Skin involvement can be the initial symptom of hematologic diseases in children.1 Lymphoblastic leukemia/lymphoma (LBL) is a neoplasm of the precursor lymphoid cells considered to be the lymphomatous variant of acute lymphoblastic leukemia (ALL). Distinction between LBL and ALL relies on the massive presence in the latter of blast cells in the bone marrow (>25% blasts) and in the blood. According to the Revised European-American Classification of Lymphoid Neoplasms (REAL classification), ALL and LBL account for 80% and 20% of neoplastic proliferation of lymphoblasts, respectively.2 LBL affects predominantly children and young adults with occasional older patients have been reported.3,4

A 2-month-old female presented with multiple subcutaneous swellings on the scalp, shoulders and upper and lower limbs for the previous week. The swelling progressed to the eyelids and lips with intermittent stridor after receiving treatment for a common cold (Figure 1). Angioneurotic edema was suspected. She received steroids treatment and frequent adrenaline nebulization, but the condition progressed. On examination, she appeared ill, of average weight and height (50th percentiles). There were multiple subcutaneous masses in the scalp, face, shoulders, and upper and lower limbs. These masses were about 1×2 to 2×4 cm, firm, nontender, and fixed to the skin, with purplish unhealthy overlying skin with lost hair for scalp lesions (Figure 1a,b).

Complete blood count showed only thrombocytopenia (hemoglobin 9.8gm/dL, WBC 12.8×10⁹/L and platelets 27×10⁹/L), prothrombin time 16.8 seconds, APTT 46 seconds, and INR 1.8). The value for fibrin degradation products was 20 µg/mL (normally <10 µg/mL). The blood film was normal. HIV antibodies test were negative, blood cultures were also negative. X-rays of the chest, skull and long bones were normal. CT skull showed multiple subcutaneous masses in scalp (white arrows) with normal brain and heavy infiltration of the skin over the orbit (Figure 2). Bone marrow aspiration was normal.

Bacillary angiomatosis was suspected and she received macrolide antibiotics orally and cefotaxime injection for 1 week, but with no improvement. A biopsy from the largest mass, located in the shoulder, revealed...
images and diagnosis

Figure 2. CT skull shows multiple subcutaneous masses in scalp (white arrows) with normal brain and heavy infiltration of the skin over the orbit.

Figure 3a. Pathological examination of the biopsied mass shows an intact overlying skin with underlying infiltration by transformed lymphocytes, arranged in vague nodular pattern (star-sky appearance) with Indian-file arrangement.

Figure 3b. Immunostaining by monoclonal antibodies shows positive CD3 and LCA with negative CD20.

Figure 4. The patient after 3 weeks treatment showing disappearance of subcutaneous mass and facial swellings. (Permission to publish photo granted by mother).

A 2×2×1 cm irregular mass, firm with a grayish white and cut surface. Microscopy showed intact overlying skin with underlying infiltration by transformed lymphocytes, arranged in a vague nodular pattern (star-sky appearance) with Indian-file arrangement (Figure 3a). There were mostly moderate to large size cleaved vesicular nuclei and occasional nucleoli. Mitosis was frequent with atypical forms. There was a perivesicular and perianodenial targetoid arrangement that was highly suggestive of non-Hodgkin lymphoma or lymphoblastic lymphoma (LBL). Immunostaining by monoclonal antibodies against CD3, CD20 and LCA (DAKO USA En Vision System) showed positive CD3 and LCA with negative CD20 (Figure 3b), which supported T cell LBL. The patient was started on the BFM protocol (oncovin IV, adriamycine IV, prednisolon oral). After 3 weeks of treatment, there was marvelous improvement with disappearance of the subcutaneous mass and facial swellings (Figure 4).

LBL seems to occur more frequently in males than in females. The majority of cases of B cell LBL presents as acute leukemia with bone marrow and pe-
Peripheral blood involvement, whereas patients with T cell LBL usually have rapidly enlarging, symptomatic mediastinal mass and/or peripheral, mainly supradiaphragmatic, lymphadenopathy. In both B and T cell LBL, a small proportion of patients have solid tumors in the skin, with or without bone marrow and peripheral blood involvement. The clinical appearance of skin lesions includes single or multiple papular or nodular lesions preferentially located on the head and neck.4-6

REFERENCES