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Mannar Mannan
DrNB Surgical Oncology Resident, Apollo cancer hospital, Teynampet Chennai, India.

Ayappan s
Senior Consultant Surgical Oncologist, Apollo Cancer Hospital, Teynampet, Chennai, India.

Correspondence:
Mannar Mannan
DrNB Surgical Oncology Resident, Apollo cancer hospital, Teynampet Chennai, India.

Primary Burkitt's Lymphoma of The Stomach Presenting with Complete Dysphagia - Case Management

Mannar Mannan, Ayappan s

Abstract
The primary gastrointestinal Burkitt's lymphoma is a rare entity. which is endemic in Africa, sporadic cases are reported in the rest of the world. Only around 50 cases have been reported till now. Here we present a 37/male from Oman with progressive dysphagia and weight loss for 2 months duration. On evaluation, he was found to have a large mass in the gastroesophageal junction and proximal stomach and body with marrow involvement in the right trochanter. OGD confirmed the growth and the biopsy did reveal NHL-Burkitt lymphoma. He received CODOX-M/IVAC + Rituximab. And his symptoms resolved in 2 days. Gastric Burkitt's lymphoma is an aggressive tumor with high proliferation. Chemotherapy, rituximab and prophylactic CNS treatment should be included as part of the treatment regimens. Early diagnosis with aggressive and early treatment can give very good long-term survival rates for the patients.

Keywords: Burkitt's lymphoma, gastric lymphoma.NHL.

1. Introduction

Burkitt's lymphoma, a disease frequently found in children of Tropical Africa, was first described by an Irish surgeon "Denis Burkitt" in Kampala, Central Africa in the mid-1900s. It was initially believed to be a sarcoma, frequent in children of Tropical Africa. In 1961, a British pathologist and academic by the name of Michael Anthony Epstein discovered a particular virus in tissue samples he took from this lymphoma. He named this virus as Epstein-Barr virus (EBV). This was the first time a viral pathogen was found to be involved in a human tumor. Burkitt's lymphoma remains to be the most frequent childhood malignancy in Africa to date. [1,2]

According to the classification given by the World Health Organization, there are three types of Burkitt's lymphoma based on clinical grounds; Endemic, sporadic and immunodeficiency associated. Nearly all cases are caused by EBV. [3] Endemic Burkitt's affects children between 4-7 years of age, affecting males more with a 2:1 male-to-female ratio. Sites affected include; the bones of the jaw and face, the gastrointestinal tract, the kidneys, the ovaries, breasts, and other extranodal sites. In Africa, the endemic Burkitt's has the highest incidence. [4,5]

Sporadic Burkitt's lymphoma occurs globally. It accounts for 1-2% of all adult lymphomas and 40% of all childhood lymphomas in the U.S. and Western Europe. The sites most commonly affected are; the abdomen, the ileocecal region, the ovaries, the kidneys, the omentum, Waldeyer's ring and the central nervous system. The central nervous system (CNS) involvement at presentation has been reported in 13-17% of the sporadic cases Immunodeficiency virus-associated BL occurs mainly in patients infected with HIV. BL accounts for 30-40% of non-Hodgkin's lymphoma in HIV-positive patients. [6]

2. Case report

Our patient is a 37-year-old male patient who is a businessman, resident of OMAN. he presented to us with complaints of weight loss of 20kg in 2 months, progressive dysphagia and epigastric pain for 2 months. the patient was able to tolerate oral clear liquids. On arrival,

he was not able to tolerate thick liquids or solid foods. He was evaluated in his country with CECT abdomen and OGD scopy. The biopsy report revealed necrotic tissue with few lymphocytes with CT abdomen showing growth involving the proximal stomach and body. He presented to us with dysphagia and the patient was admitted and resuscitated. The patient was dehydrated. Iv fluids and parenteral nutrition were started. Routine blood investigations are normal. Serum CEA and CA19-9 are within normal limits. LDH was found to be 450 IU/ml.

