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Case Report

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A PRIMARY GASTRO-INTESTINAL BURKITT LIKE LYMPHOMA: CASE REPORT AND REVIEW OF THE LITERATURE

Imen Jemni[1], Imen Akkari*[1], Soumaya Mrabet[1], Atef Ben Abdelkader[2], Kamira Zahra[3] and Elhem Ben Jazia[1]

[1]Imen Jemni, Imen Akkari, Soumaya Mrabet, Elhem Ben Jazia, Gastroenterology Department, Farhat Hached University Hospital, 4000 Sousse, Tunisia.

[2]Atef Ben Abdelkader, Anatomopathology Department, Farhat Hached University Hospital, 4000 Sousse, Tunisia.

[3]Kmira Zahra, Clinical Hematology Department, Farhat Hached University Hospital, 4000 Sousse, Tunisia.

***Corresponding Author: Imen Akkari**

Imen Jemni, Imen Akkari, Soumaya Mrabet, Elhem Ben Jazia, Gastroenterology Department, Farhat Hached University Hospital, 4000 Sousse, Tunisia.

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ABSTRACT

Primary non Hodgkin gastro intestinal lymphomas are rare and represent only 5 to 10% of gastro intestinal tract malignancies. Burkitt like lymphoma or atypical Burkitt lymphoma is not a well distinct entity because it represents a continuum between Burkitt lymphoma and diffuse large B-cell lymphoma. It's a highly aggressive B cell malignancy. The distinction is crucial because of the therapeutic repercussions on the management of the disease. We report a case of a primary gastro intestinal Burkitt like lymphoma (gastric and jejunal localization) in a 64 year-old men revealed by prolonged fever and weight loss.

KEYWORDS: Burkitt-like lymphoma; gastrointestinal tract; chemotherapy protocol.

INTRODUCTION

The gastrointestinal (GI) tract is one of the most frequent sites of extra-nodal non-Hodgkin's lymphomas (NHL) (30-50%). Although secondary GI involvement is relatively common in lymphoma, primary gastrointestinal lymphoma (PGL) is rare (5-10% of GI malignancies).[1]

NHL represents 1 to 4% of the malignancies arising in the gastrointestinal tract and stomach is the most common site of involvement.[2] The primary gastrointestinal non-Hodgkin's lymphoma, although rare, is among the most common extra-nodal lymphomas.[1] It is extremely rare for Burkitt-like lymphoma (BLL) and Burkitt lymphoma (BL) to occur in the alimentary tract of adults.[3]

BL and BLL are aggressive B-cell malignancies with a high proliferative rate that may be fatal within months if not treated promptly.[4] BL has been rather well characterized, whereas the other lymphomas morphologically resembling it are more heterogeneous. The cases classified as atypical BL or Burkitt-like lymphoma by the 2001 World Health Organization (WHO) Classification of Tumors of Hematopoietic and Lymphoid Tissue were thought to represent a continuum between BL and diffuse large B-cell lymphoma (DLBCL).[5]

Here we present a case of a primary gastro-intestinal Burkitt like lymphoma in a 64 year old men revealed by prolonged fever and weight loss.

CASE REPORT

A 64 year-old men with no medical history presented with persistent fever for the last 3 months associated with anorexia, weight loss, abdominal pain, arthralgia, xerostomia and xerophthalmia.

Physical exam showed a fever, no signs of hepatomegaly or splenomagaly and no peripheral lymph nodes enlargement. Biology findings revealed a bicytopenia (white blood cells: 2810/mm3, hemoglobin level: 7, 5 g/dl) with low reticulocytes count, high sedimentation rate at 125 in the first hour, high lactate dehydrogenate (LDH) level: 657 IU/ml, hypoalbuminemia and a slight elevated liver enzymes: 1,5*UNL.

Bone marrow biopsy showed a normal density with no signs of infiltration. A body scan was performed and revealed diffuse gastric wall thickening of 3, 9 cm with nodular infiltration of the surrounding fatty tissue, multifocal circumferential jejunal and ileal wall thickening, with intra abdominal enlarged lymph nodes (32*24mm), a liver metastasis in segment II and signs of peritoneal carcinomatosis.

