Rare Location of Primary Non-Hodgkin's Lymphoma in the Rectum

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ABSTRACT

Lymphomas of the gastrointestinal tract are usually secondary. Primary rectal lymphoma is very rare by virtue of its location. We present here a 60 years old lady diagnosed as primary rectal diffuse large B-cell Non-Hodgkin's lymphoma managed operatively. The optimum management of this entity is still much debated.

Keywords: *Non-Hodgkin's lymphoma; primary; rectum.*

INTRODUCTION

Lymphomas usually affect the gastrointestinal tract as part of a secondary process.[1] Primary lymphomas of the gastrointestinal tract constitute only 5% of all lymphomas.[2] Colorectal lymphomas account for 10-20% of all gastrointestinal tract lymphomas, most are usually localized in the cecum.[3] Primary rectal lymphoma accounts for 0.1-0.6% of colonic malignancies and 0.05% of primary rectal tumours.[4] We present here a case of primary rectal diffuse large B-cell Non-Hodgkin's lymphoma at stage I in a 60 years old lady.

CASE REPORT

A 60 years old Nepali lady of Mongolian origin was admitted to the Department of Surgery, B. P. Koirala Institute of Health Sciences with history of loss of weight and bleeding per rectum during defecation for last 6 months. Physical examination showed her to be pale and the examination of rectum showed a diffuse involvement of the rectum by a circumferential growth. The rest of the clinical examination was normal. Computed tomography scan showed a circumferential growth in the rectum involving about 8 cm length; there was no perirectal involvement (Figure 1).

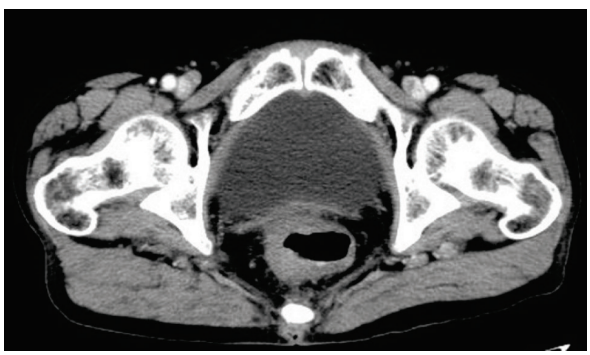

Figure 1. Contrast-enhanced computed tomography of the abdomen (axial section) shows asymmetric thickening of the wall of the rectum; no perirectal lymph nodes are seen.

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The white blood cell counts were within normal limits. Chest radiography and thorax CT were normal. Rectal biopsy was performed that showed B-cell Non-Hodgkin's lymphoma; peripheral cytology and bone marrow aspiration were also unremarkable. As a result of these findings, the patient was considered as having primary rectal lymphoma on stage 1 (according to the Ann Arbor Staging System). The patient was subjected to an abdominoperineal resection, and a permanent colostomy; there was the growth in the rectum extending up to proximal part of the anal canal with an area of ulceration (Figure 2).

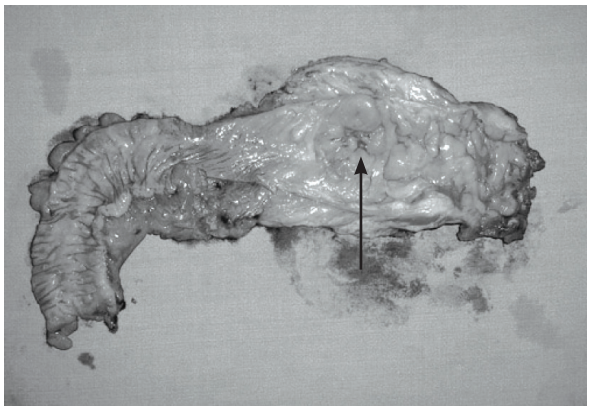

Figure 2. Specimen of abdominoperineal resection of the rectum showing the growth with an area of ulceration at the middle third of the rectum (arrow).

The resected specimen was sent for biopsy. Microscopic examination of the lesion revealed massive infiltration of the whole bowel wall by diffuse uniformly arranged large lymphoid cells causing effacement of the normal architecture (Figure 3).

