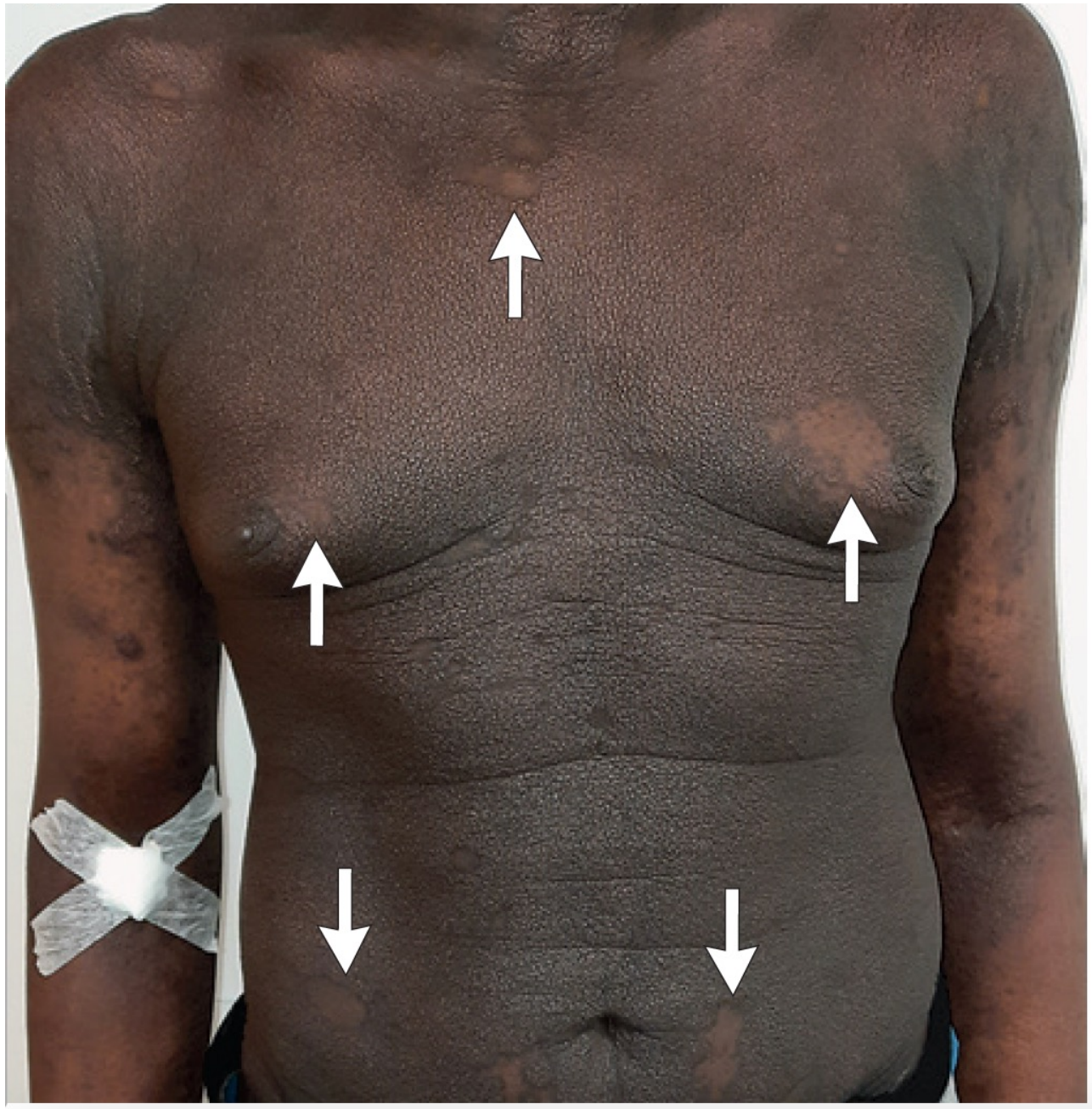
Secuenciando tratamientos en Micosis Fungoide y Síndrome de Sezary

Danielle Leao

LEX

03 abril 2025



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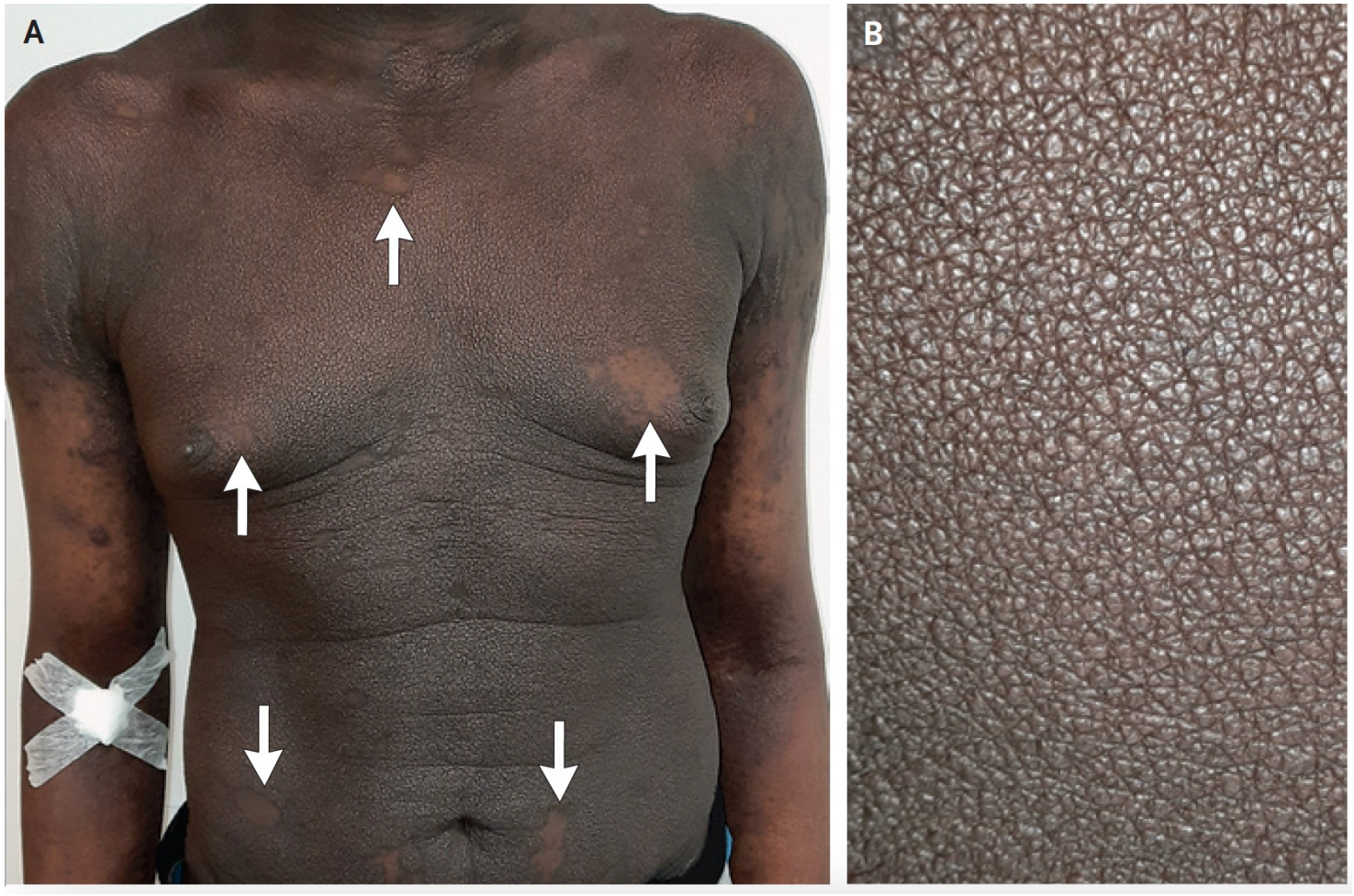
IMAGES IN CLINICAL MEDICINE



