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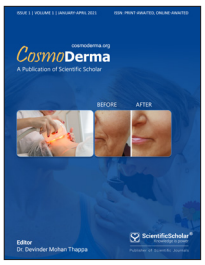

Letter to the Editor

Leukaemia cutis manifesting as leonine facies

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Quick Response Code:

Dear Sir,

T-cell prolymphocytic leukemia (T-PLL) is a rare, aggressive mature T-cell neoplasm. Cutaneous involvement of T-PLL may manifest in the form of purpura, livedoid vasculopathy, vasculitis, and erythema gyratum-like lesions. We report a case of T-PLL with leukemia cutis manifesting as leonine facies.

A 61-year-old male patient presented with firm, infiltrated papules coalescing to form plaques thrown into folds and furrowed creases over the lower forehead, periorbital area, cheeks, nasolabial folds, and perioral area [Figure 1] for 12 months. There was the involvement of the lobule of the pinna and preauricular area in the form of ulcerated nodules [Figure 2]. The lesions were pruritic. There was no hepatosplenomegaly. Histopathological examination revealed dense infiltration of the dermis by small to medium-sized atypical lymphoid cells with a high nuclear: cytoplasmic ratio [Figure 3a and b]. Some of these lymphoid cells had an irregular nucleus with conspicuous nucleoli. In addition, there was epidermotropism of atypical lymphoid cells [Figure 3c]. On immunohistochemistry, these cells were positive for CD3, CD4, and CD5, and negative for CD20 [Figure 3d-f]. The small, reactive T-cells were positive for CD8 compared to the medium-sized atypical lymphoid cells that were negative. The total leukocyte count was 40,500 leucocytes/mm^3. Peripheral blood smear revealed small- to medium-sized lymphoid

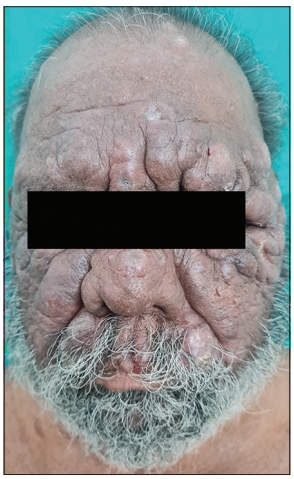

Figure 1: Firm, infiltrated papules coalescing to form plaques thrown into folds and furrowed creases over the lower forehead, periorbital area, cheeks, nasolabial folds, and perioral area.

cells with round to oval nuclei with irregular nuclear membranes. These cells also had cytoplasmic blebs. Bone marrow biopsy revealed a cellular marrow with trilineage hematopoiesis and moderate T-lymphocytosis suggestive of T-cell prolymphocytic leukemia. Based on the history, examination, histopathological findings, peripheral blood smear, and bone marrow biopsy, we made a diagnosis of leukemia cutis secondary to T-cell prolymphocytic leukemia. We referred him to the medical oncology department for further management.

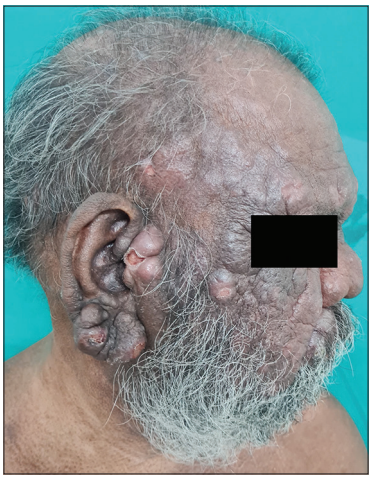

Figure 2: Involvement of the lobule of the pinna and preauricular area in the form of ulcerated nodules.

T-PLL is a rare and aggressive neoplasm presenting in elderly males aged more than 65 years. T-PLL typically presents with fever, weight loss, and lymphadenopathy. In a large case series of T-PLL, splenomegaly was seen in 73% of patients, lymphadenopathy in 53%, and hepatomegaly in 40% of patients.[1] Cutaneous involvement was seen in one-fourth of patients with T-PLL.[1] The cutaneous involvement presents in the form of maculopapular rash, purpura, papules, nodules, and erythroderma. The distribution is varied with facial preference documented.[2] Leonine facies is the infiltration of the facial skin resulting in creases and convexities. Leonine facies has been described in leprosy, mastocytosis, sarcoidosis, amyloidosis, and cutaneous T-cell lymphomas. It can also be a rare manifestation of leukemia cutis in T-PLL.[3,4]

