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Primary Non Hodgkin Lymphoma of Liver: A Rare Diagnosis

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ABSRECT
Primary non-Hodgkin lymphoma (NHL) of liver is a very rare malignancy. Here we report a case of a 50 year old man presented to us with history of pain upper abdomen for last 2 months. A computed tomography of the abdomen revealed enlarged liver with multiple hypodense lesion in 4th segment of liver. Diagnosis of primary NHL liver was made using ultrasound guided FNAC and biopsy. The patient is currently being treated with a CHOP (cyclophosphamide–doxorubicin–vincristine–prednisolone) regimen. The case has many unique features, including normal liver function tests; no type B symptoms; and negative serology for viruses. The case demonstrates that primary hepatic lymphoma should be considered in the differential diagnosis of space-occupying liver lesions in presence of normal levels of alpha-fetoprotein and carcinoembryonic antigen.

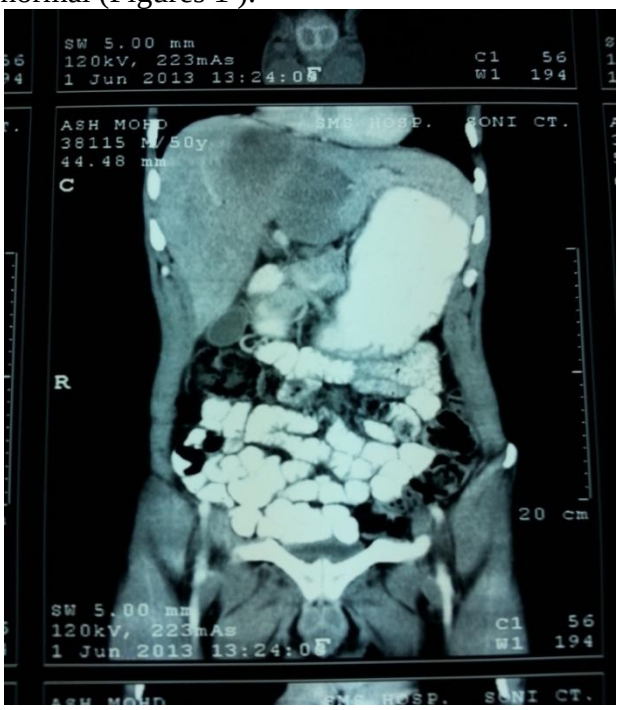
Key words*: non hodgkin ,liver , lymphoma ,extranodal*

INTRODUCTION

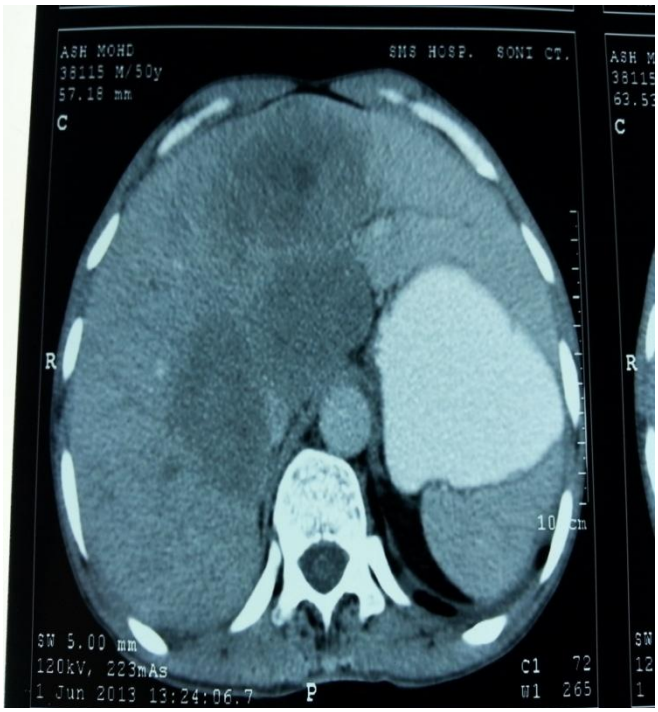
Primary hepatic lymphoma (PHL) is a very rare malignancy [1]. Although the liver contains lymphoid tissue, host factors may Make the liver a poor environment for the development of malignant lymphoma [2].

Case presentation

A 50 year old man presented to us with chief complaints of pain upper abdomen and epigastric fullness for two months and occasional back pain. Patient denied any history of fever,night sweats , vomiting , diarrhea ,weight loss and blood in stools. There was no significant past history regarding any chronic medical illness except for pulmonary tuberculosis for which he had taken anti tubercular drugs for 9 months. On physical examination, the liver was enlarged in size. No superficial lymphadenopathy was present. Laboratory results revealed hemoglobin 10.2g/dl and a white cell count of 8600/cumm, with a normal differential count . Alanine aminotransferase, aspartate aminotransferase, alkaline phosphatase (ALP), and lactate dehydrogenase (LDH) were within normal limits. Levels of serum alpha-fetoprotein and carcinoembryonic antigen (CEA) and PSA were not elevated. Serology was negative for HIV and for the hepatitis C (HCV) and B (HBV) viruses. Serum calcium was within normal limits. A CT scan of abdomen revealed liver enlarged in size with presence of multiple hypodense mass lesion , largest one measuring 84x52x82mm in segment four of liver-likely matastases. There was no lymphadenopathy. The pancreas, spleen, and biliary tract were normal (Figures 1).