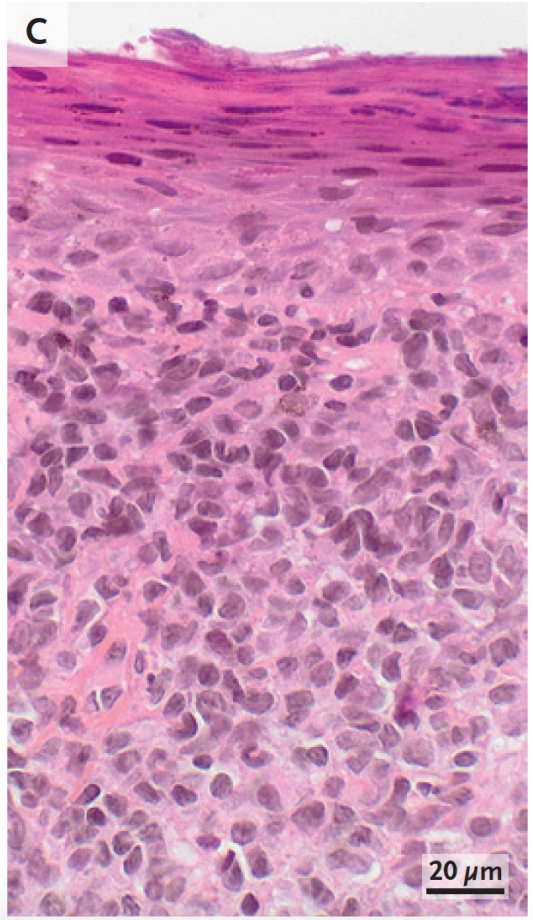
- *Confluent, well-demarcated, hyperpigmented plaques across the torso and upper arms*
- *Coalescence of small monomorphic papules*
- *Axillary and inguinal lymphadenopathy*

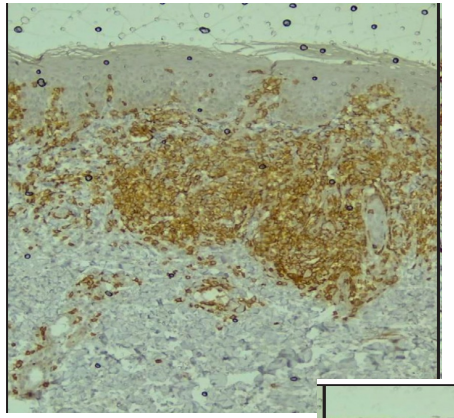
Un hombre de 27 años se presentó en la clínica de dermatología con un historial de 6 meses de prurito y erupción cutánea.

Heloise Mazoyer & Bachmeyer C. **Sorbonne Université, Paris.**
N Engl J Med 392;12 March 27, 2025

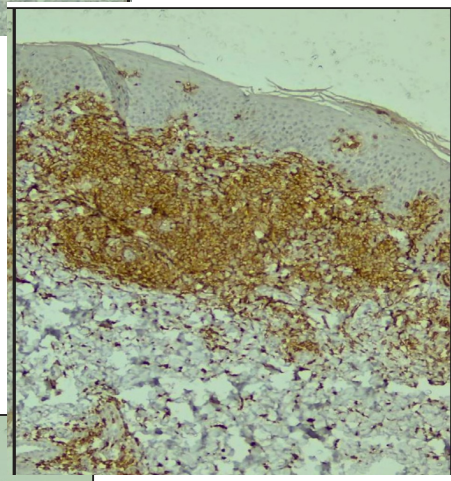
27y, male

- *Skin-biopsy: dense, perivascular, and pilotropic superficial dermal infiltrate of small T cells*
- *Positive: CD2, CD3, CD4, MUM1 and PD1*
- *Negative: CD7 and CD8.*

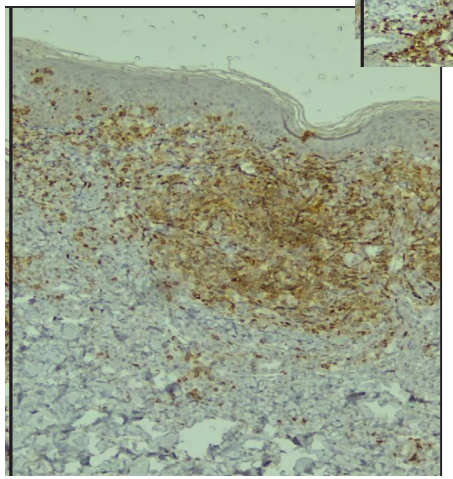




CD3+



CD4+



Relative loss of CD7

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N Engl J Med 392;12 March 27, 2025

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Limph node biopsy

This patient: dermatopathic lymphadenopathy with interfollicular atypical T cells.

