

Primary Rectal Lymphoma: A Case Report and Review of Literature

Abstract

Lymphomas represent common hematological malignancy, and depending on site, they are classified as nodal or extranodal lymphoma. The term extranodal disease refers to lymphomatous infiltration of anatomic sites other than the primary lymphatic sites; however, the diagnosis of primary versus secondary extranodal lymphoma remains challenging. Among the extranodal locations, gastrointestinal system is the most frequent site. The involvement of the stomach, small intestine, and colon is noted. Rectum as primary site for lymphoma is rare in adults and extremely rare in children. We describe a case of primary rectal lymphoma (high-grade B-cell non-Hodgkin's lymphoma) in 11-year-old child. We believe that reporting this case will add to the data about clinical presentation, radiological, nuclear medicine findings, and treatment approaches of primary rectal lymphoma.

Keywords: Fluorodeoxyglucose positron emission tomography-computed tomography, high-grade non-Hodgkin's lymphoma, pediatric, recto-sigmoid

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Introduction

The cancers arising from the cells of immune system comprise lymphoma and it is the third most common pediatric malignancy after leukemia and central nervous system tumors, constituting 10%–12% of childhood malignancies.^[1,2] Predominantly, lymph nodes are involved; however, any organ can be involved with gastrointestinal system (GIS) being frequent extranodal site.^[3] The involvement of GIS by lymphoma can be either primary or secondary. Dawson *et al.*^[4] established criteria for primary gastrointestinal lymphoma in 1961. In non-Hodgkin's lymphoma of primary GIS, the stomach is most commonly affected (50%–60%) followed by the small intestine (approximately 30%), colon, anorectum, and very rarely pancreas, liver.^[5,6] Primary colorectal lymphoma comprises around 10% of gastrointestinal lymphomas^[7,8] and 0.5%–2% of large bowel malignancies.^[8,9] The presentation, treatment, and prognosis of colorectal lymphoma differ from lymphoma of rest of gastrointestinal system, and only few case reports of pediatric rectal lymphoma have been described;^[10–12] hence, it is important to study this entity in detail.

Case Report

A girl child of 11-year age presented with increased frequency of defecation for 3 months. There were no history of fever, no micturition difficulties, and no swelling anywhere. Menarche was not attained. Magnetic resonance imaging showed a large circumferential mass involving the entire rectum and the recto-sigmoid junction with major bulk of the mass seen along posterior half of the bowel circumference with a left common iliac node. Colonoscopy revealed a recto-sigmoid growth. Biopsy showed intestinal glands with stroma displaying monotonous lymphoid population arranged in sheets. The atypical cells had high N/C ratio, oval to irregular indented nuclei, coarse chromatin, 1–3 prominent nucleoli, and scant cytoplasm [Figure 1a]. On immunohistochemistry, tumor cells were diffusely positive for CD20 [Figure 1b]. Atypical lymphoid cells were positive for CD10, MUM-1, and BCL-6 and negative for Tdt and BCL-2. C-Myc was positive in ~20% cells and Ki-67 proliferation index was ~95%. Findings were suggestive of diffuse large B-cell lymphoma, activated B-cell type. Bone marrow was uninvolved on biopsy. For staging purpose, ¹⁸F fluorodeoxyglucose positron emission tomography/contrast enhanced computed tomography (¹⁸F-FDG PET/CECT) scan was done which showed FDG-avid mass

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involving the rectum and recto-sigmoid [colored arrow in Figures 2a, 3a and d, 4a and d] with left common iliac node and marrow involvement [Figure 2b]. There was no evidence of active disease elsewhere. The patient was started on multiagent chemotherapy protocol. An interim ^{18}F FDG PET/CT showed reduction in metabolic activity and morphology of rectal mass [Figures 3b, e and 4b, e] with the post therapy ^{18}F FDG PET/CT showing complete metabolic resolution with morphological regression [Figures 2c and d, 3c and f, 4c and f]. At present, the child is on follow-up.

Discussion

Primary rectal lymphomas are rare and usually occur in patients aged 55–75 years. There have been few case reports of rectal lymphoma in children.^[10-12] The distribution of primary gastrointestinal tract lymphomas is different in adults and children, with the stomach being the most common site in adults while they occur most frequently in the distal ileum or ileocecal region in children. On abdominal CT, the lesion may be bulky with concentric thickening of the bowel wall or it may manifest as multiple polyposis or mucosal ulcerations. However, it may be difficult to distinguish the primary adenocarcinoma of the colon/rectum on radiology and biopsy is therefore crucial.

