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African American and Caucasian patients with Sézary syndrome have no differences in outcomes at an ethnically diverse urban medical center

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Abstract

Sézary syndrome (SS) is an aggressive cutaneous T-cell lymphoma with poor survival. We performed a retrospective review of SS patients at Emory University from 1990 to 2020. We collected data on race, clinical characteristics, therapy, and social determinants of health. Clinical endpoints were overall survival (OS) and time to next treatment (TTNT). Univariate association and multivariable analyses were assessed by Cox proportional hazards models. Among 62 patients, 45.2% were AA. The median OS and TTNT were 3.1 years and 6.3 months, respectively, with no difference by race. AA patients had a higher median baseline LDH (360 *vs.* 232, $p = 0.002$) and a longer delay in initiation of systemic therapy compared to CC patients (3.17 *vs.* 2.14 months, $p = 0.039$), but a shorter commute (<10 miles) and no difference in insurance coverage ($p = 0.260$). AA patients at an academic center had unique clinical features and treatment patterns, but similar survival to CC SS patients.

Keywords

Mycosis fungoides; Sézary syndrome; extracorporeal photopheresis; African American; racial disparities

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Ethical approval

This study was reviewed and approved by IRB; approval # 0001489.

Presentations

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Disclosure statement

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Introduction

Sézary syndrome (SS) is an aggressive, leukemic subtype of cutaneous T-cell lymphoma (CTCL) that classically presents with erythroderma and diffuse lymphadenopathy [1]. Sézary syndrome (SS) has a poor prognosis with a median survival of 2–4 years from the time of diagnosis [2]. SS is defined as a Sézary cell count >1000/microliter (stage B2) with erythroderma covering 80% of body surface area [3]. Sézary cells are atypical circulating lymphocytes that are identified by flow cytometry as having a postthymic, helper phenotype ($CD3^+$, $CD4^+$, $CD8^-$) with variable expression of CD7 and CD26 [4].

Given that SS can involve the skin, lymph nodes, blood, and viscera, effective treatment typically requires a multimodal approach with skin-directed therapy (SDT) and systemic therapy [5]. SS is typically poorly responsive to chemotherapy, therefore non-chemotherapy systemic treatment regimens, such as interferon-alpha or gamma, oral retinoids, and extracorporeal photopheresis (ECP) are often recommended for initial therapy. Other treatments include histone deacetylase (HDACi) inhibitors, chemotherapy, and mogamulizumab. Currently, there is no consensus on first-line treatment of SS, and treatment recommendations are largely based on expert opinion [6].

Importantly, there are limited data on prognostic biomarkers and racial disparities in patients with SS. A subgroup analysis of outcomes for AA patients in the MAVORIC trial, an international phase III clinical trial comparing mogalizumab *vs.* vorinostat in relapsed/refractory mycosis fungoides (MF) and SS, showed significant clinical differences compared to non-AA patients [7]. AA patients had a younger median age at enrollment, higher rates of early-stage disease (IB-IIA) at enrollment, and were more likely to have MF than SS. In a single-center retrospective study of outcomes in AA patients with MF/SS, Geller et al. found that hypopigmentation was associated with improved outcomes, while plaque disease, nodal disease, and elevated lactate dehydrogenase (LDH) were significantly associated with poor clinical outcomes [8]. It should be noted, however, that both aforementioned studies included early-stage disease and patients with MF.

Given its multi-compartment involvement, evaluation of response and decisions to change therapy in SS remain challenging [9]. In this study, we report treatments patterns and a comparison of disease characteristics and healthcare access between Caucasian (CC) and African American (AA) patients with SS at our institution.

Methods

Patients and data

We conducted a retrospective review of patients seen at the Winship Cancer Institute and Emory University Hospital diagnosed with SS between the years 1990 and 2020. This study was reviewed and approved by the Emory University Institutional Review Board (approval # 0001489). Patients were selected from our internal cutaneous lymphoma database, which identified patients from physician schedules and data warehouse queries using ICD-10 codes associated with Sézary Syndrome. Patients were eligible if they had histopathologic confirmation of B2 blood involvement at any point in their disease course. Of 650 patients