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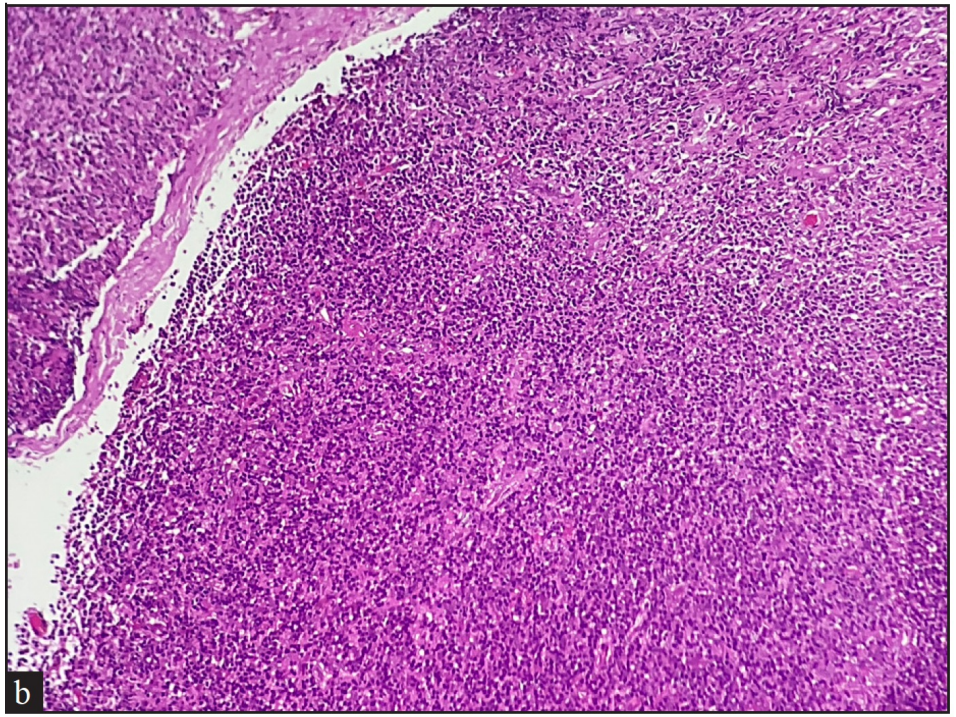
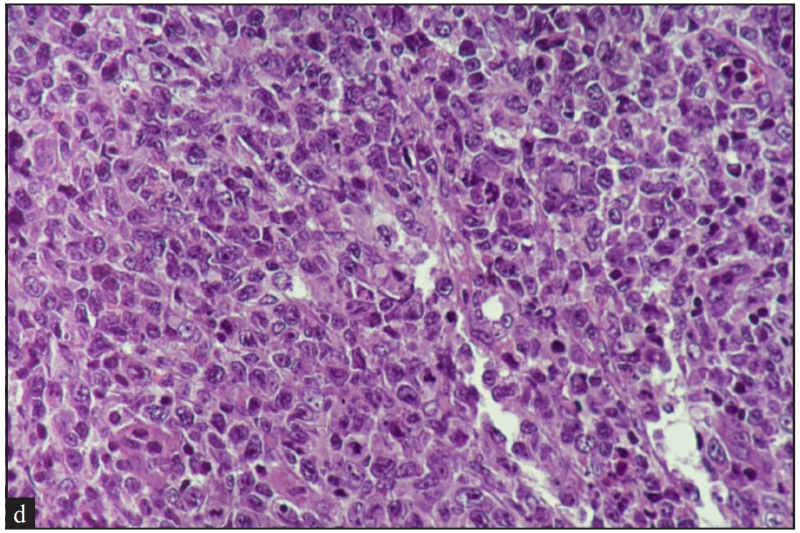
Another patient
to illustrate infiltration



Lymph node: complete effacement of the architecture (HE 20x).



Lymphocytes with atypical nuclei with coarse vesicular chromatin (HE, 40x)

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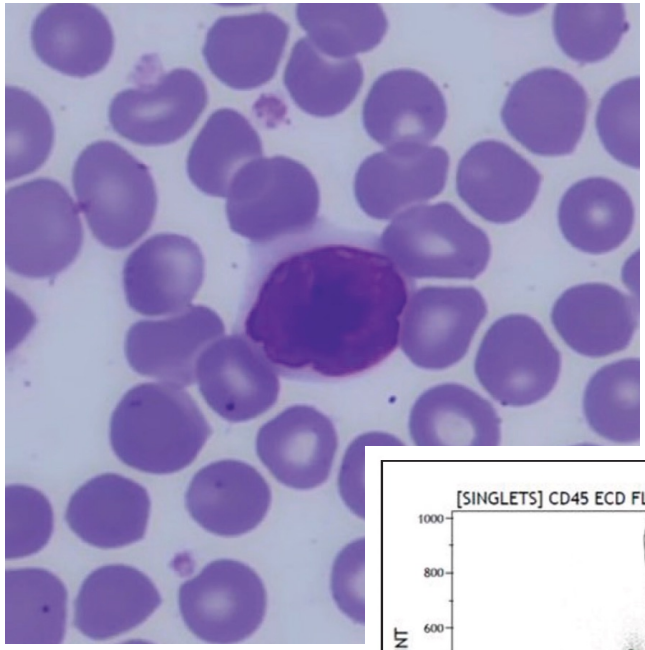
They called a doctor… Hematologists came!

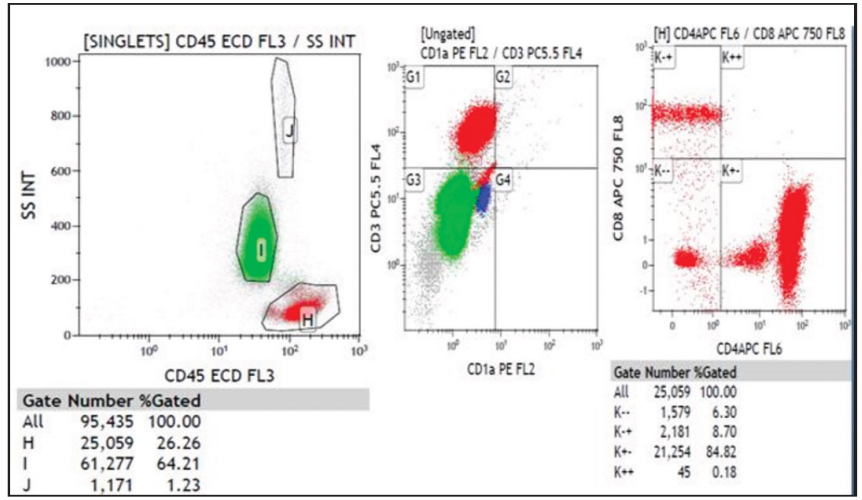
Peripheral-blood smear:

