

A rare case of Burkitt's lymphoma of the duodenal bulb

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Abstract

Gastrointestinal tract involvement in immunodeficiency-related Burkitt's lymphoma is not common and the duodenal involvement is very rare. We report the case of a 35-year-old man admitted because of abdominal pain, vomiting and weight loss. Human immunodeficiency virus infection was diagnosed and upper digestive tract endoscopy showed marked edema and hyperemia of the duodenal bulb with some violaceous areas. Immunohistochemical study of the bulbar tissue samples confirmed the diagnosis of Burkitt's lymphoma. To our knowledge, duodenal Burkitt's lymphoma affecting only the bulb has not been previously reported in the medical literature. In patients with human immunodeficiency virus infection who present with upper gastrointestinal symptoms, upper endoscopy may be diagnostic of malignancy and biopsies should be obtained from abnormal areas.

Keywords Burkitt's lymphoma, duodenal bulb, non-Hodgkin's lymphoma, human immunodeficiency virus

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Introduction

Non-Hodgkin's lymphoma (NHL) of B cell type is the second most common neoplasm after Kaposi's sarcoma, among patients with human immunodeficiency virus (HIV) infection [1,2]. Burkitt's lymphoma (BL) is one of the highly aggressive B-cell NHL that is characterized by the translocation and deregulation of the *c-myc* gene on chromosome 8 [3]. Besides the immunodeficiency-associated subtype, there are other two distinct forms of BL: endemic (African) and sporadic that share the same aggressive clinical behavior and are histologically identical [1]. Immunodeficiency-related BL cases commonly present with nodal and bone marrow involvement. Gastrointestinal presentations are uncommon and most often affect the bowel (ileum and cecum) or abdominal lymph nodes [1,4]. BL of the small intestine primary involving the duodenum is very rare and most of the tumors are found in the second portion of the duodenum [1].

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Conflict of Interest: None

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Case report

A 35-year-old man was admitted to our hospital because of weight loss, nausea, vomiting and diffuse abdominal discomfort with one-week duration. He had no known previous diseases and did not take any medication. He had lost approximately 10% of weight during the last year prior to admission. His abdomen was enlarged and tender with spontaneous epigastric and right hypochondrium pain without rebound tenderness. The relevant laboratory findings were: hemoglobin 13.2 g/dL; leukocytes 5,450/mm³; platelets 138,000/mm³; lactate dehydrogenase 2,448 U/L, aspartate aminotransferase 183 U/L, alanine aminotransferase 80.9 U/L, total bilirubin 0.4 mg/dL, albumin 3.1 g/dL, amylase 2 U/L, γ -glutamyl transferase 160 U/L and C-reactive protein 6.4 mg/dL. Coagulation and kidney function tests were normal. HIV serology was positive and the CD4 T cell count was 99 cells/ μ L. Abdominal computed tomography (CT) scan showed hepatosplenomegaly and diffuse peritoneal thickening with small cystic areas (Fig. 1). Small bowel wall thickening was also seen. Upper digestive tract endoscopy was performed: the esophagus showed non-detachable white plates (Fig. 2A); the stomach did not show abnormal findings (Fig. 2B, C); the inspection of the duodenal bulb revealed marked edema and hyperemia with some violaceous areas (Fig. 2D, E); finally, the second and third portions of duodenum showed no abnormal findings (Fig. 2F). Several biopsy samples were taken. Histopathological examination of the esophagus biopsies showed multiple spores and hyphae of *Candida albicans*. The duodenal bulb biopsies showed a diffuse infiltrate of

