

Precursor B-cell Lymphoblastic Lymphoma with only Cutaneous Involvement

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Sir,

Lymphoblastic lymphoma (LBL) belongs to a high-grade form of non-Hodgkin's lymphoma, and accounts for only 5% of all non-Hodgkin's lymphoma (1). Precursor B-cell LBL (B-LBL) is a rare neoplasm composed of immature lymphocytes that demonstrate lymphoblastic morphology and express precursors and B-cell markers. In contrast to the more common LBL of T-cell lineage, B-LBL can be an extranodal disease with a propensity to involve skin and bone (2). We report here a case of precursor B-LBL involving only a cutaneous site at the time of diagnosis, occurring in a 10-year-old Japanese boy. We provide a brief review of the literature.

CASE REPORT

In February 2005, a 10-year-old Japanese boy presented with a 4-month history of skin tumour on his scalp, but no accompanying pain. According to his mother, the cutaneous tumour had not spread during the 2 months prior to his visit. He was healthy, with no history of trauma or other illness. On examination, the parietal region had a firm, non-fluctuant, non-tender and smooth-surfaced tumour (approximately 35 mm in diameter), with no sign of inflammation (Fig. 1a). Brain magnetic resonance imaging results revealed enlargement of the parietal soft tissue in the area of swelling, without evidence of bone involvement (Fig. 1b). There was no evidence of superficial lymphadenopathy in any other region including the neck, axilla, and groin. Neurological examination was normal. Past medical history and family history were negative. A biopsy specimen from the scalp demonstrated a diffuse infiltrate of atypical lymphoid cells in the entire dermis, with a grenz zone in the papillary dermis and invasion of the skin adnexal structures. The lymphoid cells were medium to large-

sized and uniform in morphological appearance and had fine chromatin, inconspicuous nucleoli, scant, faintly basophilic cytoplasm and a high mitotic rate. The infiltrating atypical lymphoid cells were positive for CD10, CD79a, terminal deoxynucleotidyl transferase (TdT) and leukocyte common antigen, but negative for CD3, CD20, and CD34, according to immunohistochemical studies on paraffin sections (Fig. 2). The findings of the infiltrate lymphoid cells indicated precursor B-cell type.

The complete blood count showed normal values for white blood cell count and platelet count. A peripheral blood smear showed no blasts. Biochemical analysis revealed high lactate dehydrogenase (273 IU/l; normal range 115–230 IU/l), and alkaline phosphatase (1048 IU/l; normal range 115–360 IU/l), and low uric acid (2.4 mg/dl; normal range 3.8–7.5 mg/dl). Other serological results were negative, including rheumatoid factors, anti-nuclear antibodies, anti-neutrophilic cytoplasmic antibodies, and C-reactive protein. Echocardiography and chest X-rays were within the normal limits. Computerized tomography results for the chest, abdomen, and pelvis were normal. Several bone marrow aspirations revealed no evidence of any neoplastic populations. The patient was diagnosed as having precursor B-LBL with solely cutaneous involvement. He was started on the Japanese Pediatric Leukemia/Lymphoma Study Group treatment protocol, which consists of a course of cyclophosphamide, doxorubicin, vincristine, L-asparaginase, and prednisone. In addition, he was treated with intrathecal methotrexate, combined with prednisone. His tumour disappeared during the treatment course, and there was no recurrence of disease during the 3.5-year follow-up period. Repeat bone marrow evaluations showed normocellular results, with no evidence of myeloid or erythroid precursors.

DISCUSSION

Precursor B-LBL is an uncommon form of lymphoblastic lymphoma that accounts for less than 10% of all lymphoblastic lymphoma cases (2). Our case involved a skin site as the primary extranodal lesion, but did not involve any other lesions. There are 18 cases of precursor B-LBL with only skin involvement in the English literature to date, including the present report (2–9). The age at presentation of the reported patients ranged from 14 months to 66 years, with a mean age of 14 years. Female patients accounted for 12 of the 18 patients (67%), and they ranged in age at presentation from 14 months to 13 years (mean 7.5 years). The remaining 6 patients (33%) were male, and ranged in age from 9 to 66 years (mean 27 years). With the exception of one patient, all cases presented as a solitary tumour, usually approximately 2–5 cm in diameter. The tumours were most frequently located on the head ($n=7$; 39%). Immunohistochemical staining of the skin specimen showed that the cells were positive for TdT in all patients. Detection of TdT is widely used as a maker for the neoplasms of LBL. All

