

## Case Report

# Primary Hepatic Burkitt's Lymphoma in a Patient With Acquired Immunodeficiency Syndrome

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### ABSTRACT

Primary non-Hodgkin lymphoma of liver is a very rare malignancy. Here we report a case of primary hepatic Burkitt's lymphoma in a 34-year old man who was known case of acquired immunodeficiency syndrome (AIDS) and presented with weight loss, abdominal pain, and fever. Positive laboratory findings were mild anemia and elevated levels of lactate dehydrogenase (LDH) and alkaline phosphatase (ALP). Abdominal computed tomography (CT) showed multiple large low-density mass lesions in both lobes of liver with no evidence of regional or distant lymphadenopathy or metastasis. Liver needle biopsy was done and both histological and immunohistochemical findings were compatible with Burkitt's lymphoma.

**Keywords:** Burkitt's Lymphoma, Liver, AIDS

### Introduction

Non-Hodgkin-lymphoma (NHL) of the B-cell type is the second most common neoplasm in patients with human immunodeficiency virus (HIV) infection (1). The majority of NHL associated with acquired immunodeficiency syndrome (AIDS) involves extra-nodal sites, most frequently gastrointestinal tract, and central nervous system (CNS) (1).

Primary liver lymphoma (PLL) is an uncommon

neoplasm representing only about 0.4% of all extra nodal lymphomas (2).

Primary hepatic Burkitt's lymphoma (PH-BL) a highly aggressive subset of NHL is a very rare entity. There have been seven cases of PH-BL reported in adults, but only five cases had confirmed immunophenotypic diagnosis (3).

Here we report a case of (PH-BL) in a 34-year-old patient infected by HIV, presented with multiple space occupying hepatic masses.

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## Case report

A 34 year-old man, known case of AIDS, presented with abdominal pain, weight loss and night sweats for 3 months. There was also a 3-day history of fever with cough and sputum. He was a heavy smoker; with positive history of drug injection and alcohol consumption.

Physical examination revealed massive hepatomegaly and no palpable lymph node or spleen. On chest examination, fine crackles in lower zone of left hemithorax with generalized wheezing were found. Chest x-ray revealed a segmental air-space consolidation in the left lower lobe without pleural effusion or mediastinal adenopathy; compatible with infectious process.

Significant laboratory findings were Hemoglobin 12 gr/dL, Platelet 253000/mm<sup>3</sup>, WBC 3700/mm<sup>3</sup>, alanin aminotransferase (ALT) 23 Iu/L, aspartate aminotransferase (AST) 35 Iu/L, alkaline phosphatase (ALP) 1018 Iu/L, total bilirubin 6.8 mg/dL, direct bilirubin 0.3 mg/dL and lactate dehydrogenase (LDH) 2300 Iu/L. blood and urine cultures were both negative. Viral markers including HBsAg, Anti-Hbc, and Anti-HCV were all negative. He had never received antiretroviral therapy for HIV infection.

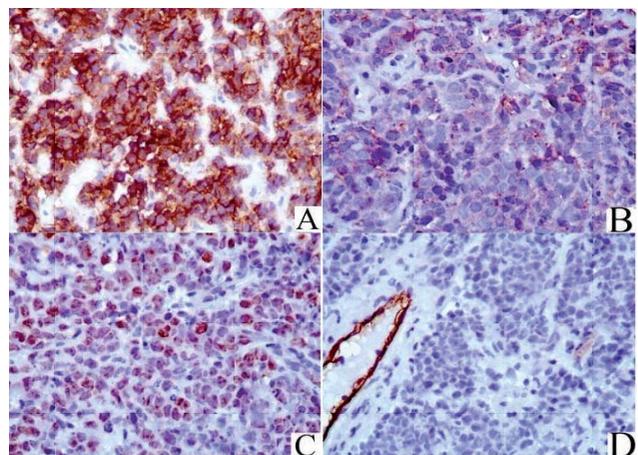
Abdominal computed tomography (CT) scan with intravenous and oral contrasts was done and showed multiple large low-density mass lesions in both lobes of liver with no enhancement (Fig. 1). These lesions encase liver vessels but no thrombosis was evident. No bile duct dilatation was noted.



**Fig. 1:** Abdominal CT shows multiple low-density mass lesions in both lobes of liver.

No cervical, abdominal and chest lymphadenopathy was revealed on the chest, abdominal, and pelvic CT. Ultra sonography-guided liver biopsy was done. Microscopic evaluation of H&E stained slides showed diffuse infiltration of monotonous small to medium sized cells with vesicular nuclei, dense chromatin pattern, prominent nucleoli, and moderate amount of cytoplasm. Mitotic figures were frequent. There were many scattered tangible macrophages creating starry-sky pattern.

