SKIN CANCER (OTHER THAN MELANOMA)

SEZARY SYNDROME: A 10-YEAR SINGLE-CENTER EXPERIENCE

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Introduction: Primary cutaneous lymphomas (PCL) are a group of extranodal non-Hodgkin lymphomas presenting with no evidence of extracutaneous disease at the time of diagnosis. Few longitudinal studies of PCL have been conducted.

Objective: To describe the behavior of PCL and evaluate patient survival in a cohort of patients from our institution.

Materials and Methods: We conducted a retrospective study of all patients with a histopathological diagnosis of PCL receiving care at a singular academic center from 2007 to 2017.

Results: Four patients with PCL were included, with a mean age at diagnosis of 56 years. Sezary syndrome (SS) accounted for all cases. Three patients with SS were male. The mean diagnostic delay was 5 years overall, 37.3 months for cases involving erythroderma at the initial presentation (3 cases), and 132 months for cases not involving erythroderma at the initial presentation (one case). Diagnosis was made by biopsy and immunohistochemistry in all cases. Median time from diagnosis of SS to the date of death was 2 years (range, 0.5-7 years). Treatment relied on PUVA, methotrexate, interferon and UVB phototherapy respectively for patients 1 to 4.

Conclusions: Recent advances in research have greatly improved our characterization and treatment of SS. There remains, however, a great need for better tools for early diagnosis, risk stratification, and more effective therapy.