LINFOMAS

Primary cutaneous T-cell lymphoma: experience from the Peruvian National Cancer Institute.

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Abstract

BACKGROUND: Primary cutaneous T-cell lymphomas constitute a heterogeneous and rare group of diseases with regional particularities in Latin America. OBJECTIVE: To determine the clinicopathological features, relative frequency and survival among patients from a Peruvian institution. METHODS: Primary cutaneous T-cell lymphomas were defined based on the absence of extracutaneous disease at diagnosis. Classification was performed following the 2008 World Health Organization Classification of Neoplasms of the Hematopoietic and Lymphoid tissues. Risk groups were established according to the 2005 World Health Organization-EORTC classification for cutaneous lymphomas. Data of patients admitted between January 2008 and December 2012 were analyzed. RESULTS: 74 patients were included. Mean age was 49.5 years. In order of frequency, diagnoses were: mycosis fungoides (40.5%), peripheral T-cell lymphoma not otherwise specified (22.95%), adult T-cell lymphoma/leukemia (18.9%), CD30+ lymphoproliferative disorders (6.8%), hydroa vacciniforme-like lymphoma (5.4%), extranodal NK/T-cell lymphoma (4.1%) and Sézary syndrome (1.4%). Predominant clinical patterns were observed across different entities. Mycosis fungoides appeared mainly as plaques (93%). Peripheral T-cell lymphoma not otherwise specified and adult T-cell lymphoma/leukemia presentation was polymorphic. All patients with hydroa vacciniforme-like lymphoma presented with facial edema. All cases of extranodal NK/T-cell lymphoma appeared as ulcerated nodules/tumors. Disseminated cutaneous involvement was found in 71.6% cases. Forty-six percent of patients were alive at 5 years. Five-year overall survival was 76.4% and 19.2%, for indolent and high-risk lymphomas, respectively ($p<0.05$). High risk group (HR: 4.6 [2.08-10.18]) and increased DHL level (HR: 3.2 [1.57-6.46]) emerged as prognostic factors for survival. STUDY LIMITATIONS: Small series. CONCLUSION: Primary cutaneous T-cell lymphomas other than mycosis fungoides or CD30+ lymphoproliferative disorders are aggressive entities with a poor prognosis.