monotonous medium-sized lymphoid cells, with round nuclei, multiple small, basophilic nucleoli and basophilic cytoplasm (Fig. 3). The atypical cells were reactive to CD20, CD10, c-myc and bcl-6 and not reactive to bcl-2 and CD3. The Ki67 antibody was consistent with a very high proliferation index (close to 100%) (Fig. 4). Epstein-Barr virus was detected by *in situ* hybridization. This histopathological findings were consistent with the diagnosis of BL of the duodenal bulb. The patient was also submitted to ultrasound-guided peritoneal biopsy and the peritoneal samples were also compatible with the diagnosis of BL. Cranial CT scan and lumbar puncture excluded central nervous system involvement. Chest CT scan showed no relevant aspects. Bone marrow biopsy revealed 80% of atypical cells with Burkitt phenotype and the translocation (8,14). On the basis of those findings, the patient was diagnosed with BL stage IV, using the modified Ann Arbor staging system. He was started on highly active antiretroviral therapy and received

systemic chemotherapy with CHOMP (cyclophosphamide, doxorubicin, vincristine, methotrexate and prednisolone). One week later, he developed a tumor lysis syndrome but he recovered and is asymptomatic two months after the diagnosis.

Discussion

Immunodeficiency-associated BL accounts for about 30% of lymphomas in HIV patients [6] and is a condition that defines acquired immunodeficiency syndrome. While endemic BL most commonly involves the jaw and facial bone, and sporadic BL presents with abdominal tumors, immunodeficiency-related BL cases often involve bone marrow and lymph nodes [4,6]. Sometimes, the liver, spleen, kidney, and ovary may be involved [1,7]. Typically, patients report "B" symptoms such as weight loss and abdominal symptoms like abdominal pain, nausea and vomiting [4,7], as seen in this case. Presentation as pancreatitis or biliary obstruction is also reported [8]. Only a few cases of duodenal BL were reported in the medical literature all referring to the second and third portions of the duodenum [7,8]. To our knowledge, duodenal Burkitt's lymphoma affecting only the bulb has not been previously reported in the medical literature. Endoscopic findings are not specific but the documented cases reported a polypoid pattern, donut-shaped lesions, thickened gastric folds and raised ulcerated nodules [1,7,9].

The exact cause and pathophysiologic mechanisms leading to the development of BL are not known but Epstein-Barr virus has been implicated and can be detected in 25-40% of immunodeficiency-associated cases of BL [5], as seen in this case. Evidence exists for a significant interaction between viral and cellular microRNA interfering with normal gene



Figure 1 Abdominal computed tomography scan showing hepatosplenomegaly

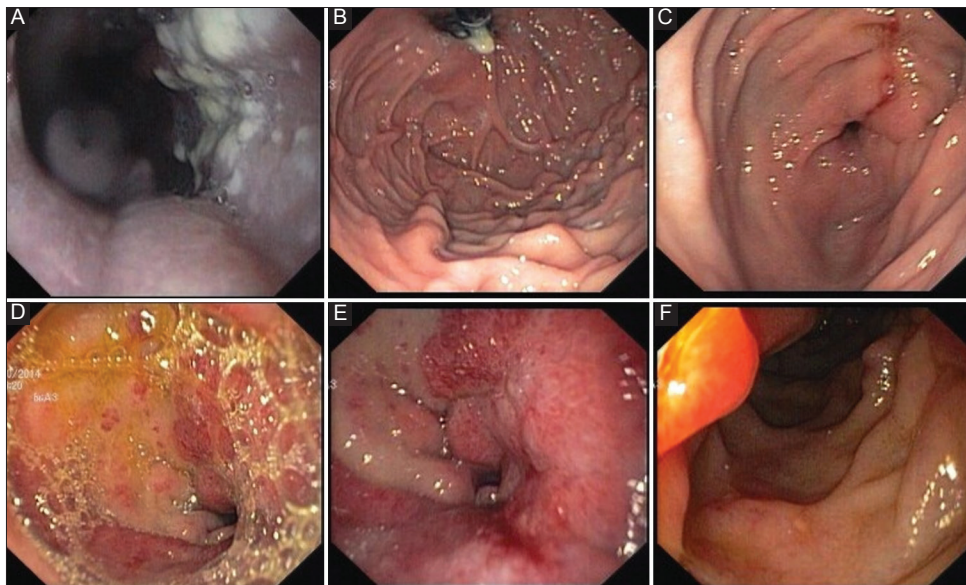


Figure 2 Upper digestive tract endoscopy: (A) esophagus showing non-detachable white plates; (B, C) stomach with no abnormal findings; (D, E) duodenal bulb revealing marked edema and hyperemia with some violaceous areas; (F) second and third portions of duodenum showing no abnormal findings