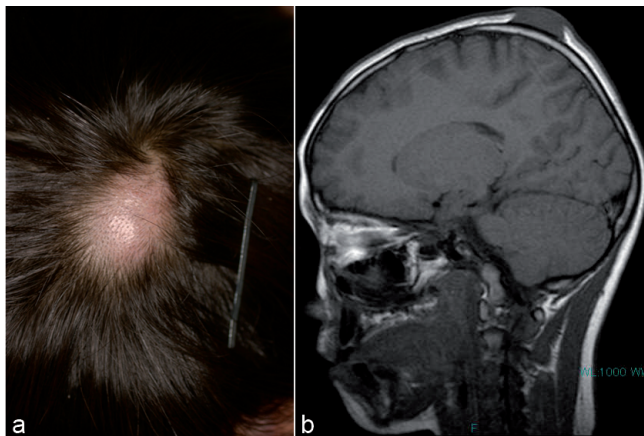


Fig. 1. (a) Physical examination showed a firm, fixed tumour mass, measuring approximately 35 mm in diameter, over the parietal area. (b) Brain magnetic resonance imaging results revealed enlargement of the parietal soft tissue.

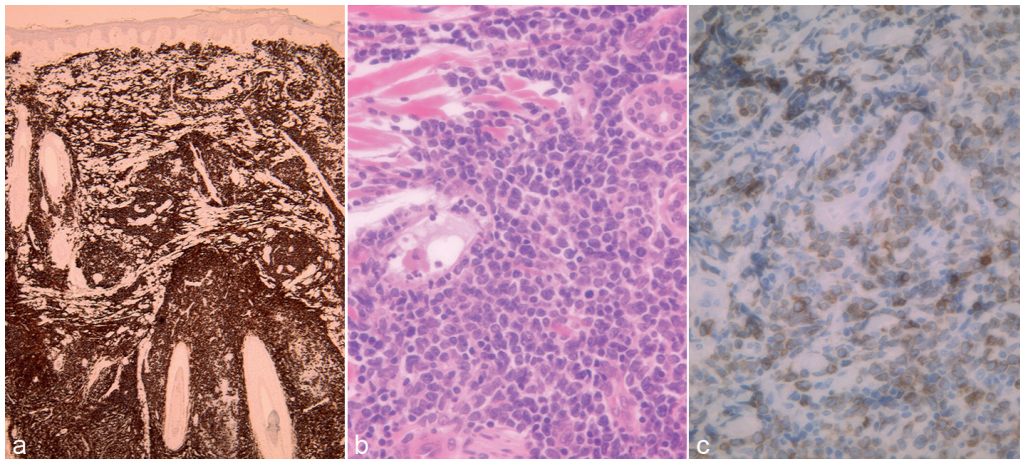


Fig. 2. (a) Skin biopsy specimen showed a dense lymphocytic infiltrate in the entire dermis composed of uniform atypical cells. A grenz zone was present below the relatively normal-appearing epidermis. The infiltrate cells stained positive for CD10 (original magnification: $\times 40$). (b) The medium to large cells were uniform in morphological appearance, with scant cytoplasm, and irregular nuclear contour (haematoxylin and eosin (H&E) $\times 400$). (c) Immunohistochemical analysis of the dense atypical lymphocytes was positive for terminal deoxynucleotidyl transferase (H&E $\times 400$).

patients were treated with multi-agent chemotherapy. In the 15 patients whose follow-up course was described, 5 (33%) patients died (2 male and 3 female) with follow-up periods ranging from 2 months to 72 months, with a mean duration of 32.8 months. The remaining 10 (67%) patients survived with follow-up periods ranging from 12 months to 18 years.

Cutaneous LBL comprises a smaller percentage (less than 20%) of all LBL cases at the time of presentation (10, 11). Precursor B-LBL, especially its cutaneous involvement, is rare, but we reviewed 18 patients with the cutaneous form at onset reported in the literature. B-LBL is included in the World Health Organization Classification of Tumors: Pathology and Genetics of Skin Tumors as a secondary tumour involving the skin (2). However, primary cutaneous precursor B-LBL is not included in the current World Health Organization European Organization for Research and Treatment Cancer cutaneous lymphoma classification (12). Primary cutaneous precursor B-LBL may have a poor prognosis according to our review, and therefore we recommend that dermatologists be aware of this rare cutaneous entity and keep primary cutaneous B-LBL in mind as a potential differential diagnosis for head tumours.

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