Rectal Lymphoma: Report of a Rare Case and Review of Literature

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Abstract

Rectal Lymphoma is a seldom disease, presenting in less than 0.5% of all primary rectal neoplasms. Due to the disease’s primary non-specific symptoms, the diagnosis is usually made at advanced stages. Clinically the disease is usually indistinguishable from rectal carcinoma. The optimal approach for managing rectal lymphoma has still not been identified. We report a case of stage IV which was staged by the hematologist and as per Ann Arbor staging primary rectal diffuse large B-cell lymphoma in a 71-year-old Omani female.

Keywords: Rectal Lymphoma Lymphoma.

Introduction

Lymphomas are neoplasms arising from lymphoid tissues, diagnosed by biopsy as either Hodgkin or Non-Hodgkin lymphomas. The vast majority are of B cell origin [1]. They present as either a primary lesion or a part of a malignant process. One-third of non-Hodgkin’s lymphomas are extra-nodal [2]. The gastrointestinal tract is the most common site among all extra-nodal diseases [3]. The most frequent gastrointestinal tract (GIT) location is the caecum (35.7%), followed by the ileum (20.3%), then the rectum (9.1%) [4]. There’s a male predominance, with the highest reported incidence in the 50-70 years age group [5].

Dawson et al [6] has established a criteria for the diagnosis of primary colorectal lymphomas in 1961 (these include: no palpable superficial lymph nodes at presentation, no enlarged mediastinal lymph nodes on chest X-ray, the normal range for white blood cell count including total and differential, at surgery, only the regional lymph nodes are involved, the liver and spleen are disease free).

A dilemma is still present in treating GIT lymphomas. The optimal approach is yet to be established. Surgical intervention remains a controversial approach. In this paper, we report a case of an elderly female presented to our institution with altering bowel habits and weight loss, diagnosed later on as stage IV primary rectal diffuse large B-cell lymphoma, treated with chemotherapeutic agents.

Case Report

The patient is a 71-year-old female with Hepatitis B, hypothyroidism, hyperlipidemia, and osteoporosis (all her medications directed on treating the patient comorbid were given during this period), presented to the Armed Forces Hospital, Muscat, Sultanate of Oman, with altering bowel habit of 7 months duration, weight loss of 10 KGs over the same period, vague epigastric pain and on/off fatiguability. No fresh rectal bleeding, or melena.

Upon examination, the patient looked emaciated, pale, and dehydrated. Abdominal examination demonstrated a soft non-tender abdomen, with bilateral enlarged inguinal lymph nodes; the left side was evident for 2 enlarged nodes 2x1 cm, discrete with no overlying skin changes. The right side showed one enlarged inguinal lymph node of 3x5 cm in size, rubbery, mobile, non-tender, with no skin changes.
**Figure 1:** Endoscopic picture of tumor at the lower rectum.

**Figure 2:** (a) Extension of the tumor to the anal canal. (b) CT abdomen with contrast, showing thickening of the lower rectum.
Per-rectal (PR) revealed perianal swelling with an ulcer. Circumferential hard mass up to 5-6 cm from the anal margin. Flexible sigmoidoscopy was done, demonstrating Indurated circumferential lesion 5 cm from the anal margin, for which a biopsy was taken.

The patient was accordingly transferred to the oncology center at SQUH where she had a CT scan that reported a circumferential asymmetrical enhancing thickening of the rectum, extending inferiorly involving the anal canal with mild perirectal inflammatory changes. A PET CT was also performed before the initiation of treatment [figure 5].
MRI was also planned, unfortunately, the patient was not cooperative enough for this type of imaging.

Other than hypochromic microcytic anemia (which was approached conservatively, the patient didn’t require a blood transfusion, IV fluids were given, Hb ranged between 10-11 g/dL), laboratory investigations including Tumor Markers (CEA, CA19.9, AFP) were within the normal ranges (done prior to diagnosis, LDH was checked and found to be high; 315 U/L).

Biopsy revealed an ulcerated rectal mucosa with underlying atypical lymphoid infiltrate composed of diffuse sheets of large atypical cells with vesicular nuclei, multiple prominent nucleoli and clear cytoplasm. The background shows abundant vasculature admixed with occasional small lymphocytes and neutrophils. There’s no evidence of epithelial dysplasia.

