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Case Report

An unusual presentation of Burkitt lymphoma as a neck mass in immunocompetent adult male: a case report

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ABSTRACT

Burkitt lymphoma (BL) is a highly aggressive B-cell non-Hodgkin lymphoma defined by MYC gene translocation. The World Health Organization (WHO) and International Consensus Classification (ICC) recognize BL and Burkitt leukemia as manifestations of the same disease and differentiate BL from other high-grade B-cell lymphomas. The three clinical types-endemic, sporadic, and immunodeficiency-related-show differing geographic distributions and presentations. We report and unusual presentation of BL presenting as a left sided neck mass in a immunocompetent adult male. This case highlights the aggressive nature of BL and the importance of early and accurate diagnosis of through imaging and biopsy.

Keywords: Burkitt lymphoma, Neck mass, Non-Hodgkin lymphoma, Case report, Immunocompetent

INTRODUCTION

BL is a very aggressive non-Hodgkin lymphoma of the B cell. It is defined by chromosome 8 MYC gene translocation and dysregulation. Three recognized clinical types of BL are immunodeficiency related, sporadic (nonendemic), and endemic (African). These types differ in their epidemiology, clinical appearance, and genetic characteristics, although sharing the same histology and exhibiting comparable clinical behavior.[1] BL and Burkitt leukemia are considered different manifestations of the same disease in the WHO 5th edition (WHO5) of the classification of hemato lymphoid tumor and the International Consensus Classification (ICC) of mature lymphoid neoplasms. Both classification systems also recognize three aggressive B cell lymphoma entities that resemble Burkitt-like lymphoma with 11q aberration (BL), high-grade B cell lymphoma with MYC and BCL2 rearrangements, and high-grade B cell lymphoma, not otherwise specified.[2,3] Since the resource-poor countries with the highest apparent prevalence-such as equatorial Africa-are unable to gather the epidemiologic data required for exact diagnosis and case ascertainment, the precise global incidence of BL remains unknown. Cases of BL are typically classified into three different clinical types for epidemiologic and diagnostic purposes: endemic (African), sporadic (nonendemic), and immunodeficiency related. The geographic distribution of endemic and sporadic clinical forms of BL varies.[4] The standard of care has yet to be defined and our preferred treatment is enrollment in a clinical trial. However, for patients who are not candidates for such trials or for those who choose not to participate, we suggest intensive, short-duration combination chemotherapy with central nervous system prophylaxis. Our preferred regimen is R-CODOX-M/IVAC (Rituximab, Cyclophosphamide, Doxorubicin, Vincristine, Methotrexate/ Rituximab, Ifosfamide, Etoposide, Cytarabine). CODOX-M/IVAC is highly toxic, primarily to the hematopoietic system, and most patients will have a prolonged hospital stay.

Therapy should be initiated promptly and dose reduction should be avoided, if possible. For older or less fit patients who may not tolerate more aggressive regimens, we suggest in fusional chemotherapy with dose-adjusted EPOCH plus rituximab. We recommend the addition of rituximab to combination chemotherapy. Although practice varies, we generally wait until the second cycle of chemotherapy to add rituximab in order to minimize tumor lysis.[1] Since, the recurrence rate of BL is very rare we could not find any data regarding any previous case reporting of the same. Thus, we hereby present the case reporting of recurrence of BL following treatment.

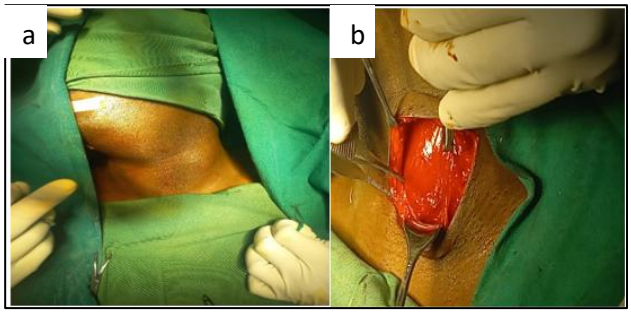

Figure 1: (a) Left sided neck swelling 3×4 cm noted behind the left sternocledomastoid (b) Exposure of neck swelling.

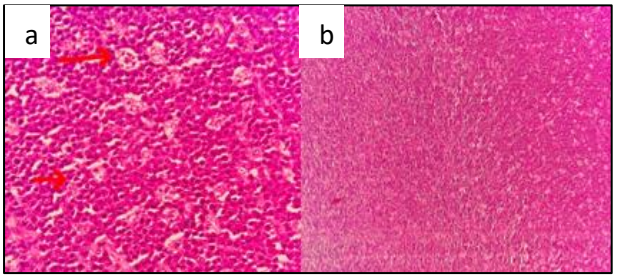

Figure 2: (a) H&E 10X: Lymph nodes showa starry sky pattern. (b) H&E 40X: Small to intermediate lymphocytes admixed with tingible body macrophages.