- *Enlarged, atypical lymphocytes with convoluted nuclei,*

Immunophenotyping and assays of T-cell clonality in blood and skin samples: "the same clone"



Sézary cells



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Cutaneous T-cell lymphoma (CTCL): S Sézary

 # Sindrome de Sézary

- 5% de los linfomas cutáneos primarios
- 80% en piel

 # Criterios:

- Eritrodermia (>80% superficie corporal)
- (às vezes) Linfadenopatía generalizada
- Células de Sézary en sangre (>1,000/µL)
- Relación CD4/CD8 ≥10 en sangre

- Supervivencia a 5 años; 36%*

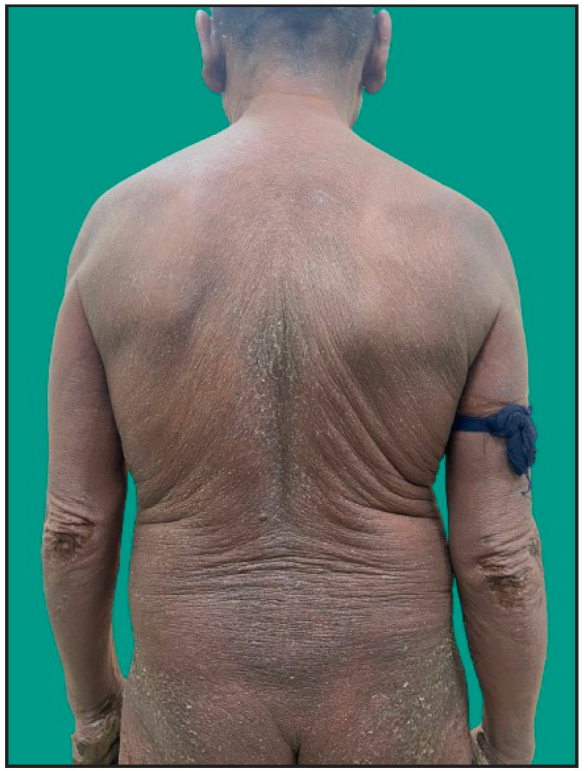


Figure 1b: Sézary syndrome presenting as erythroderma.

•Sanches et al., *An Bras Dermatol*, 2021; Stuver & Geller, *Front Immunol*, 2023 Willemze et al., *Blood*, 2019*Quadri I, et al Front. Immunol 2023* *Photo:Singh GK et al. Indian J Derm Ven Lepr 2025*

Cutaneous T-cell lymphoma (CTCL): MF

 # Mycosis Fungoides

-60% de los linfomas cutáneos primarios

- Incidencia global: 5-6 casos/millón habitantes/año

- Supervivencia a 5 años; 88%

- Patches y placas limitadas a la piel en áreas protegidas del sol

- En etapas avanzadas, puede haber afectación de los ganglios linfáticos, sangre y/o órganos viscerales.

- Comportamiento indolente.

- Recaídas frecuentes; las etapas avanzadas requieren terapias sistémicas.

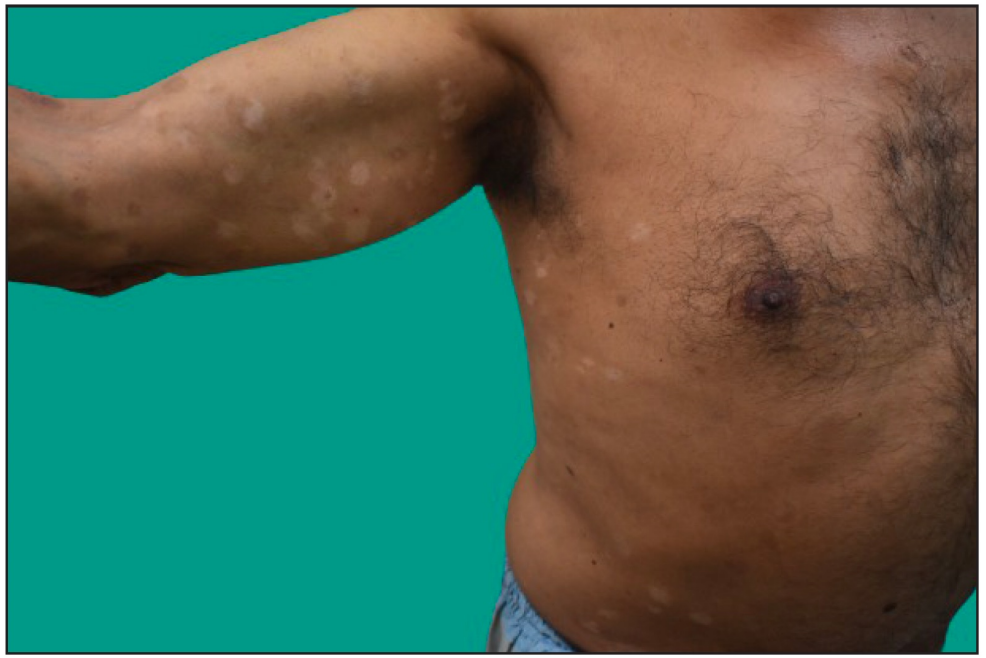


Figure 1a: Patch stage of mycosis fungoides: multiple asymptomatic hypopigmented macules mainly over the photoprotected areas.

