Extranodal Natural Killer/T Cell Lymphoma in 11-year-old Child

Takeshi Ninchoji[1], Junya Fujimura[2], Suguru Uemura[2], Nobuyuki Yamamoto[2], Kandai Nozu[3], and Kazumoto Iijima[3]

[1]Kobe University Graduate School of Medicine School of Medicine
[2]Affiliation not available
[3]Kobe University Graduate School of Medicine

July 1, 2020

Abstract

Here we report a case of advanced extranodal natural killer/T cell lymphoma (ENKTL) in 11-year-old child. Positron emission tomography-magnetic resolution image is a crucial image to diagnose ENKTL in this case. Additionally, a strong regime combined with stem cell transplantation possibly improve prognosis of ENKTL.

Key Clinical Message

Advanced extranodal natural killer/T cell lymphoma is reported to have the worst prognosis. Positron emission tomography is a crucial image to diagnose. A strong regime including stem cell transplantation possibly improve its prognosis.

Key words

extranodal natural killer/T cell lymphoma, hemophagocytotic syndrome, SMILE therapy

Case

An11-year old girl presented with vision deterioration and was diagnosed with uveitis. Chest computed tomography showed infiltration in her lung field, for which she was referred to our hospital. She also had fever(38.5°C). After the patient's fever persisted, laboratory findings fulfilled criteria for hemophagocytotic syndrome (HPS) [1]. Positron emission tomography-magnetic resolution image (PET-MRI) scanning demonstrated several abnormal accumulations (Figure 1). Pathological investigation from sinusoid, lungs and kidney biopsies showed atypical lymphoid cells with positive for CD3,4,7,8,56, and negative for CD19,20 with positive for EBV-encoded RNA in situ hybridization. Final diagnosis was extranodal natural killer /T cell lymphoma, nasal type (ENKTL).

The patient was treated with dexamethasone, methotrexate, ifosfamide, L-asparaginase, and etoposide (SMILE) for six courses following umbilical cord blood stem cell transplantation (UCB-SCT) and has been in complete remission for 3 years.

ENKTL is rare in children. Previous reports have documented involvement of multiple organs, including the nasal cavity, skin, gastrointestinal tract, liver and lungs, but only a few cases have reported involvement of the eyes and kidneys. The most distinctive symptom was deterioration of sight. In terms of treatment, advanced ENKTL with HPS has been reported to have the worst prognosis. A strong regime with SMILE in addition to SCT can possibly improve [2], and proved successful in this patient.

Authorship

TN: made substantial contribution to the preparation of this manuscript and approved the final version for submission. JF, SU, NY, KN, and KI: did the

literature search, revised the manuscript for critically important intellectual content and approved the final version for submission

Informed Consent

Informed consent was obtained for publication of this clinical images

Competing Interest

This study was supported by Grants-in-Aid for Scientific Research (KAKENHI) from the Ministry of Education, Culture, Sports, Science and Technology of Japan (subject ID: 19K08726 to Kandai Nozu, 17H04189 to Kazumoto Iijima and), by the Japan Agency for Medical Research and Development (AMED) (Grant Number JP19ek0109231h0003 to Kandai Nozu and Kazumoto Iijima).

Kandai Nozu has received consulting fees from Kyowa Kirin Co., Ltd.; and lecture fees from Kyowa Kirin Co., Ltd., Novartis Pharmaceuticals Corporation, and Chugai Pharmaceutical Co., Ltd.

Kazumoto Iijima has received grant support from Daiichi Sankyo Co., Ltd.; consulting fees from Kyowa Kirin Co., Ltd., and Boehringer Ingelheim; and lecture fees from Kyowa Kirin Co., Ltd., Chugai Pharmaceutical Co., Ltd., Takeda Pharmaceutical Company, Integrated Development Associates, and Novartis Pharmaceuticals Corporation.

The other authors declare that no conflict interests exsist.

References

[1] Henter, J.-I., Horne, A., Aricó, M., Egeler, R. M., Filipovich, A. H., Imashuku, S., Ladisch, S., McClain, K., Webb, D., Winiarski, J., & Janka, G. (2007). HLH-2004: Diagnostic and therapeutic guidelines for hemophagocytic lymphohistiocytosis. *Pediatric Blood & Cancer* ,*48* (2), 124–131.

[2] Kim, S. J., Park, S., Kang, E. S., Choi, J. Y., Lim, D. H., Ko, Y. H., & Kim, W. S. (2015). Induction treatment with SMILE and consolidation with autologous stem cell transplantation for newly diagnosed stage IV extranodal natural killer/T-cell lymphoma patients.*Annals of Hematology* , *94* (1), 71–78.

Figure legend

Figure 1 PET-MRI images

Whole scan showed abnormal accumulations in sinusoid, lungs, and kidney

B-D. MRI coronal images for each organs. B: sinusoid, C: lungs, D: kidney

Ninchoji T. et al. Extranodal Natural Killer/T Cell Lymphoma in an 11-year-old Child

Figure 1