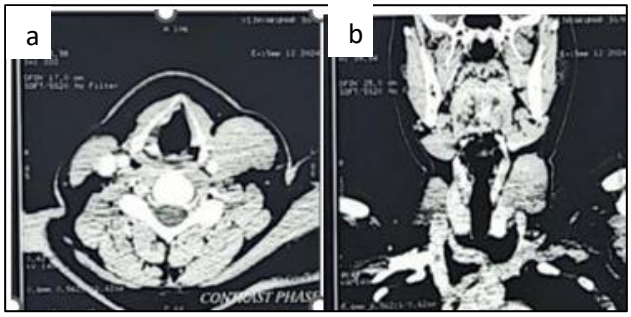

Figure 3: Contrast enhanced CT scan (a) axial cut showing well defined homogeneous minimally enhancing isodense lesion measuring 3.6×2.1×2.5 cm in the upper cervical region. (b) coronal cut showing swelling compressing left IJV.

CASE REPORT

A 30-year-old male patient reported to the ENT department with the complaints of left sided neck swelling. He noticed the swelling for 1 week which gradually progressed in size with no other associated ear, nose and throat symptoms. On local examination of neck, a diffuse swelling of size 3×4 cm (Figure 1a) was noted 3cm behind and above the clavicular head of sternocleidomastoid extending 1cm below the angle of mandible. Skin over the swelling is normal and pinchable. It is mobile, non-pulsatile, non-tender swelling with transillumination negative.

On radiological investigation USG Neck shows an ill-defined heterogeneous lesion of approximately 3.41.7 cm noted adjacent to the left lobe of thyroid with significant internal vascularity. On doppler arterial waveform was noted. CECT Neck shows a fairly well defined homogenous minimally enhancing isodense lesion measuring 3.6×2.12.5 cm noted in the upper cervical region. The isodense lesion is located between left lobe of thyroid gland superolaterally and left common carotid artery inferomedially causing mild displacement of left CCA, however left common carotid artery is well opacified. Left sternocleidomastoid laterally and fat planes is maintained. The lesion is seen compressing left internal jugular vein, however there is normal enhancement and no evidence of proximal and distal dilatation of vein noted. Multiple homogenous subcentrimetric bilateral cervical lymph nodes noted at level I, II, III, IV and V Figure 3 (a, b). FNAC suggestive of suspicious of lymphoproliferative disorder. Excision and biopsy done Figure 1(b). Histopathological examination is suggestive of non-Hodgkin lymphoma Figure 2 (a, b). Then patient started in R-COPADM regimen and no recurrence after 6 months of follow up.

DISCUSSION

BL is a highly aggressive non-Hodgkin lymphoma with a characteristic translocation involving the MYC gene located on chromosome 8.[5] The disease's classification into three types-endemic, sporadic, and immunodeficiency-associated-reflects its diverse clinical and epidemiologic presentations, though all share the same histopathologic findings.[6]

This case of left-sided neck swelling highlights the diagnostic importance of clinical examination, imaging, and histopathology. Ultrasound and CECT imaging showed a well-defined, vascularized lesion without evidence of thrombosis or calcification. Such findings align with previously reported cases of BL, where rapid cellular proliferation may cause vascular compression without complete occlusion.[7] FNAC provided initial suspicion of a lymphoproliferative disorder and Histopathological examination is suggestive of non Hodgkin lymphoma later confirmed as Burkitt lymphoma STAGE 4/ GROUP B on LMB 89 protocol. IHC supports

the diagnosis. Further, fluorescence in situ hybridization (FISH) for MYC gene rearrangement, as suggested by the World Health Organization (WHO) classification and improves the diagnostic specificity of the disease.[6] The standard treatment for BL includes combination chemotherapy with CNS prophylaxis. The R-CODOX-M/IVAC regimen are most commonly used in these scenarios combined with high-intensity chemotherapy and rituximab to improve survival outcomes.[8] However, the high toxicity of these regimens, particularly to hematopoietic tissues, often causes various symptoms like tumor lysis syndrome and myelosuppression which necessitates close monitoring for the same. In this case, recurrence of BL despite treatment highlights the potential for residual disease, even in cases initially deemed responsive.

Recurrence in BL is rare, with limited data available regarding its clinical course post-treatment. Current studies emphasize the need for long-term follow-up to detect potential relapses.[9] This case report adds to the limited pool of evidence for recurrent BL, highlighting the need for continued surveillance beyond initial remission. Additional research on predictive factors for relapse and salvage therapies is critical to improving long-term outcomes.

CONCLUSION

This case highlights BL, a rare and aggressive non-Hodgkin lymphoma, presenting as a neck mass in a young adult. The patient's clinical presentation, imaging findings, and histopathological diagnosis underscore the importance of maintaining a high index of suspicion for BL in cases of rapidly progressive, painless neck masses with atypical imaging characteristics. Early recognition and initiation of treatment are crucial for improving patient outcomes. The findings align with existing literature, which emphasizes the role of comprehensive imaging and biopsy in diagnosing lymphoproliferative disorders. The case further underscores the importance of long-term follow-up in detecting recurrences, even though recurrence rates in BL are rare.

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