

# A Rare Presentation of Primary Hepatic Lymphoma: A Case Report

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## ABSTRACT

Primary hepatic lymphoma (PHL) was a rare malignancy usually presenting with abdominal pain, malaise, Hepatomegaly, B-symptoms, fatigue, nausea, vomiting and jaundice. It mostly involves liver without any palpable lymphadenopathy and leukemia in peripheral blood smear. On July 22, 2014, a 64 year old man presented with abdominal pain localizing in Right upper quadrant and fullness from 2 years ago without any weight loss or constitutional symptoms. His physical examination revealed no mass or lymphadenopathy and lab data showed rise in Alkaline phosphatase(ALP) and Gamma-glutamyl transferase(gamma-GT).

Abdominal computed tomography(CT) scan with contrast showed a calcified lesion in the left lobe of liver and ill-defined hypodense area in medial segment of the left lobe of liver adjacent to gallbladder associated with mild central intra hepatic bile ducts dilation showing more enhancement in delay phase suggested peripheral cholangiocarcinoma. Finally surgical core needle biopsy of the liver confirmed malignant lymphoma of B cell type and patient was referred to oncologist for chemotherapy. His chemotherapy regimen consisted of rituximab 600 milligram (mg), endoxan 1250 mg, adriamycin 80 mg, vincristine 2 mg, prednisolone 100 mg (during five days) for 6 courses. After 5 months chemotherapy, on December 22, 2014 a follow up CT scan with IV and oral contrast was done. There was no evidence of previous mass lesion in the liver. In the follow up on May 9, 2015, he had no specific symptoms and all of his lab data were in normal range.

**Keywords:** Hepatic Lymphoma; Abdominal pain; Hepatomegaly

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### INTRODUCTION

Lymphomas originating from the liver were very infrequent. These lymphomas could be either primary hepatic lymphoma (PHL) or without any systemic and lymphatic involvement consisting about 0.4% of extra nodal non-Hodgkin's lymphomas (NHL) and 0.016% of all patients with non-Hodgkin's lymphomas (NHL) OR secondary involvement of the liver which occurs in 16-40% of patients in advance course of systemic non Hodgkin lymphoma(1-16).

This disease was usually seen in middle-aged

**Table 1: First Laboratory Test results**

CBC/diff	Biochemistry	Hormone	Immunity markers
WBC: 6.37 Neu:4.38 Lymph: 1.33	Urea: 38	CEA: 0.9	HBS Ag: non reactive
RBC: 4.47	Cr: 0.95	CA 19-9:1.6	HCV Ab EIA: non reactive
HB:14.6	ALK-p: 361		CRP <0.6
PLT: 293*103	Gamma glutamyl tranferase: 280		
Pt:13	B2 microglobulin: 2.5		
INR: 1.16	LDH: 337		
	Cholesterol:175		
	FBS: 110		
ESR: 13	Triglyceride: 192		
	SGOT:16		
	SGPT:13		

male patients (4,5,13,17). Abdominal pain (right upper quadrant or epigastric), discomfort, malaise, hepatomegaly, tender and palpable mass were the majority of clinical presentations. B-symptoms (fever, weight loss and night sweats) fatigue, nausea, vomiting, jaundice, anorexia, malaise, fulminant hepatic failure, hepatic encephalopathy, pleural effusion, thrombocytopenia constitute the rare presentations of the disease(1,3-5,9-11,17). Small percentage of the patients were asymptomatic and accidentally diagnosed(1,9).

Here we described a rare case with primary hepatic lymphoma only presented with chronic abdominal pain.

## CASE REPORT

On July 22, 2014, a 64 year old man presented with abdominal pain localizing on Right upper quadrant and fullness from 2 years ago without any weight loss or constitutional symptoms. He didn't have any history of fatigue, nausea, vomiting, jaundice, anorexia, malaise and B-symptoms including fever and night sweats.