The histology of T-PLL reveals dermal infiltrate with a band-like pattern seen in a few cases. A grenz zone separating dermal infiltrate from the overlying epidermis has also

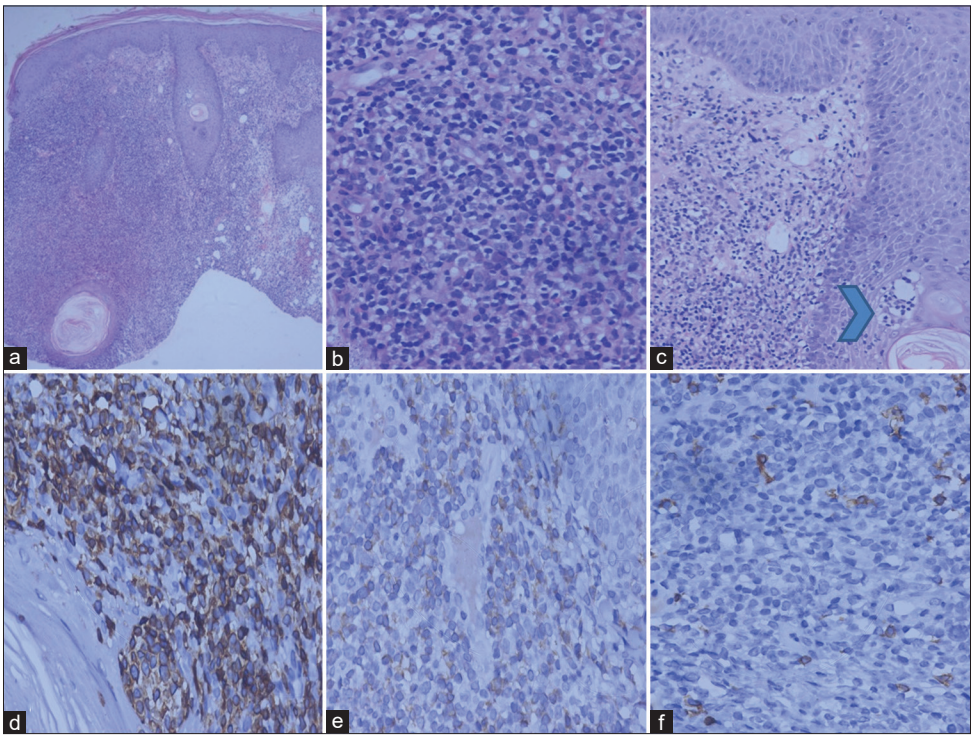

Figure 3: Histopathology demonstrating (a) sheets of atypical lymphoid cells in the dermis and subcutaneous fat (H&E, ×40), (b) medium-sized lymphoid cells with scant cytoplasm and inconspicuous nucleoli admixed with small reactive lymphocytes (H&E, ×400), and (c) epidermotropism of tumor cells (arrowhead) (H&E, ×200). Immunohistochemistry demonstrating strong positivity with (d) CD3 (DAB, ×400) and (e) CD5(DAB, x200). Immunohistochemistry with (f) CD20 positivity in reactive lymphocytes and negative in medium-sized lymphoid cells (DAB, ×200).

been seen in some cases. Peripheral blood smear of T-PLL reveals characteristic cytoplasmic protrusions or blebs. The small- to intermediate-sized cells have a finely dispersed heterochromatin and a single small nucleolus. The nuclei are indented, and the cytoplasm is agranular and eosinophilic. T-PLL is positive for the T-cell markers, including CD2, CD3, CD5, and CD7. T-PLL may be CD4+/CD8- (65%), CD4+/CD8+ (21%), or CD4-/CD8+ (13%).[5] T-PLL expresses CD52, which can be targeted by alemtuzumab. T-PLL may result from cytogenic abnormalities, the most common of which is the inversion of chromosome 14. The other cytogenic abnormality observed is tandem translocation of chromosome 14 between 14q11 and 14q32.

T-PLL has an aggressive course with a median survival of 1–2 years. CHOP chemotherapy and alemtuzumab have been tried with varying results. Autologous stem cell transplantation is useful in patients who experience initial remission with alemtuzumab. A high degree of clinical suspicion is necessary in elderly patients presenting with leonine facies to identify skin involvement with leukemic cells in such hematological malignancies. Dermatologists should be aware of this presentation to avoid misdiagnosis.

Declaration of patient consent

Patient's consent is not required as patient's identity is not disclosed or compromised.

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Conflicts of interest

Dr DM Thapa is the editor of this journal. He does not have any competing interest.

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