•Sanches et al., *An Bras Dermatol*, 2021; Stuver & Geller, *Front Immunol*, 2023 Willemze et al., *Blood*, 2019*Quadri I, et al Front. Immunol 2023* Photo:*Singh GK et al. Indian J Derm Ven Lepr 2025*

Heterogeneous clinical presentation of MF

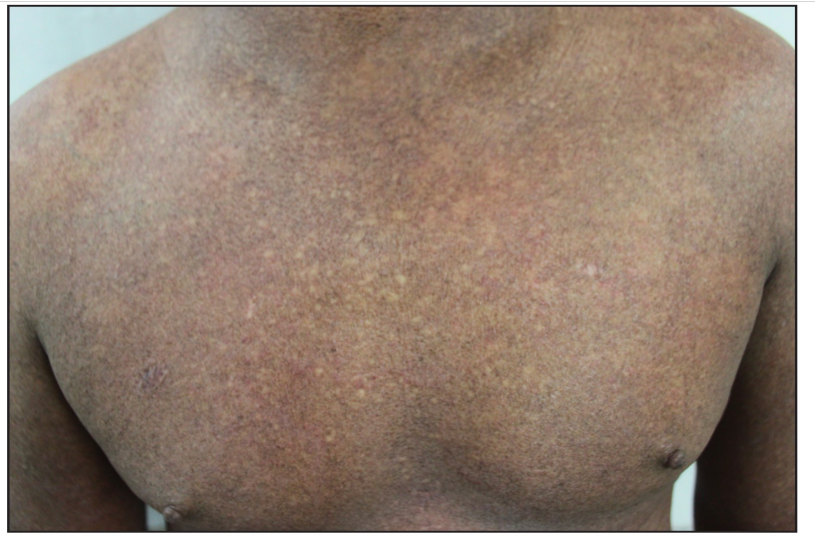
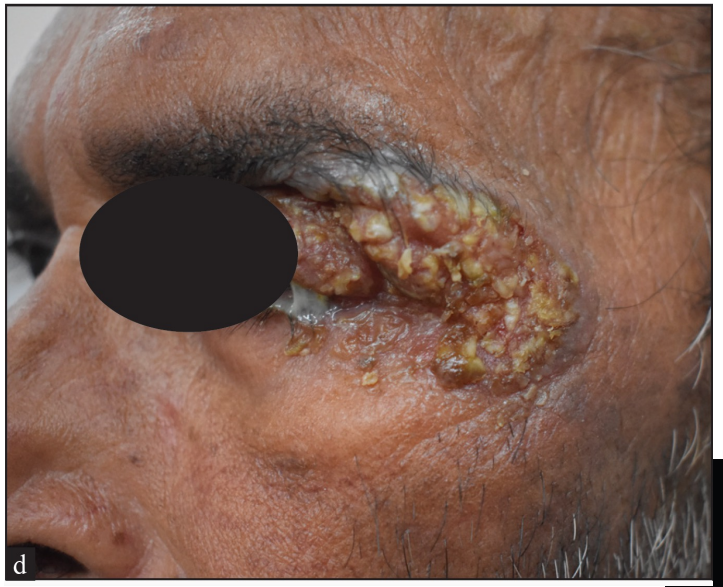
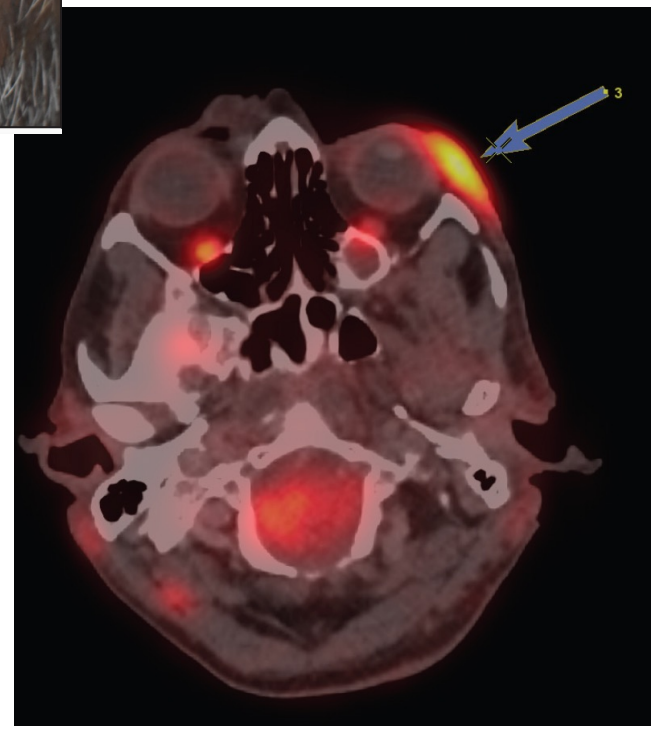


Figure 3b: Poikilodermatous MF presenting as diffuse poikilodermatous changes all over the body.



Plaque stage of MF on the left peri-orbital area and Increased 18F-FDG uptake in the plaque.



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Heterogeneous clinical presentation of MF

