

Materials and Methods

Setting and Patients

The study was a retrospective analysis which included data from patients with a clinicopathological diagnosis of mycosis fungoides (MF), folliculotropic MF (FMF), or Sézary syndrome (SS) diagnosed between 1970 and 2016 and treated at Bispebjerg Hospital in Copenhagen, Denmark. Anonymized data were extracted from the CLDBase registry. The CLDBase registry and data extraction were approved by the Danish Data Protection Agency. All patients had confirmed clinicopathological diagnosis of MF, FMF, or SS. The main clinical and demographic features of the patients are listed in Table 1. Patient-related data, including age, sex, diagnosis, disease distribution, duration of disease, disease stage, response to initial treatment course of PUVA, and date of last contact or death, were retrieved from the web-based clinical database for cutaneous lymphomas (www.cldbbase.org). PUVA monotherapy was the first choice of treatment for all patients with MF and FMF, except for patients with limited body surface area involvement in whom expectant observation or superficial radiotherapy was preferred, as well as erythrodermic patients. In patients with stage IIB or IV disease, radiation of 12–18 Gy (40–50 kV) of thick plaques or tumors was often used as supplementary treatment. Satisfactory response to PUVA was defined as complete response (no clinically visible lesions), very good partial response (<1% involvement), or partial response (>50% decrease in affected surface area). The results of directed tumor radiation were not included in the definition of PUVA responses.

Analysis

Baseline characteristics were summarized using descriptive statistics. Predictors of prognosis were determined using Cox regression. The following variables were controlled for: age, disease stage, sex, and diagnosis. Survival was calculated from the date of diagnosis to the date of death or last follow-up. To compare survival between FMF and MF groups, 24 patients with each diagnosis were matched for age (± 5 years), sex, and stage. The Kaplan-Meier method was used to determine survival for the matched control groups. All statistics were done using SPSS and results with $p \leq 0.05$ were considered significant.

A meta-analysis was performed using Review Manager (RevMan, version 5.3; Copenhagen: The Nordic Cochrane Centre, The Cochrane Collaboration, 2014). A random effects model was applied to calculate pooled risk ratios and 95% confidence intervals for the survival of FMF patients compared to MF patients (control) at 5 and 10 years. Study heterogeneity was assessed by estimating an I^2 value, where values >50% were heterogeneous. A risk ratio >1 favors conventional MF (control), suggesting an increased risk for survival compared to patients with FMF. Risk ratios <1 favor FMF, suggesting a decreased risk for survival compared to patients with MF.