Immunohistochemistry showed the neoplastic lymphoid cells are positive for CD20, BCL2, BCL6, and MUM-1. They are negative for CD3, CD10, CD5, and CD30, AE1/AE3, chromogranin, and synaptophysin. EBER-ISH is negative, Ki67 is around 50%. The final impression was high-grade lymphoma consistent with diffuse large B-Cell lymphoma, non-germinal centre (activated B-Cell) type [figure 3 & 4].
The patient was transferred to an oncology center, patient had pre-chemotherapy investigations including full blood count, urea and electrolytes, coagulation profile, CSF study (no abnormal large lymphocytes were seen), bone profile, liver function test, thyroid function test, magnesium, C-Reactive Protein (CRP), Hepatitis B PCR, along with an intimal PET CT scan, accordingly, patient, therefore, received 6 cycles of R-CHOP 3-weekly [Rituximab Injection 500 mg/50 ml, Cyclophosphamide injection 500 mg/vial, Hydroxydaunorubicin hydrochloride (Doxorubicin hydrochloride injection 50 mg/25 ml vial), Vincristine injection 1 mg/vial (Oncovin) and Prednisone 20 mg tablet], and intrathecal Methotrexate 50 mg/5 ml; 4 cycles in total (no malignant cells were found in the CSF, intrathecal Methotrexate was given prophylactically). Marrow support was established by giving granulocyte colony-stimulating factors; Filgrastim 300 microgram/0.5 ml. Complications of chemotherapy were mainly nausea and vomiting which were approached by giving Aprepitant capsules 125 mg to prevent those episodes, Granisetron injection of 1 mg/mL was given to terminate such episodes. The patient had a second PET scan (PET scan performed post 4th cycle of R-CHOP, before the fifth cycle) which in comparison to the initial scan showed persistent mild fluorodeoxyglucose uptake in the rectum at the site of the previous soft tissue lesion which is likely physiological, suggestive of Complete Metabolic Response [figure 5 & 6]. A subsequent colonoscopy was carried out, rectum had a circumferential wall stricture at 5 cm from the anal verge, the stricture was dilated using a 15 mm balloon during the procedure. The Sigmoid, Descending, and Transverse Colon had normal mucosa with no inflammatory changes. After treatment, the patient responded rather well, was clinically stable, with no palpable lymph nodes, with a normal overall physical examination. The patient was discharged on granulocyte colony-stimulating factor (GCSF), had a follow-up PET scan scheduled one-month post treatment, had repeated colonoscopy and biopsy for an end-of-treatment response, colonoscopy demonstrated a circumferential wall stricture at 5 cm from anal verge at the rectum, passed by OGD. Multiple biopsies were taken from the stricture site and a polyp was found also taken for biopsy. Stricture dilated with a 15 mm balloon, biopsy showed chronic inflammation with focal mucosal fibrosis; likely secondary to treatment. No residual lymphomatous infiltrate is detected in this material. Follow-up scheduled with the oncology department at SQUH.

Figure 6: PET scan at the end of treatment.
Discussion

Several classifications are established for the diagnosis of Lymphoma [7]. According to the location, it can be nodal or extranodal. Another classification based on histopathology is Hodgkin and Non-Hodgkin Lymphoma. Rectal Lymphoma is the rarest reported rectal malignancy.

Rectal lymphoma presents with signs and symptoms indistinguishable from rectal carcinoma [8]. The patients usually seek medical advice due to alterations in bowel habits, weight loss, or rectal bleeding. Which was a similar presentation for our patient, who presented with these unspecific symptoms along with a rectal mass.

The diagnostic approach for those patients starts with a physical examination and a digital rectal examination, followed by a colonoscopy along with a biopsy for histopathological assessment and confirmation. Our patient’s histopathological examination revealed High-Grade Diffuse Large B-cell Lymphoma.

After confirming a diagnosis, staging has to be carried out by means of a PET CT.

An ideal management approach for rectal lymphoma is yet to be established. Surgical intervention is mostly indicated for localized tumors, as many consider medical treatment as the primary approach in managing such patients [9]. Surgical management along with chemotherapy and radiotherapy has offered better outcomes than medical management alone in primary rectal lymphoma. However, secondary involvement of the rectum offers only a poor prognosis and is considered better managed with radiation alone [10].

References