

# Primary hepatic Burkitt's lymphoma in an immunocompromised adult

Sunita Singh, Parveen Rana Kundu<sup>1</sup>, Parul Tanwar, Sonia Chhabra

Department of Pathology, Post Graduate Institute of Medical Sciences, Rohtak, Haryana, <sup>1</sup>Department of Pathology, Government Medical College, Khanpur Kalan, Haryana, India.

## ABSTRACT

Primary liver lymphomas are uncommon, accounting for only 0.4% of all the extra nodal lymphomas. Burkitt's lymphoma is a rare type of Non-Hodgkin's lymphoma. It is classified into three types: Endemic, sporadic and immunodeficiency associated. Endemic type is found in Africa, associated with Epstein-Barr Viral infection. Non-endemic type usually involves mesenteric lymph nodes or ileocecal area and usually presents as an abdominal mass. Primary hepatic Burkitt's lymphoma is a very rare neoplasm. Here, we present the case of a 28-year-old male presenting with right upper quadrant pain and generalized weakness. A core needle biopsy of the liver was performed, which revealed cytologic and immunohistochemical findings compatible with Burkitt's lymphoma.

**Key words:** Burkitt's lymphoma, liver, primary

## INTRODUCTION

Burkitt's lymphoma is a rare Non-Hodgkin's lymphoma, which grows rapidly requiring aggressive therapy and occurs usually in children.<sup>[1]</sup> Primary hepatic lymphoma is a rare neoplasm accounting only for about 0.4% of all extra nodal lymphomas.<sup>[2]</sup> Primary hepatic Burkitt's lymphoma, a highly aggressive subset of Non-Hodgkin's lymphoma, is a very rare entity.<sup>[3]</sup> Clinical symptoms varies from no symptoms to mild abnormal liver function to fulminant hepatic failure. Radio logically most of the lymphomas are solitary or multiple masses in the liver but rarely diffuse hepatosplenomegaly without a definite mass may occur.<sup>[1]</sup> Liver function tests are often abnormal. Elevated levels of serum alkaline phosphatase and lactate dehydrogenase are also seen.

There have been seven cases of primary hepatic Burkitt's lymphoma in adults, but only five cases had confirmed

immunophenotypic diagnosis.<sup>[3]</sup> We present the case of a 28-year-old male presenting with primary hepatic Burkitt's lymphoma.

## CASE REPORT

A 28-year-old male presented with generalized weakness, fatigue and pain abdomen of 1 month duration. There was also history of weight loss and decreased appetite. There was no history of fever, drug intake or alcohol consumption. On physical examination, the patient was conscious, oriented, and his vitals were maintained. There was no icterus, cyanosis or lymphadenopathy. Per abdominal examination revealed hepatomegaly, liver was soft, tender and was palpable 2 cm below the costal margin. Viral markers including Hepatitis B surface antigen and anti-hepatitis C antibodies were all negative. Human immunodeficiency virus enzyme-linked immuno sorbent assay was reactive.

On ultrasound abdomen, liver measured 18.5 cm and showed multiple heteroechoic lesions in both lobes, with the largest measuring 8 cm × 7cm in diameter. Spleen measured 12.5 cm. Gall bladder, pancreas and bilateral kidneys were normal.

Fine needle aspiration cytology and core needle biopsy from the largest lesion was carried out, and smears were made. Cytological smears showed cellular smears

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#### DOI:

10.4103/2278-0513.119258

**Address for correspondence:** Dr. Parul Tanwar, Department of Pathology, Pt. B.D. Sharma PGIMS, Rohtak - 124 001, Haryana, India.  
E-mail: drparultanwar@gmail.com

comprising of monomorphic population of lymphoid cells revealing high mitotic activity in a background of macrophages and lymphoglandular bodies. These cells had round nuclei, granular nuclear chromatin, multiple nucleoli and a moderate amount of vacuolated dense blue cytoplasm [Figure 1]. Diagnosis was confirmed on histopathology [Figure 2]. On immunohistochemistry, tumor was positive for leucocyte common antigen, cluster differentiation (CD) 20, and CD 10 [Figure 3] and was negative for B cell lymphoma 2 (BCL-2) and CD 34.

