An unexpected case of primary rectal diffuse large B-cell lymphoma

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Abstract

Primary rectal lymphoma (PRL) is the rarest primary rectal cancer. Clinical presentation is similar to rectal carcinoma. The role of surgery is still controversial and the benefit of chemotherapy and radiotherapy is not well established. We here report the case of female patient suffering from PRL completely disappeared with chemotherapy.

Case Report

An unexpected case of primary rectal diffuse large B-cell lymphoma

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Consent:

Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

Keywords:

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Introduction:

Primary colorectal lymphoma (PRL) accounts for 1.4% of all cases of non-Hodgkin's lymphoma (NHL) and less than 1% of all colorectal malignancies. Although the gastrointestinal (GI) tract is the most common site of extranodal lymphoma, colorectal lymphoma is rare [1]. Dawson and al established criteria for the diagnosis of PRL in 1961 including the absence of clinically enlarged lymphadenopathy in physical examination, the absence of enlarged mediastinal lymph nodes on chest x-ray, normal white blood cell count and bone marrow biopsy, and the absence of abnormalities of the liver and spleen [2]. While treatment strategies in nodal NHL are well established, management of PRL is still unclear [3]. We present the case of a 38-year-old woman, diagnosed with a stage IIE PRL

Patient and observation:

Patient information: A 38-year-old woman presented with constipation, a feeling of incomplete evacuation and weight loss.

Clinical findings: A fixed mass was identified on digital rectal examination. No palpable superficial lymph nodes were identified and chest x-ray was normal.

Diagnostic Assessment: The colonoscopy revealed a circumferential ulcerated non obstructive mass about 5 cm from the anal verge extended up to 10 cm (Figure 1). Rectal biopsy concluded in non-Hodgkin's diffuse large B-cell lymphoma (DLBCL) CD20(+), CD5(+), CD3(+), Bcl-2(–), CD56(–), CD117 (–), cyclin D1 (-), and Ki-67 at 80% (Figure 2). Cervical and thoraco-abdomino-pelvic computed tomography (CT-scan) showed irregular circumferential non-stenosing parietal thickening of the rectum located at 4 cm from the anal verge and extended over 9cm. Mesorectal and bilateral hypogastric lymph nodes were identified, the largest one measured 26 x 20 mm (Figure 3). CEA, LDH, white blood cell count and the bone marrow biopsy were normal. According to the Ann Arbor staging classification, tumor was classified stage IIE.

Therapeutic intervention: The patient received six cycles of R-CHOP chemotherapy including Rituximab, cyclophosphamide, doxorubicin, vincristine and prednisolone with good tolerance.

Follow-up and outcomes : Radiological and endoscopic evaluation concluded in a complete remission confirmed with rectal biopsy. The patient is monitored each 3 months and is still in complete remission after 18 months of follow up.

Discussion:

The most common histologic subtype of NHL affecting the GI tract and colon is DLBCL. PRL usually presents with signs and symptoms suggestive of primary rectal carcinoma: weight loss, abdominal pain and lower gastro-intestinal bleeding [4]. Whatever the presentation, endoscopy with biopsy are the most valuable diagnostic tests. The majority of colorectal lymphomas are localised in the cecum or ascending colon [5]. As reported in the literature, colonoscopy showed in our case a budding rectal mass, and biopsy confirmed the diagnosis of DLBCL. Immunohistochemistry usually used to confirm diagnosis is CD20, CD79a, and CD10 [6]. CT scan is usually used to study tumor extension. Concentric rectal wall thickening with or without a regional lymph node involvement are frequently reported. Positron emission tomography (PET) is currently indicated in the diagnosis and disease follow-up [7]. The Ann Arbor staging system modified by Musshoff is widely used in tumor classification [8]. Given the rarity of the rectal localization, there is no standard treatment protocol. Chemotherapy remains the the main therapeutic modality [9].R-CHOP protocol including Rituximab, cyclophosphamide, doxorubicin, vincristine and prednisone remains the most used protocol [10]. Several studies reveal a trend toward improved survival when surgery is used in combination with chemo- or radiotherapy [11]. In our case, we have treated patient with chemotherapy with a complete response. Jeong and al noted a significant difference in survival between patients with stages I-II and stage IV disease [5].

Conclusion:

Management of rectal lymphoma is not well codified because of its rarity. Chemotherapy +/- radiotherapy is currently the standard treatment. Surgery is reserved for extensive post-chemotherapy stenosis. Individ-

ualized treatment strategies need to be developed based on specific clinical situations. Currently we could not make formal conclusions apart from the effectiveness of chemotherapy and anti CD20 treatment.

Competing interests

The authors declare no competing interest.

Figures:

Figure1: Rectal circumferential ulcerated tumor

Figure2: Histopathology showing an H&E stained section of a rectal biopsy× 40 (A); IHC showing CD20 + rectal cells (B) and CD3+ rectal cells (C); IHC analysis of Ki-67 expression (Ki-67 =80%) (D).

Figure3: Pelvic CT-scan: parietal thickening of the rectum and mesorectal and bilateral hypogastric lymph nodes

Authors' contributions:

I confirm that all the authors of the manuscript have read and agreed to its content.

Sondes BIZID , Fatma BEN FARHAT, Hela GHEDIRA, Sonia BENNASER,

Beya CHELLY, Hela STAMBOULI wrote the paper.

Fehmi Msadek contributed to the quality control and correction of the manuscript before submission.

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