Burkitt's Lymphoma As Primary Adrenal Lymphoma Diagnosed on Fine Needle Aspiration Cytology: A Rare Case Report

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Abstract

Introduction: Primary adrenal lymphoma presenting as Burkitt’s lymphoma is an extremely rare entity. Although biopsy or excision is often needed for definitive diagnosis computed tomography guided fine needle aspiration has great diagnostic value in detection of primary adrenal lymphoma.

Presentation of case: A case of a 20 yr old male with a right adrenal mass diagnosed as Burkitt's lymphoma on Fine Needle Aspiration Cytology (confirmed by bone marrow and cytogenetics) has been reported.

Conclusion: Fine Needle Aspiration Cytology as an initial investigation tool can be very effective for detection of unusual presentation like an adrenal mass in a case of Burkitt's lymphoma.

Keywords: Burkitt’s Lymphoma; Primary Adrenal Lymphoma; Fine Needle Aspiration

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Introduction

Burkitt’s lymphoma is a distinct pathologic entity characterized by a diffuse undifferentiated malignant lymphoma of B cell origin. It is very rare disease but probably the fastest growing tumor in humans, with exuberant proliferation. Cure rate is 70-80% of both children and adults when effective therapy is initiated precisely. Salvage therapy has been generally ineffective in patients failing the initial treatment, emphasizing the importance of the initial treatment approach [1]. Although secondary involvement of the adrenal glands by Non-Hodgkin’s lymphoma is not uncommon, primary adrenal lymphoma is extremely rare and accounts for <1% of all Non-Hodgkin's lymphoma cases [2]. Patients usually do not have disease elsewhere and this makes the diagnosis more difficult. Symptoms of the disease are variable and patients are most commonly older males. Presenting clinical features include pyrexia, lumbar pain, and/or symptoms of adrenal insufficiency. It may also be diagnosed incidentally on abdominal imaging [3]. Zhang et al [4] has presented a case of 80 year old man with primary adrenal lymphoma with burkitt like morphology and adrenal insufficiency diagnosed by CT guided Fine needle aspiration cytology. Here we report a case of a 20 yr old male with primary adrenal lymphoma diagnosed as Burkitt’s lymphoma on CT guided Fine needle aspiration.

Case Presentation

A 20 yr old male nonhypertensive, nondiabetic patient presented with history of intermittent fever for 3 months duration with recent onset paraparesis. General physical examination showed presence of icterus, pedal edema, mild hepatomegaly. Lymph nodes were not palpable. Initial laboratory investigations showed Haemoglobin-11.5g%; Total White blood cell count was raised (12000/cmm). Erythrocyte Sedimentation Rate (1st hour)-62mm was raised. Liver function test was deranged- Serum protein (Total)- 5.9 gm/dl( Albumin-3.5gm/dl), Bilirubin-12.9gm/dl (conjugated-8.6gm/dl), Alkaline phophatase-145.2 U/L, Alanine aminotransferase-140.7U/L, Aspartate aminotransferase -170.9 U/L. Patient had already been initiated on dialysis as Urea and Creatinine were high 170gm/dl and 6gm/dl respectively. Investigations for HCV, HIV 1 and 2, HBV were negative.

On computed tomography scan a well defined heterogeneous right adrenal space occupying lesion was seen(Fig 1).To exclude Pheochromocytoma, Morning serum cortisol(13.3mcg/100ml), serum ACTH(60.5pg/ml), Urinary Vanillylmandelic acid (8.5mg/24hr) were done which were within normal limits. Then Fine needle aspiration was planned in order to exclude any adrenal pathology.

Computed tomography guided fine needle aspiration cytology from the adrenal mass showed cellular smear with relatively uniform intermediate sized cells having rounded nuclei, granular chromatin with prominent nucleoli and thin rim of dense blue cytoplasm with small vacuoles. Plenty of smudge cells and lymphoglandular bodies were seen in hemorrhagic background. Cytomorphological features were suggestive of Non-hodgkin's lymphoma, more in favour of Burkitt's lymphoma (Fig 2). Subsequent bone marrow aspiration showed 76% blast cells predominantly of L3 morphological type (Fig 3).