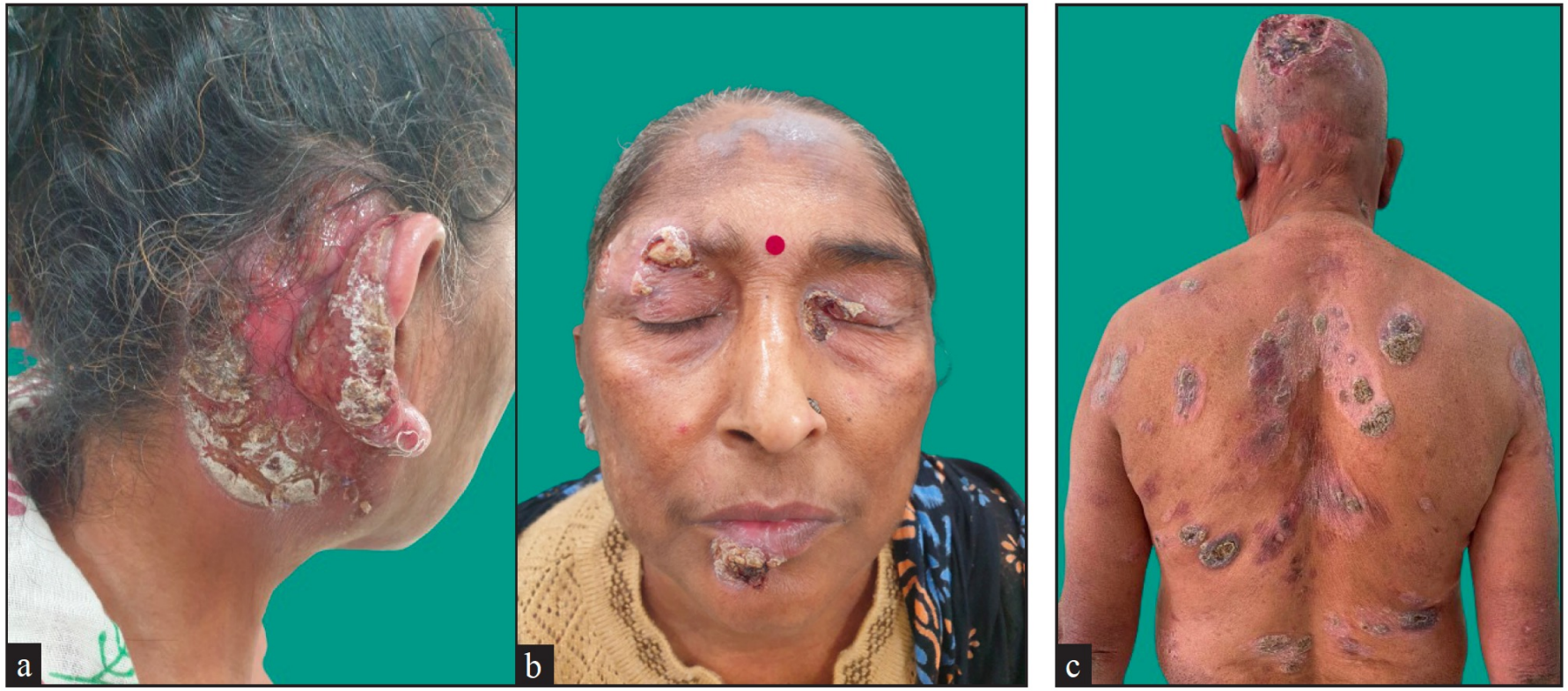

Figure 2: (a and b) Folliculotropic MF involving retroauricular area, scalp, face and trunk. (c) Large cell transformation presenting as ulcer on scalp.

•Other: patches, placas, tumores, eritrodermia). Variantes raras como foliculotrópica y granulomatosa laxa.

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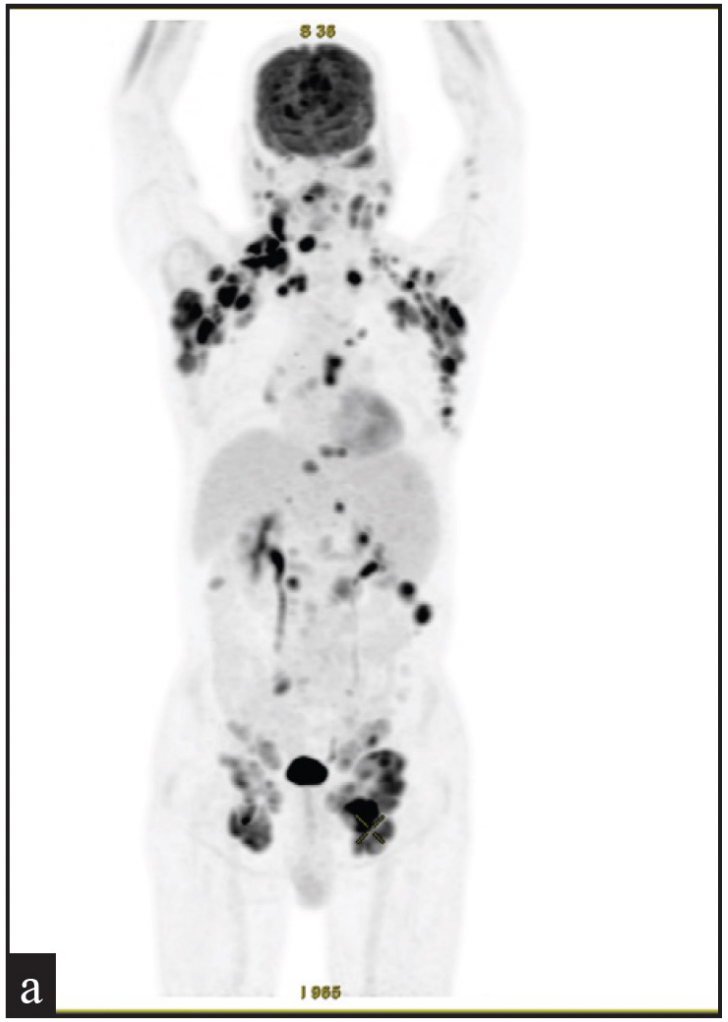
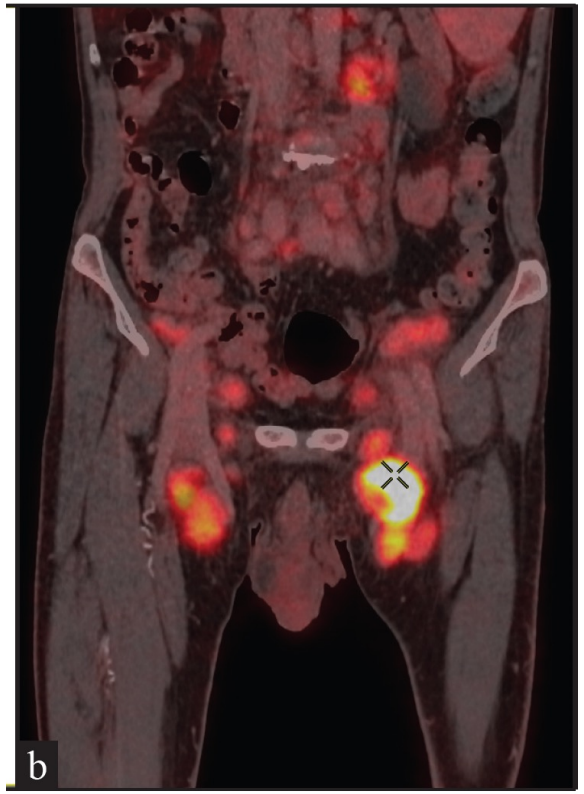
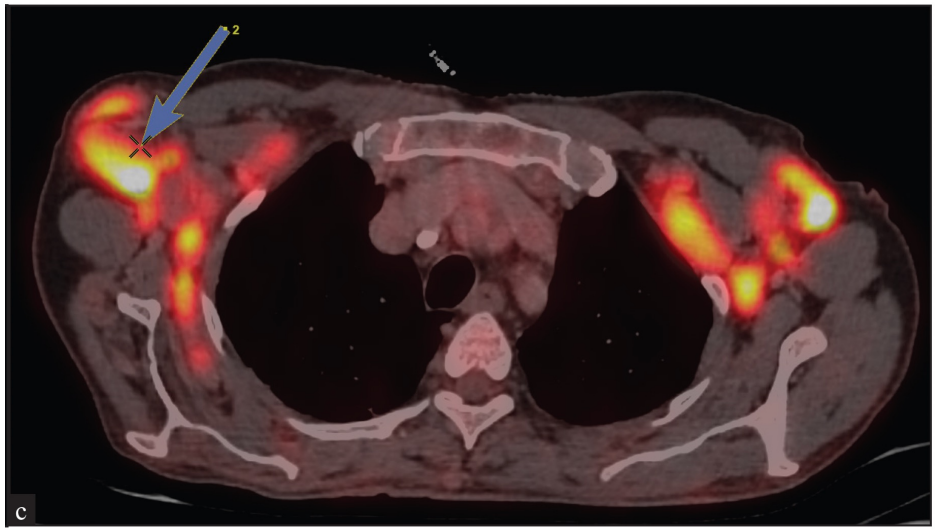
Heterogeneous clinical presentation of MF