with CTCL in our database, 62 patients with Sézary syndrome were analyzed. The date of diagnosis was chosen as the date of SS diagnosis, regardless of the institution where the initial diagnosis was made. Clinical data collected from the electronic medical record included demographics, baseline laboratory values, disease characteristics, zip code, type of health insurance, and therapy. Distance to the cancer center was calculated as the straight-line distance in miles from a patient's home address zip code centroid to the cancer center address. Patients were categorized as having a short (<10 miles), intermediate (10–50), or long (≥50 miles) commute to our cancer center. We used the date of histologic confirmation of CTCL as the date of diagnosis. We collected Sézary cell counts, CD7 expression, CD26 expression, and CD4:CD8 ratios from flow cytometry samples at baseline and these values were excluded if the sample was collected after initiation of the first-line systemic therapy. Regular dermatology follow-up was defined as at least two outpatient visits over a 1-year period or longer. The date of death was abstracted from medical records or public obituary notices.

Statistical analysis

Clinical outcomes were measured by overall survival (OS) and time to next treatment (TTNT). The date of SS diagnosis was used for all survival analyses, including for patients who had a prior diagnosis of CTCL. OS was measured from the time of diagnosis to the date of death or last follow-up. TTNT was used as a surrogate for the duration of clinical benefit and was defined as the time from the start of the first line of therapy until the initiation of the subsequent therapeutic regimen [10]. Descriptive analysis was performed for each variable and a comparison between AA and CC patients was performed using ANOVA for numerical covariates and the Chi-square test or Fisher's exact test for categorical covariates. Kaplan-Meier curves for OS and TTNT were generated for the whole cohort. A Kaplan-Meier curve was also generated to compare the time from diagnosis to initiation of first systemic therapy stratified by race along with the log-rank p-value. The univariate association of baseline variables with OS and TTNT was assessed by Cox proportional hazards models and the multivariable analyses were performed on variables that had p-values <0.05 on univariate association. Statistical analysis was conducted using SAS 9.4 (SAS Institute Inc., Cary, NC, USA), and statistical significance was assessed at the 0.05 level.

Results

Demographic information and disease characteristics

Most patients were males (58.1%) and the median age at diagnosis was 65.9 years (Table 1). Nearly one-half (45.2%) of patients were AA, which is a similar proportion of AA patients in our existing cutaneous lymphoma database (42%). The median follow-up was 2.1 years (range, 0.3–13.2 years). A total of eight patients developed SS after an initial diagnosis of CTCL; the stage at diagnosis for these patients was 1 A (n = 1), 2 A (n = 1), 3 A (n = 1), and 3B (n = 5). Of these, four were AA and four were Caucasian. The median Sézary count at diagnosis was 1320 cells/uL. Among patients with available T-cell receptor (TCR) rearrangement results, 27 (43.5%) had clonal TCR in the skin and 35 (56.5%) had clonal TCR in the blood. A minority of patients (n = 8, 12.9%) had large cell transformation (LCT) (six skin, one lymph node, one both skin and lymph node). Lymph node involvement

was common. Abnormal lymph nodes clinically or pathologically were noted in 67.7% of patients (Nx, N1, or N2 in 33, and N3 in 9). Visceral metastasis was rare ($n = 2$).

Clinical outcomes and prognostic markers

The median OS and TTNT for the overall cohort were 3.1 years and 6.3 months, respectively. A subset of patients ($n = 10$, 16.1%) passed away within 1 year of diagnosis but most patients ($n = 34$, 54.8%) had a TTNT > 6 months. Patients who did not receive ECP showed a trend toward shorter OS (HR: 1.67, 95% CI: 0.91–3.06, $p = 0.098$) and TTNT (HR: 1.65, 95% CI: 0.98–2.80, $p = 0.062$) in univariate association (Table 2). Patients who did not have regular dermatology follow-up had significantly shorter OS (HR: 2.75, 95% CI: 1.27–5.93, $p = 0.010$) and TTNT (HR: 2.72, 95% CI: 1.34–5.51, p 0.005). On multivariable analysis, elevated WBC (10^3 cells/μL) and LDH (units/L) were significantly associated with shorter OS (WBC HR: 1.05, 95% CI: 1.01–1.08, $p = 0.01$; LDH HR: 1.003, 95% CI: 1.001–1.005, $p = 0.011$) and shorter TTNT (WBC HR: 1.04, 95% CI: 1.002–1.08, $p = 0.041$; LDH HR: 1.002, 95% CI: 1.001–1.004, $p = 0.048$).