Physical examination revealed no mass, lymphadenopathy and splenomegaly. Lab data revealed rise in Alkaline phosphatase (ALP): 361 and Gamma-glutamyl transferase (gamma-GT): 280 and normal level of lactate dehydrogenase (LDH): 337, Carcinoembryonic antigen (CEA): 0.9 and Carbohydrate antigen 19-9 (CA 19-9):1.6. Hepatitis B surface antigen (HBs Ag) and Hepatitis C virus antibody (HCV antibody) were both non-reactive in this patient (Table 1).

Ultrasonography of abdomen shows a 79×46 mm coarse calcified heterogenic and hypo echoic mass

with poorly defined borders in the hilum of the liver and mild dilation of hepatic bile ducts of the left lobe of liver suggesting peripheral cholangiocarcinoma. Thickening of gall bladder was also noted. Several cortical cysts in the left and right kidneys were observed. Abdominal computed tomography (CT) scan with contrast (Figure 1) showed a calcified lesion measuring 45 × 42 mm in the left lobe of liver suggestive of an old hydatid cyst. Ill-defined hypo dense area measuring 79 × 87 mm in medial segment of the left lobe of liver and adjacent to gall bladder associated with mild central intra hepatic bile ducts dilation showing more enhancement in delay phase, suggested peripheral cholangiocarcinoma. Spiral CT scan of thorax (Figure 2) showed varicoid bronchiectasis in lingual left lower lobe and right middle lobe. There were also pulmonary scars in apices. Finally surgical core needle biopsy of the Liver was performed (Figure 3). A tiny island of liver parenchyma adjacent to an extensive zone of destructive fibrosis had replaced the normal liver tissue and was heavily infiltrated by lymphocytes, few eosinophils and many crushed cells. Lymphoid cells were mostly positive for CD 20(Figure 4) and No CD10, ckAE1/AE3 or CD 30 were detected. CD5 and CD3 and bcl-2 markers were positive on included small T lymphocytes. Ki 67 marker showed 12–15 % proliferative activity (Figure 5) and CD 23 was focally positive. All of these data confirmed malignant lymphoma of B cell type. So the patient was referred to an oncologist for chemotherapy. His chemotherapy regimen consisted of rituximab 600 milligram (mg), Endoxan 1250 mg, Adriamycin 80 mg, vincristine 2 mg, prednisolone 100 mg (during five days) for 6 courses. After 5 months on December 22, 2014 a



