An unusual presentation of non-hodgkin’s lymphoma

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Abstract

Background: NHL accounts- 60% of all lymphomas.

Case Characteristics: 8 year, female hospitalized with c/o fever* 1 month, lump in abdomen associated with pain* 15 days, was diagnosed with high grade NHL-burkitt type confirmed by Sr. LDH, CD79α, CD20 and Ki-67.

Outcome: Prognosis is bad.

Message: It is essential to completely evaluate to clinche the diagnosis.

Keywords: NHL, BL, LDH

Introduction

Non hodgkins lymphoma accounts for approximately 60% of all lymphomas in children and adolescents. It represents 8-10% of all malignancies in children between 5-19 yr of age. While most of the children and adolescents with NHL present with de novo disease,a small number of patients develop NHL secondary to etiologies, including inherited or acquired immune deficiencies, viral etiologies (e.g. HIV,EBV) or as a part of genetic syndromes (e.g. ataxia telangiectasia, Bloom syndrome).Most children who develop NHL, however, have no obvious genetic or environmental etiology [1].

Burkitt’s lymphoma (BL) is a highly malignant, aggressive and rapidly growing B cell non-Hodgkin’s lymphoma, which has a low long-term survival rate [2, 3]. According to the WHO Classification, there are three clinical variants of Burkitt’s lymphoma: endemic, sporadic and immunodeficiency [4, 5]. The endemic variant is related to EBV or malaria infection and occurs in children,4-7 years old, involving the jawbone and other facial bones, kidneys, ovaries, gastrointestinal tract, breasts and other extranodal sites [4, 5]. Sporadic Burkitt’s lymphoma is an extranodal disease associated with no specific geographic or climate area and accounts for 40% of all lymphomas in children and only 1-2% in adults. Although almost localisation is intra-abdominal, affecting the intestinal tract, ovaries, kidneys, omentum and Waldeyer’s ring,but very rarely lymph nodes of any localisation [5]. Immunodeficiency-associated Burkitt’s lymphoma occurs mainly in patients with HIV, but also occurs in acquired and congenital immunodeficient patients.

Methods Case Report

A 8-year old female came to pediatric OPD with complaints of fever x 1 month, pain in abdomen x 15 days and swelling in the right hypochondrium x 15 days. The swelling was initially small but progressive in nature and attained current size 5 x 4 cm. No h/o bowel/bladder complaints, loosestools, vomiting. No h/o respiratory distress/chest pain.

No h/o any contact of tuberculosis, any radiological exposure. On examination Temp-98.6 degree F, HR-110/min, RR-24/min, BP- 118/86 mmhg, abdominal girth- 48 cm. On P/A examination abdomen was distended, umbilicus- everted and tenderness was present in right hypochondrium and epigastric region. On palpation-hepatomegaly 4-5 cm, firm in consistency.

Precussion: fluid thill was present.
Rest other systems-wnlCBC was done Hb- 6.8gm%, TLC – 10200, P66%, L18%, E10%, M06%, Platelet count-2.2 lakh/cumm, LFT’s-Sr.bilirubin-1.0mg%, direct-0.7mg%, serum proteins (total) -6.5gm%, albumin-3.0gm%, globulin-2.5gm%. Serum uric acid- 5.6mg %.

USG abdo/pelvis shows hepatomegaly with multiple liver abscess, pelvic mass lesion mostly LN in origin causing mild obstructive changes in renal system.

Initially provisional diagnosis of ovarian tumour was made for which CA-125 levels were done-222.9U/ml but then Liver biopsy was done which showed hepatic involvement by High grade Non Hodgkin Lymphoma of B cell type-

Suggestive of Burkitt type with clinical correlation of serum LDH levels-713IU/L confirming the diagnosis.

Results Histopathology

On the histopathology report, the enlarged lymphoid node completely lost its anatomical structure and was infiltrated with the malignant cells, which gave it a “star-ry sky” appearance. Immunocytochemical stained cells were positive for CD79a, CD20 and Ki-67 (100%), and slightly positive for CD43 and CD10, which confirmed the diagnosis of high risk non-Hodgkin’s lymphoma, B cell phenotype -Burkitt’s lymphoma type. Bone marrow examination confirmed only non-specific reactive changes.

Discussion and Conclusions

In 1958, Dennis Burkitt, a surgeon, first described a disorder presenting with jawbone tumours in African children. He noted children with huge facial tumours unilateral or bilaterally involving mostly jawbones and other facial bones, and sometimes accompanied by enormous abdominal masses. A few years later, the neoplasm was identified as a form of malignant lymphoma, which initially emerged as a clinical syndrome became a pathological entity called Burkitt’s lymphoma.

Histologically, Burkitt’s lymphoma is composed of monomorphic, medium-sized neoplastic cells of lymphocyte origin with round nuclei, multiple nucleoli and relatively abundant basophilic cytoplasm. These cells typically possess an extremely high proliferation rate and high rate of programmed cell death (apoptosis). Morphological tumour characteristics are numerous, including admixed body macrophages phagocytosing abundant apoptotic debris, creating a typical “starry-sky” pattern. The exact origin of the malignant cells is still unknown.

Generally, the disease presents with mild, non-specific gastrointestinal symptoms and “B” symptoms, such as weight loss, unexplained fever and night sweats. These symptoms are identical to those of numerous opportunistic infections and may delay the diagnosis of lymphoma.

In the extreme, Burkitt’s lymphoma can initially manifest with aggressive gastrointestinal symptoms and signs, which demonstrate an advanced disease requiring surgical treatment.

Although the prognostic features have not yet been determined, some features that have been associated with adverse outcome in adults and children include older age, advanced disease, poor performance status, bulky disease, high level of LDH and involvement of central nervous system or bone marrow. There are different manifestations of Burkitt’s lymphoma, which overlap with other intra-abdominal illnesses in symptoms and signs. Although the presented case may not be a typical example of Burkitt’s lymphoma, it undoubtedly suggests an aggressive lymphoma that requires an intensive and broad diagnostic approach and appropriate therapy, especially in many intrinsic entities such as acute abdomen. The prognosis is excellent for most forms of childhood and adolescent NHL. Patients with localised disease have a 90-100% chance of survival, and patients with advanced disease have a 60-95% chance of survival. The variation in survival depends on pathological subtype, tumor burden at diagnosis as reflected in serum LDH level, presence or absence of CNS disease and specific sites of metastatic spread.

![Fig 1: A 8-year old female with swelling in the right hypochondriac region diagnosed as non Hodgkin’s lymphoma.](image1)

![Fig 2: Histopathology Report. Burkitt’s lymphoma cells found on the Light microscopy, found in our patient (H/E, x40)](image2)

Reference