Critical Care Issues in Management of High-Grade Lymphoma

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KEYWORDS

• Burkitt lymphoma • Renal failure • HIV • AIDS • Tumor lysis syndrome
• Inferior vena cava obstruction

KEY POINTS

- Lymphoma is a heterogeneous malignancy of the lymphatic system with presentations ranging from insidious to fulminant.
- Although very aggressive, BL can be highly treatable.
- Patients with BL often present with conditions that warrant critical care.
- TLS is a phenomenon seen in rapidly proliferating malignancies, usually in response to rapid cell death in response to cytotoxic therapies.
- Follicular lymphoma is unique because it has the potential to transform into the more aggressive DLBCL, with an estimated risk at 3% per year after initial diagnosis.
- Transformed DLBCL have poorer outcomes compared with de novo (newly diagnosed) DLBCL and therefore accurate diagnosis with prompt treatment is critical.

INTRODUCTION

Lymphoma is a heterogeneous malignancy of the lymphatic system with presentations ranging from insidious to fulminant. Based on the 2008 World Health Organization classification, there are close to 70 subtypes of lymphoma, generally divided into Hodgkin and non-Hodgkin lymphoma (NHL), both of which are further subdivided based on clinical and pathologic behavior.[1] Lymphoma is not rare; NHL is the fifth most common malignancy in the United Kingdom. In the United States, NHL

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accounted for more than 65,000 new cases in 2007 and more than 20,000 deaths in 2008.[2] This article compares the presentation of two patients who were seen in the emergency department (ED) on the same day at our institution (a National Comprehensive Cancer Network site) with life-threatening complications arising from high-grade lymphomas of very different origin, highlighting the varied presentations of high-grade lymphoma. Additionally, the cause, evaluation, and management of critical care issues in aggressive lymphoma are described.

CASE PRESENTATION #1
Burkitt Lymphoma

The first patient is Mr A, a 42-year-old Hispanic man who presented with a massive right axillary mass and abdominal pain. His medical history is significant for hypertension, non–insulin dependent diabetes, and morbid obesity. He denies known history of renal failure. For 10 days before his presentation at our ED, the patient had noticed a rapidly enlarging, painless mass in his right axilla. Over the last 4 days, the patient has complained of anorexia, early satiety, and progressive right upper quadrant pain for which he has been taking 12 to 16 ibuprofen tablets daily without relief.

Eventually, the pain led the patient to present to a community ED, where he underwent computed tomography (CT) of the chest, abdomen, and pelvis and was informed that he had widespread metastatic disease of unknown type. Mr A was advised to seek treatment from an oncologist and self-referred to our institution. On further questioning in our ED, the patient endorses increasing abdominal girth, malaise, and multiple episodes of drenching night sweats over the last 2 weeks. He denies other constitutional symptoms including weight loss, cough, dyspnea, nausea, vomiting, or diarrhea. The patient denied a history of sick contacts, foreign travel, history of intravenous drug abuse, tobacco, or alcohol abuse beyond one to two drinks weekly. The patient states he is in a long-term monogamous heterosexual relationship, has no tattoos, and has never had sex with another male.

The combination of localized painless lymphadenopathy, drenching night sweats, and abdominal symptoms raises concern for development of lymphoma. Coupled with the sudden onset and rapid progression of symptoms, the patient's presentation suggests a very aggressive disease. Clinical presentation led to high suspicion of aggressive lymphoma, such as Burkitt lymphoma (BL). Mr A urgently underwent a fine-needle aspiration of the axillary mass, which revealed BL, BCL-6$^+$, CD19$^+$, CD20$^+$, CD22$^+$, FMC7$^+$, and kappa light chain–positive, BCL-2, and Epstein-Barr virus negative. Ki-67, a measure of cell proliferation, is 100%.

BL is a rare, highly aggressive mature B-cell lymphoma accounting for less than 1% of all NHL.[3] BL is considered the "fastest growing human tumor," and is invariably fatal without treatment.[4] Survival is reckoned in weeks to months without therapy.[4] Symptoms are related to rapid turnover of mature B lymphocytes, extent of extranodal involvement, or invasion into organs. BL has three variants: (1) endemic, (2) sporadic, and (3) immunodeficiency related.[5] Endemic BL, a childhood disease, is usually found in equatorial Africa and presents as painless neck, jaw, or facial masses.[5] Abdominal, gonadal, and skeletal presentations are also seen. Sporadic forms of BL generally present with bone marrow involvement and abdominal disease, with diagnosis often precipitated by bowel obstruction or intussusception caused by mass effect. Other presentations include intracardiac disease, pancreatitis, ovarian or breast involvement, and occasional skin lesions. Generalized lymphadenopathy is less common.[5] Finally, immunodeficiency-related BL usually presents with localized lymphadenopathy and usually spares the bone marrow.[5] Whether innate or acquired, immune