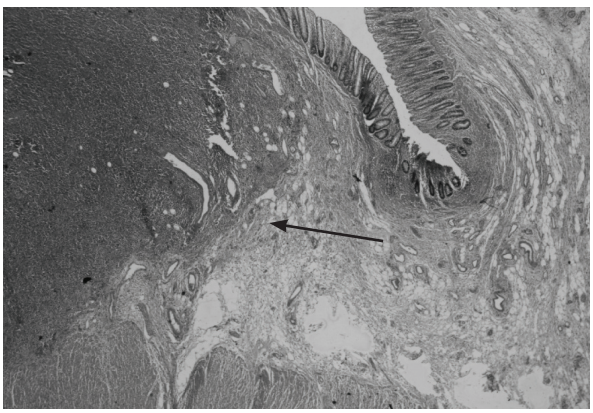

Figure 3. Photomicrograph showing mucosa, submucosa and muscularispropia of the rectum with diffuse infiltrates of submucosa with tumor cells (arrow) extending upto mucosa and muscularispropria. (H&E stain, 20X)

Few residual epithelial glands were observed in the mucosa. The tumor cells had oval to round to indented, irregularly folded vesicular nuclei with multiple nucleoli and scant to moderate cytoplasm (Figure 4). Numerous mitotic figures including atypical ones are observed. The tumor cells expressed positivity for B-Cell marker, i.e., CD 20 on immunohistochemical study (Figure 5,6). Based on these findings on histopathology and immunostaining, she was diagnosed as primary diffuse large B-cell Non-Hodgkin's lymphoma of the rectum. Twelve months after the surgery, she remains disease-free and there is no evidence of involvement at other sites till date.

DISCUSSION

Dawson et al 1961 first proposed the criteria for definition of primary GI lymphomas which include: 1) absence of palpable peripheral lymphadenopathy at the time of first clinical presentation, 2) no mediastinaladenopathy on chest radiography, 3) a normal peripheral blood smear, 4)

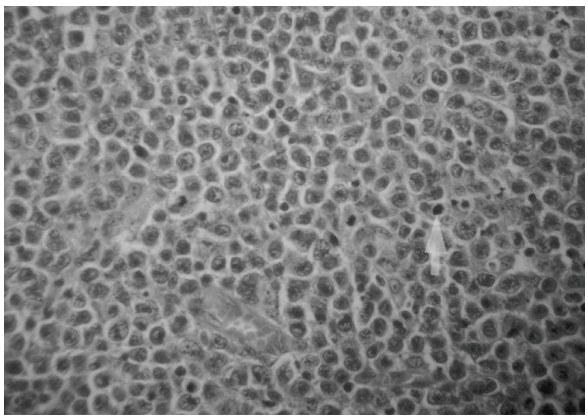

Figure 4. Photomicrograph showing high power view of the lymphoma cells: they are large with high nucleocytoplasmic ratio, oval to round to indented, having irregularly folded nuclei, vesicular chromatin, one to two nucleoli and moderate amount of cytoplasm. Numerous mitoticfigures (green arrow) and few karyorrhexis are observed (H&E stain, 40X).

at laparotomy, involvement of only the gastrointestinal tract or only the regional lymph nodes (excluding retroperitoneal lymphadenopathy), 5) no involvement of liver and spleen except by direct spread from a contiguous site.[5] In the present case, the first presentation of the patient with rectal involvement fulfilled Dawson et al's criteria for primary rectal lymphoma. In the modern era, these criteria have been expanded to new diagnostic tools. Krol et al in a paper from 2003 proposed a more liberal definition of primary extranodalNon-Hodgkin's lymphoma that includes all patients who present with Non-Hodgkin's lymphoma

that apparently originated from an extranodal site, even in the presence of disseminated disease, as long as the extranodal component is clinically dominant.[6]

The World Health Organization (WHO) classification subtypes lymphomas into diffuse large B-cell lymphoma, extranodal marginal zone lymphoma (mucosa-associated lymphoid tissue ([MALT]-associated lymphoma), mantle cell lymphoma,Burkitt's lymphoma, and follicular lymphoma.[7] The most common histologic subtype affecting the gastrointestinal tract and colon is diffuse large B-cell lymphoma.[8,9] These are generally aggressive and are composed of rapidly proliferating cells of B-cell origin.

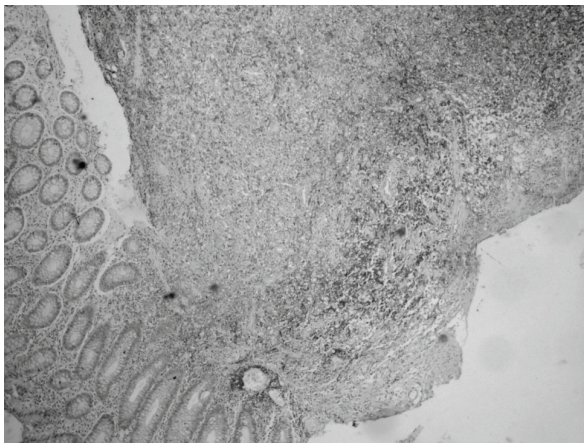

Figure 5. Photomicrograph showing membranous and cytoplasmic staining of the tumor cells by CD-20 (B-cell marker) (20X).

