Long Term Outcomes of Juvenile-Onset Mycosis Fungoides

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Disclosures

- I do **not** have any relevant financial relationships.

This presentation and/or comments will provide a balanced, non-promotional, and evidence-based approach to all diagnostic, therapeutic and/or research related content.

Objectives

- To assess survival outcomes in juvenile-onset mycosis fungoides (jMF)
- To assess stage progression in jMF patients
- To assess treatment failure in jMF patients

Methods

Included **119** patients diagnosed with biopsy-proven MF before age 20, seen between 1980-2023

Retrospective chart review of clinical data, including stage of disease and treatments

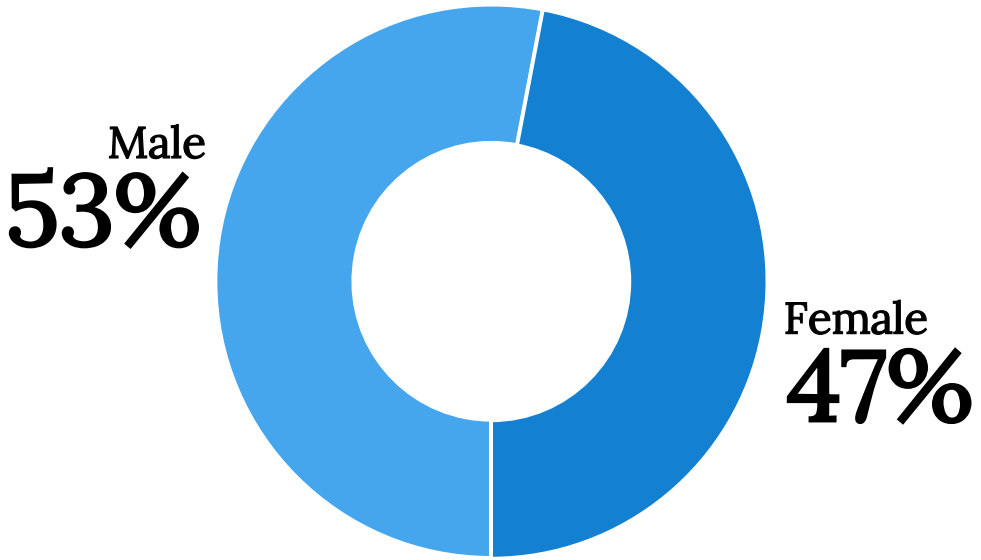
Prospective phone calls attempted to all patients to assess survival

Survival

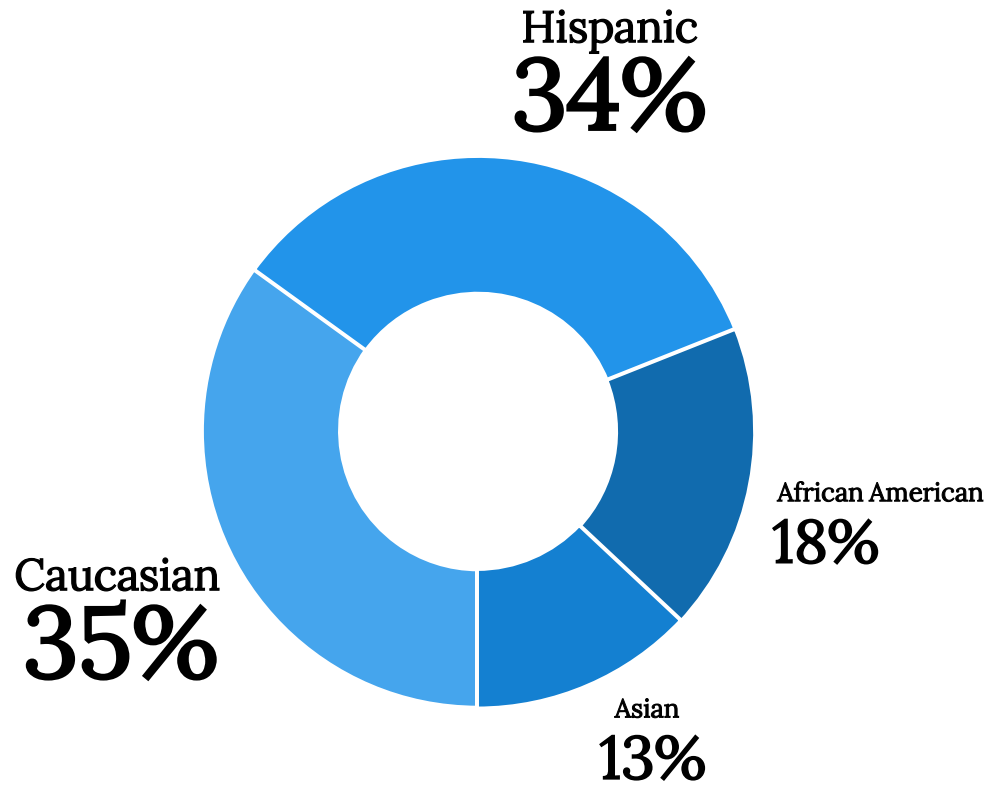
- Five-and 10-year overall survival (OS) was 99% (76 of 77 patients alive) and 97% (37 of 38 patients alive) respectively.
- One patient died of EBV reactivation in the setting of common variable immune deficiency syndrome (not related to MF).

