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# Five Years Survival of Patient with Primary Hepatic Non-Hodgkin's Lymphoma

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**Abstract:**

Primary Non-Hodgkin's Lymphoma of liver is an extremely rare case. We describe a case 21-year old female who presented with right groin pain, no hepatosplenomegaly, weight loss, night sweats or lymphadenopathy with elevated lactate dehydrogenase. She had mildly enhancing hypodense mass at porta hepatis as observed on contrast enhanced CT (CECT) scan of whole abdomen (CECT. FNAC, biopsy and ultimately immunohistochemistry revealed high-grade B cell non-Hodgkin's lymphoma. She was treated with chemotherapy by CHOP schedule and followed up for a period of five years. During that period, she was found well, and no lesion was observed in liver on CECT of whole abdomen. The patient was successfully treated. She overcame the 5-year survival rate and is still alive without any complaints. We conclude her treatment was successful, although her presentation was unusual.

**Key Words:** Primary Hepatic NHL, 5-year relative survival rate

## Introduction

Primary lymphoma in liver is found in about 0.4% of extranodal lymphoma and 0.01% of cases of Non-Hodgkin's Lymphoma (NHL); secondary involvement is not uncommon [1-3]. Extranodal lymphoma, especially NHL, is more commonly seen than Hodgkin's Lymphoma. NHL can affect persons of any age; the incidence increases from childhood and mostly found in adult men at the 5th decade of life [4]. On imaging, hepatic NHL presents as a single mass at porta hepatis on USG, contrast enhanced CT scan (CECT), and MRI [1]. Hepatic NHL can be successfully treated by surgery or chemotherapy [1, 5, 6]. Usually, more than 70% of primary hepatic lymphoma cases present as a single mass in USG, CECT, and MRI of whole abdomen [1, 7]. Patients with primary hepatic NHL present with fever, right upper quadrant pain, weight loss, night sweats [8]. On examination, hepatosplenomegaly is usually found [2].

## Case Report:

Our case, a 21-year old female patient had history of occasional right groin pain with no fever, weight loss, night sweats or any significant co-morbidity. On physical examination, liver and spleen were not palpable. There was no enlarged cervical, axillary, intraabdominal or inguinal lymph node. Previous reports on complete blood count, liver function tests, kidney function tests, serum electrolytes, and total protein were within normal range. Her serology for hepatitis B and C viruses was negative, and the chest x-ray was also normal. Ultrasonography of whole abdomen demonstrated an ill-defined isoechoic mass at porta hepatis compressing the portal vein (Figure 1). Her lactate dehydrogenase (LDH) was 823 U/L, Alpha-fetoprotein level was 2.01 ngm/mL and Beta-hCG was less than 0.10 ngm/mL. She was advised for CECT of whole abdomen on 12 July 2011), and a large low-density mild-to-moderately enhancing lesion was revealed arising at porta

hepatis occupying IV, V and VI segments of liver with compression and displacement of right hepatic and portal veins (Figure 2). This lesion caused posterior displacement of gall bladder, pancreas, and kidneys. Rest of the abdominal organs was unremarkable, with no ascites or any enlarged lymph node. FNAC from the lesion on 15 July 2011 suggested malignant round cell tumour with the possibility of NHL as there were large sheets of cells having distorted nucleus and inconspicuous cytoplasm. Histopathology report from the lesion confirmed intermediate grade of NHL on 24 July 2011. Immunohistochemistry (Figure 3) revealed that CD 10, CD 20, BCL 6, and Ki 67 were positive in majority of lesional cells whereas CD3, CD5, Mum 1, and Tdt were negative, which further confirmed high-grade B cell NHL with differential diagnosis of Burkitt lymphoma and diffuse large B cell lymphoma with high proliferation index. From the above diagnoses, the patient received chemotherapy with CHOP (cyclophosphamide, doxorubicin, vincristine, and prednisone) regimen. She was treated with eight cycles of CHOP regimen. From 2013 to 2015 (Figure 4), regular imaging by USG was performed with LDH, Beta-hCG and Alpha-fetoprotein which were always normal. Finally, the last CECT on 9 December 2016, five years after initial diagnosis, revealed her disease-free state (Figure 5).