For further confirmation, Fluorescence In Situ Hybridization (FISH) analysis of 200 interphase cells was done which was positive for rearrangement of the cMYC on chromosome 8, band q24 in 73% cells. Chromosomal analysis in bone marrow following unstimulated cell culture and GTG banding revealed complex rearrangement with 46,XY,der(1)-13,-21,+mar x2 chromosomal pattern in 16% of cells examined (Fig 4).These deletions carry poor prognosis. The chromosomal study was corroborative with the Fine needle aspiration cytology finding of Burkitt’s Lymphoma. The patient was severely ill and he passed away before initiation of chemotherapy.
**Figure 1** Contrast Enhanced Computed Tomography Showing Right Adrenal Space Occupying Lesion

**Figure 2** Fine Needle Aspiration Showing uniform intermediate sized cells having rounded nuclei, granular chromatin with prominent nucleoli and thin rim of dense blue cytoplasm with small vacuoles. Cytological Features suggestive of Burkitt’s Lymphoma (Leishman Stain, 100X)
Figure 3 Bone Marrow Aspiration Showing Blast Cells of L3 Morphological Subtype

Figure 4 Chromosomal Analysis of Bone Marrow

Discussion

Three different clinical variants of Burkitt’s Lymphoma have been described: endemic, sporadic, and immunodeficiency Burkitt’s Lymphoma. Adult patients with sporadic or immunodeficiency associated
Burkitt’s Lymphoma typically present with extranodal disease, with the abdomen being the most frequent site of involvement. Intra-abdominal presentations usually affect the bowel or intra-abdominal lymph nodes, although kidney, pancreas, liver, spleen, breast or ovarian involvement can occur. At diagnosis, patients usually have bulky disease and elevated serum lactate dehydrogenase and uric acid levels. Bone marrow and central nervous system (CNS) involvement is reported in 30% to 38% and 13% to 17% of adults, respectively [5-7]. Most of the cases have MYC translocation at band 8q24 of the IG heavy chain region, 14q32 or, less commonly, at the lambda. 22q11 or kappa, 2p12 light chain loci [8]. In this case Fluorescent In situ Hybridization analysis was done which was positive for cMYC chromosomal rearrangement. Several laboratory findings are consistently abnormal in Burkitt’s Lymphoma patients. Increased activity of alanine aminotransferase, serum alkaline phosphatase has been reported [9]. Anaemia and leukocytosis are common. In addition, the erythrocyte sedimentation rate and blood urea nitrogen may be elevated [10]. This patient showed increased levels of serum alkaline phosphatase, erythrocyte sedimentation rate, and leucocytosis.

Primary adrenal lymphoma is a rare disorder and more common in men from the sixth decade onwards [11]. The majority of cases involve both adrenal glands and bulky disease is more common [12]. The main clinical features are local symptoms such as lumbar pain or systemic symptoms such as fever and weight loss. Symptoms of adrenal insufficiency such as vomiting, fatigue, skin hyperpigmentation, and hypotension can also occur especially if there is extensive bilateral adrenal involvement [13,14]. Computed Tomography scan will generally show homogenous masses with predominantly low density and slight to moderate enhancement [12]. These features are however not pathognomonic.

The most common histology in Primary Adrenal Lymphoma is diffuse large B-cell lymphoma, usually activated B-cell phenotype. T-cell and angiocentric large cell lymphoma type B are the exception [15]. Primary adrenal lymphoma with Burkitt’s like morphology is rare but has been reported [4].

Although dispersed cell populations cytologically favour lymphoma, metastatic poorly differentiated carcinoma and adrenal cortical carcinoma can manifest similarly. Integrated histological, immunohistochemical, and flow cytometric immunophenotyping would provide an accurate and definitive diagnosis [4].

Histological diagnosis is a gold standard in evaluation of lymphoma. Prognosis is usually poor and the median survival is usually less than 1 year [12]. Poor prognostic factors include advanced age, large tumor size, bilateral involvement, high Lactate Dehydrogenase levels, involvement of other organs, and initial presentation with adrenal insufficiency [12]. The therapeutic modalities include combination chemotherapy and radiotherapy in order to prevent local recurrence.

**Conclusion**

The diagnosis of Burkitt lymphoma as primary adrenal lymphoma requires a combination of ancillary techniques as there is no single diagnostic gold standard but Fine needle aspiration cytology as an initial investigation tool can be very effective for detection of such rare presentation which will help in initiation of early therapeutic intervention.
Consent

Consent was taken from the patient and the patient’s father (after patient’s death) for publication of this case report.

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