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Extranodal Diffuse Large B-Cell Lymphoma of the Small Bowel in Female Patient Causing Intestinal Obstruction: A Case Report

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Abstract. Diffuse large B cell lymphoma is the most common extranodal non-Hodgkin lymphoma of the small intestine accounting for more than 50% of cases. Forty percent of these cases initially present with small bowel obstruction. Therefore, the diagnosis is usually established after surgery for bowel obstruction. The treatment is then continued with a certain chemotherapy regimens. We present a case of a 46-years-old female patient with signs of small bowel obstruction due to previously undiagnosed diffuse large B-cell lymphoma. Postoperatively, the patient was treated with 7 cycles of R-CHOP protocol and complete response was achieved in the short follow-up period.

Key words: diffuse large B-cell lymphoma, extranodal, small bowel obstruction.

Introduction

The most frequent extranodal presentation of non-Hodgkin lymphoma (NHL) is the gastrointestinal tract (GIT), accounting for more than 30% of cases [1]. Additionally, diffuse large B-cell lymphoma (DLBCL) is the most common non-Hodgkin lymphoma and it is also frequently diagnosed with extranodal presentation in the gastrointestinal tract [2]. They are difficult to diagnose due to their insufficiently defined symptoms and

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the diagnosis is often established in an urgent settings [3]. The small intestine is the second most common place in GIT where extranodal NHL occurs (20% of all gastrointestinal lymphomas) [4]. Few reports in the literature exist on DLBCL causing acute small bowel obstruction [3, 5, 6]. We present a case of small bowel obstruction caused by an aggressive type of DLBCL in a female patient with previous absence of symptoms regarding malignant lymphoma. Written informed consent has been obtained from the patient.

Case presentation

A 46-years-old female patient presented to the emergency department with clinical signs of intestinal obstruction. The patient reported cramp and abdominal pain, and absence of flatus and stool passage one day prior to the exam. She had no previous abdominal surgery nor visible groin herniation on physical examination. The serum analysis showed C-reactive protein value of 5.80~(0.0-5.0~mg/L) and serum Iron level of $5.80~(6.6-28.3~\mu mol/L)$. Plain abdominal upright X-ray image confirmed the suspicion for intestinal obstruction (Figure 1).

Contrast-enhanced Computerized Tomography (CECT) of the abdomen was the next diagnostics tool. The small intestine was presented with dilated loops, formed "air fluid levels" and free fluid in the Douglas pouch. The tumor formation itself was not detected by the radiologist on this scan (Figure 2).

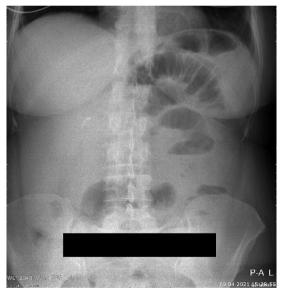


Figure 1. Plain abdominal X-ray with small bowel airliquid levels



Figure 2. CECT of abdomen with signs of small bowel obstruction

Admission in hospital and exploratory laparotomy followed. Intraoperatively, a tumor with the size of an egg was found in the ileal wall causing the obstruction. Resection of the affected loop was done with a termino-terminal anastomosis. Postoperative period was uneventful and the patient was discharged on postoperative day 4. Superficial Surgical Site Infection (SSI) occurred at the first scheduled checkup and it was treated in the outpatient ward.

The macroscopic histopathology analysis revealed the presence of a tumor with dimensions of 6x5x2.5 cm (ulcerative type) that spreads in the surrounding adipose tissue. The microscopic aspect of hematoxylin and eosin staining presented with infiltrative growth of lymphoid cells with large nuclei and frequent mitoses. Immunohistochemical staining showed high proliferation over 90% of Ki67 and CD20 (Figures 3a, 3b, and 3c). The overall pathology report concluded the presence of aggressive diffuse large B-cell lymphoma.

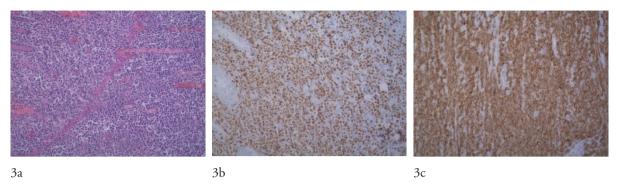


Figure 3. 3a – Hematoxylin Eosin stain with infiltrative growth of Lymphocytes, 3b – Ki67 stain with high proliferation, 3c – CD20 stain with proliferation over 90% (100x magnification)

The patient was referred to a hematologist for proper staging and therapy. The positron emission tomography-computed tomography (PET/CT) scan showed a normal findings with low metabolic activity in the area of the wound infection (Figure 4). Bone marrow biopsy was negative. According to the Lugano score [7], the patient was diagnosed with Stage I of the disease.

Seven cycles of R-CHOP (rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisolone) regimen were given to the patient as the first line of treatment. Post-chemotherapy PET/CT scan presented with normal findings. The patient is still in complete remission after the combined treatment with surgery and chemotherapy and regular future checkups are scheduled.

Discussion

DLBCL remains to be the most common extranodal NHL in the small intestine in 56.8% of patients [5]. More than two-thirds are diagnosed in Stage I/II (70.7%) [8], but nevertheless, initial presentation with small bowel obstruction is reported in 40% of them [9, 10]. However, only a few case reports are published [3, 6, 7, 11]. According to gender distribution, the male to female ratio is 2:1 [12]. The diagnosis is challenging in the emergency setting and it is often established postoperatively [3].

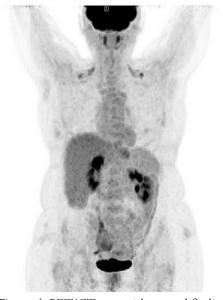


Figure 4. PET/CT scan with normal finding

In these patients, there is still a debate which treatment modality offers the best results regarding the complete remission. The proposed first-line regimen for extranodal gastrointestinal DLBCL is the use of CHOP or R-CHOP chemotherapy protocols. Most of the reports give a significant advantage of the rituximab addition to the CHOP protocol in terms of significantly greater overall survival and progression-free survival [13, 14].

Several studies have shown that the combination of surgery (first) followed by chemotherapy provides better survival benefit in patients with DLBCL [15, 16]. According to Zhang, the combination of surgery and R-CHOP offers significantly better event-free survival, better response rate, lower relapse rate, and higher overall survival rate, but without statistical significance in older patients [12]. Hwang reports 5-year survival rate of 86.4% in cases treated with surgery plus CHOP or R-CHOP [17].

To conclude, an emergency abdominal condition with clinical signs of bowel obstruction in a young and/ or middle aged patients without previous abdominal surgery history should raise suspicion for extranodal GIT lymphoma. One of the reliable therapeutic options in such emergency setup is the combination of surgery and chemotherapy and one should expect a complete therapeutic response in such patients.

Conflict of interest

The authors declare that there is no conflict of interest regarding the publication of this article.

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