**Discussion:**

Non-Hodgkin's Lymphomas consist of a heterogeneous group of malignant neoplasms and originate from B or T cell and also rarely from Natural Killer Cell. Extranodal NHL of liver is unknown; however, some viruses, such as HBV, HCV, and Epstein-Barr, have been implicated [2, 5, 9-11]. A few occupational exposures, like herbicides, pesticides, wood dust, forestry, painting, carpeting, and tanning play roles in the development of NHL. This case presented with only groin pain but no upper abdominal pain or weight loss as well as no lymphadenopathy or hepatosplenomegaly. Like other hepatic NHL patients, Beta-hCG and Alpha-fetoprotein were not raised [1, 12, 13] and must be considered in the differential diagnosis of liver metastasis when no other primary tumor is present [14]. In the present case, lesion of liver was situated at porta hepatis evident from imaging. Similar findings were reported in previous studies [1, 2, 4]. The mainstay of therapy for NHL patients is chemotherapy with CHOP regimen, with the addition of Rituximab (monoclonal antibody against CD-20), which prolongs survival with minimal toxicity; so, the scheme is called R-CHOP. Our patient was treated with CHOP regimen without Rituximab. Usually, all patients with NHL are treated with six cycles of CHOP regimen but our patient was given eight cycles because, after completion of six cycles, she had residual lesion on follow-up (Figure 6). One month after follow-up, on completion of eight cycles, she had the disease-free state (Figure 7). During the follow-up period from 2013 to 2015, regular imagings by USG were performed. CECT of whole abdomen could be the best choice for imaging of her liver lesion but, due to financial constraints, we only advised USG of whole abdomen for follow-up with chest x-ray, LDH, Beta-hCG and Alpha-fetoprotein. All the parameters were always normal at follow-up. The last CECT of whole abdomen on 9 December 2016, five years after initial diagnosis, also revealed her disease-free state. Masood et al. [9] reported that their patient (65 years old male) with hepatic NHL had disease-free state up to completion of chemotherapy. A pervious study by Myoteri et al. [8] in 2014 reported 3 years relative survival of their patient (70 years old male) with primary hepatic NHL. Nagral et al. [2] reported 6 months relative survival of their hepatic NHL patient (53 years female). Actually, success of treatment depends on advanced age, constitutional symptoms, bulky disease, unfavourable histologic subtype, elevated levels of LDH and β2-microglobulin, a high proliferation rate, cirrhosis, and co-morbidity [1, 5]. Our patient was fortunate enough to achieve 5 years relative survival time as she was young and had no associated co-morbidity.

**Conclusion:**

The findings of this case study can make the oncologists aware that NHL can affect the liver (although rare,), and proper management can ensure the five years survival (or more) of patients with primary hepatic NHL.

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**Figures and Illustrations:**