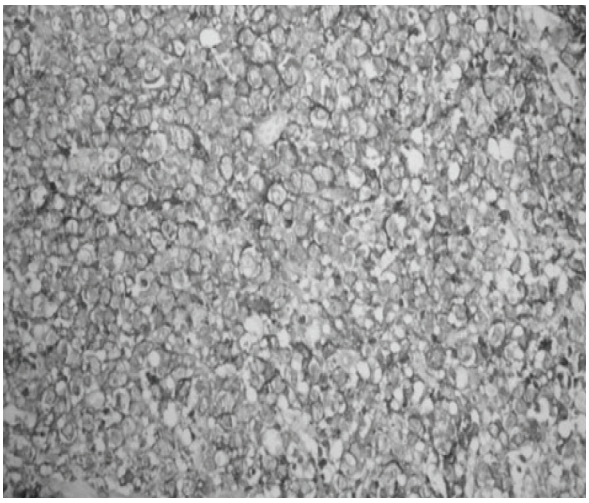

Figure 6. Photomicrograph showing membranous and cytoplasmic staining of the tumor cells by CD-20 (B-cell marker) (40X).

The second most common colorectal lymphoma is MALT-associated low-grade B-cell lymphoma. MALT-associated lymphomas are low-grade tumors arising from B cells associated with mucosal immunity. MALT-associated lymphomas are most commonly seen in the stomach, where they are often preceded by a chronic inflammatory state, such as Helicobacter pylori infection. Gastric MALT-associated lymphomas can be successfully treated by H. pylori eradication alone.[9] Colorectal MALT-associated lymphomas do not have the same association with H. pylori infection and, therefore, behave as a different clinical entity.

Primary colorectal lymphoma most often affects patients in the fifth to seventh decades of life.[1]Males are affected more commonly than women. As with other types of lymphoma, patients with chronic immunosuppression like those with inflammatory bowel disease, human immunodeficiency virus infection, and transplant recipients are more prone to have colorectal lymphoma. The most common presenting symptoms are weight loss and abdominal pain.[2,9] Fan et al reported that 62% of patients presented with pain and 43% presented with weight loss in their series of 37 patients with colorectal lymphoma,[2] and Zighelboim and Larson reported a 40% rate of abdominal pain and weight loss in their series of15 patients with colorectal lymphoma.[9] Lower gastrointestinal bleeding occurs in approximately 20% of patients.[2,9] In frequency of these complaints suggests that, despite the large size of the lymphoma, mucosal ulceration does not usually occur.[2] Up to half of patients present with a palpable abdominal mass, suggesting that these tumors can be present for a long period of time without causing symptoms. Conversely, obstruction is a rare event and occurs much less often in patients with colorectal lymphoma than those with colorectal adenocarcinoma, probably due to the more pliable nature of colorectal lymphomas and the absence of a desmoplastic response. In addition, patients with colorectal lymphoma rarely present with bowel perforation.

The lack of specific complaints makes the diagnosis hard to establish. Unfortunately, for some patients, a surgical procedure is the only diagnostic tool. As a rare lesion, it requires a high index of suspicion, and, the optimal treatment for this disease is still controversial. The role of surgery in its management is relatively well-defined. Surgical resection was the only statistically significant prognostic for patients with intestinal lymphomas in the study by Gobbi et al.[10] In another study, 5-year survival rates were 46% for patients who underwent surgery whereas it was 0% for patients managed without surgery.[11] Authors recommending primary surgical treatment defend the opinion that surgery provides information on the distribution and histology of the tumor, hence prognostic information, and decreases the probability of complications as

obstruction and perforation, and may offer a chance for cure with or without chemotherapy.[9-11] Hence, there is a universal agreement that in the absence of disseminated disease, surgical resection is generally performed.[9-11] If complete excision is not possible, then non-operative therapy becomes the treatment of choice. Other surgeons have published reports of remission with primary chemotherapy and radiotherapy, and these reports have been increasing in number lately.[12-15] Those authors primarily recommending medical treatment suggest that a residual tumor following surgery would worsen the course of the disease.[2,12] In the present case, we opted for surgical intervention, because of the uncommon presentation with ulceration and continuous bleeding that she had presented with. In conclusion, considering the less number of individual cases reported in the literature rather than extensive trials, it is difficult to establish medical management as primary treatment modality. Primary rectal lymphoma is in itself a rare entity and a rare cause of bleeding per rectum; larger studies with longer follow-up periods are needed to support a definite treatment opinion.

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