Patient Demographics for 119 jMF patients

Sex

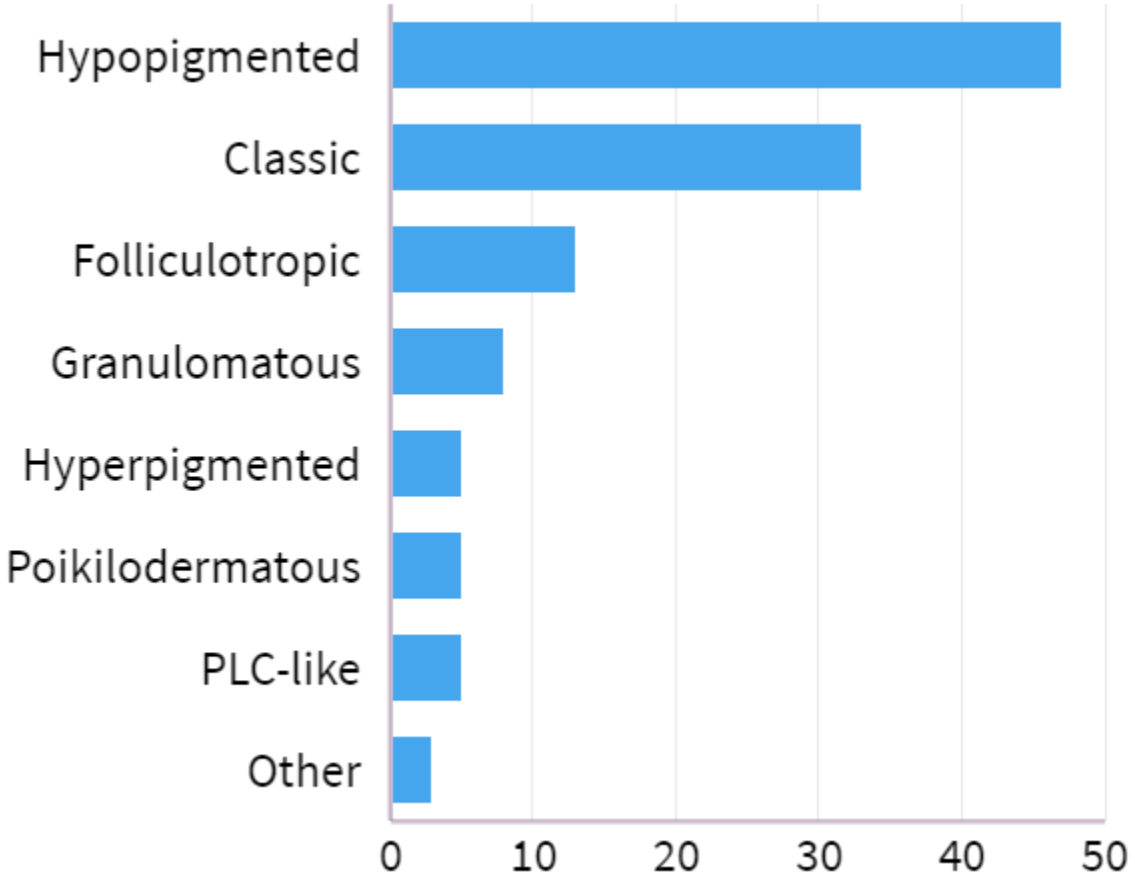


Race

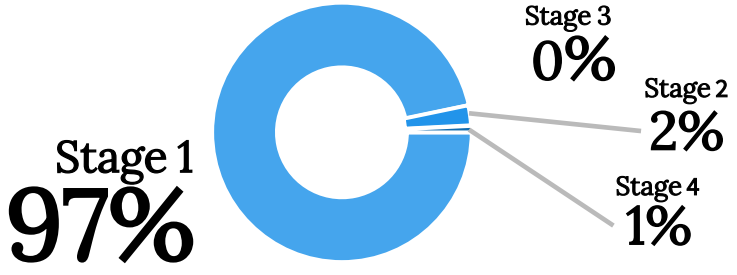


Clinical Characteristics at Diagnosis

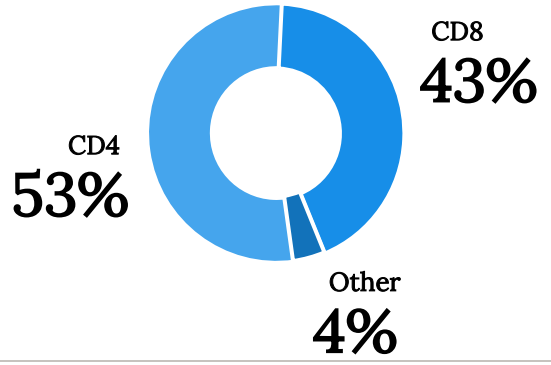
Variant at Diagnosis



Stage at Diagnosis



Phenotype at Diagnosis



Stage Progression

- Stage Progression (SP) was defined as per Olsen's criteria[1]. SP was seen in 8% of patients and median time to progression was 3.5 years (range:1-13 years).
- Sex and stage at diagnosis were not associated with SP ($p>0.05$).
- Granulomatous MF, Caucasian race, and CD4 dominant phenotype were associated with SP ($p<0.05$).
- Hypopigmented MF and CD8 dominant phenotype were associated with a non-progressive course ($p<0.05$).

Treatments Used	Narrow band UVB	50 (42%)
	Topical corticosteroids	30 (25%)
	Tazarotene	16 (13%)
	Nitrogen mustard	14 (12%)
	Local electron beam therapy	12 (10%)
	Psoralen plus UVA	11 (9%)
	Pegylated interferon 2-alpha	9 (8%)
	Bexarotene oral	6 (5%)
	Acitretin	5 (4%)
	Methotrexate	5 (4%)
	Broadband UVB	5 (4%)
	Bexarotene gel	4 (3%)
	Imiquimod	2 (2%)
	Total skin electron beam therapy	2 (2%)
	Extracorporeal photopheresis	2 (2%)
	Other systemic therapy*	4 (3%)
	Allogeneic Stem Cell Transplant	3 (3%)

*Romidepsin, brentuximab vedotin, vorinostat, pralatrexate, CART cell therapy

Treatment Failure

- Treatment failure (TF) was defined as regimen change secondary to lack of satisfactory response and not due to intolerance or treatment logistics. TF was seen in 15% of patients.
- Sex and race were not associated with TF ($p>0.05$).
- Advanced-stage disease (stage IIB and above), granulomatous and folliculotropic MF, LCT, CD4 phenotype were associated with TF ($p<0.05$).

Conclusion

- JMF is associated with an excellent prognosis with a **5-year and 10-year OS** of **97% and 95%** respectively (97% of patients were stage I).
- Stage progression was seen in **8%** of patients:
  - **Granulomatous MF, CD4-phenotype, & Caucasian race** were associated with progression. **CD8 phenotype and hypopigmented MF** were associated with a non-progressive course.
- Treatment failure was seen in **15%** of patients:
  - **Folliculotropic MF, Granulomatous MF, LCT, & advanced-stage disease** were associated with treatment failure.

Acknowledgements




From the left: Dr. Julia Dai, Dr. Madeleine Duvic, Dr. Auris Huen, Dr. Seda Purnak