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Case Report

Gastric Burkitt Lymphoma, an Infrequent Cause of Hematemesis

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Abstract

Among gastric malignancies, lymphomas rank as the second most common type, following gastric adenocarcinoma. The majority of gastric lymphomas are either mucosa-associated lymphoid tissue lymphomas or diffuse large B-cell lymphomas. Primary gastric Burkitt lymphoma, is a subtype of non-Hodgkin's lymphoma, represents an exceptionally rare and aggressive malignancy, with only a limited number of reported cases globally. We present the case of a male patient with primary gastric Burkitt lymphoma revealed by digestive hemorrhage. The diagnosis was confirmed by endoscopy and gastric biopsies. The gastric localization of Burkitt's lymphoma is unusual. Our work is a review of the literature on Burkitt's lymphoma in its gastric localization.

Keywords: Hematemesis, Burkitt Lymphoma.

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INTRODUCTION

Burkitt's lymphoma (BL) was first described by Denis Burkitt, an Irish surgeon, in the middle of the last century, in Kampala, Central Africa. In 1961 Burkitt met Epstein, an experimental pathologist who identified a specific virus in the samples of tissues from this lymphoma. The virus became known as Epstein Barr virus (EBV). This was the first time ever that a virus was identified as a contributing factor in the development of a tumor in humans [1].

Burkitt's lymphoma is a malignant proliferation of B lymphocytes. It is an aggressive lymphoma that most often has an extra-nodal lymph. When it involves the digestive tract, it is located electively in the ileocoecal region, gastric involvement is extremely rare [2].

The endoscopic appearance of this type of lymphoma, due to its rarity, remains poorly understood. We take this opportunity of this observation to clarify the clinical presentation and the Endoscopic features of gastric Burkitt's lymphoma.

CASE PRESENTATION

An 81-year-old man, Operated for benign prostatic hypertrophy by trans ureteroprostatic resection, smoker and chronic alcoholic, who presented to the emergency unit for epigastralgia evolving for a month with intermittent vomiting complicated on the same day of his hospitalization by the occurrence of hematemesis and melena evolving in a context of altered general condition with weight loss not quantified. Physical examination revealed pallor and epigastric tenderness.

Initial laboratory investigations revealed hemoglobin was 9, 7 g/dL, the white blood count was 7, $2\times10^9/L$, and the platelet count was $324\times10^9/L$. Other findings included creatinine 24 mg/dl, Carbohydrate antigen (CA) 19-9 and carcinoembryonic antigen (CEA) were within normal limits, HIV test was negative.

Proceeding with investigations, the upper gastrointestinal endoscopy revealed an antral pre-pyloric ulcerative process, bleeding easily on contact, as well as two fundic ulcerations with raised edges. The duodenum and the esophagus were normal. (Figure 1)

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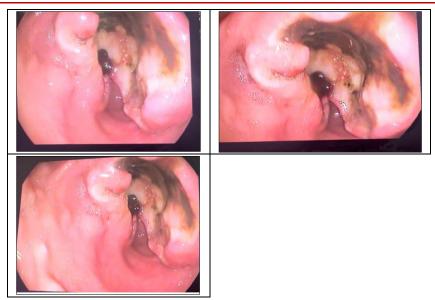


Figure 1: Upper gastrointestinal endoscopic images

Histological examination of the endoscopic gastric biopsy revealed diffuse infiltration of the gastric mucosa by a lymphomatous tumoral process (Figure 2A and 2B), the immune histochemical evaluation revealed that lymphoid cells were positive for CD20, CD10, and

Bcl6, negative for CD23, CD5 and Bcl2. All of the neoplastic cells were positive for Ki-67. These findings pointed toward a diagnosis of gastric Burkitt's lymphoma. (Figure 3)

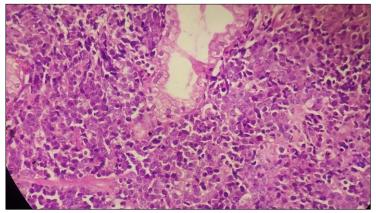


Figure 2A: H and E section at 10x magnification showing atypical lymphoid cells surrounding and infiltrating the gastric glands

