



T-immunophenotype lymphoblastic lymphoma with secondary cutaneous involvement associated with rapid regression followed up with positron emission tomography

Carla de F. P. Favaro, MD,^a Anna Paula V. O. Bomtempo, MD,^b Bruno S. dos Santos, MD,^a Luciana T. C. de M. Tucunduva, MD,^c Silmara da C. P. Cestari, MD, PhD,^a and Reinaldo T. Filho, MD, PhD^a
São Paulo, Brazil

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INTRODUCTION

Lymphoblastic hematopoietic neoplasms, according to the World Health Organization classification, may present as leukemia or lymphoma; by definition, lymphoma is associated with less than 20% of blasts in the bone marrow and a mediastinal mass or a mass in another location.¹ Lymphoblastic lymphoma is a rare type of non-Hodgkin lymphoma that is subdivided according to the precursor cell line into B and T immunophenotypes.²

T-lymphoblastic lymphoma, which affects adolescents and young adults, has the main clinical manifestations of peripheral lymphadenopathy and a mediastinal mass. Secondary cutaneous involvement is uncommon and is characterized by multiple nodules in the neck and trunk.²

We present a case in which cutaneous involvement was the first indication of T-lymphoblastic lymphoma and emphasize the rare nature of the presentation and the imaging examinations performed for patient follow-up.

CASE REPORT

The patient was a 20-year-old man with asymptomatic lesions on the scalp for 60 days; these lesions increased in size over time, and similar new lesions appeared on the face and trunk. Dermatologic examination revealed nodules and

hard, erythematous, infiltrated plaques of varying size on the scalp, left ear, left side of the brow, chin, cheeks, right shoulder, left side of the lumbar region, and presternal region (Fig 1).

Clinical hypotheses that were considered included cutaneous lymphoma, Sweet's syndrome, mycosis fungoides, sarcoidosis, and leprosy. Excisional biopsies were performed for anatomopathologic examination of lesions in the right shoulder and left side of the lumbar region, which revealed an infiltrate of immature neoplastic hematology cells with an interstitial, perivascular, periadnexal, and subcutaneous pattern and numerous mitoses (Fig 2). Immunohistochemical results were CD3⁺, CD4⁺, CD5⁺, CD7⁺, CD8⁺, CD10⁺, Ki-67⁺, TdT⁺, and CD79A⁺ (Fig 3); these results, in combination with the morphologic findings, were consistent with cutaneous infiltration by T-immunophenotype lymphoblastic lymphoma/leukemia.

The patient was referred to the hematology department. Immediate hospital admission was indicated, and additional tests for investigation were requested. Positron emission tomography—computed tomography showed glycolytic hypermetabolism in the cervical lymph nodes, mediastinal mass, and nasopharynx, as well as in multiple areas of cutaneous and subcutaneous thickening (mainly in the scalp), bone marrow, the pancreas, and the renal cortex.

From Dermatology, Sírio-Libanês Hospital, São Paulo^a; Sírio-Libanês Teaching and Research Institute, São Paulo^b; and Bone Marrow Transplant Center at Hospital das Clínicas of São Paulo University and Sírio-Libanês Hospital, São Paulo.^c

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Correspondence to: Anna Paula V. O. Bomtempo, Prof. Daher Cutait Street, 69 Bela Vista, São Paulo/SP, Brazil. E-mail: annapaula_bomtempo@hotmail.com.

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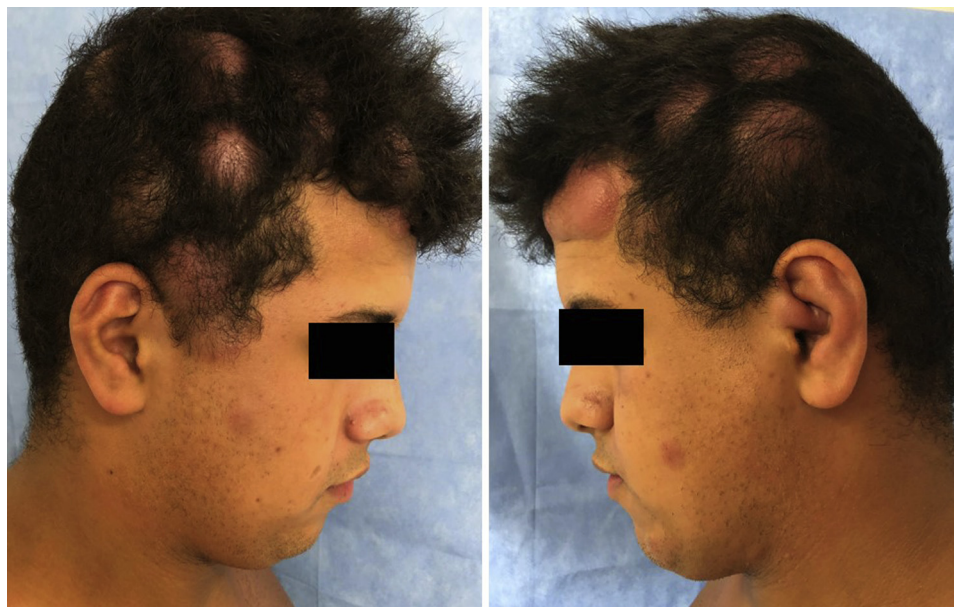


