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*Net Letter*

# Erythema–multiforme like leukemia cutis in T-cell acute lymphoblastic leukemia – A case report

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**Quick Response Code:**

Sir,

Leukemia cutis is infiltration of neoplastic leukocytes or their precursors into the epidermis, dermis, or subcutis resulting in cutaneous lesions. Leukemia cutis is rare in acute lymphoblastic leukemia and is often a poor prognostic marker.[1] Here, we report a case of leukemia cutis as an initial manifestation of T-cell acute lymphoblastic leukemia (T-cell-ALL) in a young male.

A 24-year-old male patient presented to our department with pruritic red-raised lesions associated with fever and cough of 1-month duration. General examination revealed multiple enlarged hard, mobile, non-tender, and non-matted cervical lymph nodes on both sides. He also showed multiple erythematous plaques distributed over the face, anterior and posterior aspects of trunk, both arms, forearms, and thighs. Few of them had central necrosis [Figures 1 and 2]. There were multiple hard nodules over the back of the trunk. Complete hemogram and urine microscopy were normal. Serology for antinuclear antibodies and peripheral smear examination did not reveal any abnormalities. Chest X-ray detected mediastinal widening and left-sided pleural effusion. Mantoux test was negative. Serology for human immunodeficiency virus, human T-cell lymphotropic virus-1, and hepatitis B and C viruses was negative. Differential diagnoses of lymphoma and erythema multiforme were considered. Skin biopsy from the hard nodules and the erythema multiforme like lesions showed diffuse infiltrate of atypical cells in the papillary and reticular dermis extending to the subcutis. Immunohistochemistry analysis showed positivity for CD3 and CD4. The cells were CD8, CD20, and CD30 negative [Figure 3a-c]. Bone marrow aspiration and trephine biopsy showed monomorphic population of atypical lymphoid cells, [Figure 4] constituting 90% of the infiltrate. Flow cytometry of bone marrow aspirate showed the cells to be positive for CD1a, CD3, CD4, CD5, CD7, and CD99 and negative for CD34, CD117, CD13, CD33, CD19, and CD22. Tumor cells were positive for terminal deoxynucleotidyl transferase (TdT). Staining for myeloperoxidase (MPO) was negative. The pleural fluid analysis showed protein 3 g/dl, sugar 106 mg/dl, and total count 17,000/$mm^3$ with 85% lymphocytes. Cytological examination of pleural fluid did not reveal any atypical cells. The patient was diagnosed to have T-cell-ALL with leukemia cutis and was treated with Berlin-Frankfurt-Munster-95 (BFM-95) protocol. Skin lesions showed complete subsidence in 7 days. Pleural effusion also responded to chemotherapy in 3 weeks. However, the patient manifested pruritic erythematous and purpuric macules and plaques (a few showing vesiculations) over the anterior and posterior aspects of the trunk, both arms and forearms [Figure 5] 4 months after initiation of chemotherapy. He was treated with injection dexamethasone 4 mg, $8^{th}$ hourly for 5 days. However, the general condition of the patient deteriorated, and he succumbed to illness 5 months after the diagnosis.

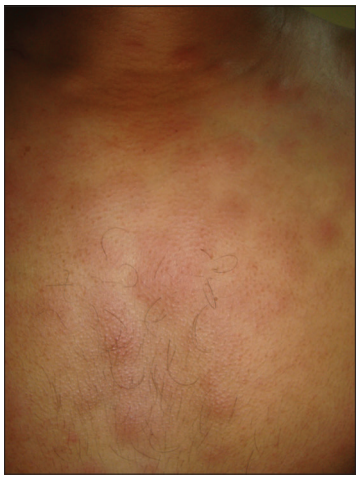

**Figure 1:** Multiple erythematous plaques over the chest.

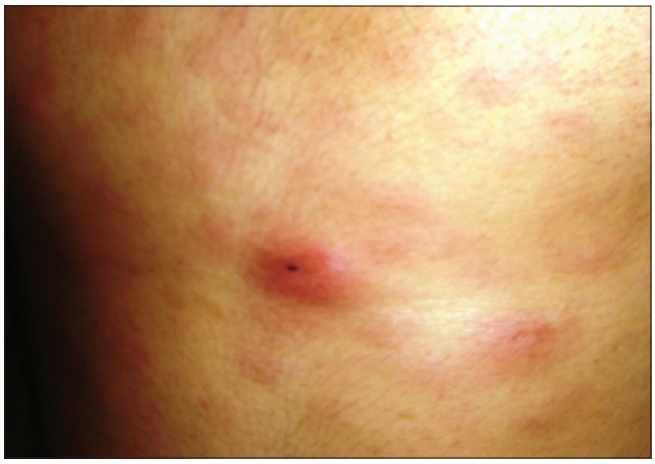

**Figure 2:** Erythematous plaque with central charring over the back.

T-cell-ALL is characterized by the proliferation of lymphoblasts involving bone marrow. T-cell-ALL represents around 25% of all adult cases of acute lymphoblastic leukemia.[2]

The World Health Organization defines T-cell-ALL and T-cell lymphoblastic lymphoma (T-LBL) as the same disease. The terminology T-cell-ALL is used when there is extensive bone marrow involvement, whereas T-cell-LBL is primarily a mass lesion with 20–25% blasts in the marrow.[3]

Leukemia cutis refers to the migration of leukemic cells into the skin. On most occasions, leukemia cutis manifests along with or succeeds the diagnosis of systemic leukemia; rarely, leukemia cutis may precede the development of leukemia (mostly acute myeloid leukemia), when it is termed as "aleukemic" leukemia cutis.[4]