Analysis by self-reported race

In an analysis by self-reported race, AA patients had a higher female: male predominance compared to CC patients (53.6% female *vs.* 28.1% female, respectively, $p = 0.045$, Table 1). AA patients also had lower median hemoglobin (12.6 *vs.* 14.3, $p = 0.036$) and higher median LDH (360 *vs.* 232, $p = 0.002$) at diagnosis. Though not statistically significant, there was a higher rate of large cell transformation (21.4 *vs.* 6.3%, $p = 0.084$), a lower median WBC count (10.4 *vs.* 16 × 10^3/μL, $p = 0.849$), and a lower median monocyte count (1.01 *vs.* 0.6 × 10^3/μL, $p = 0.061$) among AA patients. There were no differences in median age at diagnosis or nodal stage. AA patients were significantly more likely to have a short commute (<10 miles) to our cancer center compared to CC patients (39.3 *vs.* 6.25%, $p = 0.006$). There was no significant difference in health insurance patterns by race (AA: 7.1% uninsured, 35.7% public health insurance, 57.1% private health insurance; CC: 15.6% uninsured, 18.8% public health insurance, 65.6% private health insurance, $p = 0.260$).

There was no significant difference in median OS (Figure 1, CC: 30 months, AA: 48 months, $p = 0.227$) or median TTNT (Supplemental Figure 1, $p = 0.183$) by race per Kaplan–Meier estimation.

Treatment information

Systemic treatments are summarized in Table 3. The median number of systemic therapies was 3.0 (range: 0–11 lines). The median time from diagnosis to first systemic therapy was 2.4 months. The most common first-line systemic therapies were oral retinoids (43.5%), ECP (32.3%), and interferon (30.6%) (Table 3). More than half of patients ($n = 32$, 52%) received ECP within the first 3 lines of systemic therapy. The median time from diagnosis to treatment with ECP was 28.0 months. HDAC inhibitors and total skin electron beam (TSEB) radiation were other common treatments (46.8% received HDAC inhibitors, 38.7% received TSEB), but these were rarely used in the first-line setting.

Some differences in treatment patterns emerged by race. AA patients had a longer delay in time to first systemic therapy compared to CC patients (3.17 *vs.* 2.14 months, $p = 0.039$, Figure 2) and were more likely to receive HDACi (AA 64.3 *vs.* 28% for CC, $p = 0.005$). There was a trend toward decreased ECP treatment (AA 39.3 *vs.* CC 64.5%, $p = 0.053$), and a significantly longer time to ECP initiation (37.7 *vs.* 8.0 months, respectively, $p = 0.009$, Supplemental Figure 2). There were no baseline differences among patients who did *vs.* did not receive ECP in terms of median age, nodal involvement, WBC, LDH, or LCT. Additionally, there was no difference in receipt of systemic therapy overall, TSEB, or lines of therapy by race.

Discussion

In this study, we highlight racial differences in SS patients treated at our institution over a 30-year period. We found AAs had elevated LDH, longer delay to systemic treatment, lower rates of ECP, but higher rates of HDACi treatment compared to CC patients; this was offset by a shorter commute distance and no difference in health insurance. The median overall survival was 3.1 years and did not differ by race.

African Americans are underrepresented in clinical datasets among patients with CTCL and Sézary syndrome [11,12]. Large US-based registry studies have demonstrated that AA CTCL patients have up to twice the risk of death, coupled with inferior social determinants of health as indicated by decreased insurance coverage, a lower median income, and a lower likelihood of treatment in academic settings [13,14]. This is the first study, to our knowledge, to present data suggesting that AA patients with Sézary syndrome may have different treatment patterns than Caucasian patients. We found a significantly longer delay in systemic treatment initiation and decreased ECP yet increased rates of HDACi among AA patients with SS. ECP was less frequently received (63% in CC *vs.* 38% in AA, $p = 0.053$), and significantly delayed in AA, with a median time to ECP of 3.2 years in AA compared to 8 months in CCs (Supplemental Figure 2). Conversely, over twice as many AA received HDAC inhibitors (64.3 *vs.* 28.1%, respectively). There were no differences in systemic therapy, lines of therapy, receipt of TSEB, or likelihood of consistent dermatology follow-up at our institution by race. ECP is only offered at specialized institutions and is time-intensive, while HDAC inhibitors are more easily administered. Differences in ECP usage may be clinically relevant given its possible association with improved survival and TTNT [15]. Patients at our institution who received ECP showed a trend toward improved OS and TTNT. It should be noted, however, that guarantee bias may be contributing to this observation. On the other hand, HDACi is associated with a significantly shorter TTNT compared to ECP [16].