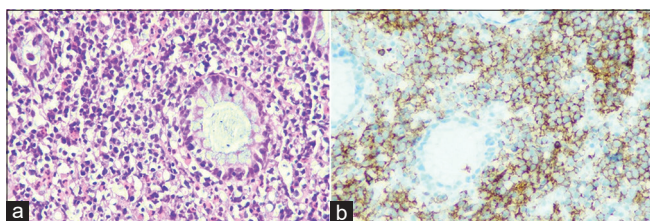


Figure 1: Photomicrograph of rectal mass shows intestinal gland with stroma displaying monotonous lymphoid population arranged in sheets (a) and immunohistochemistry (b) shows tumor cells diffuse positive for CD20

^{18}F -FDG PET/CT is the optimal modality for staging, restaging, and evaluation of treatment response in cases of ^{18}F -FDG-avid lymphoma. In our case, the primary lesion showed a maximum standard uptake (SUVmax) of 34.2, and focal areas of FDG uptake suggesting bone marrow involvement were detected on PET/CT, which also has prognostic implications. Follow-up ^{18}F -FDG PET/CT demonstrated complete metabolic resolution and significant morphological regression.

CHOP (cyclophosphamide, doxorubicin, vincristine, prednisone) with supportive care remains a mainstay for treatment of lymphoma. In our institute, the child was treated with the Modified Multicentric Protocol (MCP-842 regimen). Modified MCP-842 regimen consists of cyclophosphamide, adriamycin, vinblastine (instead of vincristine), cytarabine, methotrexate.^[13,14] Multiple studies are required to define the role of radiotherapy in the management of rectal lymphoma in the pediatric population. In cases of gastrointestinal lymphoma, surgery may be required for conditions such as intestinal obstruction, intussusception, perforation and to obtain biopsy. Cai *et al.*^[15] and Zhai *et al.*^[16] published case series which discussed the role of surgery in emergency conditions in cases of lymphoma.

As rectal lymphoma is extremely rare in pediatric patients, prospective randomized clinical trials would be difficult to conduct to define the optimal therapy for primary rectal lymphoma in this population. Treatment decisions are mainly based on the opinion of specialists and the consensus-based on joint clinic or tumor board discussions on the basis of disease stage and the general condition of the patient.

Conclusion

Primary rectal lymphoma is extremely rare in childhood, though diagnosis needs to be considered in patients presenting with rectal mass. The individualized treatment

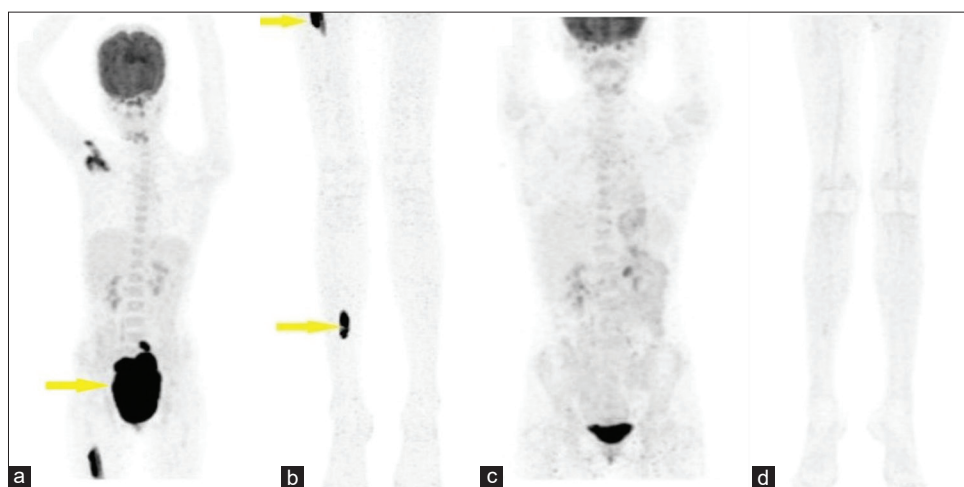


Figure 2: ^{18}F -fluorodeoxyglucose positron emission tomography/computed tomography scan done for initial staging shows maximum intensity projection image (a) with fluorodeoxyglucose-avid rectal mass and image (b) indicates lesion in right femur and tibia (colored arrow). The posttreatment scan shows complete metabolic resolution of rectal mass (c) and marrow activity in femur and tibia (d) (confirmed on fused axial images)

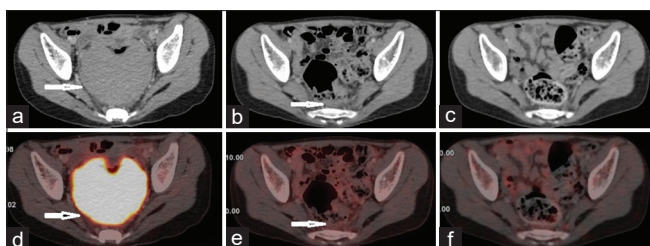


Figure 3: Axial computed tomography image (a) shows bulky rectal mass (arrow). Postchemotherapy follow-up image shows significant morphological regression (b) and image (c) shows complete morphological regression of rectal mass. Corresponding positron emission tomography/computed tomography image shows fluorodeoxyglucose-avid mass (arrow in image d). Postchemotherapy follow-up positron emission tomography/computed tomography image shows significant metabolic resolution (arrow in image e) and image (f) shows complete metabolic resolution

