Case Report

A rare tumor with unusual clinical presentation detected by positron emission tomography-computed tomography

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ABSTRACT

Primary hepatic lymphoma represents <1% of extranodal lymphoma and predominantly seen in men older than 50 years of age. Exact etiology for these tumors is not certain yet, but presumed to be caused by certain viruses like Epstein-Barr virus and hepatitis C virus due to the frequent association of these viruses with disease. Most of these tumors are diffuse large B cell non-Hodgkin lymphoma. Direct tissue histopathology with immunochemistry may give clues about diagnosis and prognosis up to certain extent. The rituximab-based chemotherapy is the mainstay of therapy for these tumors; the role of radiotherapy is still not clear but used for management for bulky tumors.

Keywords: High-grade B cell lymphoma, positron emission tomography-computed tomography, primary hepatic lymphoma

INTRODUCTION

Primary hepatic lymphoma (PHL) is one of the rarely encountered malignant tumors of the liver and a rare form of extranodal lymphoma. The disease is more common among men and presents with vague symptoms. Traditional tumor markers for hepatocellular carcinoma are generally negative for PHL and histopathology with immunohistochemistry is the only way to prove disease and may be helpful in predicting prognosis. We are presenting a case of 77-year-old man presented with left flank pain and conventional imaging failed to clinch the diagnosis; however, positron emission tomography-computed tomography (PET-CT) paved a clue to diagnose the high-grade B cell PHL.

CASE REPORT

A 77-year-old man admitted to our hospital with pain on the left flank region, moderate intensity, and nonradiating colicky in nature. The patient did not mention any fever, hematuria, night sweats, vomiting, chest pain, abdominal pain, diarrhea, blood in stools, jaundice, or weight loss. His past medical history was significant for arterial hypertension, coronary artery disease, diabetes mellitus, and hypothyroidism. On physical examination, he had pallor, mild epigastric, and lumber tenderness. His complete blood counts were normal apart from mild anemia with hemoglobin of 9.8 g%, and peripheral blood smear was showing features of microcytic hypochromic anemia. His liver function tests were total bilirubin: 1 mg/dl, aspartate aminotransferase: 94 U/L, alanine aminotransferase: 58 U/L, gamma-glutamyl transferase: 44 U/L, alkaline phosphatase: 434 U/L, total protein: 6.0 mg/dl, and albumin: 1.8 g/dl. Lactate dehydrogenase (LDH) level was found to be 1266 U/L. His prothrombin time was 19.2 s and international normalized ratio (INR): 1.98. His serology tests for hepatitis B virus, hepatitis C virus (HCV), and human immunodeficiency virus were negative. His stool for occult blood was negative. The results of the iron profile were serum ferritin: 1495 ng/ml, serum iron: 14 μg/dl, transferrin saturation: 9.3%, and total iron binding capacity: 150 μg/dl. His esophagogastroduodenoscopy, capsule endoscopy, and

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colonoscopy were normal. During hospital stay, he developed the complaint of epistaxis which managed by conservative means but INR failed to come down despite transfusion of Vitamin K and fresh frozen plasma. Ultrasonography (U/S) of abdomen did not reveal any hepatic mass lesion or chronic liver disease changes but shown hepatosplenomegaly. His contrast-enhanced CT scan of the abdomen showed multiple splenic hypodense areas mimicking splenic infarcts likely due to a mycotic aneurysm. PET-CT was ordered in view of inconclusive diagnostic clues which revealed homogenous increased hepatic fluorodeoxyglucose (FDG) uptake with splenic hypoattenuating lesions, and no other site increased uptake was noted on PET [Figure 1]. The transjugular liver biopsy was obtained for high suspicion of infiltrating liver disorders which revealed dense lymphoplasmacytic infiltrate, high-grade B cell non-Hodgkin lymphoma (NHL) CD20+/CD5+/CD38−, no activity for Tdt or cyclin D1 with CK7+/CK20 − and high Ki-67 index (approximately 80% cells) on biopsy specimen [Figure 2]. Bone marrow aspiration and biopsy examination showed no evidence of lymphoma. Cerebrospinal fluid examination was normal. The diagnosis of primary large cells NHL was made. The patient received R-CHOP treatment (cyclophosphamide, doxorubicin, vincristine, prednisone, and rituximab) and was on regular follow-up of medical oncology.