We also found racial differences in baseline clinical characteristics. AA patients at our institution had a female predominance, higher LDH, lower hemoglobin, and CD7 expression loss on flow cytometry [17,18] compared to CCs. However, these observed differences in laboratory values, particularly hemoglobin, may be related to natural differences between races rather than an indicator of more advanced disease at diagnosis [19]. Unlike prior studies, we did not show differences in age or nodal involvement [20]. Likewise, we noted increased numbers of LCT in AA patients ($n = 6$) compared to Caucasians ($n = 2$). While

this did not reach statistical significance, it is likely clinically significant and suggests biological differences.

Despite increased risk factors in the AA patients, such as elevated LDH and delay in systemic therapy, overall survival was not significantly different from CCs. This equity in survival outcomes may be related to improved access in our population: AA patients had a shorter commute (<10 miles, $p = 0.006$) to our cancer center and no differences in health insurance coverage compared to CC patients. The lack of difference in survival contrasts with registry studies which have shown AA race to be independently associated with worse OS and disease-specific survival (DSS) independent of stage [21–23]. However, AA patients treated at large urban centers have may have improved outcomes. A recent study of 157 Black MF/SS patients from Memorial Sloan Kettering Cancer Center found no association between socioeconomic parameters and prognosis [11]. Similarly, black-white disparities were diminished in a prior study of CTCL patients treated at our institution [24]. The population of Georgia is 32% AA, and also has the largest AA middle class in the nation [25]. These factors suggest that improved healthcare access may have contributed to the outcomes of AA patients at our institution.

This study had several limitations. First, this was a retrospective analysis and was inherently vulnerable to selection bias. Furthermore, the retrospective nature of this study made it challenging to accurately determine the duration from symptom onset to diagnosis. Our study was small with only 62 patients, hence, this is an exploratory analysis that is primarily descriptive in nature. Despite its size, this study represents the largest AA SS cohort to date [7,8,26,27]. Statistical power in treatment pattern analysis was limited by a large number of unique therapies and response assessments were inconsistent. This study spans a wide era of treatment of disease choices and supportive care options. Brentuximab vedotin and mogamulizumab were only approved recently and only a small number of patients received these agents. This may not have greatly affected our results as a *post-hoc* analysis of the MAVORIC study demonstrated no difference in outcomes or response between AA and CC patients receiving mogamulizumab [20]. Although we noted that AA patients were less likely to receive ECP, we did not account for reasons, such as the risk of bacteremia, venous access issues, or patient preference [28]. Furthermore, differences in treatment patterns may reflect differences in presentation. For example, AA patients more frequently present with tumor stage and large cell transformation and may receive TSEB therapy in the first line for SS. This may be because erythema in early-stage disease may be more difficult to appreciate in AA patients compared to CC patients. We identified an association between consistent dermatology follow-up and improved outcomes. Community and some academic dermatologists are more apt to follow CTCL patients with limited-stage, confounding the association with outcomes. Another major limitation was missing or incomplete data. We were missing baseline laboratory data for approximately half of the patients in our study, limiting the power and validity of our assertions on prognosis. Lastly, social determinants of health, such as income level, education, and neighborhood socioeconomic status were not included and would greatly aid future studies. Nevertheless, our robust data involving treatment patterns and assessments of racial differences are important novel contributions to the field.

Conclusions

We describe treatment patterns, risk factors, and outcomes in an ethnically diverse cohort of SS patients followed over 30 years. AA patients with SS had distinct treatment patterns and high-risk features including elevated LDH and delayed initiation of systemic therapy, yet no difference in survival compared to Caucasians. Lack of survival differences among AA patients may be related to improved healthcare access in our population. These data are hypothesis-generating and should be validated in larger, prospective studies.

Supplementary Material

Refer to Web version on PubMed Central for supplementary material.

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Data availability statement

Data were available within the article or its supplementary materials.