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MF X SS

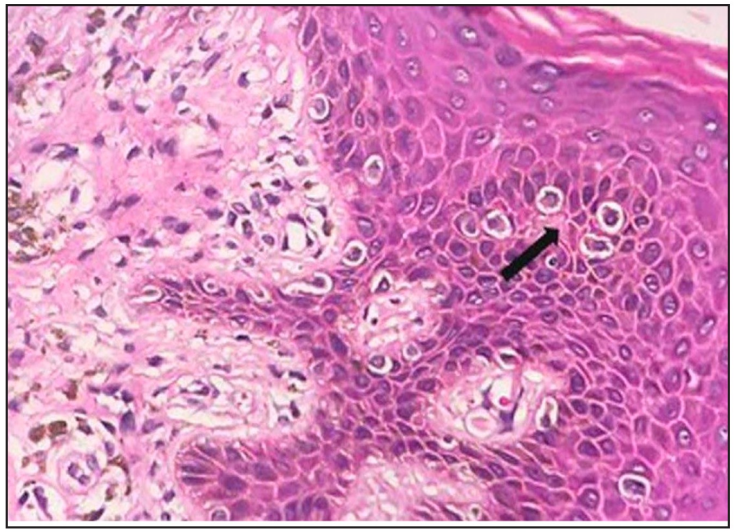


Figure 4a: Black arrow shows discrete lining of atypical lymphocytes at the dermo-epidermal junction. (Toy soldier appearance in the patch stage of MF). (Haematoxylin & eosin, 40x).

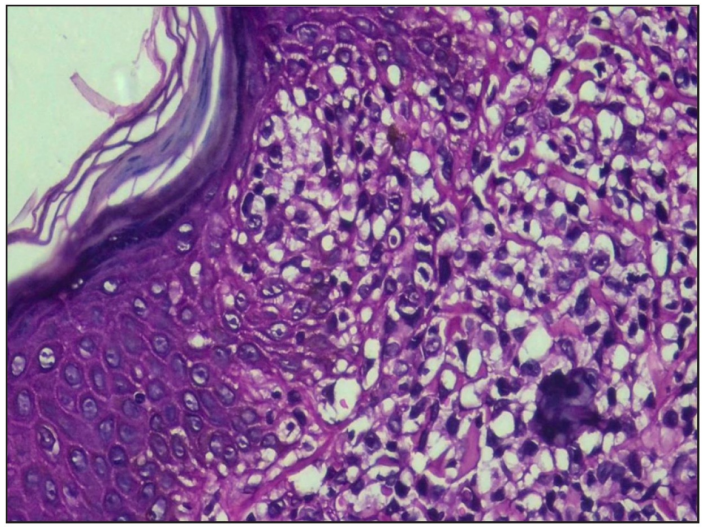
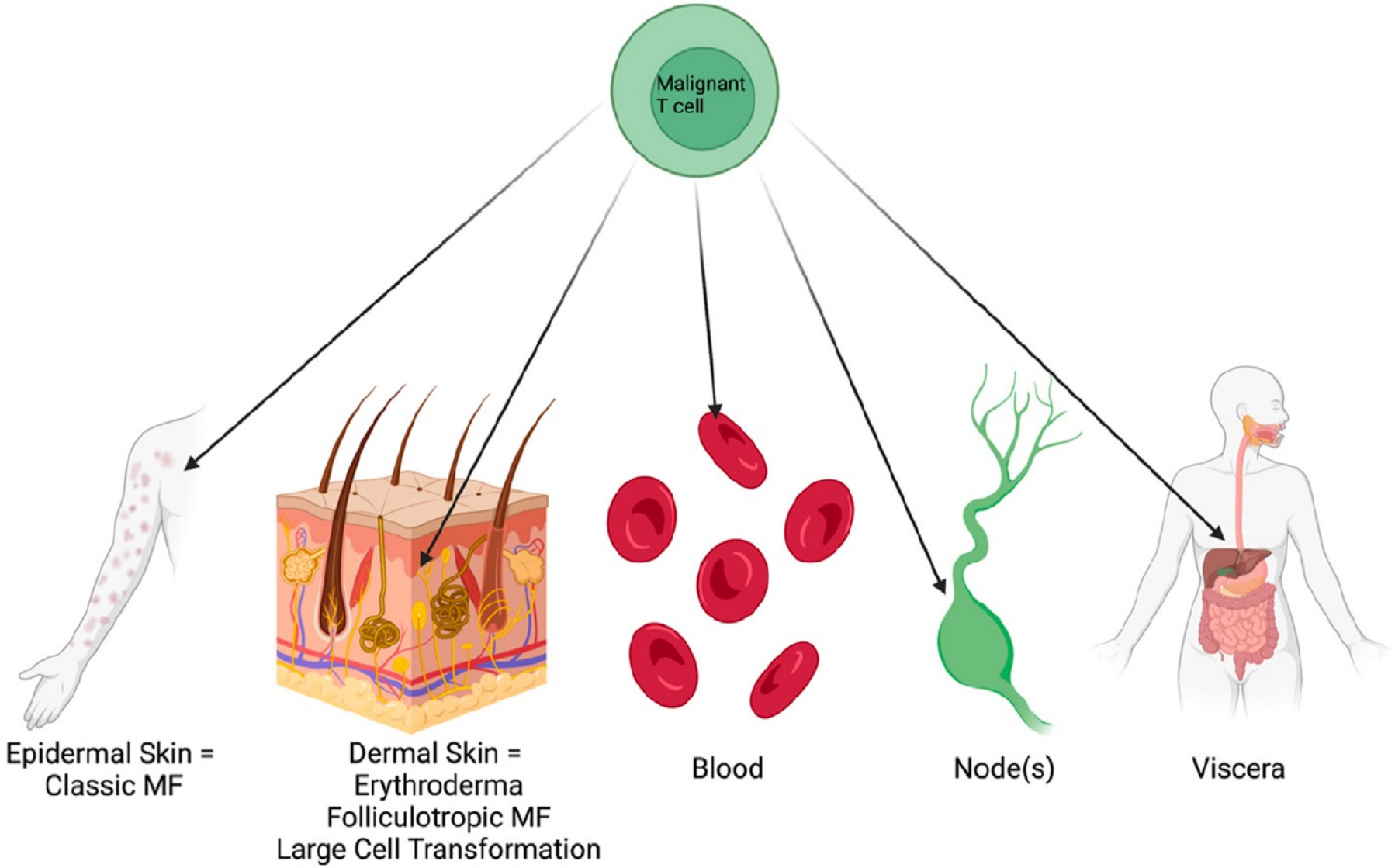