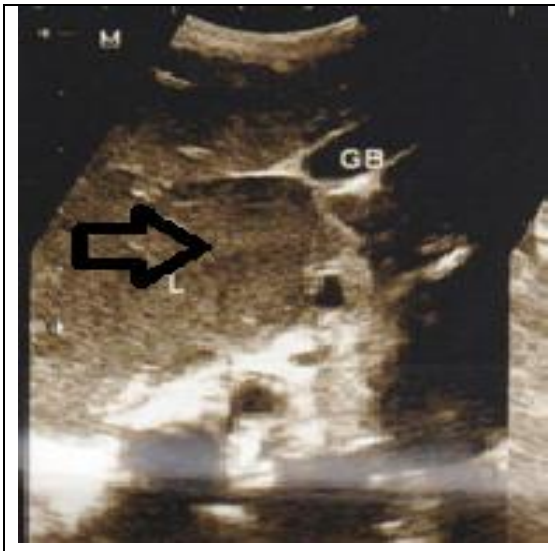



**Figure 1:** USG of liver before treatment demonstrate an isoechoic mass (open black arrow) at porta hepatis.

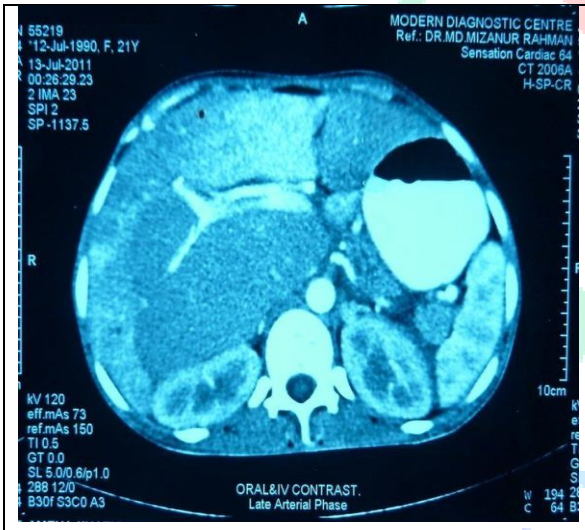
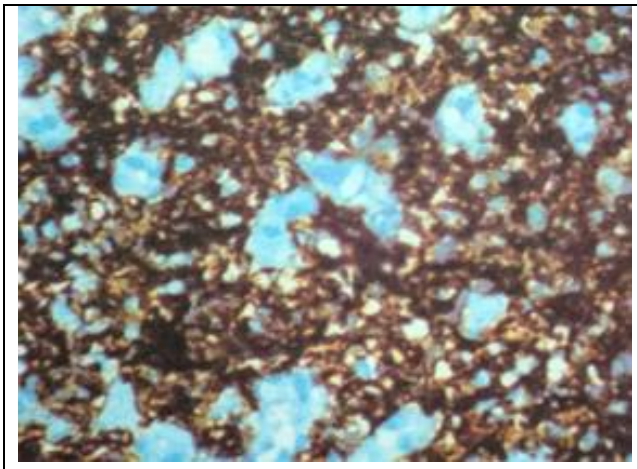
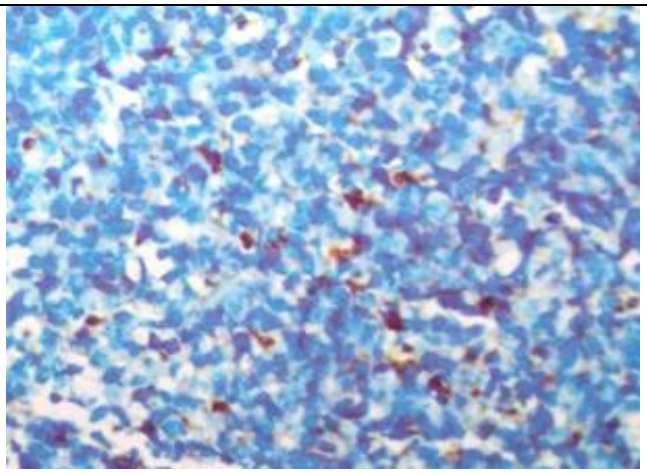


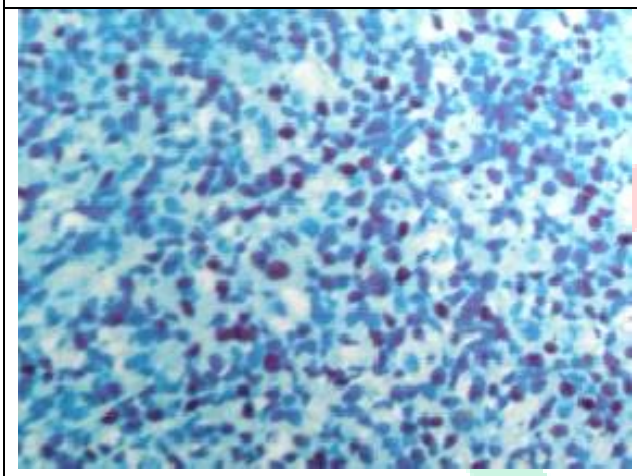

**Figure 2:** CECT before treatment showing large low density moderately enhancing lesion at porta hepatis occupying IV to VI segments of liver with compression and displacement of right hepatic, portal veins.