Upper digestive endoscopy revealed three suspect ulcers in the fundus. The pathology report of these lesions biopsy showed signs of Burkitt like lymphoma with

strong expression of CD20, Bcl 6 and 100% Ki 67 expression (fig 1, Fig2). The diagnosis of primary gastro-intestinal burkitt like lymphoma with no central nervous system or bone marrow involvement was established.

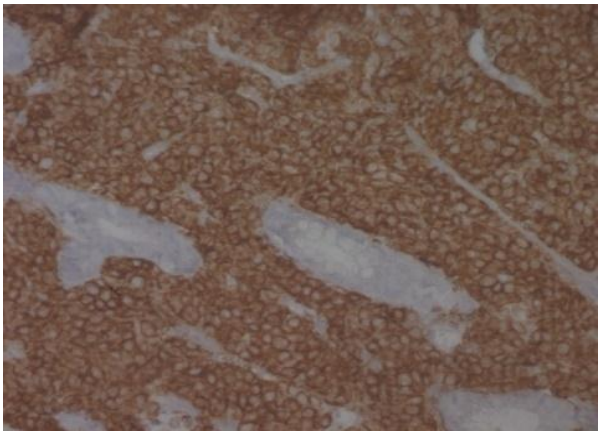

Fig 1: Histopathology aspects of the gastric ulcers biopsy (x 400): Expression of CD 20.

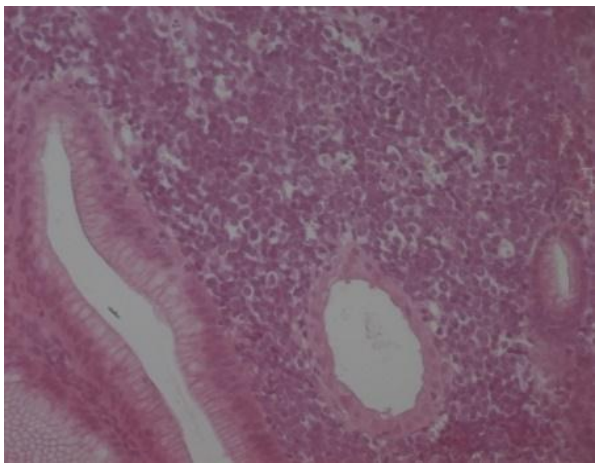

Fig 2: Histopathology aspects of the gastric ulcers biopsy (HE stain x 400): Diffuse infiltrate of atypical lymphoid cells showing mild pleomorphism admixed with tangible body macrophages (starry sky pattern).

The patient was treated with multiagent chemotherapy based on the LMB regimen:
RCOP (Rituximab –vincristine-cyclophosphamide – prednisone) followed by 2 courses of COPADEM (vincristine, cyclophosphamide, Adriamycin or doxorubicin, high dose methotrexate, prednisone), then 2 sessions of CYM (ARA-C: cytarabin, methotrexate).

Last chemotherapy course was in March 2016. The Last body scan control was in January 2017, the patient has been in a complete sustained remission.

DISCUSSION

Burkitt-like lymphoma (BLL) is a rare and rapidly progressive type of lymphoma that usually occurs in young males.[5] In the Revised European– American Lymphoma (REAL 1994) classification, BLLs are described as diffuse B-cell lymphomas with a high proliferative rate and molecular biological characteristics considered to be borderline between those of classic Burkitt lymphoma (BL) and those of diffuse large B-cell lymphoma (DLBCL).[5]

BLL can be recognized by its combined morphological and phenotypic features and by the high grade lymphoma that it presents, BLL exhibits morphological features intermediate between BL and diffuse large B-cell lymphoma: a heterogeneous population of medium to large cells with prominent central nucleoli, greater nuclear pleomorphism, smaller number of prominent nucleoli, a ‘‘starrysky’’ appearance are distinctive morphological features, but similar features of nearly 100% growth fraction and “strong presumptive evidence” of *MYC* translocation.[5] BLL is pathologically less well defined because it can present either with or without the c-myc translocation. It can sometimes also be associated with other chromosomal translocations, such as the bcl-2translocation in 33% of cases.[4]

The question arises whether BLL is a similar disease entity to BL, or whether it lies within a continuum between BL and DLBCL, with sufficiently different clinical behavior and morphologic, immunophenotypic, or genetic findings to warrant segregation from classic BL.

