Case Report

Primary hepatic lymphoma mimicking hepatocellular carcinoma: case report and review of literature

Digvijoy Sharma*, Gandadhar Rao Gondu, Nirjhar Raj, Suryaramachandra Varma Gunturi, Venu Madhav Thumma, Kunduru Navakishore, Jagan Mohan Reddy, Nagari Bheerappa

Department of Surgical Gastroenterology, Nizams Institute of Medical Sciences, Hyderabad, Telangana, India

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*Correspondence:

Dr. Digvijoy Sharma,

E-mail: digz.sarma@yahoo.in

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ABSTRACT

Primary hepatic lymphoma (PHL) is a very rare malignancy presenting with non specific symptoms leading to late diagnosis. Due to its ability to masquerade other liver tumors, misdiagnosis is common. Here we present a case of a 52 years old man who presented with an incidentally detected SOL in liver with features of atypical hepatocellular carcinoma on imaging and fine needle aspiration cytology. He underwent a left hepatectomy and the final histology was a surprise in the form of hepatic lymphoma. He underwent appropriate chemotherapy post surgery and is asymptomatic at follow up.

Keywords: Chemotherapy, Primary hepatic lymphoma, Surgery

INTRODUCTION

Primary Hepatic Lymphoma(PHL) is one of the rare primary tumors of liver that constitutes about 0.016% of Non Hodgkin's Lymphoma and 0.4% of extranodal lymphomas.¹ The etiopathogenesis, features and natural history is not clearly understood.^{2,3} The published literature has not been able to establish consensus on the exact definition of PHL.⁴⁻⁶ However Lei defined the following characteristics for diagnosis of disease: 1) Clinical signs and symptoms due to liver involvement 2) No evidence of distant lymphadenopathy (clinical or radiological) 3) Peripheral blood smear showing absence of leukemic picture.⁷ Prognosis of PHL is not promising with high recurrence rate and low survival rate. Herein we report this case for its rarity and ability to masquerade hepatocellular carcinoma.

CASE REPORT

A 52 year old man was under evaluation for urethral stricture with urinary tract infection when he was

incidentally found to have a space occupying lesion in left lobe of liver. There was no history suggestive of chronic liver disease, though he was a chronic alcoholic. His routine blood investigations including liver function tests and viral markers were within normal limits. Tumor markers, AFP and CA19-9 were normal. He was further evaluated by triphasic CECT abdomen to characterise the mass, which showed a 7x9 cm predominantly hypodense lesion in segment 4A and 2 of liver with faint enhancement in arterial and portovenous phase with no wash out of contrast (Figure 1).

Figure 1 : CECT abdomen showing hypodense lesion in left lobe of liver with faint peripheral enhancement in portovenous phase.

Since AFP was normal and CECT could not provide a concrete diagnosis, image guided FNAC from the lesion was done which showed presence of malignant cells. Hence a diagnosis of atypical hepatocellular carcinoma was made and patient was planned for a major hepatectomy after assessment of fitness for the same. Intraoperatively the tumor was located in segment 2, 3 and 4A of liver with no evidence of extrahepatic disease (Figure 2) He underwent left hepatectomy.

Figure 2: SOL (black arrow) in segment 2, 3, 4A of liver. Line of demarcation (white arrow) after inflow control.

Figure 3 : Uniform population of lymphoid cells of large size with mitotic figures.

Intraoperative and postoperative periods were uneventful. Final histopathology revealed T cell histiocyte rich B cell lymphoma positive for CD3, CD20 and LCA (Figure 3). Patient was referred to medical oncology where he underwent PET-CT which ruled out presence of nodal and other extranodal disease, hence a final diagnosis of primary hepatic lymphoma was established. He received chemotherapy in the form of CHOP-R regimen. Patient was symptom free on follow up at 9 months post surgery.

DISCUSSION

The literature on PHL is sparse due to less number of cases reported. This could probably be due to ambiguous presentation and late diagnosis or more often misdiagnosis. The disease is believed to be more prevalent in males in fifth decade. Right upper abdominal pain is the most frequent complaint. Fatigue, weight loss, jaundice, anorexia, nausea, malaise, night sweats, and vomiting may be present.⁸ About 50 % present with hepatomegaly.⁹

Proposed etiological factors include preexisting chronic hepatitis, cirrhosis, Epstein Barr Virus (EBV), Hepatitis B and C and those on immunosuppressives.¹⁰⁻¹⁴ Radiologically predominantly hypodense solitary lesion is the most common presentation, although multiple lesions and diffuse infiltrative forms with variable enhancement patterns are known.² In the present case report the tumor presented as a solitary lesion with faint enhancement and FNAC suggestive of hepatocellular carcinoma, that led to the misdiagnosis. Lactate dehydrogenase, alkaline phosphatase and bilirubin levels are usually elevated while alpha fetoprotein and carcinoembryonic antigen typically remain normal.¹⁵

Pathologically most PHL are diffuse large B cell type (46-68%), less than 5% of cases include diffuse mixed large and small cell, diffuse immunoblastic, lymphoblastic and small non-cleaved or Burkitt lymphoma.^{2,5,9} Imaging helps in identifying the extent of the lesion and assess the respectability. Ultrasound or CT followed by guided biopsy is the usual diagnostic course. MRI though helpful is generally not required.¹⁹ Immunohistochemistry (CD10, BCL6 and MUM1) is indicated to distinguish germinal center type from activated B cell type.²⁰

Optimal therapy is not standardized, however a combination of chemotherapy, radiotherapy and surgery in selected cases is often employed.² Chemotherapy is the treatment of choice for PHL in general. CHOP regimen (cyclophosphamide, doxorubicin, vincristine, and prednisone) is followed. Addition of rituximab (monoclonal antibody against CD-20) prolongs survival with minimal toxicity (R-CHOP).¹⁶ Surgery has shown favorable results in localized resectable tumors.¹⁷ Previously surgery was also indicated prior to chemotherapy for debulking of the growth.¹⁵ Adjuvant radiotherapy has also been used in a few cases.¹⁸

Poor prognostic factors include extrahepatic disease, higher mitotic rate, elevated liver enzymes, advanced age at presentation, comorbidities. The median survival ranges from 3-123.6 months.¹⁵ Low haemoglobin levels and absence of fever are considered independent adverse prognostic factors.

CONCLUSION

Primary hepatic lymphoma is amenable to therapy, hence better diagnostic modalities for early diagnosis are warranted. Meanwhile a high index of clinical suspicion would go a long way till more standardised guidelines are formulated.

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