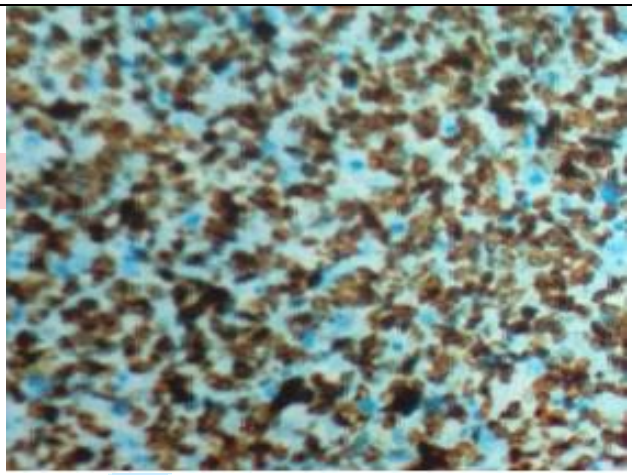
CD20



CD10

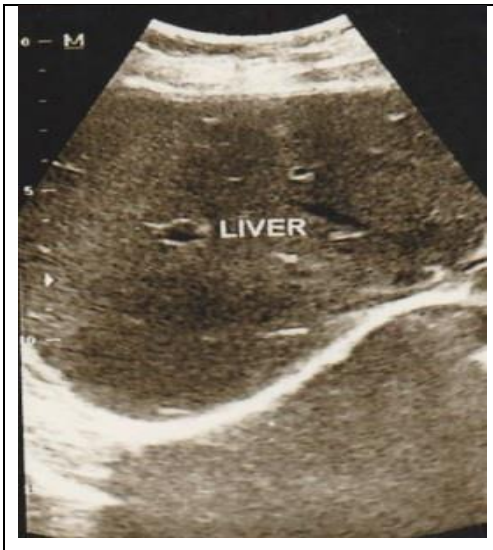


BCL6

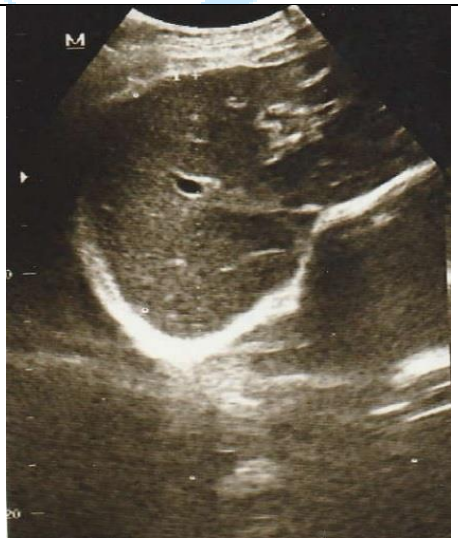


KI67

**Figure 3:** Photomicrographs showing majority of lesional cells positive for CD20, CD10, BCL6, KI67.




In the year 2013



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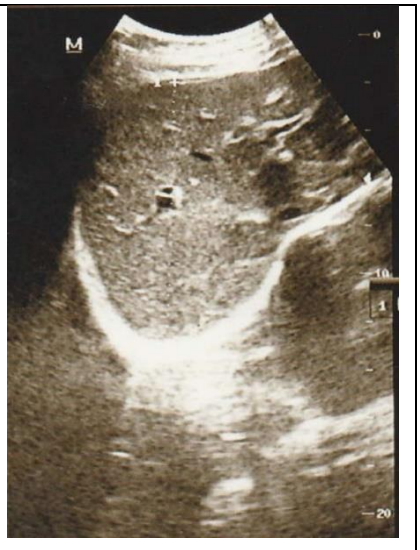

