Primary Hepatic Peripheral T-Cell Lymphoma Treated with Corticosteroid

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Abstract

Primary hepatic lymphoma is a very rare condition, and the majority of the cases reported are of B-cell origin. We report a case of a 65-year-old man with primary hepatic peripheral T-cell lymphoma, not otherwise specified (PTCL-nos) who presented with 15% weight loss and general fatigue over the previous 9 months. Imaging studies and bone marrow examination could not confirm a diagnosis of lymphoma. Liver biopsy was performed because of an elevated soluble interleukin-2 receptor (sIL-2R) level (17,000 U/I) and hepatomegaly. After the diagnosis of primary hepatic PTCL-nos, treatment with low-dose corticosteroid was initiated, and the sIL-2R level decreased. Discontinuation of corticosteroid treatment resulted in the re-elevation of the sIL-2R level, and subsequently, treatment with low-dose corticosteroid was reinitiated. The sIL-2R level decreased rapidly, and the patient is alive with no evidence of lymphoma for 50 months after diagnosis. Thus, we found that a low-dose corticosteroid was effective in the long-term control of the disease, whereas many previous studies reported that primary hepatic PTCL-nos has a poor prognosis.

Key words: primary hepatic lymphoma, T-cell lymphoma, sIL-2R, low-dose corticosteroid

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Introduction

Primary hepatic lymphoma (PHL) comprises only approximately 0.016% of all cases of non-Hodgkin’s lymphoma (1) in spite of the fact that the liver is a common site for secondary extranodal lymphoma in advanced non-Hodgkin’s lymphoma. In the majority of the cases of PHL originates in B cells, and T-cell lymphoma is less common as only a few cases of primary hepatic T-cell lymphoma have been reported (2). Multi-agent chemotherapy was applied in most of the cases with a confirmed pathological diagnosis of lymphoma. We report a case of primary hepatic peripheral T-cell lymphoma, not otherwise specified (PTCL-nos) treated with corticosteroid alone.

Case Report

In 2005, a 65-year-old man presented with 15% weight loss and general fatigue experienced in the previous 9 months. He had a history of chronic alcoholism, and a prior medical history of acute pancreatitis and idiopathic osteonecrosis of the femoral head. A physical examination revealed hepatomegaly, palpable 3 cm below the right costal margin without lymphadenopathy or splenomegaly. The results of the complete blood count were normal apart from mild leukocytosis (white blood cell count, 11,700/μL). The results of the liver function tests were normal. Lactate dehydrogenase (LDH) level was found to be within the normal range. The level of soluble interleukin-2 receptor (sIL-2R) was elevated to 17,000 U/I. The results of the liver function tests were normal. Lactate dehydrogenase (LDH) level was found to be within the normal range. The level of soluble interleukin-2 receptor (sIL-2R) was elevated to 17,000 U/I. The results of the serologic tests for hepatitis B virus, hepatitis C virus, human immunodeficiency virus, and human T-lymphotropic virus-I were negative. A computed tomography scan of the abdomen demonstrated mild hepatosplenomegaly without intra-abdominal lymphadenopathy. Fluorine-18 fluorodeoxyglucose positron emission tomography showed no abnormal uptake in the entire body. Bone marrow aspiration and biopsy examination showed no
Figure 1. Pathologic examination. A photomicrograph of the liver biopsy specimen shows (a) infiltration of small- and intermediate-sized lymphoid cells (Hematoxylin and Eosin staining, 200×); (b) tumor cells positive for CD3 (CD3 immunohistochemistry, 200×); (c) tumor cells positive for CD8 (CD8 immunohistochemistry, 200×); and (d) tumor cells positive for TIA-1 (TIA-1 immunohistochemistry, 300×).

evidence of lymphoma.

An ultrasonography-guided needle liver biopsy was performed to evaluate the possibility of intravascular lymphoma. The liver specimen showed an infiltration of small- and intermediate-sized lymphoid cells with irregular nuclear contours (Fig. 1a). Immunohistochemical studies showed that the abnormal lymphoid cells were positive for CD3, CD8, and TIA-1 (Fig. 1b, c, and d); negative for CD4, CD20, CD56, and Epstein-Barr virus-encoded small RNA (EBER); and the MIB-1 index was only 9%. Thus, the diagnosis of PTCL-nos was established. Later we analyzed the rearrangement of TCRγ gene by polymerase chain reaction in the paraffin-embedded specimen of the liver and detected a monoclonal band. This supported the diagnosis of T-cell lymphoma.