The 2008 revisions to the WHO Classification of Tumors of Hematopoietic and Lymphoid Tissue no longer formally recognize the atypical BL or BLL and plasmacytoid variants of BL.[1] Cases of high-grade, mature B-cell NHL not readily classifiable as BL or DLBCL are now designated “B-cell lymphoma, unclassifiable, with features intermediate between DLBCL and BL”.[1] Cases previously classified as atypical BL or BLL for morphologic features slightly deviating from typical BL (such as slight nuclear pleomorphism and/or more prominent single nucleoli), are now classified as BL if the immune phenotypic and genetic criteria are otherwise fulfilled. Cases of otherwise typical BL in which an *MYC* rearrangement cannot be demonstrated should be still classified as BL.[1]

BLL occur in various gastrointestinal sites and 30–80% of patients have involvement of the small bowel or colon and rectum, especially the ileocecal region. Involvement of the stomach occurs in only 10% of the patients and almost all of the cases were detected in the late stage as a bulky mass located in the abdomen or other gastrointestinal sites.[6]

Symptoms, such as abdominal pain, weight loss, bowel obstruction, GI bleeding and acute appendicitis are nonspecific. At diagnosis, patients usually have bulky disease and elevated LDH and uric acid levels. Bone marrow and central nervous system (CNS) involvement are reported in 30% and 13% of adults, respectively.[4]

Sonographic diagnosis of BLL is based on: marked wall thickening, absence of stratification with extremely low echogenicity and preserved compressibility. 3 the combination of all 3 of these mentioned findings yielded a sensitivity of 92% and a specificity of 92%. Thus, the sonographic criteria may be useful in the diagnosis of gastrointestinal lymphoma.[3]

Endoscopic features of rare B cell lymphoma including Burkitt like lymphoma were described in the study of Calogero V et al[7]: a burgeoning lesion in 54% of cases, an ulcero- burgeoning aspect in 27%, an infiltrative form in 8%, an ulcero infiltrative form in 6% and finally an ulcerative aspect in 5% of cases. On colonoscopy, endoscopic signs of the lesion may manifest in a variety of ways, ranging from solitary burgeoning masses to multiple colonic polyps, ulceration is the major endoscopic form.[4] The digestive exploration realized in our case revealed a rare localization and aspect of lymphoma.

The distinctions between these different entities (BL, DLBCL and BLL) have significant therapeutic implications, since a high proportion of cases of BL can be cured with short, intensive multi agent chemotherapy (usually in combination with rituximab: an anti-CD20 monoclonal antibody) and CNS prophylaxis, whereas CHOP-R (cyclophosphamide, doxorubicin, vincristine, prednisone, rituximab) is considered inadequate therapy for BL but represents a standard of care regimen for DLBCL.[5]

Treatment of BLL is based on intensive multi agent chemotherapy, such as the Magrath regimen CODOX-M+IVAC (cyclophosphamide, vincristine, doxorubicin and high-dose methotrexate/ ifosfamide, etoposide and high dose cytarabine and intrathecal methotrexate), or the BFM regimen (cyclophosphamide, ifosfamide, high dose methotrexate, and CNS prophylaxis).[4]

Another similar regimen for adults with Burkitt-like lymphoma published by Soussain et al[8] is the LMB (B-cell non-Hodgkin's lymphoma and B-ALL) pediatric protocol (initially set up for children with small non-cleaved cell lymphomas). It is based on the same kind of agents: cyclophosphamide, vincristine, prednisone, doxorubicin (Adriamycin), high-dose methotrexate, high-dose cytarabine (cytosine arabinoside) and intra-thecal ara-C.[8]

Surgery is generally not indicated, except in selected cases where there is bowel obstruction or perforation. In some institutions, treatment includes the use of consolidative bone marrow transplantation.[4]

Untreated BLL has a poor prognosis, the median survival is 6 months and 90% of patients with gastrointestinal BLL die within1 year of diagnosis.[3] With LMB regimen, a complete remission rate can be attained in 89% and a 74% overall survival at 3 years.[8] Our patient was treated with this regimen with complete remission.

CONCLUSION

The importance of the report is based on the fact that BLL is a rare entity. This case also underscores the importance of early recognition of highly aggressive lymphoma in order to secure optimal and timely treatment.

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