DISCUSSION

PHL is unusual and rare extranodal lymphoma, accounting for <1% of all extranodal lymphomas.[1] Etiology for PHL is not certain yet, but its association with HCV seen occasionally and thought to be one of the implicating factors.[2] PHL usually presents with constitutional symptoms, hepatomegaly, and signs of cholestatic jaundice. However, disease spectrum varies from nonspecific constitutional symptoms to acute liver failure, coma, and death.[3] Depending on lymphoid cell infiltration into the liver, PHL is subdivided into nodular and diffuse variants. In literature, nodular variants of PHL present as a hypoechoic mass on U/S and a hypoattenuating lesion on CT scan. Contrast injection leads to rim enhancement of the lesion and on FDG-PET nodular lesions present with more FDG uptake than the surrounding parenchyma.[4] However, diffuse variant of PHL is difficult to diagnose on imaging modalities like U/S and CT scan that will show only hepatic enlargement without any abnormal enhancing pattern, while FGD-PET will show diffuse increased avidity of hepatic parenchyma as demonstrated in our case report.

Most PHLs are the diffuse large B cell type, other major subtypes include: Lymphoblastic lymphoma, Burkitt's lymphoma, and follicular lymphoma.[5] Elevated LDH may be a clue for diagnosis, but traditional tumor markers for hepatocellular carcinoma are negative in this context. Apparently, there is no definitive diagnostic imaging modality to confirm the diagnosis; tissue histopathology is the only means to prove the diagnosis, and immunohistochemistry is useful to predict the prognosis.[6] Chemotherapy with R-CHOP prolongs survival with minimal toxicity.[7] Advanced age, bulky disease, high proliferative index, elevated LDH, β2-microalbumin, B cell lymphoma-2 expression, and cirrhosis of liver indicate poor prognosis in these patients.[8]

CONCLUSION

PHL is a rarely encountered extranodal lymphoma where traditional tumor markers and radio imaging are not fair enough to prove disease as opposed to primary hepatocellular carcinoma. However, PET-CT imaging technique helped by adding the precision of anatomic localization to functional imaging, on the basis of high degree of suspicion of disease, which was previously lacking from pure CT. Hence, PET/CT is useful in making the diagnosis by excluding extrahepatic sites of lymphoma with high degree of sensitivity, can also be used for staging and to assess posttreatment response by complete resolution of FDG uptake. Tissue histology and immunohistochemistry are helpful in fulfilling the diagnosis, predicting the prognosis up to some extent, and high clinical suspicion, and vigilance is warranted, due to nonspecific and vague disease presentation.

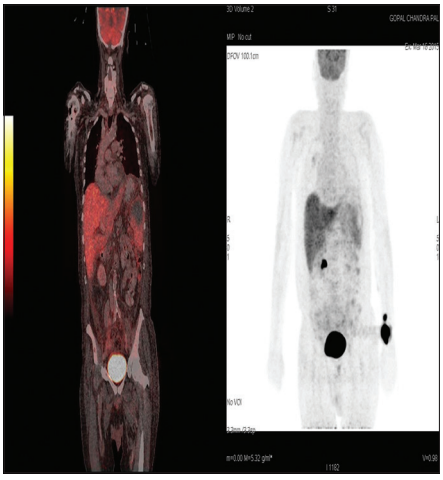

Figure 1: Fluorodeoxyglucose-positron emission tomography scan showing diffuse hepatic glucose uptake without any other site increased uptake

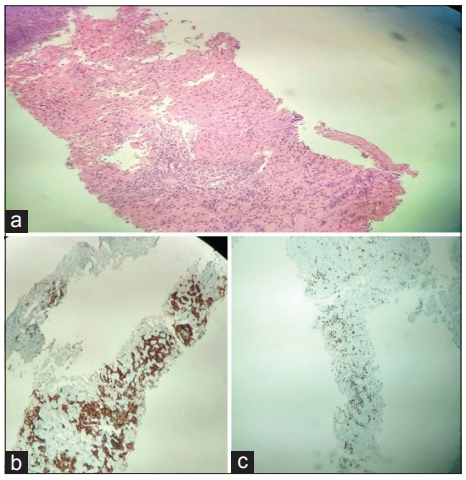

Figure 2: (a) H and E staining showing dense lymphocytic infiltrate. (b) Flow cytometry showing high CD20 positivity of cells. (c) Ki-67 activity on infiltrating cells

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Conflicts of interest

There are no conflicts of interest.

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