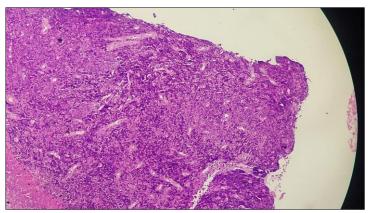


Figure 2B: H and E section at 10x magnification showing lymphomatous infiltration in diffuse sheets made up of medium-sized cells with ovoid nuclei that are mostly crushed

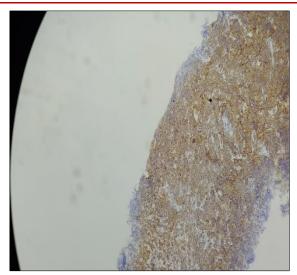


Fig. 3A

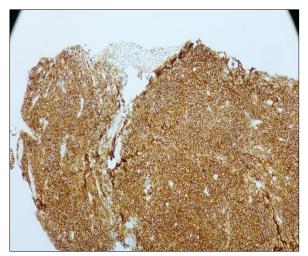


Fig. 3B

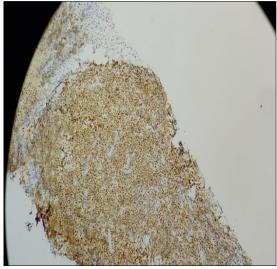


Fig. 3C

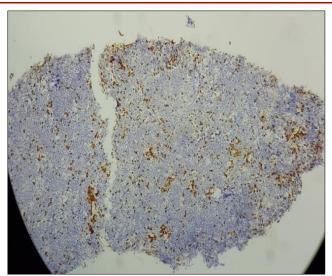


Fig. 3D

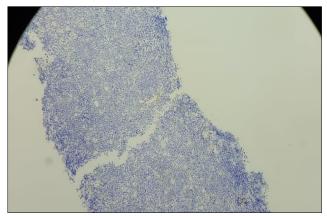


Fig. 3E

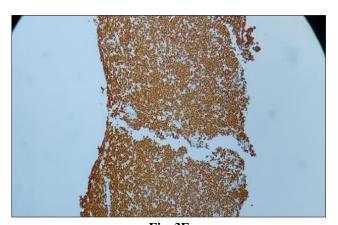


Fig. 3F

Figure 3: Immunohistochemical study showing positive labelling of tumors cells with cd20 (Fig 3A), cd10 (Fig 3B), bcl6 (Fig 3C), negative labelling of tumors cells with cd3 (Fig 3D) and bcl2 (Fig 3E), and present a Ki67 index of 100% (Fig 3F)

CT scan of the abdomen showed diffuse, irregular, endoluminal thickening of the antro-pyloric gastric wall (maximum thickness 27 mm, extending over 65 mm). Lymph nodes measuring 1 to 2 cm were present around the stomach, with no secondary localization (Figure 4)

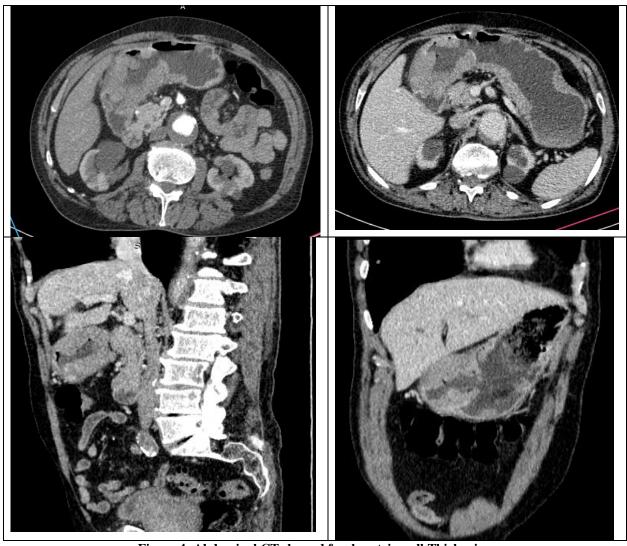


Figure 4: Abdominal CT showed focal gastric wall Thickening

Considering all the clinical, radiological, and pathological findings, a final diagnosis of Burkitt's lymphoma arising from the stomach was made. The case was discussed in the multidisciplinary clinic and with the family members and it was decided to go ahead with intensive systemic chemotherapy with cyclophosphamide and rituximab