In the year 2015

**Figure 4:** USG of abdomen of the patient at follow up period.

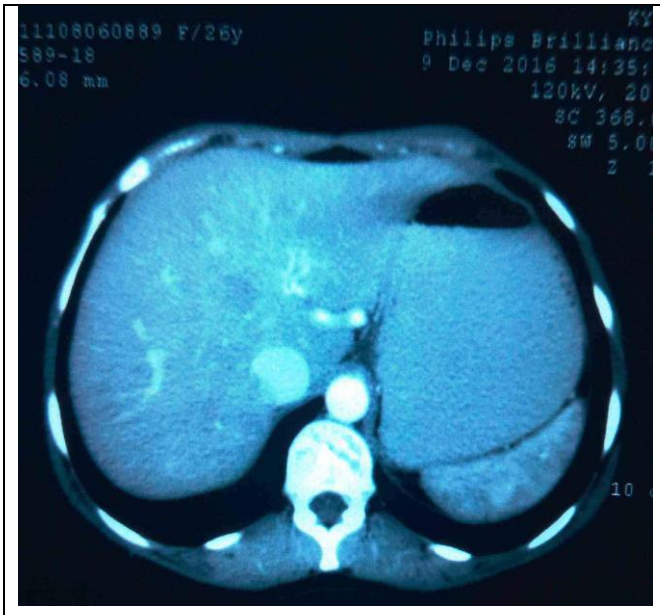

**Figure 5:** Follow up image 5 years after initial diagnosis revealing no residual disease.

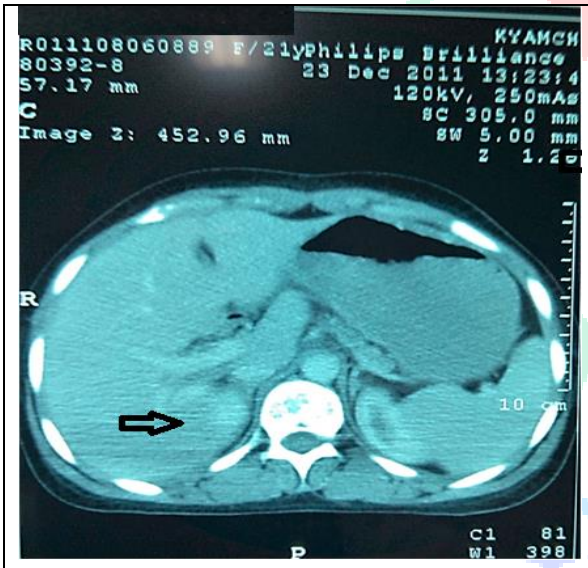


**Figure 6:** CECT after six cycle CHOP showing residual lesion at porta hepatis of liver (Open black arrow).

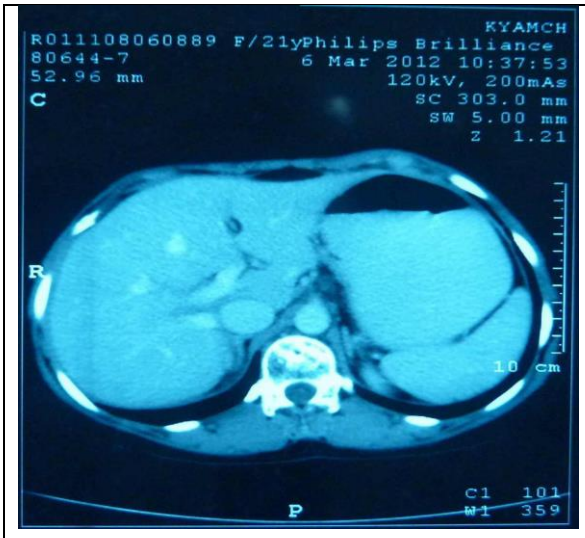


**Figure 7:** CECT after eight cycle CHOP showing no residual lesion at porta hepatis of liver.