Leukemia cutis is typically seen in myeloid or mature lymphoid malignancies and is very rare in T- and B-cell ALL.[5]

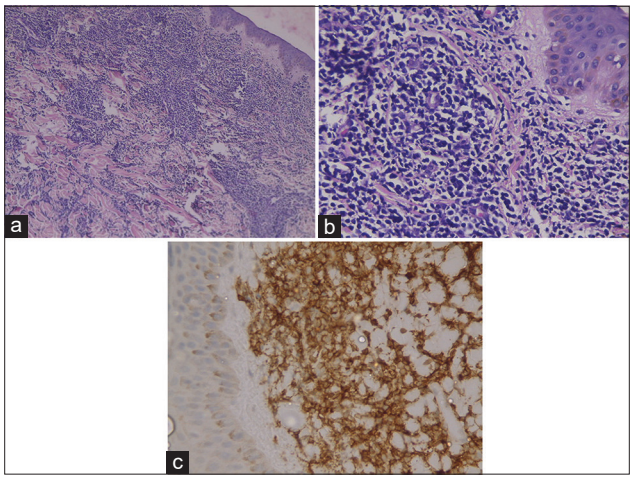


**Figure 3:** (a) Diffuse infiltrate of atypical cells in the dermis extending to subcutis (H&E, ×100). (b) Dense infiltration of atypical cells with pleomorphic nuclei in the upper dermis (H&E, ×400). (c) Diffuse infiltrate of CD4+ cells in the dermis (DAB chromagen, ×200).

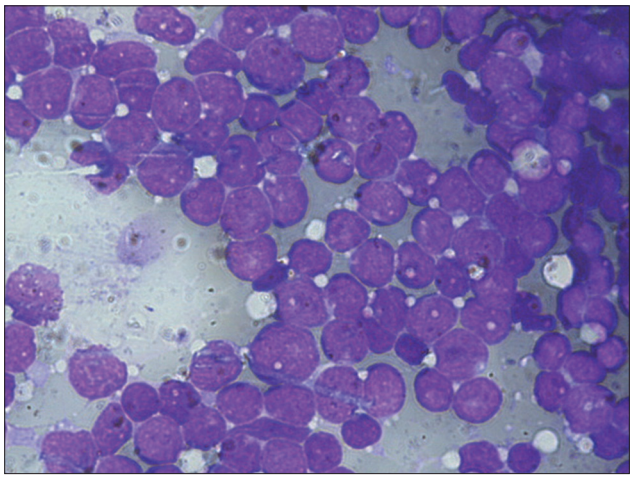

**Figure 4:** Bone marrow imprint shows monomorphic population of atypical lymphoid cells (Leishman stain, ×100).

The usual presentations include erythematous to violaceous papules or firm, rubbery nodules or plaques. Rarely, it may present as ulcers, vesicles, diffuse erythema, purpura, and erythroderma. Sweets syndrome like lesions, butterfly-like rash, urticaria, leonine facies, urticaria pigmentosa-like lesions, erythema annulare centrifugum, erythema nodosum, stasis dermatitis, guttate psoriasis, macular erythema, chronic paronychia, subungual lesions, umbilicated lesions, cauliflower-like lesions, granuloma annulare like lesions, Grover's disease-like lesions, and chemical burn-like lesions have also been reported as manifestations of leukemia cutis. The lesions tend to be localized or disseminated. A combination of lesions with different morphology may also occur. Mucosal involvement in the form of ulceration or gingival hyperplasia may be seen.[6,7] Our patient had

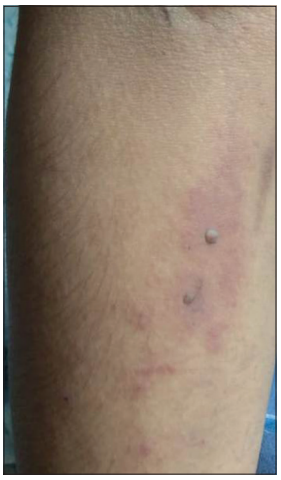

**Figure 5:** Purpuric macule with vesiculation over the medial aspect of the right forearm.

erythematous plaques and nodules and a few erythema multiforme like lesions. Ohtani *et al*. reported erythema multiforme like lesions as a specific manifestation in adult-T-cell leukemia, though usually the former is considered as a non-specific manifestation of leukemia.[8]

In our patient, histopathology analysis of erythema multiforme like lesions showed leukemic infiltrates; hence, we consider it a specific lesion of leukemia (leukemia cutis). The patient developed similar lesions during relapse as well.

Confirming the diagnosis of leukemia cutis in the absence of systemic leukemia is challenging. In view of the skin lesions and lymphadenopathy, we had considered cutaneous lymphoma as a differential diagnosis. However, the presence of atypical lymphoid cells positive for CD3 and CD4 in skin biopsy, TdT positive, and MPO negative cells in bone marrow biopsy and the flow cytometry findings helped us to reach the final diagnosis of T-cell-ALL. Our patient was treated with BFM-95 protocol (prednisolone, vincristine, daunorubicin, and L-asparaginase). Although he responded to the treatment in the initial months, his condition rapidly deteriorated, along with the relapse of the skin lesions.

Leukemia cutis can be the first sign of systemic leukemia. T-cell-ALL in adults is a highly aggressive disease with inferior survival outcome. The presence of cutaneous involvement as the initial manifestation of T-cell-ALL predicts still poorer prognosis.

### Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

### Conflicts of interest

Dr. Anza Khader and Dr. K. Abdul Samad are on the editorial board of the Journal.

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