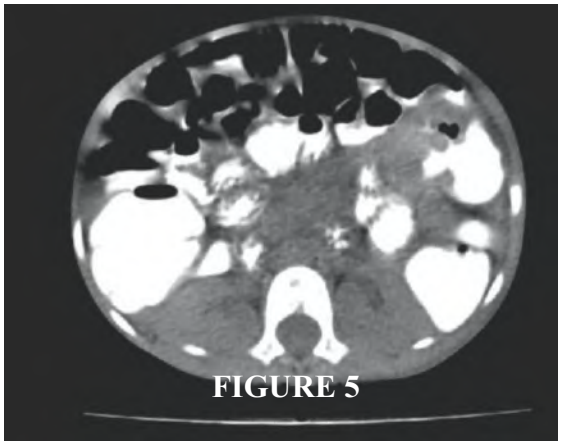

FIGURE 5

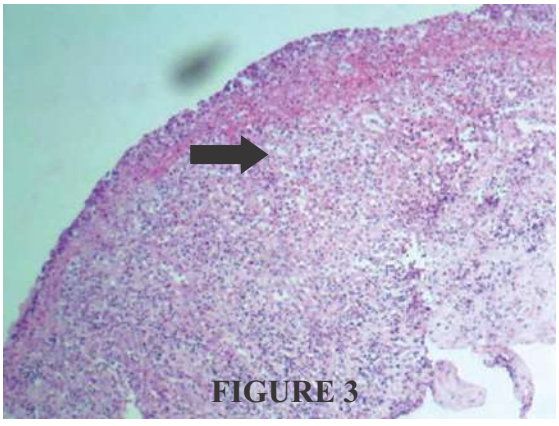

FIGURE 3

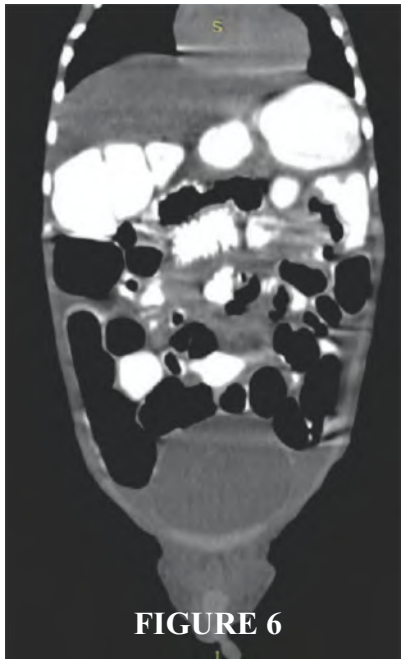

FIGURE 6

lymphoma(DLBCL)-large intestine (TABLE 2)

CD40	3+(neoplastic cells)
CD3	1+(scattered mature T -lymphocytes)
CD68	2+(background infiltrating histiocytes)
CD4	1+(normal T -lymphocytes)
BCL -8	3+(neoplastic cells)
CD30	0
CD10	0
PAX-5	3+(neoplastic cells)
CD20)	3+ (neoplastic cells)
VIMENTIN	4+(neoplastic cells)

As per Dawson criteria, patient was diagnosed to have Primary Gastrointestinal lymphoma involving large bowel with extension into small bowel causing chronic diarrhea and protein losing enteropathy.[3]

Patient was referred to medical oncologist. As there were no features of intestinal obstruction and chemotherapy being a good curative option, patient was advised chemotherapy.

Discussion:

Dawson et al. were the first to describe colorectal lymphoma in 1961.[4] Involvement of the large intestine is rare (10%-20% of all gastrointestinal lymphomas) in comparison to the stomach or small bowel. Primary NHL accounts for 0.1%-0.5% of all malignant tumors of the colon and rectum which makes it the third most common large bowel malignancy after adenocarcinoma and carcinoid.

Malignant lymphomas are the third most common type of childhood cancer. Children typically present with diffuse extranodal disease in contrast to adults among whom primary nodal disease is common. Primary GI malignancies are a rarity in children, with limited information from Asian population. The peak age for NHL of GI tract in children is 5–15 years.

The ideal treatment approach in GI lymphoma is debatable as per literature[5]. Radical tumor resection followed by chemotherapy in early disease(St. Jude stage I and II), and limited or no resection followed by polychemotherapy in advanced disease(St. Jude stage III and IV) may be the justified approach. However, recent studies have proposed the use of chemotherapy alone as an effective treatment option in primary GI lymphoma in all stages. A study performed at AIIMS, New Delhi comparing chemotherapy vs chemotherapy plus surgery in non Hodgkins lymphoma patients showed that five years EFS(event free survival) and OS(overall survival) were 72% and 67% for CT only group compared to 60% and 64% for CT+surgery group [6].

A study from Asian population which was done in Pakistan to identify the primary GI lymphomas under 19 years age group had shown higher prevalence of DLBCL among GI lymphomas[7].

Due to rarity of presentation and non specific symptoms we are reporting the above case which was diagnosed with Primary NHL of colon (DLBCL) with extension into small bowel causing chronic diarrhea.

Conclusion:

Primary GI lymphomas is a multifarious disease varying in its staging, site of involvement, histological subtype and type of treatment offered.

Primary GI lymphomas especially of the large bowel are rare and diagnosis can be delayed (in upto 35-65 % of cases) due to variable clinical presentation , while the options for cure exist.

Among lymphoma patients, children present predominantly with extranodal disease unlike adults. Even the presentation of pediatric GI lymphomas varies with adult group as the primary organ involved is small and large intestine unlike stomach in adults.

References

1. Morsi A, Abd El-Ghani Ael-G, El-Shafiey M, Fawzy M, Ismail H, Monir M. Clinico-pathological features and outcome of management of pediatric gastrointestinal lymphoma. J Egypt Natl Canc Inst. 2005;17:251–9.
2. Ladd AP, Grosfeld JL. Gastrointestinal tumors in children and adolescents. Semin Pediatr Surg. 2006;15:37–47.
3. Dawson IM, Cornes JS, Morson BC. Primary malignant lymphoid tumours of the intestinal tract. Report of 37 cases with a study of factors influencing prognosis. Br J Surg. 1961;49:80–89
4. Barbaryan A, Ali AM, Kwatra SG, et al. Primary diffuse large B-cell lymphoma of the ascending colon. Rare Tumors. 2013;5(2):85–88.
5. Bandyopadhyay R, Sinha SK, Chatterjee U, et al. Primary pediatric gastrointestinal lymphoma. Indian J Med Paediatr Oncol. 2011;32(2):92–95. doi:10.4103/0971-5851.89786.
6. Raina V, Sharma A, Vora A, Shukla NK, Deo SV, Dawar R. Primary gastrointestinal non-Hodgkin lymphoma chemotherapy alone an effective treatment modality: Experience from a single center in India. Indian J Cancer. 2006;43:30–5.
7. Khurshed A, Ahmed R, Bhurgri Y. Primary gastrointestinal malignancies in childhood and adolescence – An Asian perspective. Asian Pac J Cancer Prev 2007;8:613-7