Radiography and CT scan of chest did not reveal any mediastinal lymphadenopathy. Ultra sound guided FNAC showed morphology consistent with non Hodgkin's lymphoma. Ultrasound guided core needle biopsy further confirmed the diagnosis by demonstrating diffuse infiltrates and small-to-intermediate atypical cells consistent with lymphoma. Immunostaining of the tumour cells showed reactivity for CD20, CD5, CD45, and Bcl6 (figure 2).



The patient was diagnosed with primary large B-cell non Hodgkin lymphoma stage IE of liver, given that no additional foci of lymphoma were found anywhere else in the body. To date, the patient has received 2 cycles of cyclophosphamide–doxorubicin–vincristine–prednisone (CHOP).

Discussion

Although secondary liver involvement of lymphoma in the advanced stage is common, primary liver lymphoma (PLL), which is defined as lymphoma either confined to the liver or having major liver involvement, is extremely rare [3,4]. PLL constitutes 0.4% of cases of extranodal non-Hodgkins lymphoma, and comprises approximately 0.01% of non-Hodgkin's lymphomas [3].

The exact cause of PHL is still not clear. A number of recent reports showed a higher prevalence of HVC or HBV infection in PLL patients. Hepatitis C is found in 40% - 60% of patients with PHL; however, our patient was not positive for HCV or HBV. Other reports de-scribed that it might be associated with cirrhosis and immunosuppressive drugs.

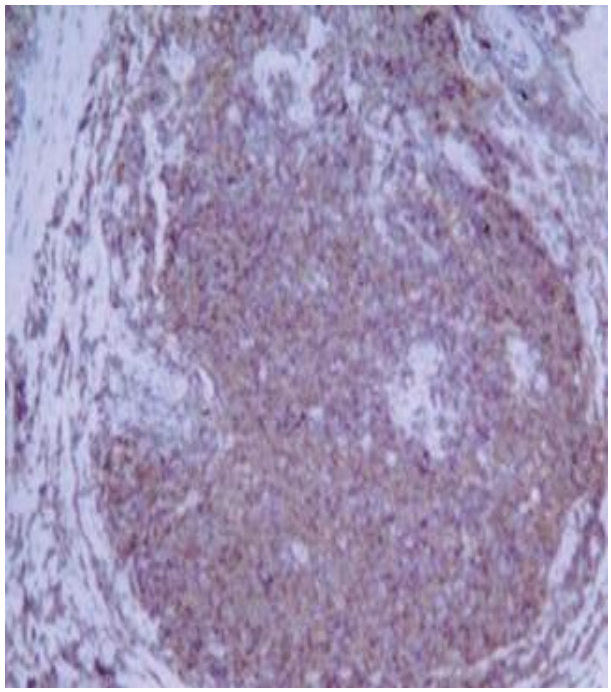
Clinical presentation of PHL is nonspecific. Most often fever, loss of weight and night sweats (also known as 'B' symptoms) occur. Alternative symptoms described are: right upper abdominal pain, epigastric pain, abdominal distension, nausea, vomiting, asthenia or itch. No specific physical complaints are typical for PHL. Abdominal pain, jaundice and hepatomegaly are the only physical findings described for various patients. Blood count can show abnormal aspartate aminotransferase, alanine aminotrans-ferase, alkaline phosphatase, total and direct bilirubin and LDH[5]. Hypercalcemia and Bence Jones protein peak are rare but have been described[5].

At initial presentation a third of patients present with a solitary liver nodule while another third have multiple lesions, and the remaining cases have diffuse infiltration of the liver[5]. Radiological investigation consists of an ultrasound of the liver, on which the tumor is hypo-attentuating or iso-attentuating[6]. On tri-phasic liver CT scan PHL usually presents itself as a hypodense lesion, with possible areas of inhomogeneity. Occasionally local areas of rim enhancement or calcifications may be seen[6].

The majority of PHL consist of B-cell NHL (63%) and T-cell lymphoma (25%)[8]. Diagnosis is often made upon histopathological investigation of the resection specimen. This is due to the hesitance in obtaining tissue biopsies of suspect liver lesions and risking needle track metastases [8]. Further differentiation can be done by immunohistochemical investigation [5,7]. Chemotherapy is the recommended therapeutic treatment for all extranodal diffuse large B-cell lymphoma and T-cell lymphoma, making it the treatment of choice when PHL is diagnosed preoperatively. Indications for surgical treatment are localized disease, which can be resected completely, or surgical debulking[8].

Conclusion

PLL confined to the liver without lymph node or bone marrow involvement is extremely rare. If the clinical picture is suspicious for PHL, a liver biopsy should be obtained, because the disease is treatable, and with new therapeutic drugs such as rituximab, overall survival has improved for these patients. The prognosis is variable, with good response to early aggressive combination chemotherapy.



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