

Primary liver lymphoma (PLL) with isolated CNS relapse

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ABSTRACT

Background: Primary liver lymphoma (PLL) is a rare disease. Patients usually present with abdominal problems and liver mass without any extrahepatic tumor. PLL usually responds well to chemotherapy, while chemotherapy±radiation is the standard treatment of choice.

Case report: A 42 year-old man was presented because of abdominal pain, 8.5kg weight loss and fever for 3 months. There was not lymphadenopathy, splenomegaly, abdominal mass or hepatomegaly in physical examination. A computed tomography (CT) scan of the abdomen disclosed multiple large liver masses without adenopathy. Patient responded well to CHOP chemotherapy regimen. Three months later, he was visited because of headache, dizziness and convulsion. A brain CT scan showed a mass in brain. He was treated by brain radiation plus intrathecal injection of methotrexate (MTX) (15mg) followed by 6 cycles of ICE protocol.

Conclusion: PLL is a rare disease that responds well to multi-agent chemotherapy. CNS metastasis should be considered in the affected patients.

Keywords: Primary liver lymphoma, CNS relapse, Chemotherapy.
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INTRODUCTION

Primary liver lymphoma (PLL) is a rare disease and only 0.016% of all cases of non-Hodgkin lymphoma are PLL (1). It was first reported by Ata and Kamal in 1965 (2). Totally, 251 cases of PLL have been described until 2003 (3). The most common presenting symptom is abdominal pain or discomfort, which occurs in 39–70% of patients. Other presenting symptoms include fatigue, jaundice, anorexia, malaise, nausea, and vomiting (4-7).

Lei defined the following criteria for diagnosis of PLL: 1- At the time of disease presentation, the patient symptoms are caused mainly by the liver involvement; 2- There is an absence of palpable lymphadenopathy, and no radiologic evidence of distant lymphadenopathy; 3- There is an absence of leukemic blood involvement in the peripheral blood smear (4).

Due to its infrequent occurrence, the disease is poorly understood and few clinical studies have been conducted to elucidate the natural course of disease, aetiology, optimal therapy, response to therapy and survival (3). The median survival for all patients is 15.3 months, however, it varies widely and reported survival ranges from 3 to 123 months (5).

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Patient was treated by CHOP (cyclophosphamide, Adriamycin, Vincristin, Prednisolon) chemotherapy regimen every 3 weeks and responded well to this regimen. After 6 cycles of chemotherapy, abdominal CT scan was repeated and showed only a small mass in liver. Three months later, he was visited because of headache, dizziness and convulsion. A brain CT scan showed a mass in brain (figure 3). CSF examination was positive for lymphoma cells. He was treated by brain radiation plus intrathecal injection of methotrexate (MTX) (15mg) followed by 6 cycles of ICE (Ifosfamide, Carboplatin, Etoposide) protocol. He was followed for 3 months after the last chemotherapy.

Seventeen months later, liver sonography was normal without mass and physical examination was unremarkable. He enjoys normal life thereafter.

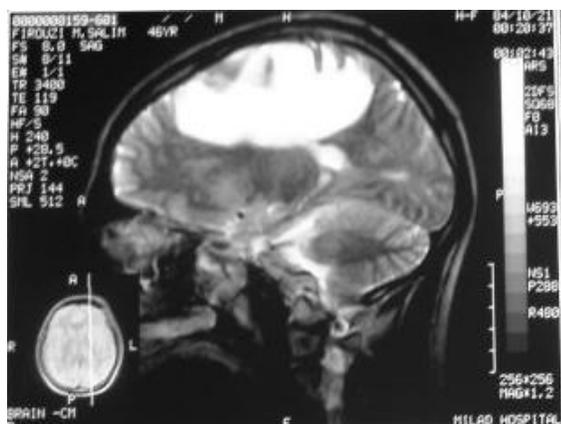


Figure 3. Brain metastasis in patient with primary liver lymphoma

DISCUSSION

PLL occurs in a wide range of ages (17 to 84 years) and has been reported mainly in male patients (2). It is usually B-cell type.

Due to the rarity of this disease entity, the non-specific clinical presentation, laboratory, and radiologic features, PLL may be confused with hepatitis, primary hepatic tumors, carcinoma with hepatic metastases, and systemic lymphoma with

secondary hepatic involvement. The pathologic appearance of PLL may also be misleading, and initial misdiagnoses of poorly differentiated carcinoma, embryonal sarcoma, granulomatous cholangitis, inflammatory pseudo-tumor and granulomatous hepatitis have been reported (5). Our case was also confirmed by IHC.

Treatment options for PLL include surgery, chemotherapy, radiation or varying combinations of these modalities (3). Page et al. performed a retrospective cohort review of 24 patients treated at MD Anderson Cancer Center between 1974 and 1995. They noted an overall complete remission rate of 83.3% (100% of patients treated with alternating triple therapy) and a 5-year relapse-free survival rate of 83.1% (6). In contrast to the exceedingly favorable results of chemotherapy noted by Page et al., other authors have not had similar experiences (3).

Thus, it seems reasonable to treat these patients first with systemic chemotherapy. If disease persists or only partially regresses in the liver, surgical resection can be advised if no evidence of extra hepatic involvement exists.

Relapse, as an isolated CNS involvement, is unique for our patient since to our knowledge, no other cases have been reported yet.

Our patient responded well to brain radiation and intrathecal MTX as well as systemic chemotherapy.

Although our patient was HCV Ab negative but a number of recent reports have described an increased incidence of PLL in patients with hepatitis C virus (HCV) infection (3).

In conclusion, PLL is a rare disease that responds well to multi-agent chemotherapy. CNS metastasis should be considered in the affected patients.

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