Introduction: Primary cutaneous anaplastic large cell lymphoma (pcALCL), the second most common cutaneous T-cell lymphoma (CTCL), generally has a good prognosis with the 5-year disease-specific survival (DSS) ranging between 85% and 95%. There have been several papers on prognostic factors, most of which report on Caucasian populations. Exploring the prognostic factors in cohorts with other races is important because the frequency and clinical presentation in cutaneous lymphomas are different among ethnicities. In addition, clinical implications of *DUSP22* rearrangement and the association between *DUSP22* rearrangement and lymphoid enhancer-binding factor 1 (LEF1) expression pattern in pcALCL remains to be elucidated, although they have been studied in ALK-negtive systemic ALCL. From this perspective, we conducted a multicenter observational study to examine prognostic factors of pcALCL and the clinical implication of *DUSP22* rearrangement in pcALCL, lymphomatoid papulosis (LyP), and CD30-positive mycosis fungoides (MF) with large cell transformation (MF-LCT) in Japan.

Methods: Multicenter retrospective study including patients with pcALCL, LyP, and MF-LCT diagnosed between January 1, 2000 and December 31, 2018 at six large tertiary hospitals in Japan. Baseline data at diagnosis, treatment course, overall survival (OS), and DSS were collected. Immunohistochemical analysis including LEF1 expression and fluorescence in situ hybridization to detect *DUSP22* and *TP63* rearrangement were performed using skin samples at diagnosis. We investigated the association between staining pattern and these gene rearrangements. We also assessed the prognostic implications of clinical status, immunohistochemical results and the presence of gene rearrangements.

Results: *DUSP22* rearrangement was detected in 50% of pcALCL cases (11/22), but not in any cases with LyP (0/14) or MF-LCT (0/11). *TP63* rearrangement was not detected in any case. Clinically, pcALCL patients with DUSP22 rearrangement did not tend to form ulcers ($p = 0.081$). There was no significant association between DUSP22 rearrangement status and immunohistochemical results including *LEF1* expression pattern. T3 stage and the presence of lower limb lesions were significantly associated with shorter OS ($p = 0.012$ and 0.021, respectively by log-rank test). Similarly, they were significantly correlated with

shorter DSS (p = 0.016 and 0.0001, respectively). The presence of *DUSP22* rearrangement tended to be associated with longer OS (p = 0.061), but it had no effect on DSS (p = 0.751).

Conclusion: *DUSP22* rearrangement is relatively specific to pcALCL among CD30-positive CTCLs in Japan, but not associated with the prognosis in pcALCL. Moreover, different from ALK-negative systemic ALCL, LEF1 expression pattern was not related with DUSP22 rearrangement in pcALCL. We confirmed that T3 stage and the lower limb involvement were significantly associated with the decreased OS and DSS similar to the results of previous reports. Presence or absence of lower limb lesions should better be included in T stage subcategory in the future.