Fig 1. Lymphoblastic lymphoma. Nodules and hard, erythematous, infiltrated plaques on the face and scalp.

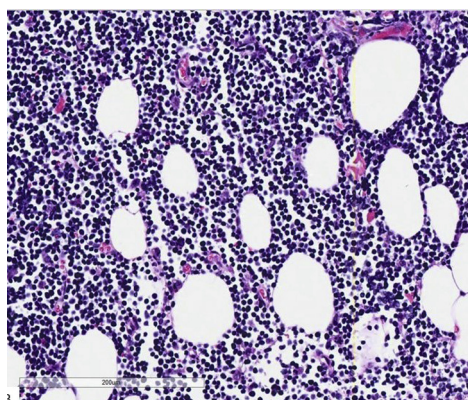


Fig 2. Lymphoblastic lymphoma. Anatomopathologic examination showing intermediate-sized immature lymphocytes, dense chromatin, and scant cytoplasm. (Hematoxylin-eosin stain; 200 μ m).

Normal results for hemogram, myelogram, immunophenotyping, fluorescence in situ hybridization for acute lymphoblastic leukemia, and bone marrow karyotype tests were obtained; there were no signs of lymphomatous infiltration. Cerebrospinal fluid testing and immunophenotyping were performed to investigate infiltration of the central nervous system; the results were negative. A diagnosis of stage IV T-immunophenotype lymphoblastic lymphoma with secondary cutaneous involvement was therefore determined.

Chemotherapy with methotrexate, daunorubicin, vincristine, and cyclophosphamide was initiated. On the seventh day after the start of chemotherapy, the patient's lesions exhibited

almost complete involution, maintaining only residual hyperchromia.

The patient was followed up by the hematology department and underwent another positron emission tomography–computed tomography scan to assess the therapeutic response after induction chemotherapy (Fig 4); this scan confirmed the disappearance of the cutaneous lesions after 36 days. During the first year of follow-up, the patient continued to be treated and is in remission of the disease.

DISCUSSION

Secondary cutaneous involvement in T-lymphoblastic lymphoma is rare. Although the exact frequency of such involvement is unknown, Lee et al³ analyzed 203 cases and found that cutaneous manifestations were observed in 4.3% (7/163) and 15% (6/40) of patients with the T and B immunophenotypes, respectively.

In the medical literature from 1991 to 2019, 16 cases of T-lymphoblastic lymphoma associated with skin lesions were reported. The affected individuals varied from aged 8 to 75 years (mean 33 years) and included 12 male and 4 female patients. Primary involvement for which the lesion is limited to the skin is even more rare and was described in 2 of the 16 aforementioned reports. In the majority of the 16 reported cases, cancer was diagnosed at an advanced stage.^{2,4}

Despite technologic advances in the understanding of hematologic tumors, imaging tests during the follow-up of patients with cutaneous T-

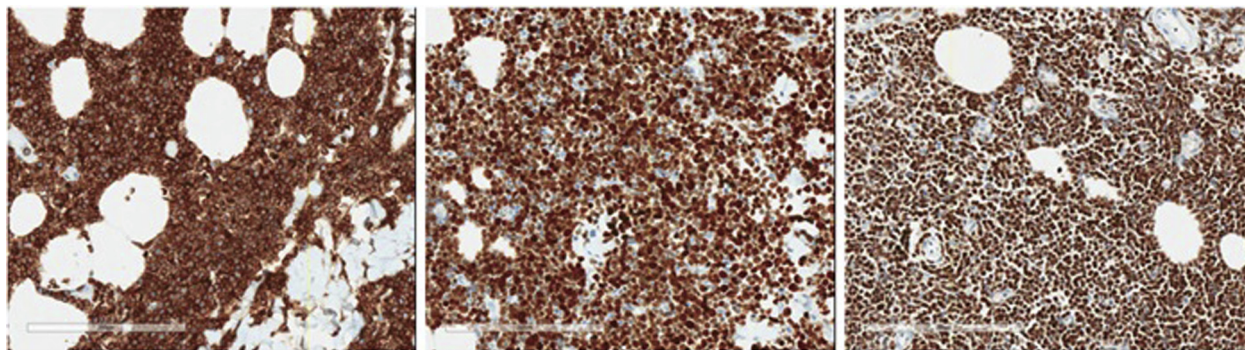


Fig 3. Lymphoblastic lymphoma. Immunohistochemistry for CD3⁺, Ki-67⁺, and TdT⁺, respectively.