Fig. 1: Abdominal CT scan with contrast

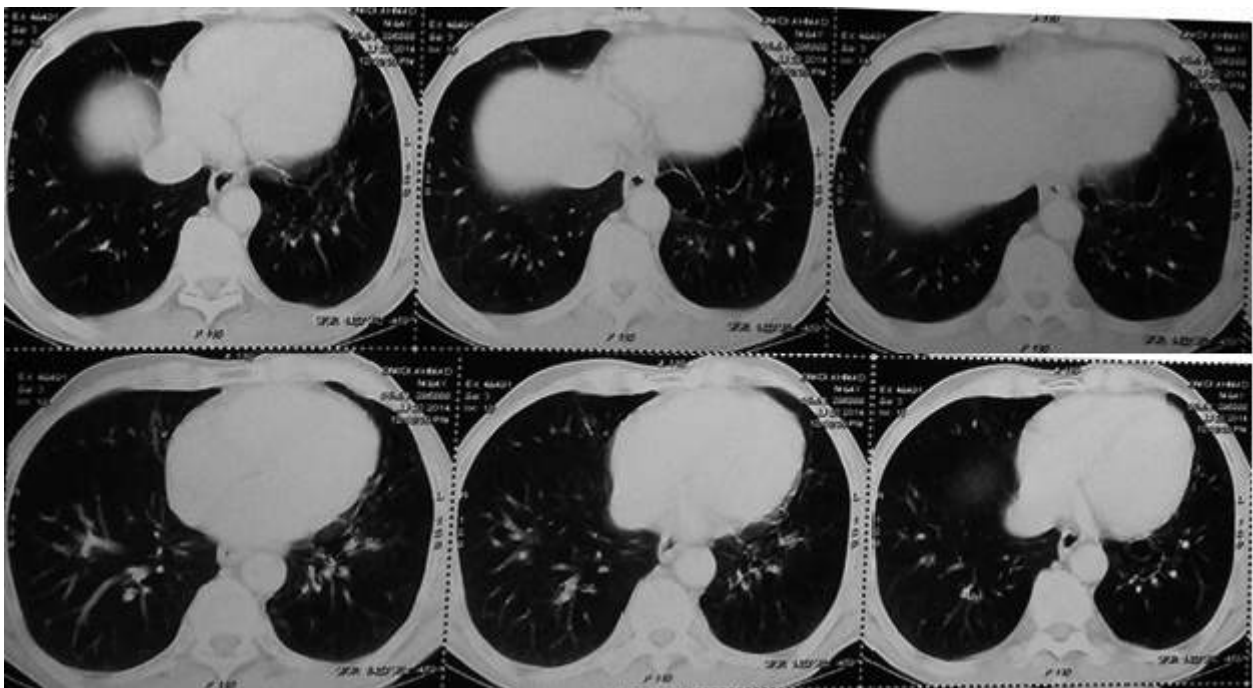


Fig. 2: Spiral CT scan of thorax

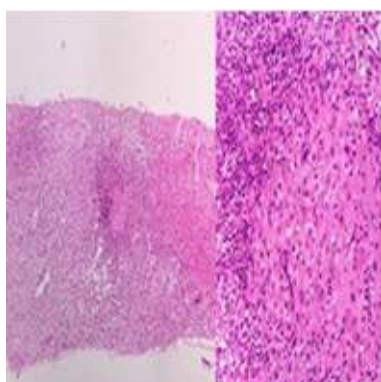
follow up CT scan with IV and oral contrast was done. There was a dense calcified lesions measuring 44×41 mm in medial segment of the left lobe of liver that was suggestive of an old hydatid cyst. There was no evidence of previous mass lesion in the liver. Few simple cortical cysts were seen in both kidneys. In the follow up on May 9, 2015, he had no symptom and all

of the patient's lab data were in normal range (Table 2).

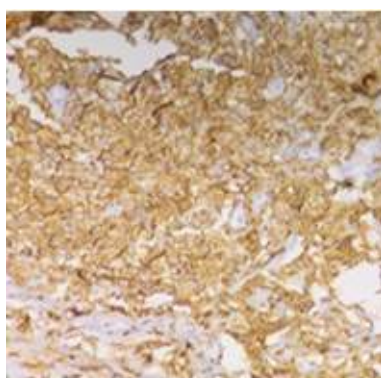
## DISCUSSION

Large B cell lymphomas constitute the vast majority of cases with primary hepatic lymphomas. Other less common subtypes include primary low-grade hepatic B-cell lymphoma of extra nodal marginal zone B

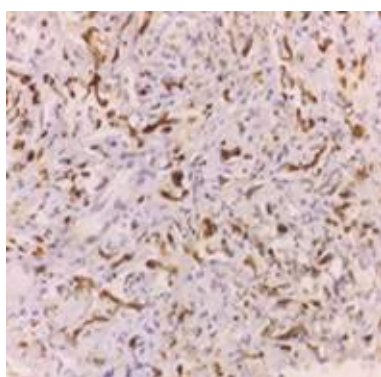




**Fig. 3:** Pathological result of core needle biopsy of the liver



**Fig. 4:** Positivity of lymphoid cells for CD20 markers



**Fig. 5:** Ki 67 immunohistochemistry test

**Table 2:** Follow up laboratory test results

CBC /diff	Biochemistry
WBC: 6.1 Neu: 56% Lymph: 30%	Urea: 38
RBC: 4.91	Bilirubin T: 0.9
HB:14.3	Bilirubin D: 0.4
PLT: 311*103	LDH: 381
ESR: 8	ALK-p: 165
CRP= negative	Cholesterol:187 HDL:41 LDL:123
Ca: 9.5	FBS: 86
P: 3.6	Triglyceride: 116 SGOT: 10 SGPT: 24

