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Primary Diffuse Large B-Cell Lymphoma of the Rectum: A Case Report and Literature Review

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Abstract

Primary rectal lymphoma (PRL) is a rare form of gastrointestinal lymphoma, representing less than 0.5% of all colorectal neoplasms; Clinical symptoms are similar to those of rectal cancer. This article presents a rare case of primary rectal lymphoma discussed by a thorough review of the literature.

Keywords: Rectal lymphoma, Diffuse large B-cell lymphoma, Immunohistochemistry, R-CHOP.

INTRODUCTION

Lymphoma are a malignant lymphocyte. They can be classified as Hodgkin's lymphoma and non-Hodgkin's lymphoma [1]. Malignant non-Hodgkin's lymphoma can be divided into nodal and extranodal lymphoma. The gastrointestinal tract is the most common localization of extranodal lymphoma.

Primary rectal lymphoma is a rare disease accounting for 0,2% of all rectal malignancies. Diffuse large B-cell lymphoma is the most common form of primary colorectal non-Hodgkin lymphoma.

Dawson *et al*., established diagnostic criteria for primary colorectal lymphoma in 1961; these are: Absence of clinically lymphadenopathy on physical examination. A chest x-ray does not show enlarged mediastinal lymph nodes. Normal white blood cell count. Only the regional lymph nodes are affected during surgery. The liver and spleen are free of disease.

CASE REPORT

We present the case of a 75-year-old male presented with constipation, minor rectal bleeding and 6 kg weight loss within the past three months. On physical examination, the patient was haemodynamically stable. There were no enlarged palpable lymph nodes. Abdominal examination did not show any palpable masses. The rectal examination associated by anuscopy was normal. Laboratory analyses were normal with a Hb of 14 g /dl.

Colonoscopy showed an ulcerated process with a pseudonodular big folds, circumferential, non-stenosing of the upper rectum extended from 13 to 18 cm from the anal margin, other parts of colon, from sigmoid colon to cecum, were normal.

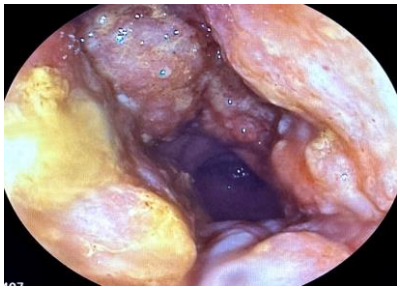

Fig 1: Endoscopic view showing a pseudonodular and ulcerated big folds in the rectum

Rectal biopsy concluded in non-Hodgkin's diffuse large B-cell lymphoma (DLBCL). Immunohistochemistry showed that the neoplastic cells were positive for CD20, Bcl 2 and Bcl 6 but negative for CD10, AE1 /AE3 and cyclin D1.

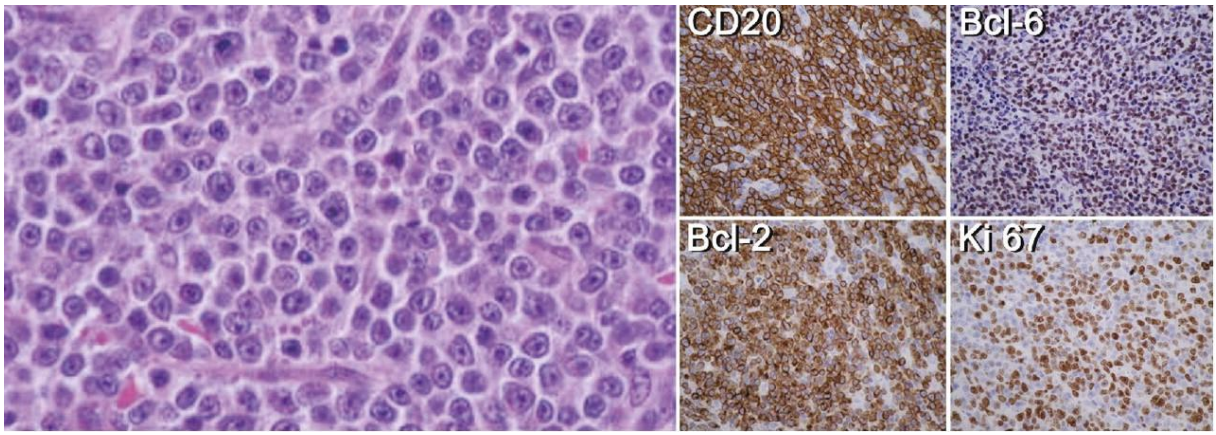

Fig 2: Morphological and immunohistochemical features of DLBCL

Upper endoscopy showed an erythematous gastritis whose anatomopathological results were in favor of a chronic antral gastritis with focal intestinal metaplasia, Helicobacter pylori infection was confirmed

Thoraco-abdomino-pelvic computed tomography showed irregular circumferential non-stenosing parietal thickening of the middle and upper rectum extending to 9 cm. here was no associated intra-abdominal lymphadenopathy. the patient is classified as having a stage I diffuse large B-cell lymphoma.

The patient was referred to clinical hematology department for R-CHOP chemotherapy including Rituximab, cyclophosphamide, doxorubicin, vincristine and prednisolone.

DISCUSSION

Gastrointestinal primary lymphomas account for 30% of extranodal lymphomas The stomach and small intestine are the most commonly affected sites [1]. They account for 0.1-0.6% of all colonic malignancies and 0.05% of all primary rectal malignancies [2]. Most of these tumors are of B-cell origin and are of the large diffuse type [3]. The majority of patients with rectal lymphoma present with nonspecific symptoms suggestive of primary rectal cancer: Weight loss, abdominal pain, lower gastro-intestinal bleeding [4].

The tumor may present as a circumferential or polypoid mass, ulceration, or a diffuse infiltration with structuring and bowel wall thickening. 86 per cent of the lesions are solitary, but may be multifocal and diffuse

Endoscopy with biopsy is the most valuable diagnostic test, Immunohistochemistry commonly used to confirm the diagnosis includes CD20, CD79a, and CD10 [5]. A CT scan is usually used to determine the extent of the tumor. Concentric thickening of the rectal wall with or without regional lymph node involvement is commonly reported. Positron Emission Tomography (PET) is currently being adapted for diagnosis and disease progression monitoring [6]. The Ann Arbor staging system, modified by Musshoff, is widely used to classify tumors [7].

There are two risk factors for developing primary colorectal lymphoma: Inflammatory bowel disease and immunosuppression (post-transplant, AIDS or immune disorders). The progressive nature of AIDS-related lymphoma can usually lead to disseminated disease at diagnosis. None of which were present in our patient [8].

Due to the rarity of rectal localization, there is no standard treatment protocol. Chemotherapy remains the most important treatment [9]. The RCHOP protocol, which includes rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisone, remains the most commonly used protocol [10]. Several studies have shown a trend towards improved survival when surgery is combined with chemotherapy or radiotherapy [11]. In our case, the patient was referred to her R-CHOP protocol.

CONCLUSION

Lymphoma of the rectum is a very rare entity, and there is still controversy about its treatment, the management is not well codified because of its rarity. Chemotherapy +/- radiotherapy is currently the standard treatment. Surgery is reserved for complications.

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