Classic Burkitt's Lymphoma in the Adult
A Double Dose of Rarity

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Introduction

- Burkitt's lymphoma is an aggressive B-cell type neoplasm that has three main variants: endemic (Sub-Sahara Africa), non-endemic (sporadic), and immunodeficiency related.
- The sporadic variant, is most commonly seen in children and comprises less than one percent of adult Non-Hodgkin's lymphomas in Western Europe and the United States.
- Adults with Burkitt's lymphoma are usually diagnosed with an atypical Burkitt's lymphoma type, rather than the classic form.

Case Description

- A 24 year old Caucasian woman with no significant past medical history transferred from outside hospital with acute abdominal pain. The patient reported a 15 pound weight loss in 2 months and excessive fatigue. On physical exam, she was pale, tachycardic, tender to palpation in all abdominal quadrants, and had +1 lower extremity edema bilaterally.
- CT scan showed free air in the abdomen, and exploratory laparotomy revealed small bowel perforation and diffuse carcinomatosis.
- She had lymphopenia, anemia, and elevated LDH and uric acid.
- Histopathological findings were consistent with Burkitt's Lymphoma (Fig 1) and 95% of the cells stained positive for Ki-67. Fluorescence in situ hybridization (FISH) revealed c-myc positivity with t(8q,24) in 73% of analyzed cells. Flow cytometry was weakly positive for CD20, Pax5, CD10, BCL-6, CD43, MUM1.
- Bone marrow biopsy was negative for lymphomatous infiltration. PET CT showed increased metabolic uptake in multiple gastrointestinal areas (Fig 2). Lumbar puncture showed no malignant cells. He was started on Rituximab with Hyper-CVAD in addition to intrathecal methotrexate and Cytarabine with a curative intent.

Objective

To demonstrate an uncommon presentation of lymphoma in the adult population and highlight the rare diagnosis of sporadic Burkitt's Lymphoma

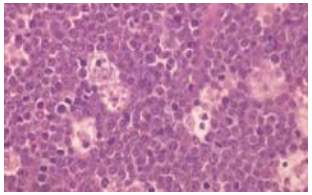

Figure1. Scattering of Macrophages containing debris derived from very rapid cell turnover contributing to the "starry sky appearance"

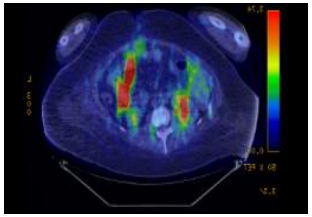

Figure 2. FDG PET Scan revealed large transverse colonic mass with increased metabolic activity.

Description

- This case highlights a rare presentation of a rare malignancy. Non-endemic Burkitt's lymphoma is uncommon, comprising less than one percent of NHL in the adult population. The presentation of this rare neoplasm in adults is usually atypical but our patient presented with all the classic features of Burkitt's lymphoma that are extremely uncommon in the adult population including abdominal pain, classic histology, and classic cytogenics, making this a rare presentation of an extremely rare hematologic malignancy.

Table 1. Characteristics of Burkitt's Lymphoma

- ❑ Population: Children > adults, male > female
- ❑ Clinical Features: Extranodal > nodal, bulky, rapidly growing masses
- ❑ Morphology: Uniform or slightly pleomorphic medium sized cells, starry-sky pattern
- ❑ Immunophenotype: CD20+, CD10+, Bcl-6+,, Bcl-2−, CD5−, TdT−, monotypic sIg+, Ki67 ~100%
- ❑ Genotype: t(8;14), t(2;8), or t(8;22) (*myc and IgH or IgL); no bcl-2 or bcl-6* translocation

References

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