A Case of Lymphoblastic Lymphoma with Pre-B Cell Phenotype

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Lymphoblastic lymphoma usually shows a T cell phenotype [1], but rarely some lymphoblastic lymphoma shows a pre-B cell phenotype. We present an uncommon case of lymphoblastic lymphoma expressing a pre-B cell phenotype and review the literature.

An 11-year-old Japanese boy complained of right cervical and bilateral inguinal lymphadenopathy in April 1986. Hepatosplenomegaly was not found. A surgical specimen obtained from the right cervical lymph node showed malignant lymphoma, lymphoblastic type. A chest X ray and computed tomographic scan of the thorax and abdomen revealed no abnormal finding. A hemogram of peripheral blood did not show leukemic picture. Bone marrow aspiration showed normocellularity with no leukemic cell. The patient received a combination chemotherapy and prophylactic cranial irradiation (2,400 rad), and went into complete remission. He is now receiving maintenance chemotherapy.

Microscopically, normal architecture of the cervical lymph node was almost effaced by a diffuse infiltration of atypical lymphoid cells consistent with lymphoblasts, although a few atrophic follicles were observed. The neoplastic cells had convoluted nuclei with dusty chromatin and small prominent nucleoli, and scanty basophilic cytoplasm. Electron microscopy revealed that the neoplastic cells had indented or convoluted nuclei with small prominent nucleoli. The chromatin was evenly distributed in the nucleus with peripheral condensation of heterochromatin along the nuclear membrane. The cytoplasm possessed numerous ribosomes and polyribosomes, several mitochondria and a few short rough endoplasmic cisternae. The neoplastic cells reacted with anti-HLA-DR, anti-CD19(B4), and anti-CD10(CALLA), but not with anti-CD20(B1), anti-CD21(B2), anti-T-cell-associated antigens and anti-myeloid-associated antigens. The neoplastic cells did not express any surface immunoglobulins. Immunoelectron microscopy demonstrated that the neoplastic cells bore the cytoplasmic μ -chain (fig. 1.). In addition, the neoplastic cells were positive for TdT activity. These findings indicate that the present case represents a neoplastic counterpart of pre-B cell. Non-T ALL with a pre-B cell phenotype is not uncommon. However, pre-B cell lymphoblastic lymphoma without leukemic blood picture is very rare; to our knowledge several cases have been reported [2-7] (table 1). These reported cases displayed the common characteristics of indented or convoluted nuclei as well as the preferential involvement of skin and lymph nodes. In addition, these cases often showed lytic bone lesions. The present case is somewhat different

from the previously reported cases because of no involvement of skin and lytic bone lesions. It still remains obscure why pre-B cell lymphoma arises from the peripheral lymphoid tissue or why pre-B cell lymphoma preferentially involves the skin, although pre-B cells are usually found in the liver, spleen and bone marrow during fetal stage and only in the bone marrow after birth [8, 9]. Further investigation is required to clarify the histogenesis and biological nature of pre-B cell lymphoblastic lymphoma.

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Table 1. Summary of reported cases with pre-B lymphoblastic lymphoma

Author

Ref.

No.

Age,
years

Sex

Site

Recurrence

Prognosis

, months

Nuclear shape

Link et al.

2

3.75

f
skin

+ *

2D

convoluted

6

f
skin, LN, SP,
BM

-

34 D

convoluted
Cossman et al.

3

16

4

m

f

LN, BM bone

—

28 A 12 A

convoluted convoluted
Smith

4

16

f

bone, BM

+ *

8D

convoluted
Meyers and Hakami Kamps and Poppema Grümayer et al.

5 6

7

0.5 6

0.5 9

m f

m m

skin, BM

LN

skin, LN skin

+ *

8 D 36 A
33 A 40 A

convoluted
nonconvoluted
convoluted
This case

11

m
LN

-

54 A

convoluted
LN = Lymph
node;
SP = spleen;
BM =
bone
marrow; +
* = hematolog
ical recurrence;
D =
dead; A =
alive.

■ :'

I

Fig. 1. The nuclear membrane and a part of rough endoplasmic reticulum of lymphoblasts revealed positivity for anti- μ -chain antibody.

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