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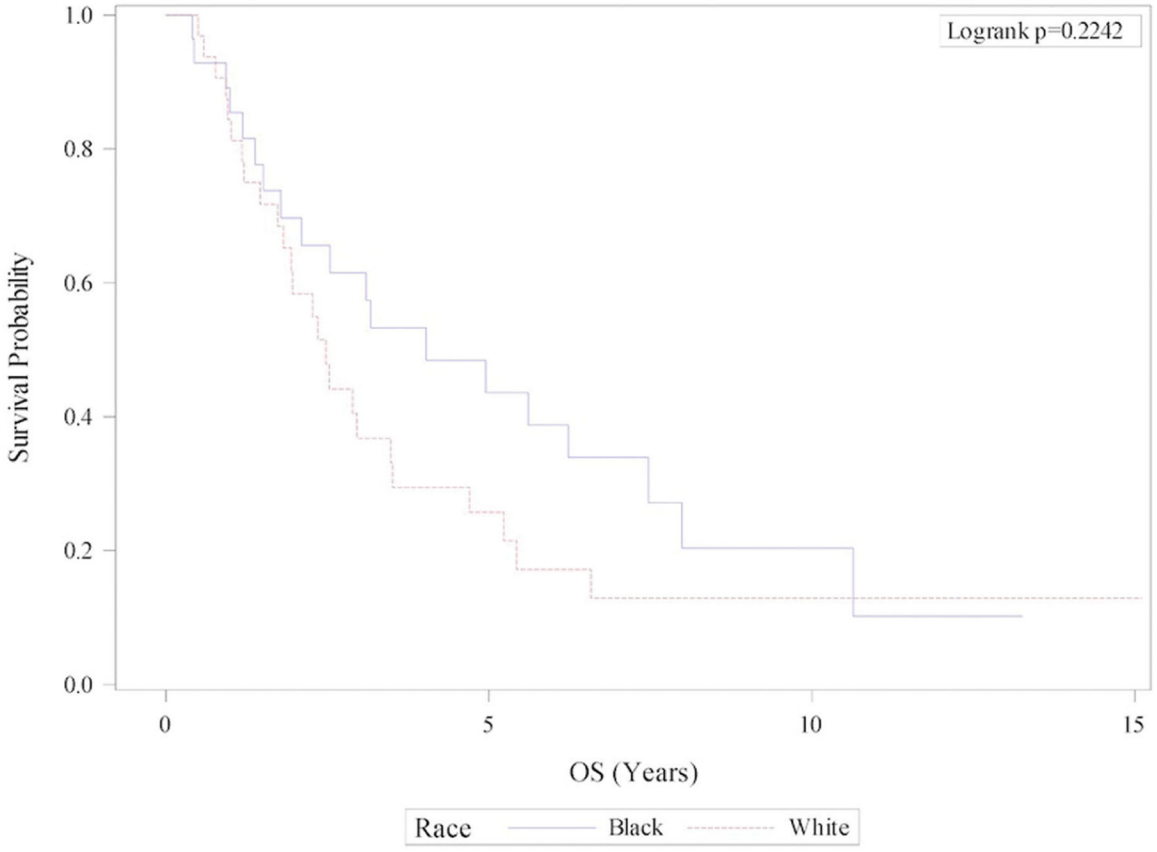

Figure 1.
Kaplan–Meier curves for overall survival (OS) stratified by race.