DISCUSSION

Burkitt's lymphoma, recognized as one of the most aggressive forms of B-cell Non-Hodgkin Lymphoma (NHL), with replication approaching 100% has three clinical forms; endemic, sporadic and immunodeficiency associated, the endemic variant is prevalent in Africa, the sporadic variant is found in the U.S. and Western Europe, and immunocompromised variant occurs mainly in HIV patients. The sporadic variant comprises 30% of pediatric lymphomas and less than 1% of adult NHL [3].

The primary involvement of BL or a small noncleaved cell lymphoma in the gastrointestinal tract, although rare, has been reported in the literature. Despite gastric lymphomas being more common than intestinal lymphomas, the primary gastric involvement is extremely rare in BL. In adults, Burkitt's lymphoma of the stomach stands out as an exceptionally uncommon occurrence [2-4].

The precise mechanism triggering Burkitt's lymphoma formation remains elusive. The Epstein-Barr virus is implicated, with its presence detected in 25-40% of immunodeficiency variant cases of Burkitt's lymphoma, Normal gene expression and translation process of cellular microRNA has been shown to be interfered with by Epstein-Barr virus interaction with the cellular microRNA. Burkitt's lymphoma affects patients with CD4 T cell counts greater than 200/mm³, which may suggest that immunity does not have a role in the matter [5].

In the imaging assessment of Burkitt's lymphoma (BL), CT scanning and three-dimensional reconstruction are more useful for illustrating gastric

thickening, secondary localizations and accurately displaying bone destruction, This is the case with our patient, where we had to use a CT scan. MRI is also superior to CT due to its lack of ionizing radiation. For superior staging and assessment of the treatment response, PET/CT is a better choice since it can evaluate the functional status of abnormally hyper metabolic tissues throughout the whole body [6].

Histologically, the tumor cells of BL are medium-sized with an abundant, basophilic cytoplasm and display the typical "starry sky" pattern. The tumor cells are positive for BCL-6, CD19, CD20, CD22, CD10 and CD79a but negative for CD3, CD5, CD23 and TdT [7], in our patient's case, immunohistochemical evaluation revealed that lymphoid cells were positive for CD20, CD10 and Bcl6, negative for CD23, CD5 and Bcl2. All neoplastic cells were positive for Ki-67.

In a case series involving 21 cases of gastric Burkitt's lymphoma (BL) [9], stage 1 and 2 were identified in five patients each, while stage 4 was observed in 11 patients. The body and the antrum of the stomach were the most commonly affected sites. All the cases were treated with intensive systemic chemotherapy and 71% (17/21) of patients achieved a complete response. As to patients in stages 1 and 2, 9/10 showed a complete response. These findings underscore the critical importance of early diagnosis and treatment in ensuring remission for gastric Burkitt's lymphoma.

Burkitt's lymphoma is a very aggressive malignancy and one of the fastest growing amongst human malignancies. It requires immediate and aggressive intervention. Fortunately, it exhibits responsiveness to aggressive chemotherapy, establishing chemotherapy as the gold standard treatment despite its exceptionally rapid growth. It is crucial to be vigilant about the potential occurrence of tumor lysis syndrome, a complication resulting from the rapid, massive, and acute destruction of tumor cells during initial chemotherapy. The more extended the disease the more the chances it will get complicated and thus harder to treat [2].

CONCLUSION

Gastric Burkitt's lymphoma (BL) is an infrequent form of non-Hodgkin's lymphoma in adults, characterized by a high proliferation rate, aggressive nature, and poor prognosis. The optimal therapy for this disease is still unknown and is mainly adapted from pediatric treatment regimens. Early diagnosis with

aggressive and early treatment can result in very favorable long-term survival rates, reaching approximately 70-80% for patients. However, if the disease progresses and affects older age groups, it can swiftly become lethal. It is a disease which deserve great attention and as we are at the edge of breaking through into its treatment, focusing on early detection efforts can significantly impact patient outcomes.

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