## DISCUSSION

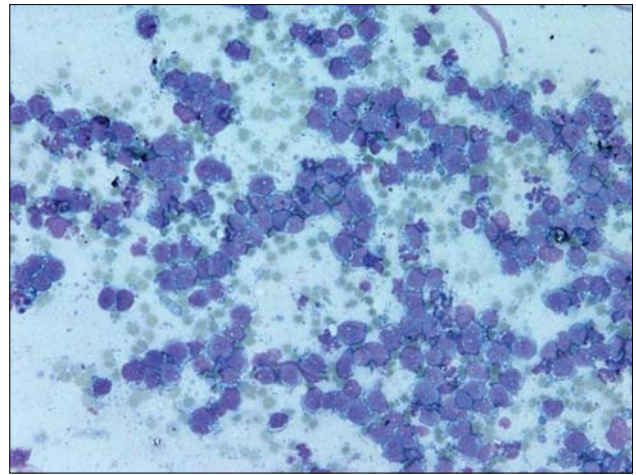
Burkitt's lymphoma is an undifferentiated malignant lymphoma of B lymphocytes. It was first described by Denis Burkitt, who named the tumor on the African children's jaw sarcoma.<sup>[4]</sup> It is classified into three types: Endemic, sporadic and immunodeficiency associated.<sup>[1]</sup>

Definite diagnostic criteria for primary hepatic lymphomas has not been established.<sup>[3]</sup> Caccano suggested strict criteria that primary hepatic lymphoma should have liver involvement only. However, many cases have regional lymph nodes, spleen and bone marrow involvement. So, Lei suggested a broader criteria that the symptom expression mainly originate from liver infiltration with no distant lymphadenopathy and no leukemoid reaction in peripheral blood.<sup>[5]</sup>

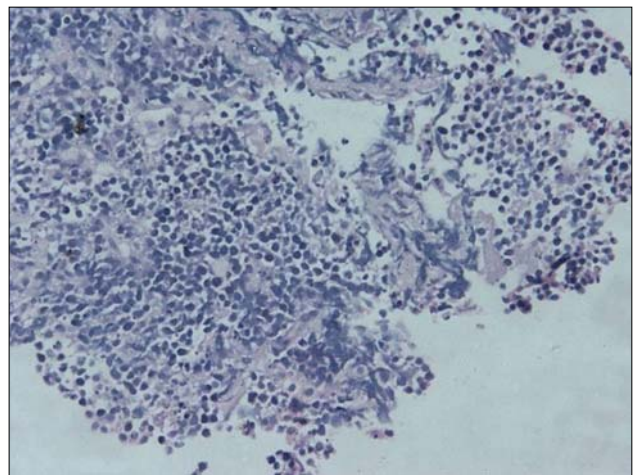
Secondary involvement of the liver is relatively common including 50-80% of Hodgkin's lymphoma or Non-Hodgkin's lymphomas at autopsies whereas primary involvement accounts for less than 1% of all extra nodal lymphomas.<sup>[3]</sup> Primary hepatic Burkitt's lymphoma is a very rare entity, accounting for only 0.4% of all extra nodal lymphomas.<sup>[2]</sup> Primary hepatic Burkitt's lymphoma make up to 3% of primary hepatic lymphomas.<sup>[2]</sup> In a study conducted by Wissam *et al.*, seven cases of hepatic Burkitt's primary in adults were described, but only five cases had confirmed immunophenotypic diagnosis.<sup>[6]</sup>

Primary liver lymphomas have many histologic types. Diffuse large B-cell lymphoma is the most common type and lymphoblastic lymphoma, Burkitt's lymphoma, follicular lymphoma, diffuse histiocytic lymphoma, mantle cell lymphoma and T-cell rich B-cell lymphoma are reported.<sup>[7]</sup>

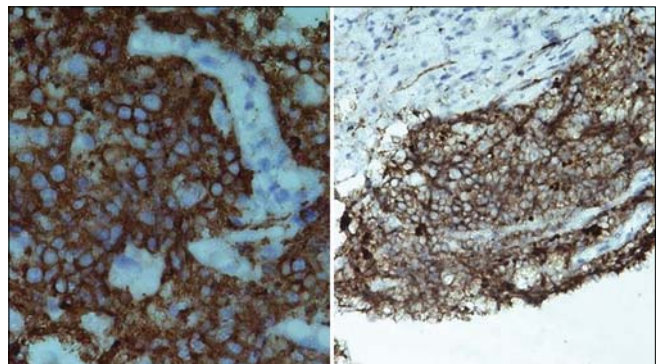
On histological examination, Burkitt's lymphoma is composed of uniform population of medium sized cells with round nuclei and multiple small basophilic nucleoli. The nuclei are approximately the same size as the nuclei of the admixed histiocytes. The nuclear contours are generally round without deep indentation. The cytoplasm is strongly basophilic with small round cytoplasmic vacuoles best observed in air dried touch imprints. Tingible body