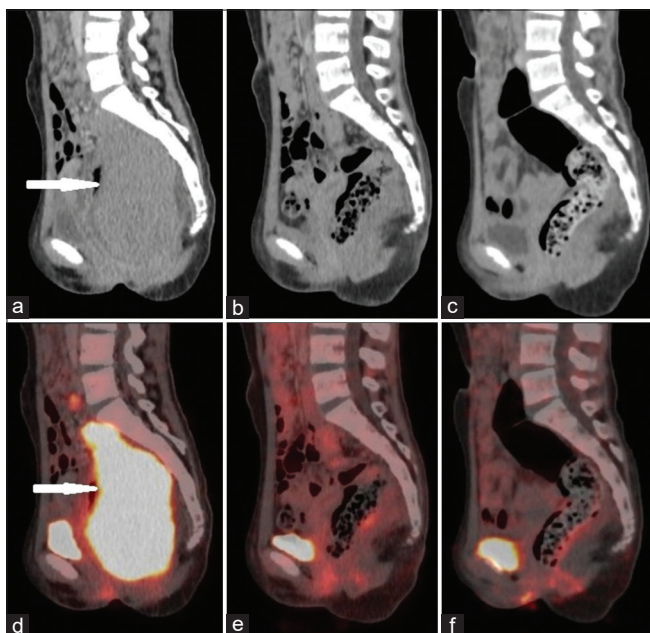


Figure 4: Sagittal computed tomography image (arrow in image a) shows bulky rectal mass. Postchemotherapy follow-up image shows significant morphological regression (b) and image (c) shows complete morphological regression of rectal mass. Corresponding positron emission tomography/computed tomography image shows fluorodeoxyglucose-avid mass (d). Postchemotherapy follow-up positron emission tomography/computed tomography image shows significant metabolic resolution (e) and image (f) shows complete metabolic resolution

approach is needed which may improve overall survival as well as disease-free survival.

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Conflicts of interest

There are no conflicts of interest.

References

1. Toma P, Granata C, Rossi A, Garaventa A. Multimodality imaging of Hodgkin disease and nonHodgkin lymphomas in children. *Radiographics* 2007;27:1335-54.
2. Linabery AM, Ross JA. Trends in childhood cancer incidence in the U.S. (1992-2004). *Cancer* 2008;112:416-32.
3. Kumar R, Xiu Y, Potenta S, Mavi A, Zhuang H, Yu JQ, *et al.* 18F-FDG PET for evaluation of the treatment response in patients with gastrointestinal tract lymphomas. *J Nucl Med* 2004;45:1796-803.
4. 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;49:80-9.
5. Koniaris LG, Drugas G, Katzman PJ, Salloum R. Management of gastrointestinal lymphoma. *J Am Coll Surg* 2003;197:127-41.
6. Weingrad DN, Decosse JJ, Sherlock P, Straus D, Lieberman PH, Filippa DA. Primary gastrointestinal lymphoma: A 30-year review. *Cancer* 1982;49:1258-65.
7. Blackledge G, Bush H, Dodge OG, Crowther D. A study of gastro-intestinal lymphoma. *Clin Oncol* 1979;5:209-19.
8. Contreary K, Nance FC, Becker WF. Primary lymphoma of the gastrointestinal tract. *Ann Surg* 1980;191:593-8.
9. Henry CA, Berry RE. Primary lymphoma of the large intestine. *Am Surg* 1988;54:262-6.
10. Zhang KR, Jia HM. Clinical images. Primary non-Hodgkin's lymphoma of the sigmoid colon in a child. *Am J Surg* 2009;197:e11-2.
11. Chen CY, Wu CC, Hsiao CW, Chen CW, Jin JS, Jao SW. Primary non-Hodgkin's lymphoma of rectum presenting with rectal prolapse. *J Pediatr Surg* 2009;44:e1-3.
12. Favini F, Massimino M, Esposito V, Maestri L, Fava G, Spreafico F. Rectal burkitt lymphoma in childhood. *J Pediatr Hematol Oncol* 2008;30:176-8.
13. Vijayasekharan K, Prasad M, Pradhan ND, Phillip D, Gujral S, Shet T, *et al.* Favorable outcomes and reduced toxicity with a novel vinblastine-based non-high dose methotrexate (HDMTX) regimen (modified MCP-842) in pediatric anaplastic large cell lymphoma (ALCL): Experience from India. *Leuk Lymphoma* 2020;61:912-8.
14. Banavali S, Arora B, Hingmire S, Vora T, Bansal S, Pai SK, *et al.* Vinblastine improves outcome in children with anaplastic large cell lymphoma (ALCL) treated with a uniform short duration intensive protocol. *SIOP XXXIX meeting Abstracts PJ038. Pediatr Blood Cancer* 2007;49:532.
15. Cai S, Cannizzo F Jr., Bullard Dunn KM, Gibbs JF, Czuczman M, Rajput A. The role of surgical intervention in non-Hodgkin's lymphoma of the colon and rectum. *Am J Surg* 2007;193:409-12.
16. Zhai L, Zhao Y, Lin L, Tian Y, Chen X, Huang H. Non-Hodgkin's lymphoma involving the ileocecal region: a single-institution analysis of 46 cases in a Chinese population. *J Clin Gastroenterol.* 2012;46:50914. <https://doi.org/10.1097/MCG.0b013e31823716c>.