origins in gastrointestinal tract with similar clinical symptoms(3,5). This differential diagnosis was firstly suggested in our patients due to the results of CT scan and ultrasonography. There were no explicit gold standard criteria for primary hepatic lymphoma(15). One criteria for diagnosis of PHL has been described by Lei KI including manifestation of disease symptoms that chiefly involve liver, nonattendance palpable lymphadenopathy, lack of radiological documents about distant lymph node involvement and lack of leukemia evidence in peripheral blood smear(1,19). Caccamo described the PHL criteria as lymphoma confined only to the liver without the involvement of any other organs like spleen, bone marrow, lymph nodes, peripheral blood or other tissues until at least six months after diagnosis(3, 10, 20). Abnormal changes in the liver function tests such as rise in aspartate aminotransferase (AST or SGOT), alanine aminotransferase (ALT or SGPT), elevation in bilirubin level, Gamma glutamyl tranferase, ALP (alkaline phosphatase), LDH(lactate dehydrogenase ), normal level of alpha-feto protein (AFP) and car-cinoembryogenic antigen (CEA) are the main laboratory results(1,7,9,10,17). LDH have been proposed as a diagnostic marker(5). hypercalcemia, coagulopathy, changes in serum proteins electrophoresis are uncommon results in laboratory examinations (1,5). Our patient had a rise in the serum level of Gamma glutamyl tranferase and ALP (alkaline phosphatase) but he also had normal levels of LDH and CEA.ESR (erythrocyte sedimentation rate), CRP(C-reactive protein) and Beta-2-microglobulin which has been known as a

cell lymphoma (MALT lymphoma)and peripheral hepatosplenic T-cell lymphoma(1-4,7,10,18). Different presentations of Primary hepatic lymphoma include single or multiple liver mass or diffuse infiltration of tumor in the liver. Some of differential diagnosis of PHL are hepatocellular carcinoma and tumors that have been metastasized from other

prognostic factor for lymphoma, also increase in the term of the disease(1). Primary hepatic lymphoma is more common in men than women, median Age of 50 to 55 years (3, 9,13,17). PHL has been known to have a poor prognosis with median survival of 15 months but ranges from 3 to 124 months and in patients who undergo just chemotherapy median survival is 6 months which is less than patients who undergo combination therapy(1,3).Risk factors that are associated with poor prognosis are increase in LDH level, rise in  $\beta$ 2-microglobulin, liver cirrhosis, older age, wide dissemination in the liver, HIV, chronic hepatitis and immune suppression(1,3,7,21). There is no strong evidence about the etiology of PHL but despite the lymphoid origin of liver tissue, liver has been known as an insignificant environment for arising lymphomas(3,10,11,17).Several factors have been posed to have role in the pathogenesis of PHL such as hepatitis B or C virus (HCV) infection, Epstein barr

virus (EBV), human immunodeficiency virus (HIV) and Human T-cell lymphotropic virus (HTLV). It also has known to be related with immune suppression that can be seen in acquired immune deficiency syndrome( AIDS), cyclosporine consumption in transplantation, systemic lupus erythematosus, hemochromatosis and liver cirrhosis which seem to have relationship with decrease of T cell progenitors and unrestrained proliferation of B cells(1,3,5,17). Nevertheless none of these etiologies were compatible with our patient that shows a rare case. Treatment of PHL is not well established, but current treatments consist of resection of tumor with or without mixture of radiotherapy and chemotherapy (1,5,13). Our patient got 6 courses of chemotherapy for 6 months and was tumor free after 5 months of treatment. On follow up CT scan there was no evidence of previous mass lesion in liver.

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