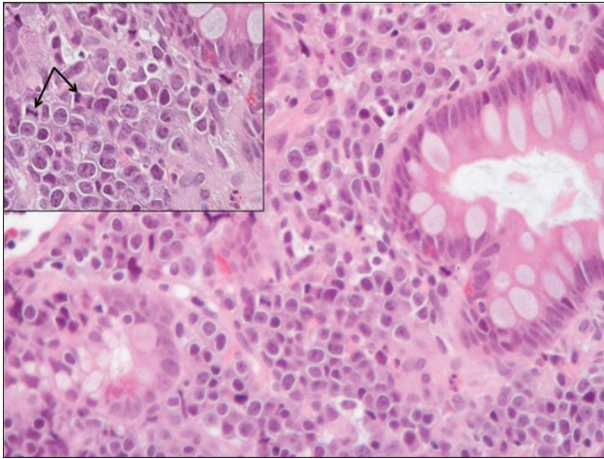


Figure 3 Between the crypts of the duodenal mucosa there is a diffuse infiltration by medium-sized lymphoid cells; they have a round or oval nuclei and a squared-off borders (H&E, x400); there are many mitotic figures (insert H&E, x1000- arrow)

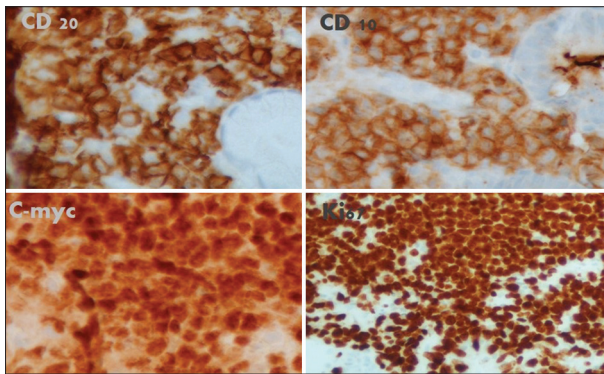


Figure 4 The cells are immunoreactive to CD20 (x400), CD10 (x400) and c-myc (x400) but were negative to bcl-2 (not shown). The Ki67 proliferative index (x200) is virtually 100%

expression and translation [10]. BL typically affects patients with high CD4 T cell counts ($>200/\text{mm}^3$) which suggests that decreased immunity is not a risk factor in this variant of BL [4]. However, our patient had low CD4 T cell counts (less than 200 cells/ μL).

The Ann Arbor staging system consist of four stages in which the stage 4 refers to central nervous system or bone marrow involvement. Patients with more than 25% bone marrow involvement are usually referred to as having Burkitt leukemia or, according to French American British classification, L3

type acute lymphoblastic leukemia [4]. The World Health Organization Classification of lymphoid neoplasms identifies BL and Burkitt leukemia as two different manifestations of the same disease based on genetic and immunohistochemical similarity [5]. Our patient was considered as having Burkitt leukemia because he had 80% bone marrow involvement.

As BL is one of the fastest growing malignancies in humans [4] requires immediate therapeutic intervention. Despite its rapid growth, it responds well to aggressive chemotherapy [4,6], the gold standard treatment. Tumor lysis syndrome can occur as a result of massive acute destruction of the tumor cells during initial chemotherapy [4]. The probability of complications increases with the extent of the disease [4].

In conclusion, in patients with human immunodeficiency virus infection who present with upper gastrointestinal symptoms, upper endoscopy may be diagnostic of malignancy and biopsies should be obtained from abnormal areas.

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