We did PET CT-WB which revealed a hyper-metabolic mass-like thickening with a predominantly exophytic component in the proximal stomach and GEJ involving the body and lesser curvature. few FDG avid and non-avid prominent splenic hilar nodes. focal hyper-metabolism in the lesser trochanter of the right femur-marrow lesion. f/s/o of the possibility of lymphoma

We repeated the OGD scopy. entry into the stomach was difficult, the lumen narrowed because of extrinsic compression and was angulated and entered with difficulty. Large ulceroproliferative growth is seen starting from the fundus and extending up to the midbody of the stomach along the lesser curvature of the stomach. The distal body was normal. antrum, pylorus and duodenum are normal. Shown in (Fig 1).

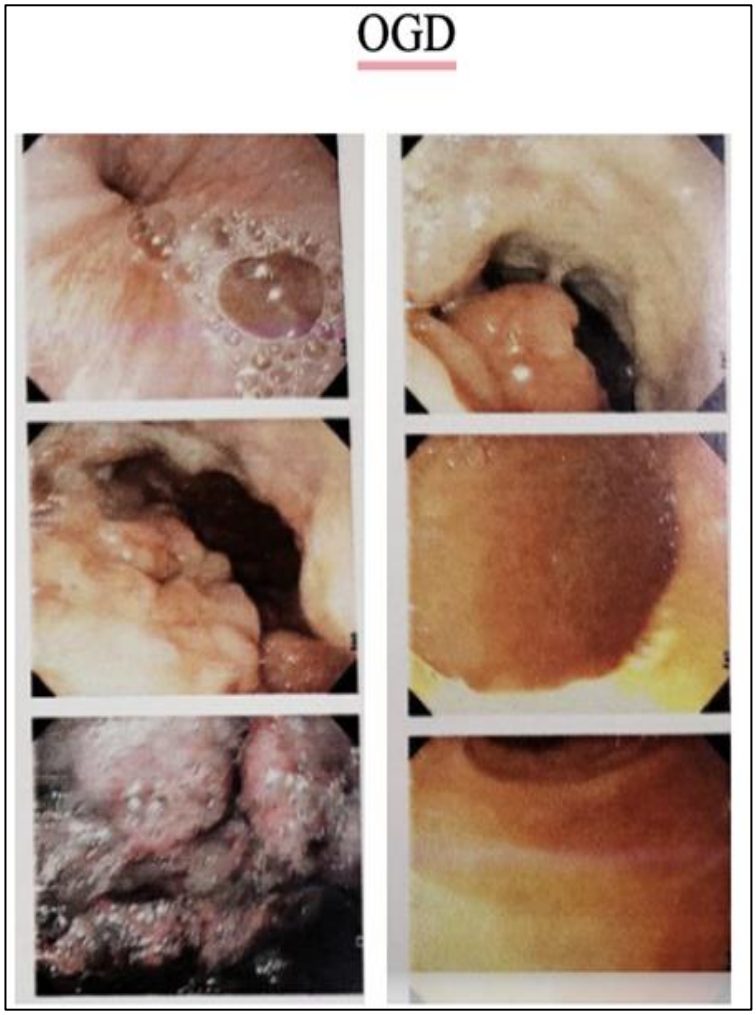

Fig.1: OGD.

Multiple biopsies were taken and revealed. NHL. IHC was suggested. In due course. the patient went into complete dysphasia. The option of central line insertion, iv nutrition and nasojejunal tube insertion was given to the patient. Despite detailed counseling, the patient refused any of the advised ways of nutrition. finally, it was decided to perform a feeding jejunostomy as he had significant weight loss and will need 4-6 cycles of multi-agent chemotherapy and the patient refused NJ insertion or central line placement

The IHC report did reveal Burkitt lymphoma. high grade with KI67 index of 98 %. CD20 was positive. CD 10 positive BCL6- positive, BCL2- negative. C-MYC was positive. EBER-ISH-negative. Shown in (Fig 2). The patient was discussed on the tumor board. It was decided any form of surgery will delay the chemotherapy. Since the patient is on peripheral parenteral nutrition (PPN) for the past 15 days. It was decided to start chemotherapy with PPN.