**Figure 1:** Cellular smears showing monomorphic population of lymphoid cells revealing high mitotic activity in a background of macrophages and lymphoglandular bodies. Cells had round nuclei, granular nuclear chromatin, multiple nucleoli and a moderate amount of vacuolated dense blue cytoplasm (Leishman stain, x100)



**Figure 2:** Monomorphic population of lymphoid cells (H and E, x200)



**Figure 3:** A-CD 20 positivity in the lymphoid cells (x200 anti CD 20); B-CD10 positivity in the lymphoid cells (x100 anti CD 10)

macrophages, phagocytosing abundant apoptotic debris creating starry sky appearance is a characteristic finding, although not always present.<sup>[3,4]</sup>

On immunohistochemical examination, the neoplastic cells

are positive for surface immunoglobulin Ig M and Ig light chains; pan B-cell antigens (CD 19, CD 20, CD 22, CD 79) and co-expresses CD 10, BCL-6, CD 43, and p53 but not CD 5, CD23, BCL-2 and CD 138. The proliferating fraction is nearly 100%.<sup>[1,3,4]</sup>

A molecular defining feature of Burkitt's lymphoma is the presence of translocation between c-myc gene and IgH gene (t(8;14)) or between c-myc gene and the gene for either the kappa or lambda light chain (t(2;8) or t(8;22)).<sup>[8]</sup>

The clinical feature of primary hepatic lymphoma varies from no symptom to fulminant hepatic failure, fever, weight loss, night sweats, right upper quadrant pain, hepatomegaly, fatigue, jaundice, nausea, vomiting, and splenomegaly are common symptoms and rarely bleeding tendency, ascitis, pleural effusion and hepatic encephalopathy can occur.<sup>[3]</sup>

There are no specific imaging criteria for diagnosing primary hepatic Burkitt's lymphoma. Large nodules and diffuse infiltrative lesions could suggest a broad range of tumors and infiltrative process.<sup>[9]</sup>

## REFERENCES

1. Lee SH, Kim HJ, Mun JS, Oh HC, Lee HW, Choi CH, *et al.* A case

- of primary hepatic Burkitt's lymphoma. *Korean J Gastroenterol* 2008;51:259-64.
2. Kuroda J, Omoto A, Fujiki H, Okugawa K, Tamai H, Yamagishi H, *et al.* Primary hepatic Burkitt's lymphoma with chronic hepatitis C. *Acta Haematol* 2001;105:237-40.
3. Saffar H, Zamani H, Sotoudehmanesh R, Taheri AP, Sotoudeh KF. Primary hepatic burkitt's lymphoma in a patient with acquired immunodeficiency syndrome. *Iran J Pathol* 2011;6:97-100.
4. Burkitt D. A sarcoma involving the jaws in African children. *Br J Surg* 1958;46:218-23.
5. Caccamo D, Pervez NK, Marchevsky A. Primary lymphoma of the liver in the acquired immunodeficiency syndrome. *Arch Pathol Lab Med* 1986;110:553-5.
6. Mattar WE, Alex BK, Sherker AH. Primary hepatic Burkitt lymphoma presenting with acute liver failure. *J Gastrointest Cancer* 2010;41:261-3.
7. Lei KI. Primary non-Hodgkin's lymphoma of the liver. *Leuk Lymphoma* 1998;29:293-9.
8. Burmeister T, Schwartz S, Horst HA, Rieder H, Gökbuget N, Hoelzer D, *et al.* Molecular heterogeneity of sporadic adult Burkitt-type leukemia/lymphoma as revealed by PCR and cytogenetics: Correlation with morphology, immunology and clinical features. *Leukemia* 2005;19:1391-8.
9. Jacobs SL, Rozenblit A. HIV-associated hypervascular primary Burkitt's lymphoma of the liver. *Clin Radiol* 2006;61:453-5.

**Cite this article as:** Singh S, Kundu PR, Tanwar P, Chhabra S. Primary hepatic Burkitt's lymphoma in an immunocompromised adult. *Clin Cancer Investig J* 2013;2:253-5.

**Source of Support:** Nil, **Conflict of Interest:** None declared.