

Childhood cutaneous natural killer/T-cell lymphoma successfully treated with only one course of chemotherapy and incomplete tumor resection

Natural killer (NK) lymphoma has been reported to occur mostly in adults and very few cases have been reported in children. NK lymphomas generally have a poor outcome. We report the case of a 13-year old boy who has maintained complete remission for 33 months after only one course of chemotherapy and incomplete resection of his tumor.

A tumor developed on the patient's right shin 6 months before admission without any other symptoms. The laboratory tests (complete blood cell counts, C-reactive protein, ferritin, and lactic dehydrogenase) were normal. The tumor was 5.5 cm in diameter, dark-brown colored, elastic hard, and painless (Figure 1A). Computed tomography and ⁶⁷Ga scintigraphy did not detect any other involvement. The biopsy of the tumor revealed diffuse infiltration of atypical lymphoid cells in the subcutaneous adipose tissue and no angiocentric features (Figure 1B). Immunohistochemically these cells were positive for CD45, CD3ε, and CD56. On the other hand, CD2, 3, 4, 7, 8, 16, 20, and cytoplasmic complete CD3 molecule were negative. TdT was very weakly positive. The cytotoxic molecules, granzyme B, and TIA-1 were also negative. Flow cytometric analysis showed only CD45 and CD56 were positive. EBV encoded small nuclear RNA (EBER-1) was not detected in tumor cells by means of *in situ* hybridization. We diagnosed him as having blastic-NK/T lymphoma.¹⁻⁴ Southern blot analysis showed germ line rearrangement of both the T-cell receptor (TCR) β and γ genes. Chromosomal analysis (G-banding) revealed normal 46, XY in all analyzed cells. The bone marrow aspiration revealed no infiltration of tumor cells and no hemophagocytic features. Induction chemotherapy consisted of prednisolone, vincristine, cyclophosphamide, daunorubicin, L-asparaginase, and thiotepa (Figure 2). Tumor resection was performed during chemotherapy, but numerous tumor clusters were observed at the margin of the removed tissue by microscopy after resectioning (Figure 1C). His family refused any additional treatment and he was discharged after one course of chemotherapy. Although he has not received any treatment for 33 months, he is well and shows no relapse. In general, NK lymphoma has an extremely poor prognosis. The extranodal NK/T lymphomas are classified under two major entities, nasal-type and blastic-type. Clinical characteristics of the nasal-type NK lymphoma are an aggressive course and poor prognosis. Most investigators report the median overall survival is range from 3.5 to 15 months.^{1,2,5,6} In these reports, the patients are treated with intensive chemotherapy, radiotherapy, and surgical resection. Only a few successfully treated cases have been reported in the literature.⁹⁻¹¹ These successful cases are usually children treated with stem cell transplantation. The prognosis for the blastic NK lymphoma is also poor. The mean survival period of blastic NK lymphoma is less than 24 months.^{1-3,5-8}

We considered our case extremely rare, because the patient remained in remission after only one course of chemotherapy and an incomplete resection. L-Asp is rarely used for NK lymphoma, and thiotepa is secreted out through the skin. The application of L-Asp and/or thiotepa might be at least in part related to the favorable outcome. The prognosis of the cutaneous disease without extracutaneous involvement may be better than that with extracutaneous manifestations.¹²

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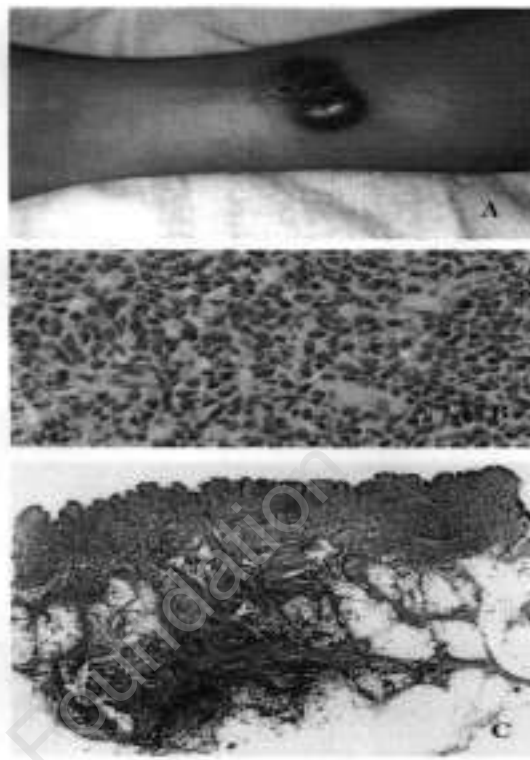


Figure 1A. The tumor was palpable on right shin: it was dark brown, elastic hard, and not mobile. The diameter of the tumor was 5.5 cm at presentation. Figure 1B. Histologic findings of the tumor biopsy (hematoxylin-eosin staining). The specimen shows diffuse infiltration of malignant lymphoid cells. There is no definite angiocentric feature. Figure 1C. Histologic findings of the resected tumor (hematoxylin-eosin staining). The malignant cells infiltrated the subcutaneous adipose tissue and spread over the border.

Key words: NK lymphoma, childhood, L-Asp, thiotepa.

Acknowledgments: We thank Dr. Shigeo Nakamura (Department of Pathology and Clinical Laboratories, Aichi Cancer Center Hospital) for his suggestion on histology. We also appreciate Drs. Tasutoshi Nakahata, Souichi Adachi, and Ken-ichiro Watanabe for their advice.

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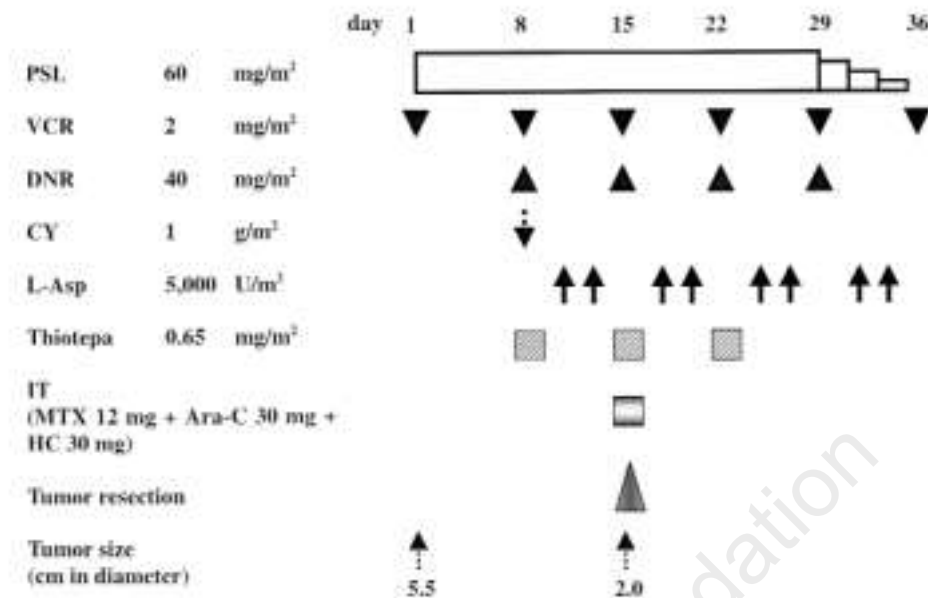


Figure 2. The sole treatment for the present case.

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