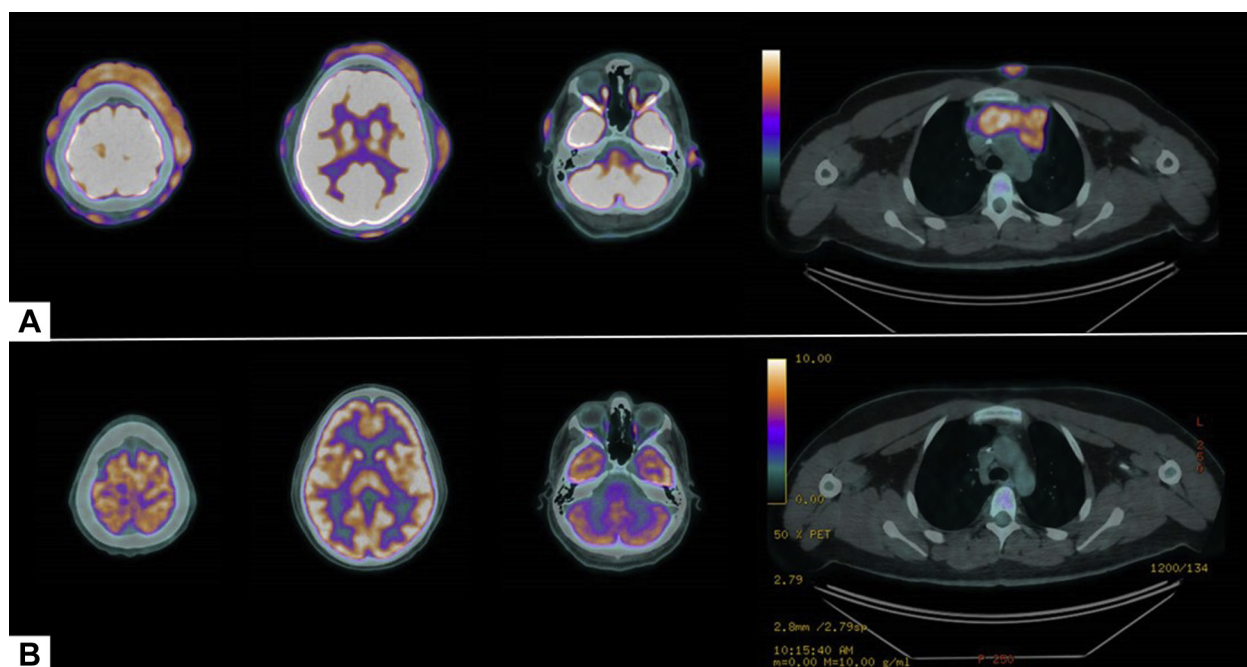


Fig 4. Lymphoblastic lymphoma. Findings from examinations performed 36 days apart. During the first positron emission tomography–computed tomography scan, cutaneous and subcutaneous hyperintense areas infiltrating the scalp, ear, and thorax were observed (**A**). Resolution of glycolytic hypermetabolism and morphologic regression were observed during the subsequent examination (**B**).

lymphoblastic lymphoma are infrequently reported because of the rarity of this disease and the high cost of certain of these tests. In the present study, it was possible to perform continuous evaluation of the lesions with positron emission tomography and to thereby provide evidence of the effectiveness of the treatment according to regression of the patient's condition.

It is well established in the literature that lymphoblastic lymphoma should not be treated as non-Hodgkin's lymphomas are but rather as acute lymphoblastic leukemia is. Therefore, CHOP (cyclophosphamide/doxorubicin/vincristine/prednisone)

or ICE (ifosfamide/carboplatin/etoposide) are not an option to treat this disease.⁵ This patient was treated according to the T-cell acute lymphoblastic leukemia (T-ALL) French Protocol Group for Research on Adult Acute Lymphoblastic Leukemia (GRAALL), which includes maintenance.

Chemotherapy regimens currently offer complete remission rates of 55% to 100% and 5-year disease-free survival rates of 45% to 65%.⁴ Delayed diagnosis can negatively affect prognosis. In this respect, it is important to consider neoplastic diseases in the differential diagnosis of cutaneous nodular lesions to ensure appropriate

and immediate treatment in cases involving such diseases.

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