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EBV-associated cutaneous NK/T-cell lymphoma: review of a series of 14 cases from peru in children and young adults.

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Abstract

We have reviewed clinically, morphologically, and immunophenotypically a series of 14 Epstein-Bar virus (EBV)+ cutaneous natural killer cell (NK)/T-cell lymphoma from Peru. Most (11 out of 14) of these cases fit well into the category of Hydroa vacciniforme-like lymphoma (HVLL), but 3 have a different clinical presentation, without facial involvement. In all 14 cases, skin lesions present in both the sun-exposed and nonexposed areas exhibited a slowly progressive relapsing course, changing from edema, to blistering, ulceration, and final scarring. The immunophenotype had a cytotoxic T or NK-cell lineage. The mean time of disease before admission to hospital was 69 months (range, 6 mo to 31 y). Only 2 patients had fever, hepatosplenomegaly, systemic lymphadenopathy, and a high lactate dehydrodenage (LDH) level at the time of diagnosis, whereas 10 had facial swelling. After treatment, only 4 patients remain alive, although with persistent disease. Ten patients died after a mean follow-up of 11.6 months after the initial diagnosis (range, 1 to 32 mo), because of concurrent infections (4 cases), disease progression (4 patients) or both (2 patients). Endemic Epstein-Bar virus (EBV)-positive cutaneous NK/T-cell lymphoproliferative disorders in childhood and early adulthood are characterized by a protracted clinical course, eventually leading to an aggressive phase characterized by concurrent infections and disease progression.