Primary hepatic lymphomas are rare hepatic malignancies which are often misdiagnosed preoperatively. Early accurate diagnosis is essential as the patients can be treated successfully with chemotherapy, eluding the need for surgery. We present a case of primary hepatic lymphoma which mimicked as focal nodular hyperplasia with normal biochemical tumor markers, and $^{18}$F-fluorodeoxyglucose ($^{18}$F-FDG) whole-body positron emission magnetic resonance showed intense FDG uptake in the large hepatic lesion. The patient subsequently underwent right hepatectomy, and histopathology revealed diffuse large B cell lymphoma.

**Case History**

A 64-year-old female presented with complaints of epigastric discomfort, difficulty in eating, loss of appetite, and loss of weight. CT showed a large space-occupying lesion in the right lobe of the liver with central scar which was suspicious for FNH. AFP (alpha-fetoprotein), CEA (carcinoembryonic antigen), and CA 19-9 were within normal levels.

A dose of 6 mCi of $^{18}$F-FDG was administered intravenously to the patient in the fasting state. After 35 minutes of injection on a 3 Tesla Siemens Biograph mMR, image acquisition was started. $^{18}$F-FDG whole-body PET-magnetic resonance (MR) showed intense FDG uptake in large lobulated tumor in the right lobe of the liver. Central portions of the
Fig. 1 18F-FDG PET maximal intensity projection image showing large focus of increased abnormal uptake of FDG in the right lobe of liver. Physiological tracer uptake is seen in the brain, kidneys, and urinary bladder. Axial section of the liver on MR and fused PET-MR showing that bright central portions of the tumor are bright on T2-weighted images, consistent with the necrosis/scar with solid lesion in the periphery. MR, magnetic resonance; PET, positron emission tomography.

Fig. 2 Right hepatic mass in diffusion-weighted imaging, ADC map, postcontrast imaging, and T1-weighted volumetric interpolated breath-hold examination (VIBE). Postcontrast MR imaging shows washout of the solid components of the tumor in delayed phase and delayed enhancement of the central T2 bright scar tissue. MR, magnetic resonance.
tumor are bright on T2-weighted images and consistent with the necrosis/scar. Solid components in the periphery have a lobulated well-defined margin. Postcontrast imaging shows washout of the solid components of the tumor in the delayed phase and delayed enhancement of the central T2 bright scar tissue. Diffusion restriction was noted in the diffusion-weighed imaging and apparent diffusion coefficient (ADC) mapping. Fused imaging demonstrated intense FDG uptake in the peripheral solid components of the lesion. SUVmax of the lesion was 24. MRI features were suspicious for fibrolamellar carcinoma or FNH. As the lesion showed intense FDG uptake in the isolated liver lesion with no other FDG-avid lesions on whole-body PET-MR imaging, the possibility of primary hepatic malignancy was considered. Thus, the patient underwent right hepatectomy. The histopathology of the specimen revealed dyscohesive cellular neoplasm arranged in lobules, nests, cords, and trabeculae admixed with lymphocytes, eosinophils, and foamy macrophages with ingested debris. The neoplastic cells are positive for CD 20, Pax 5, CD 10, bcl 6, and bcl 2. Ki index greater than 90%. The diagnosis of diffuse large B cell lymphoma was confirmed. The patient was subsequently started on rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisone (R-CHOP) regimen of chemotherapy.

Discussion

PHL is defined as lymphoma confined to the liver without any involvement of other organs, or leukemic changes in the peripheral blood for at least 6 months after diagnosis. PHL represents 0.4% of extranodal lymphomas and 0.016% of NHLs. Although the pathogenesis of PHL is not well understood, it is usually seen in immunocompromised patients like organ-transplant recipients and individuals with acquired immune deficiency syndrome receiving immunosuppressive therapy. Recently, a strong relation between hepatitis C infection and PHL has also been identified, proposed cause being stimulation of B lymphocytes and chronic polyclonal proliferation, resulting in hepatic lymphoma.

PHL shows variable clinical presentation from incidental discovery in asymptomatic patients to nonspecific symptoms such as fatigue, night sweats, weight loss, evening rise of temperature, jaundice, abdominal pain, and hepatomegaly. Laboratory tests are also not helpful in diagnosis, with tumor markers like CEA, AFP, and CA 19.9 usually within normal ranges.

PHL may be Hodgkin’s or non-Hodgkin’s, with the latter being more common. B cell type is more common than the T cell type. Microscopically, PHL may show diffuse or nodular pattern of infiltration, both resulting in destruction of liver parenchyma. Among B cell types, diffuse large B cell type is the commonest followed by chronic lymphocytic lymphoma, Burkitt’s lymphoma, and mantle cell lymphoma. Similar to peripheral T cell NHL, NOS is common among T cell type, followed by anaplastic large cell lymphoma and hepatosplenic gamma delta lymphomas.

PHL can present as a solitary lesion, multiple lesions within the liver, or as diffuse hepatic infiltration. Radiological appearance of PHL is nonspecific and can resemble a hypovascular metastasis. Fungal and tubercular infections and inflammatory pathology like sarcoidosis may also show similar imaging appearances. PHLs are usually hypoechoic in ultrasonogram, hypointensating with minimal or no enhancement in CT, hypodense in T1-weighted and variable in T2-weighted sequences with similar pattern of enhancement in MRI. As a result, PHLs are rarely diagnosed preoperatively and commonly misdiagnosed as other malignant or benign lesions. In this case, the MRI appearance was highly suggestive of FNH or fibrolamellar variant of hepatoma. However, FDG avidity as well as absence of elevated tumor markers should have served as an alert, to consider an alternate diagnosis. Biopsy should be considered in such atypical cases for accurate diagnosis to plan appropriate treatment.

Surgical resection, chemotherapy, and radiotherapy alone or in combination are the various treatment modalities. The CHOP regimen is the standard treatment given for the patients with diffuse large B cell lymphoma. Rituximab targeting the CD20 antigenic marker can be added, which results in an improved response rate and prolonged overall survival. Liver resection followed by adjuvant chemotherapy and/or radiation has also been shown to have favorable short-term and mid-term outcomes with a prolonged survival rate.

The factors which determine the treatment outcome and prognosis are advanced age, poor performance status, bulky disease, unfavorable histology, elevated lactate dehydrogenase, comorbidities, and immunosuppressed state. Since PHL is chemosensitive, there is a distinct role of
chemotherapy as an adjuvant to surgery or as a primary treatment modality.

In conclusion, PHL is extremely rare with nonspecific clinical presentation and laboratory and imaging findings. This case demonstrates that this diagnosis should be considered in any patient with atypical presentation like FDG-avid liver mass with normal tumor markers. Definite diagnosis and timely therapeutic intervention can produce better outcome in these patients than other liver tumors owing to their chemosensitivity.

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Conflict of Interest
None declared.

References