Conference Case

Intradural extramedullary relapse of peripheral T-cell lymphoma, NOS

Keywords: Intradural extramedullary relapse; peripheral T cell lymphoma; romidepsin

CASE REPORT

A 73-year-old Japanese man was referred to our clinic with general malaise. He had a history of diabetes mellitus and hypertension of longer than 10 years. He presented with lymphadenopathy and liver dysfunction. Laboratory examination demonstrated a WBC count of 23.6×109/L with 10% abnormal lymphocytes having basophilic irregular nuclei, aspartate aminotransferase of 124 U/L (normal range: 5–40 U/L), alanine transaminase of 166 U/L (5–35 U/L), total bilirubin of 4.1 mg/dL (1.0–0.3 mg/dL), creatinine of 1.46 mg/dL (0.6–1.1 mg/dL) and soluble interleukin-2 receptor (sIL-2R) of 15,889 U/mL (122–496 U/mL). Serological tests for HBV, HCV and HTLV-1 were all negative. The physical examination revealed bilateral cervical lymph node swelling, multiple abdominal skin pigmentation and peripheral edema. On computed tomography (CT) of the trunk, generalized multiple lymphadenopathy with mild splenomegaly was noted. Biopsy specimens of cervical lymph nodes and abdominal skin exhibited monotonous infiltration of medium to large-sized lymphocytes with a phenotype of CD3+, CD5+, CD10–, CD20–, CD79a–, CD30–, CD56–, Bcl6–, granzyme B–, CD45RO–, CCR4+ and TdT–. (Figure 1A–E) The Ki-67 labeling index was 80%. EBER in situ hybridization was negative. Lymphoma cells were also detected in the bone marrow. The prognostic index score for T-cell lymphoma in this case was 4, considered to be high risk.1 The final diagnosis was PTCL, NOS, stage IVB. We treated the patient using modified CHOP therapy. After one cycle of chemotherapy, the swelled lymph node shrunken and partial response was achieved. However, on day 13 after the modified CHOP therapy, his general condition deteriorated and the WBC increased to 9.6×109/L with 36% lymphoma cells. He was treated with intensified CHOP therapy, his general condition deteriorated and the response was achieved. However, on day 13 after the modified CHOP therapy, the swelled lymph node shrunk and partial response was achieved. When the sIL-2R levels decreased to 1,423 U/mL, the patient recovered rapidly. His sIL-2R levels decreased to 1,428 U/mL. When the WBC count recovered to 7.6×109/L 17 days later, 8% lymphoma cells persisted in the peripheral blood and one cycle of the monoclonal antibody mogamulizumab (1 mg/kg for every 4 weeks) was added. He received a second cycle of romidepsin, and the disappearance of lymphoma cells from the peripheral blood and all lymph node swelling was confirmed. On day 7 after the second cycle of romidepsin, the patient suddenly complained of severe lumbago with bilateral weakness of the lower limbs. Initial MRI of the entire spine detected no abnormalities. CT demonstrated complete remission of the lymphadenopathy. His sIL-2R value was stable at 1,423 U/mL. When the physical examination revealed bilateral cervical lymph node swelling, multiple abdominal skin pigmentation and peripheral edema. On computed tomography (CT) of the trunk, generalized multiple lymphadenopathy with mild splenomegaly was noted. Biopsy specimens of cervical lymph nodes and abdominal skin exhibited monotonous infiltration of medium to large-sized lymphocytes with a phenotype of CD3+, CD5+, CD10–, CD20–, CD79a–, CD30–, CD56–, Bcl6–, granzyme B–, CD45RO–, CCR4+ and TdT–. (Figure 1A–E) The Ki-67 labeling index was 80%. EBER in situ hybridization was negative. Lymphoma cells were also detected in the bone marrow. The prognostic index score for T-cell lymphoma in this case was 4, considered to be high risk.1 The final diagnosis was PTCL, NOS, stage IVB. We treated the patient using modified CHOP therapy. After one cycle of chemotherapy, the swelled lymph node shrunken and partial response was achieved. However, on day 13 after the modified CHOP therapy, his general condition deteriorated and the WBC increased to 9.6×109/L with 36% lymphoma cells. The disease progressed and we decided to use the histone deacetylase (HDAC) inhibitor romidepsin (14 mg/m2 1×/week for 3 weeks) as second-line therapy. After the first administration of romidepsin, the patient recovered rapidly. His sIL-2R levels decreased to 1,428 U/mL. When the WBC count recovered to 7.6×109/L 17 days later, 8% lymphoma cells persisted in the peripheral blood and one cycle of the monoclonal antibody mogamulizumab (1 mg/kg for every 4 weeks) was added. He received a second cycle of romidepsin, and the disappearance of lymphoma cells from the peripheral blood and all lymph node swelling was confirmed. On day 7 after the second cycle of romidepsin, the
Fig. 1. Histopathology of the biopsy specimen of the right cervical lymph node. Monotonous infiltration of medium to large-sized lymphoma cells is observed (A, low-power field; B, high-power field. Hematoxylin-eosin staining). Immunohistochemistry shows that the lymphoma cells are CD3-positive (C), CD20-negative (D) and CCR4-positive (E).

Fig. 2. Thoracolumbar MR images: T2-weighted image (A) and contrast-enhanced sagittal fat-saturated T1-weighted images (B, C). On the T2-weighted image, the CSF signal surrounding the conus medullaris is effaced. Red triangles: The leptomeningeal linear or nodular enhancement, corresponding to the intramedullary mass.
CONFLICT OF INTEREST

All procedures performed in this study involving the patient were in accordance with the ethical standards of our institutional and national research committee, and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. Informed consent was received from the patient. The authors declare no conflicts of interest in this study.

REFERENCES


Yutaka Shimazu,1) Akihiko Sakata2) and Masaharu Nohgawa1)

1)Department of Hematology, Japanese Red Cross Wakayama Medical Center, Wakayama, Japan, 2)Department of Radiology, Japanese Red Cross Wakayama Medical Center, Wakayama, Japan

Corresponding author: Yutaka Shimazu, M.D., Department of Hematology, Japanese Red Cross Wakayama Medical Center, 4-20, Komatsubara-dori, Wakayama 640-8558, Japan.

E-mail: yshimazu@kuhp.kyoto-u.ac.jp

Received: July 2, 2019.
Revised: October 27, 2019.
Accepted: November 27, 2019.
Online Published: June 20, 2020
DOI:10.3960/jslrt.19024

Copyright © 2020 The Japanese Society for Lymphoreticular Tissue Research

This work is licensed under a Creative Commons Attribution-NonCommercial-ShareAlike 4.0 International License.