Immunohistochemical stains were positive for LCA, CD20, and CD10 and negative for BCL2 and CD34 (Fig 2). Proliferation index based on the ki-67 fraction was more than 98%.



**Fig. 2:** Immunohistochemical stains are positive for CD20 (A) and CD10 (B). C) High proliferative rate in Ki-67 staining. D) immunohistochemical stain is negative for CD34; although the endothelial cells are positive. (×400)

These findings were compatible with pathologic diagnosis of Burkitt's lymphoma. The spinal fluid analysis and bone marrow biopsy results were negative. Polymerase chain reaction (PCR) for EBV DNA was inconclusive due to scarcity of specimen remained in paraffin block.

The patient was referred to oncologist for receiving chemotherapy but unfortunately he died in few days, without any treatment.

## Discussion

A definite diagnostic criterion for primary hepatic lymphoma has not been established yet. According

to Lei criteria in primary hepatic lymphoma symptom expression mainly originate from liver infiltration with no distant lymphadenopathy or leukemoid reaction in peripheral blood smear (4). Secondary involvement of the liver is relatively common including 50-80% of Hodgkin's or non-Hodgkin's lymphoma at autopsies (4) whereas primary involvement represents less than 1% of all extranodal lymphomas (5).

Hepatic NHL in patients with AIDS was first described by Reichert *et al.* in 1983 in an autopsy series (6).

PH-BL a highly aggressive subset of NHL is a very rare entity. According to study performed by Wissam *et al.* (3) there have been seven cases of PH-BL reported in adults, but five cases had confirmed immunophenotypic diagnosis. They also reported a case of confirmed PH-BL presenting with acute liver failure (ALF).

On histopathologic examination, Burkitt's lymphoma is defined as infiltration of neoplastic cells, which are markedly uniform in size and shape. The nuclei are approximately the same size as the nuclei of the admixed histiocytes and therefore are smaller than the centroblasts and contain two to four basophilic nucleoli. The nuclear contours are generally round without deep indentation. The cytoplasm is strongly basophilic with small round cytoplasmic vacuoles best observed in air-dried touch imprints (7).

The presence of tangible body macrophages, phagocytosing abundant apoptotic debris creating starry-sky appearance is characteristic finding, although it is not always present (8).

On immunohistochemical examination the neoplastic cells are positive for surface IgM and Ig light chain ( $\kappa > \lambda$ ), pan-B-cell antigens (CD19, CD20, CD22, CD79 $\alpha$ ) and the germinal center associated markers such as CD10 and BCL6 (7).

We diagnosed our case as Burkitt's lymphoma because CD10 and CD20 were positive, BCL2, CD3 and CD34 were negative and Ki-67 expression was over 98%.

A molecular defining feature of Burkitt's lymphoma is the presence of a translocation between the c-myc gene and the IgH gene [t(8, 14)], or between c-myc and gene for either the  $\kappa$

or  $\lambda$  light chain [t(2, 8) or t(8, 22)] (9). Among seven reported cases of PH-BL cytogenetic study was performed in only one case (2) which showed t(8, 14).

Of patients with HIV related Burkitt's lymphoma, 30-50% of the cases are infected by EBV (3). Reviewing seven reported cases of adult PH-BL showed that EBV DNA was positive in one case (3), negative in two cases (2, 12), and was not described in other 4 studies. As mentioned previously in our case, PCR for EBV DNA was inconclusive.

The clinical feature of primary hepatic lymphoma varies from no symptom to fulminant hepatic failure. Fever, weight loss, night sweating (known as B symptoms), right upper quadrant pain, hepatomegaly, fatigue, jaundice, nausea, vomiting, and splenomegaly are common symptoms and rarely, bleeding tendency, ascites, pleural effusion, hepatic encephalopathy can occur (8).

There is no specific imaging criterion for diagnosis of PH-BL. Large nodules and diffuse infiltrative lesions could suggest a broad range of tumors and infiltrative processes (10).

Early administration of multiple chemotherapeutic agents increases the curative potential of Burkitt's lymphoma. Patients with primary hepatic NHL without ALF have an overall survival of 87% at 5 years after oncological treatment (11). However, same admission mortality rate for primary hepatic NHL with ALF is 87% with an average survival of 11 days (3).

Reviewing seven cases of PH-BL showed that five patients died during the initial admission. One death was due to tumor lysis syndrome and adult respiratory distress syndrome on chemotherapy initiation (12). One case with presentation of ALF responded well to combination chemotherapy and was alive one year after diagnosis (3). As previously mentioned our case was expired in few days without any treatment.

In conclusion, we think that the diagnosis of primary hepatic lymphoma should be considered in evaluation of space occupying liver lesions especially in HIV infected patients. Image guided needle biopsy of the liver is the best method for the definite diagnosis of hepatic lymphoma.

## Acknowledgements

The authors declare that there is no conflict of interests.

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