

Lymphoma, Cutaneous/Mycosis Fungoides

P1700

Human T-cell lymphotropic virus type 1 (HTLV-1)-associated adult T-cell leukemia/lymphoma presenting with cutaneous papules, hypercalcemia, and osteolytic bone lesions

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A 61 year-old previously healthy Japanese-American woman was hospitalized for severe recalcitrant hypercalcemia of 3 months duration. She complained also of a generalized asymptomatic rash and fatigue. Her medical history was remarkable for multiple fractures and widespread osteolytic bone lesions of unknown etiology. The patient's calcium level on admission was 17.5 with a suppressed parathyroid hormone. Complete blood count and serum electrophoresis were within normal limits.

Physical exam revealed generalized, dark red, firm papules coalescing into plaques. Punch biopsy from the right thigh showed a dense band-like and perivascular dermal infiltrate of atypical lymphocytes, which focally abutted the epidermis. The atypical cells expressed CD2, CD3, CD4, and CD25, with diminished expression CD5 and absent expression of CD7. CD8, CD30, and CD56 were negative. PCR analysis of the paraffin-embedded skin biopsy specimen demonstrated a clonal T-cell receptor gene rearrangement. The histopathologic and immunophenotypic findings were diagnostic of lymphoma and highly suspicious for human T-cell lymphotropic virus type 1 (HTLV-1)-associated adult T-cell leukemia/lymphoma (ATCLL). HTLV-1 serology was subsequently found to be positive.

The patient was treated with cyclophosphamide, vincristine, doxorubicin, and dexamethazone. Shortly after one cycle of chemotherapy, the cutaneous involvement resolved completely. Additional cycles of chemotherapy were postponed due to prolonged chemotherapy-associated pancytopenia.

HTLV-1 is endemic in Japan, the Caribbean, Central Africa, and the southeastern US. Infection is transmitted by sexual contact, blood transfusion, or mother-to-child vertical transmission. The HTLV-1 genome is clonally integrated into the DNA of the lymphoma cells. Since the patient was raised from age 7 in the western US, the virus was most likely transmitted to her as an infant via breast feeding while living in Japan. The strong expression of CD25 by the lymphoma is a characteristic histopathologic finding. This case was remarkable for the key role that skin biopsy played in suggesting the diagnosis. ATCLL should be considered in patients presenting with cutaneous lymphoid infiltrates, hypercalcemia, and osteolytic bone lesions, especially in patients living in or derived from endemic areas.

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