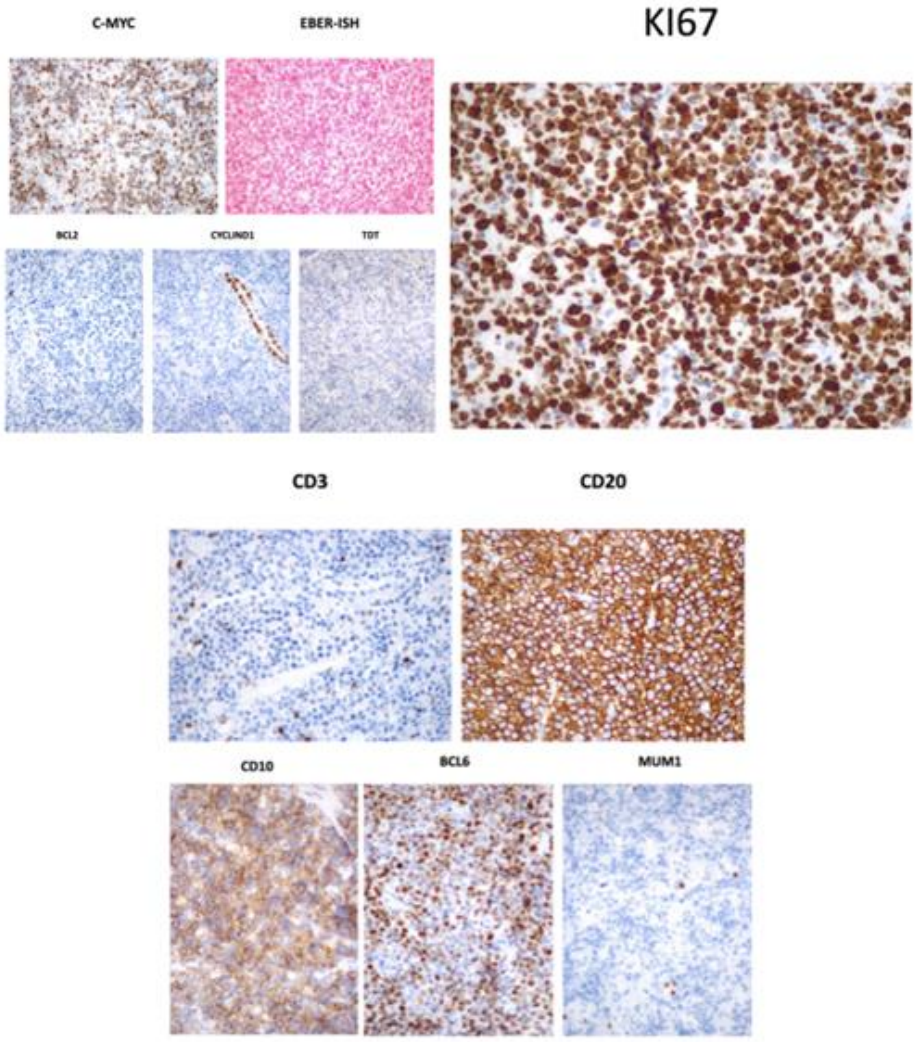

Fig. 2: IHC Markers.

The patient was started on CODOX-M/IVAC + RITUXIMAB after good hydration. Every precaution is taken as tumor lysis syndrome is expected. Two days after the initiation of chemotherapy. His dysphagia started resolving. The patient initially tolerated liquids and was finally able to take solids. He was planned for 4 cycles of the regimen with 12 sittings of intrathecal methotrexate. And with a possible cure rate of 70-80 per cent remission.

3. Discussion

Burkitt's lymphoma is one of the most aggressive forms of B-cell NHL, with replication approaching 100% has three clinical forms; endemic, sporadic and immunodeficiency associated. The endemic variant is common in Africa, the sporadic variant is present in the U.S. and Western Europe, and the immunocompromised variant occurs mainly in HIV patients. The sporadic variant comprises 30% of pediatric lymphomas and less than 1% of adult NHL. [7]

The most frequently affected site outside of lymph node involvement is the gastrointestinal tract (30-50%). Primary gastrointestinal lymphoma is rare. Secondary involvement of the gastrointestinal tract is common in lymphoma. Primary gastrointestinal lymphoma presents with symptoms localized to the GI tract or predominating mainly in the GI tract.