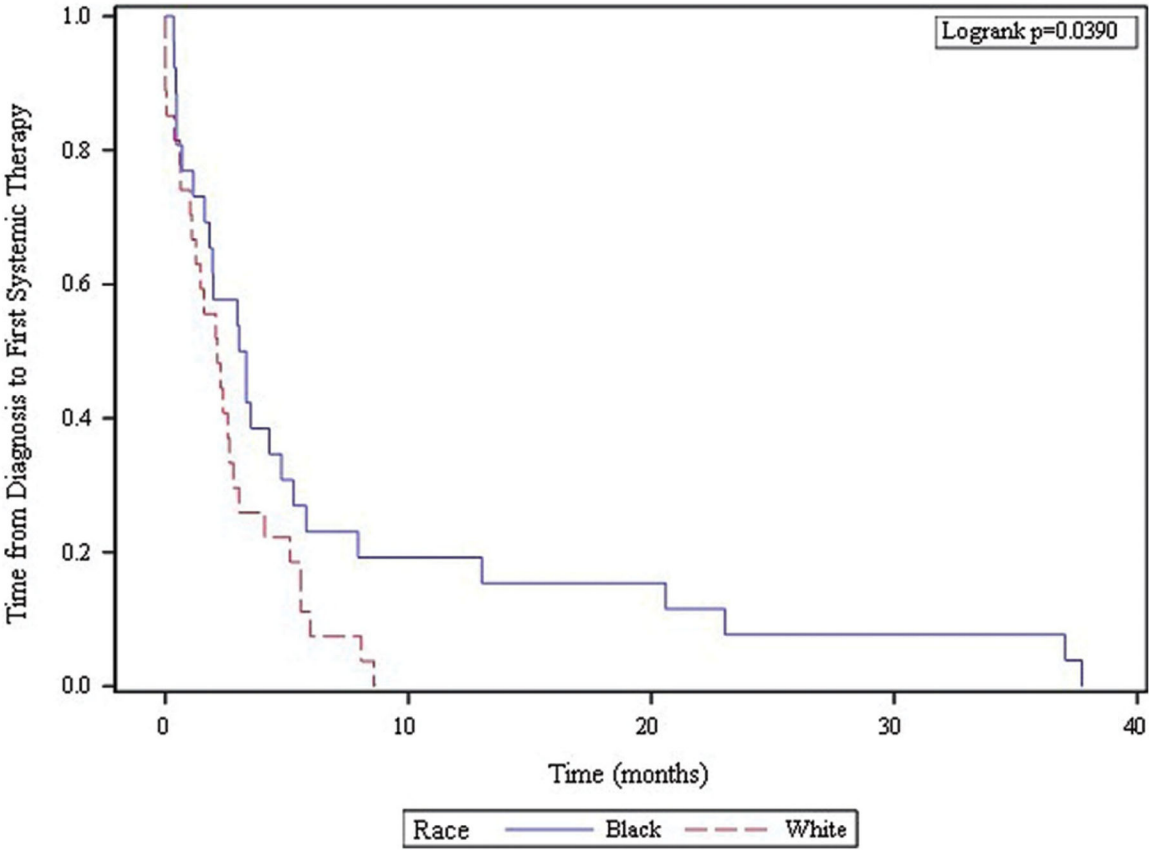

Figure 2.
Kaplan–Meier curves for time from diagnosis to initiation of first systemic therapy stratified by race.

Table 1.

Comparison of descriptive statistics by race.

	Caucasian (*n* = 32)	AA (*n* = 28)	*p*-value
Demographics and baseline disease characteristics			
Female gender	*n* = 9, 28.1%	*n* = 15, 53.6%	0.045 *
Median age (years)	66.1	64.4	0.104
Median Sezary count at diagnosis (Cells/L) [a]	1926	1320	0.764
N0 stage at diagnosis	*n* = 9, 28.1%	*n* = 8,29.6%	0.899
+ TCR rearrangement (skin) [b]	*n* = 13, 76.5%	*n* = 13, 81.3%	0.137
+ TCR rearrangement (blood) [c]	*n* = 19, 82.6%	*n* = 15, 65.2%	0.208
Large cell transformation	*n* = 2, 6.3%	*n* = 6, 21.4%	0.084
$CD7^-$ at diagnosis [d]	*n* = 14, 77.8%	*n* = 16, 100%	0.045 *
Median laboratory values at diagnosis			
Hgb [e]	14.3 g/dL	12.6g/dL	0.036 *
LDH [d]	232 U/L	360 U/L	0.002 *
WBC [f]	16.0 × 10^3/μL	10.4 × 10^3/μL	0.849
ANC [e]	5.33 × 10^3/μL	4.62 × 10^3/μL	0.450
AMC [e]	1.01 × 10^3/μL	0.6 × 10^3/μL	0.061
Platelets [e]	227 × 10^3/μL	278 × 10^3/μL	0.708
Albumin [d]	3.8 g/dL	4.0g/dL	0.503
Treatment and follow-up data			
Received systemic therapy at any point	*n* = 27, 87.1%	*n* = 25, 92.6%	0.493
Median time from diagnosis to first systemic therapy	2.14 months	3.17 months	0.039 *
Median total number of systemic therapies received	2.5	3.5	0.252
Received ECP at any point	*n* = 20, 64.5%	*n* = 11, 39.3%	0.053
Received HDACi at any point	*n* = 9, 28.1%	*n* = 18, 64.3%	0.005 *
Received TSEB at any point	*n* = 11, 35.5%	*n* = 12, 42.9%	0.562

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	Caucasian (*n* = 32)	AA (*n* = 28)	*p*-value
Dermatology follow-up **	*n* = 13, 54.2%	*n* = 14, 77.8%	0.114
Median OS	30.0 months	48.0 months	0.224
Median TTNT	6.6 months	5.8 months	0.181

AA: African American; TCR: T-cell receptor; Hgb: hemoglobin; LDH: lactate dehydrogenase WBC: white blood cell count; ECP: extracorporeal photopheresis; TSEB: Total skin electron beam radiation.

