Abstract We report on a case of CD20-positive peripheral T cell lymphoma. The lymphoma cell was positive for CD20 and T cell lineage markers such as cytoplasmic CD3, CD4, and CD5 and had a monoclonal rearrangement of the T cell receptor (TCR) γ chain gene. The clinical characteristics resembled angioimmunoblastic lymphadenopathy: spontaneous regression of lymphadenopathy and immunological abnormalities such as polyclonal hypergammaglobulinemia, positive results of direct and indirect antiglobulin tests, and a high antinuclear antibody titer. We reviewed seven cases of CD20-positive T cell malignancies including the present case. Three were immature T cell malignancies (acute lymphoblastic leukemia) and four were peripheral T cell malignancies (non-Hodgkin’s lymphoma and chronic lymphocytic leukemia). Hepatomegaly and/or splenomegaly were common features. Further cases must be evaluated to understand the clinical significance of the CD20 expression on the surface of T cell malignancies.

Keywords CD20 antigens · T cell lymphoma · T cell leukemia

Introduction

The CD20 molecule, with a molecular weight of 33–37 kDa, is a membrane protein appearing relatively late in B cell maturation, which is lost before the plasma cell stage in B cell ontogeny [12]. CD20 is classified as a pan-B cell marker and believed to be restricted to B cells and follicular dendritic cells [2]. However, it was recently reported that a small population of normal T cells in the peripheral blood or bone marrow cells expressed CD20 [1, 4, 10]. Moreover, rare cases of CD20-positive T cell malignancies have been reported [8, 10, 11, 14]. The biological and clinical significance of CD20 positivity in a T cell malignancy is unknown. We report on a case of CD20-positive peripheral T cell lymphoma and review the literature on CD20-positive T cell malignancies.

Case report

In August 1996, a 60-year-old man presented with dyspnea and systemic lymphadenopathy (axillary, cervical, and inguinal). Laboratory investigations revealed elevated serum levels of lactate dehydrogenase (LDH) (764 U/l) and hypoxemia (PO2, 65.4 mmHg), but plain X-rays and computed tomography (CT) of the chest were unremarkable. He was treated with antibiotics and theophylline, and his dyspnea and lymphadenopathy disappeared. Three months later, systemic lymphadenopathy recurred. The pathology of a biopsied specimen of the lymph node suggested non-Hodgkin’s lymphoma (NHL), but reactive lymphadenopathy was not completely ruled out. His lymphadenopathy disappeared again without any treatment. In April 1997, he developed systemic lymphadenopathy again accompanied with fatigue, lumbago, and abdominal pain. The liver was enlarged 5 cm below the right costal margin, and the spleen was 4 cm below the left costal margin. Laboratory investigations revealed a normal complete blood cell count but elevated serum levels of LDH (862 U/l), IgG (2,883 mg/dl), IgA (428 mg/dl), IgM (836 mg/dl), soluble interleukin-2 receptor (sIL-2R) (23,000 U/ml), and antinuclear antibody. The direct and indirect antiglobulin tests were positive, but apparent hemolysis was not detected.

A biopsy of a cervical node led to the diagnosis of NHL (see next section). Mediastinal and abdominal para-aortic nodes and bone marrow were also involved with the lymphoma. He received six courses of CHOP [cyclophosphamide, doxorubicin, vincristine, and prednisolone (PSL)] therapy and attained complete remission in August 1997.

In January 1998, his lymphoma relapsed, which was complicated with autoimmune hemolytic anemia (AIHA). He received an IMVP-16 (ifosfamide, methotrexate, etoposide) regimen. Although his nodal lesions responded well to therapy, he developed meningeal infiltration of the lymphoma, which was successfully
treated with intracranial chemotherapy, and muscle weakness in the limbs, which was diagnosed as peripheral motor neuropathy by an electrodiagnostic examination. He died due to leukemic transformation of the lymphoma in August 1998.

Pathological findings

The histological findings of the lymph node at the diagnosis are shown in Fig. 1 (a-c). There were diffuse infiltrates of mediumsized lymphoid cells. Scattered large-sized cells such as immunoblasts or centroblasts were also present. Mitotic figures were occasionally observed. These cells were positive not only for CD3 (cytoplasmic CD3ε) and CD45RO (UCHL-1) but also CD20 (L26) using immunohistochemical staining of paraffin-embedded sections.

Immunophenotypic findings by flow cytometric analysis are shown in Table 2. Dual fluorescence analysis of lymphoma cells from a lymph node, cerebrospinal fluid, and peripheral blood (more than 90% was lymphoma cells) showed that the cell surfaces of the malignant cells were positive for CD4, CD5, and CD20 (Leu16) and negative for CD3, CD19, CD22, and surface immunoglobulin (Fig. 2). Genotypic analysis showed a clonal rearrangement for the T cell receptor (TCR) γ chain gene but not for immunoglobulin (Ig) heavy-chain (J H and C µ ) genes and the Ig light-chain (J κ) gene. These findings were consistent with the diagnosis of NHL (peripheral T cell lymphoma, unspecified), except for CD20 positivity. Chromosomal analysis of the lymph node biopsy specimen showed that 9 of 13 cells had 51, XY, X, t(1;12) (p32; q21), +3, +4, +14, add(15)(q26), +17 karyotype.

Literature review

Our literature review identified six well-documented cases with CD20-positive T cell malignancies [three acute lymphocytic leukemia (ALL), two NHL, and one chronic lymphocytic leukemia (CLL)] [8, 10, 11, 14]. The clinical characteristics of these six patients and our patient are summarized in Table 1. The physical findings were described in six of seven patients, four of whom had hepatosplenomegaly and one splenomegaly. We could not find any other clinical findings in common among these patients. Immunophenotypic data showed that all patients exhibited typical T cell phenotype except for CD20 positivity (Table 2). Clonal rearrangements were detected with probes in the TCR β or TCR γ chain genes in three of four patients examined (Table 3).