The primary involvement of BL or a small non-cleaved cell lymphoma in the GI tract, although rare, has been reported in the literature. Despite gastric lymphomas being more common than intestinal lymphomas, primary gastric involvement is extremely rare in BL. For non-endemic Burkitt's lymphoma, the gastrointestinal tract is the most common site, followed by the retroperitoneal, kidney, ovary, and testes respectively. Burkitt's lymphoma of the stomach is an exceptionally rare disease in adults. [8]

The exact mechanism leading to the formation of Burkitt's lymphoma is not yet known. The Epstein-Barr virus has been implicated to have involvement as it can be found in 25-40% of immunodeficiency variant cases of Burkitt's lymphoma. Normal gene expression and translation process of cellular microRNA is 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, however, in our case study, it was not the case. [9]

Burkitt's lymphoma is a very aggressive malignancy and one of the fastest-growing human malignancies. It requires immediate and aggressive intervention. Fortunately, it does respond to aggressive chemotherapy regardless of it being a very rapidly growing malignancy, chemotherapy being the gold standard treatment for it. Tumor lysis syndrome which is a complication of rapid, massive and acute destruction of the tumor cells can occur during initial chemotherapy and one should remain wary of that fact. The more extended the

disease, the more the chances it will get complicated and thus harder to treat. [10]

4. Conclusion

Gastric Burkitt's lymphoma is an aggressive tumor with high proliferation; we are yet to find the optimal treatment for this disease to help its poor prognosis. We have been able to hold it at bay by aggressive chemotherapy, mainly derived from pediatric treatment regimens, unspecific to Burkitt's. Chemotherapy, rituximab and prophylactic CNS treatment should be included as part of the treatment regimens. Early diagnosis with aggressive early treatment can give very good long-term survival rates for the patients, reaching around 70-80%.

References

1. Burkitt D. A sarcoma involving the jaws in African children. Br J Surg. 1958;46(197):218–223. doi:10.1002/bjs.18004619704.
2. Burkitt DP, Charles S. Mott Award. The discovery of Burkitt's lymphoma. Cancer. 1983;51(10):1777–1786. doi:10.1002/1097-0142(19830515)51:10601777::AID-CNCR2820511003>3.0.CO;2-E.
3. Young LS, Rickinson AB. Epstein-Barr virus: 40 years on. Nat Rev Cancer. 2004 Oct;4(10):757-68. doi: 10.1038/nrc1452. PMID: 15510157
4. Ferry JA. Burkitt's lymphoma:clinicopathologic features and differential diagnosis. Oncologist. 2006;11(4):375–383. doi:10.1634/theoncologist.11-4-375.
5. Koch P, del Valle F, Berdel WE, Willich NA, Reers B, Hiddemann W, et al. Primary gastrointestinal non-Hodgkin's lymphoma:I. Anatomic and histologic distribution, clinical features, and survival data of 371 patients registered in the German Multicenter Study GIT NHL 01/92. J Clin Oncol. 2001;19(18):3861–3873. doi:10.1200/JCO.2001.19.18.3861.
6. Law MF, Chan HN, Pang CY, et al. Durable survival after chemotherapy in a HIV patient with Burkitt's lymphoma presenting with massive upper gastrointestinal bleeding. Int J STD AIDS 2016;27:690–6.
7. Magrath IT, Shiramizu B. Biology and treatment of small non-cleaved cell lymphoma. Oncology (Williston Park, NY) 1989;3(11):41–53
8. Howell JM, Auer-Grzesiak I, Zhang J, et al. Increasing incidence rates, distribution and histological characteristics of primary gastrointestinal non-Hodgkin lymphoma in a North American population. Can J Gastroenterol 2012;26:452.
9. Aquino G, Marra L, Cantile M, et al. MYC chromosomal aberration in differential diagnosis between Burkitt and other aggressive lymphomas. Infect Agent Cancer 2004;8:37.
10. Gastwirt JP, Roschewski M. Management of adults with Burkitt lymphoma. Clin Adv Hematol Oncol. 2018;16:812-822