*Statistically significant at the level of $p < 0.05$.

**Defined as at least two outpatient visits for at least 1 year.

[a]Missing data from 10 patients.

[b]Missing data from 27 patients.

[c]Missing data from 14 patients.

[d]Missing data from 26 patients.

[e]Missing data from 22 patients.

[f]Missing data from 20 patients.

Table 2.

Univariate and multivariable analysis of the association between select variables and clinical outcomes.

	OS		TTNT	
	HR (CI)	***p*-value**	**HR (CI)**	***p*-value**
Univariate association				
Did not receive ECP at any time	1.67 (0.91–3.06)	0.098	1.65 (0.98–2.80)	0.062
No dermatology follow-up	2.75 (1.27–5.93)	0.010 *	2.72 (1.34–5.51)	0.005 *
Multivariable analysis **				
WBC (10^3 cells/μL)	1.05 (1.01–1.08)	0.01 *	1.04 (1.002–1.08)	0.041 *
LDH (units/L)	1.003 (1.001–1.005)	0.011 *	1.002 (1.001–1.004)	0.048 *

OS: overall survival; TTNT: time to next treatment; HR: hazard ratio; CI: 95% confidence interval; ECP: extracorporeal photopheresis; WBC: white blood cell count; LDH: lactate dehydrogenase.

* Statistically significant at the level of $p < 0.05$.

** WBC and LDH were analyzed as continuous variables. Number of observations in original data set = 62. Number of observations used in multivariable analysis = 29.

Table 3.

Distribution of treatment regimens received after diagnosis of Sézary syndrome.

Treatment regimen	First-line (*n*, %)			Any line (*n*, %)		
	All patients *n* = 62*	**Caucasian *n* = 32**	**African American *n* = 28**	**All patients *n* = 62**	**Caucasian *n* = 32**	**African American *n* = 28**
Oral retinoids	29 (43.5%)	16 (40.6%)	13 (46.4%)	40 (64.5%)	22 (68.8%)	18 (64.3%)
Extracorporeal photopheresis (ECP)	20 (32.3%)	13 (40.6%)	6 (21.4%)	33 (53.2%)	21 (65.6%)	11 (39.3%)
Interferon	19 (30.6%)	13 (40.6%)	5 (17.9%)	31 (50.0%)	18 (56.3%)	13 (46.4%)
Methotrexate	6 (9.7%)	4 (12.5%)	2 (7.1%)	11 (17.7%)	6 (18.8%)	5 (17.9%)
Total skin electron beam (TSEB) radiation	5 (8.1%)	4 (12.5%)	1 (3.6%)	24 (38.7%)	11 (34.4%)	12 (42.3%)
Single agent chemotherapy	4 (6.5%)	0 (0.0%)	4 (14.3%)	11 (17.7%)	3 (9.4%)	8 (28.6)
Histone deacetylase inhibitor (HDACi)	3 (4.8%)	1 (3.1%)	2 (7.1%)	27 (46.8%)	9 (28.1%)	18 (64.3%)
Combination chemotherapy	3 (4.8%)	2 (6.2%)	1 (3.6%)	11 (17.7%)	6 (18.8%)	5 (17.9%)
Denileukin diftitox	1 (1.6%)	0 (0.0%)	1 (3.6%)	4 (6.5%)	1 (3.1%)	3 (10.7%)
Pralatrexate	0 (0%)	0 (0.0%)	0 (0.0%)	10 (16.1%)	4 (12.5%)	6 (21.4%)
Mogamulizumab	0 (0%)	0 (0.0%)	0 (0.0%)	4 (6.5%)	1 (3.1%)	3 (10.7%)
Brentuximab vedotin	0 (0%)	0 (0.0%)	0 (0.0%)	4 (6.5%)	2 (6.2%)	2 (7.1%)

HDACi: histone deacetylase inhibitor; ECP: extracorporeal photopheresis; TSEB: total skin electron beam.

*One patient was Asian and one had an unspecified race.

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