The patient was treated with 20 mg of oral prednisolone daily because his general condition was in Eastern Cooperative Oncology Group performance status (PS) 3 due to lymphoma and accompanying leg edema. He was presumed as intolerable to chemotherapy. The administration of prednisolone resulted in a rapid decrease in the sIL-2R level. His PS and hepatomegaly improved in a relatively rapid manner. On reducing the dose of prednisolone, the level of sIL-2R returned to within the normal range (145-519/μL). Subsequently, the patient was maintained on 5 mg of oral prednisolone because the discontinuation of prednisolone resulted in the immediate re-elevation of the sIL-2R level (8,590/μL). He had a normal sIL-2R level and no other evidence of lymphoma at 50 months after the diagnosis (Fig. 2). We obtained his informed consent for this article publication.

Discussion

In the present case, multi-agent chemotherapy could not be applied due to his poor general condition. Treatment with low-dose corticosteroid resulted in an improvement of his general condition with a decrease in the level of sIL-2R. There were no tumor markers to assess lymphoma other than sIL-2R. LDH level was not elevated throughout the clinical course. Because the standard strategy for PHL was not established at that time, we did not apply chemotherapy after prednisolone treatment decreased the sIL-2R level. Liver biopsy was used for an accurate diagnosis of lymphoma, and a good clinical course was observed. This indolent clinical course corresponded to the low MIB-1 index of the liver specimen.

The median survival time of patients with PHL is reported to be 15 months; however, it varies widely and ranges from 3 to 124 months (2). Thus far, the prognosis of PHL has not been assessed on the basis of its histologic origin. The correlation between the pattern of liver involvement and the prognosis has been reported by Emile et al (3).
They classified patients with PHL into 2 groups depending on the type of liver involvement: nodular or diffuse. The 1- and 3-year survival rates of the nodular type were 70% and 57%, respectively, and those of the diffuse type were 38% and 18%, respectively. This difference was statistically significant (P-value = 0.003). The present case was considered to have a poor prognosis as this case corresponded to the diffuse type by histological analysis.

Stancu et al reviewed 14 cases of primary hepatic T-cell lymphoma; of these, they encountered 3 cases, and 11 cases...
had been reported previously (4). They reported that primary hepatic T-cell lymphoma has a poor prognosis since an accurate diagnosis of hepatic T-cell lymphoma before death is sometimes difficult because of its aggressive clinical course.

We collected 19 cases of primary hepatic T-cell lymphoma reported previously (4-13)(Table 1). As for clinical manifestations, all of them presented with hepatomegaly. Fever occurred in 9 cases, weight loss occurred in 9 cases, and night sweats occurred in 3 cases. Abnormal dysfunction was observed in 14 of 17 described cases. In the histological analysis, 17 of 19 cases showed diffuse liver infiltration and the remaining 2 cases showed nodular liver infiltration. Of these 19 patients, 8 underwent chemotherapy; 3 received CHOP therapy (cyclophosphamide, anthracycline, vincristine, and prednisone); 1 received CHOP and DHAP therapy (dexamethasone, cytarabine, and cisplatin); 1 received COP therapy (cyclophosphamide, vincristine, and prednisolone); and data were not available for 3 patients. Of the remaining 11 patients, 5 were treated with a corticosteroid, 4 received only supportive therapy, 1 underwent hepatectomy, and data was not available for 1 patient. It is difficult to establish a generalized effective treatment for primary hepatic T-cell lymphoma because of the varying follow-up durations used in previous studies.

To our knowledge, thus far, a corticosteroid has not been used as the only treatment for primary hepatic T-cell lymphoma. This case suggests the efficacy of a corticosteroid in the treatment, however, we do not highly recommend the monotherapy of corticosteroid for patients with primary hepatic T-cell lymphoma based only on the evidence of this case. Further accumulation of the evidence of treatment by corticosteroid is needed.

The authors state that they have no Conflict of Interest (COI).

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References