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Overall and cause-specific mortality among patients with cutaneous T-cell lymphoma from the United States

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INTRODUCTION

Cutaneous T-cell lymphomas (CTCLs) are rare subtypes of Non-Hodgkin lymphoma with heterogeneous clinical courses. Though often discussed as chronic conditions with little effect on overall survival, data is lacking on long-term outcomes and causes of death.

METHODS

- Data source: November 2018 submission of the Survival, Epidemiology, and End-results (SEER) registry
- Subtypes included:
  - Mycosis fungoides (MF, ICD-O-3 9700)
  - Sézary syndrome (SS, 9701)
  - Primary cutaneous anaplastic large cell lymphoma (pcALCL, 9718)
  - Subcutaneous panniculitis-like T-cell lymphoma (SPTCL, 9708)
- Diagnosed between 2000 and 2018, age ≥ 15 at diagnosis
- Two- and 5-year OS and median OS determined using SEER*Stat Survival session
- Standardized mortality ratio (SMR) and absolute increased risk (AER) for death determined with SEER*Stat Multiple Primary-SIR session
- Incidence of cause-specific mortality estimated using cumulative incidence functions with Fine-Gray model (R Statistical Software, Version 4.3.1)
- Survival curves compared using log-rank test (R Statistical Software, Version 4.3.1)

RESULTS

CTCL was the most common cause of death among CTCL patients.

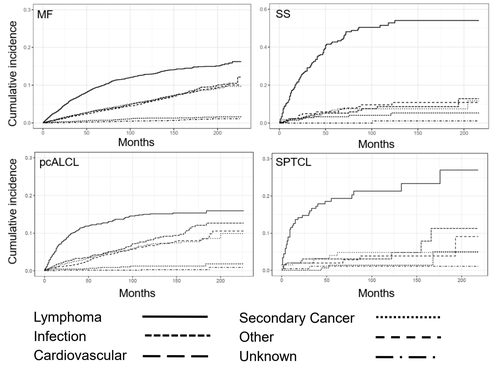


Figure 1. Cumulative incidence of cause-specific mortality by CTCL subtype

	MF	SS	cALCL	SPTCL
Patients diagnosed	7,957	272	1,452	205
Incidence (per 1,000,000)	6.1	0.21	1.1	0.16
Patient deaths	1,474	120	470	69
Follow up in months (median, range)	66, 0 - 227	26, 0-216	66, 0 - 226	48, 0 - 218
2-year OS, %	93.0	67.7	88.0	79.0
5-year OS, %	83.6	38.4	79.9	71.3
Median OS in months	NR	44.1	NR	NR

Table 1. Incidence and Kaplan-Meier analysis by CTCL subtype

CTCL patients were at increased risk of death from select secondary cancers and from infection.

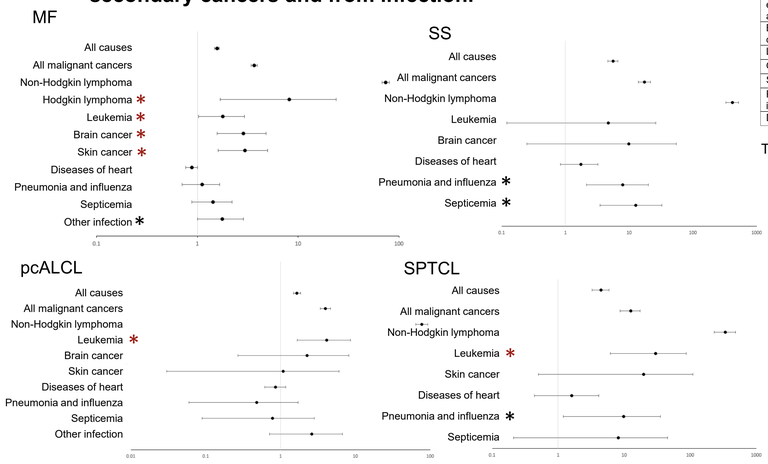


Figure 2. Standardized mortality ratio by histology

Despite increased risk of death from secondary cancers and infection, these causes contributed little to total mortality

	MF	SS
Non-Hodgkin Lymphoma	13.15	118.60
Skin cancer excluding basal and squamous	0.21	-0.16
Brain and CNS cancer	0.20	1.34
Leukemia	0.16	1.17
Other infection	0.16	-0.25
Septicemia	0.14	5.48
Pneumonia and influenza	0.06	5.19
Diseases of heart	-0.66	6.42

	pcALCL	SPTCL
Non-Hodgkin Lymphoma	14.92	31.08
Skin cancer excluding basal and squamous	0.01	0.99
Brain and CNS cancer	0.14	-0.06
Leukemia	0.68	3.01
Other infection	0.32	0.91
Septicemia	-0.07	-0.18
Pneumonia and influenza	-0.28	1.87
Diseases of heart	-0.827	1.573

Table 2. Absolute excess risk (AER) by CTCL subtype. AER is per 1,000 persons per year

CTCL accounted for a significant proportion of deaths, even in patients initially presenting with localized disease

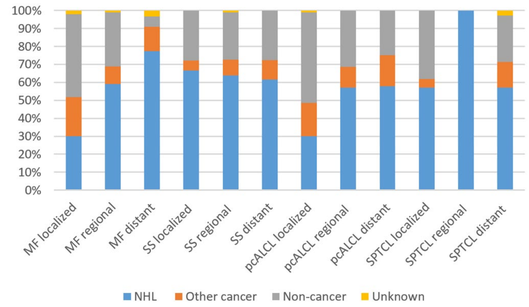


Figure 3. Cause of death by stage and CTCL subtype

CONCLUSIONS

- Patients with CTCL are at increased risk of all-cause mortality relative to age-matched population controls.
- Significant proportion of deaths are due to lymphoma even in those with initially localized disease: 30.1% (MF), 30.1% (pcALCL), 57.1% (SPTCL), 66.7% (SS).
- Even patients with localized disease at presentation have increased risk of all-cause mortality.
- Patients with CTCL had higher risk of mortality from secondary causes relative to controls - Leukemia (MF, pcALCL, and SPTCL), skin cancer (MF), infection (MF, SS, and SPTCL).
- Secondary causes made overall minor contributions to total mortality, however.
- We need better therapies and long-term follow-up of patients with CTCL regardless of stage at presentation.

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