Figure 4b: Collection in atypical lymphocytes in clusters in the epidermis (Pautrier's microabscesses) from a case of plaque stage of MF. (Haematoxylin & eosin, 40x)

- Histopatología:
  - Epidermotropismo, microabscesos de Pautrier en MF
  - infiltrado dérmico denso en SS.
- Inmunofenotipo: CD3+/CD4+/CD7- en MF; CD4+/CD26- en SS.
- Estudios moleculares: Clonalidad del TCR mediante PCR BIOMED-2.

- Sanches et al., *An Bras Dermatol*, 2021; Stuver & Geller, *Front Immunol*, 2023. Willemze et al., *Blood*, 2019.

Mycosis Fungoides (MF) / Sezary Syndrome Compartments



Brazel D & Pinter-Brown L.(SOHO) Clinical Lymphoma, Myeloma and Leukemia June 2023; *Sutver R & Horwirtz SM Hematology 2024 l ASH Education Program*

Table 2: Specific investigations with their relevance in CTCL/SS

S No	Investigations	Relevance
	Complete blood count	Anaemia, raised total leucocyte count may indicate leukaemic variant of CTCL, that is, Sézary syndrome.
	Peripheral blood smear	Sézary cell count, anaemia.
	Liver function test	Necessary for baseline, metastasis and therapeutics.
	Renal function test	Necessary for baseline and therapeutics.
	Serum lactate dehydrogenase	Independent prognostic factor.
	X-ray chest	Necessary for baseline status and metastasis.
	Ultrasound abdomen	For organomegaly and metastasis in liver and spleen.
	Skin biopsy	H&E stain for atypical lymphocytes, toy soldier appearance, epidermotropism, Pautrier's microabcesses. Typical immunohistochemistry profile is CD3+, CD4+, CD5+ and CD8− with relative loss of expression of CD7. CD30 for large cell transformation.
	Lymph node biopsy	If clinically enlarged lymph nodes, H&E stain for atypical lymphocytes, Pautrier's micro-abcesses, partial or complete effacement of lymph nodes. IHC – same as skin biopsy
	Flow cytometry	In case of suspected leukaemic involvement/Sézary syndrome CD4/CD8 ratio and percentage of CD7 and CD26 positive cells. CD4/CD8 ratio >10 is diagnostic of SS.
	FDG-PET scan	A must in any stage advanced than patch or limited plaque stage for lymph node and visceral involvement.
	T-cell receptor (TCR) gene rearrangement studies on either skin/lymph node biopsy specimen or peripheral blood	To look for clonal rearrangements of the TCR – poor prognostic factor by PCR. A tumour clone frequency of >25% in skin biopsy is a predictor for disease progression.
	Bone marrow biopsy	Bone marrow biopsy is recommended in patients with MF and SS who have B2 blood involvement or unexplained haematologic abnormalities. It is done to look for metastasis in bone marrow.
	Liver biopsy	Any suspicious lesion (metastasis) on radiology of liver (CT/FDG-PET) must be confirmed by liver biopsy.

Investigations up to serial number 8 must be done in both early and advanced MF/CTCL, whereas investigations beyond serial number 8 are indicated in advanced stages only. A clinically significant lymph node must be biopsied whenever present

MF x SS

Característica diagnóstica	MF temprana	SS avanzado
Epidermotropismo	Presente	Menos pronunciado
Células Sézary en sangre	Ausentes	>1000/µl

Característica	MF	SS
Frecuencia	39%	2%
Edad pico	55–60 años	Similar
Pronóstico inicial	Favorable	Reservado

Subtipo	Frecuencia (%)	Supervivencia a 5 años (%)
Micosis fungoide (MF)	39	88
Variantes de MF	<1	75–100
Síndrome de Sézary (SS)	2	36

Willemze R et al. The 2018 update of the WHO-EORTC classification for primary cutaneous lymphomas. *Blood*. 2019

Características Clínicas: MF x SS

Característica	Micosis Fungoide	Síndrome de Sézary
Involucramiento cutáneo	Patches → Placas → Tumores	Eritrodermia difusa
Afectación sanguínea	Rara	>1,000 células Sézary/μL
Linfadenopatías	Localizadas	Generalizadas
Otros síntomas	Prurito leve/moderado	Prurito severo, alopecia

Olsen et al., *J Clin Oncol*, 2011; Sanches et al., *An Bras Dermatol*, 2021.

DIAGNOSTICO DIFERENCIAL

Through thick and thin: confronting the aggressive cutaneous T-cell lymphomas

Robert Stuver[1] and Steven M. Horwitz[1,2]

INDOLENT:

- Inflammatory conditions like eczema/psoriasis, and sézary syndrome (SS) from other erythrodermic dermatoses
- - Dermatitis atópica
- - Pitiriasis rubra pilaris (PRP)
- -Reacciones medicamentosas

AGGRESSIVE CTCLS:

- Cases of advanced-stage mycosis fungoides/sézary syndrome
- Primary cutaneous CD8+ aggressive epidermotropic cytotoxic t-cell lymphoma (PCAETCL)
- Primary cutaneous gamma delta t-cell lymphoma (PCGDTCL)

Sutver R & Horwirtz SM
Hematology 2024 I ASH Education Program

Heterogeneous clinical presentation of MF
Tumor-stage mycosis fungoides.

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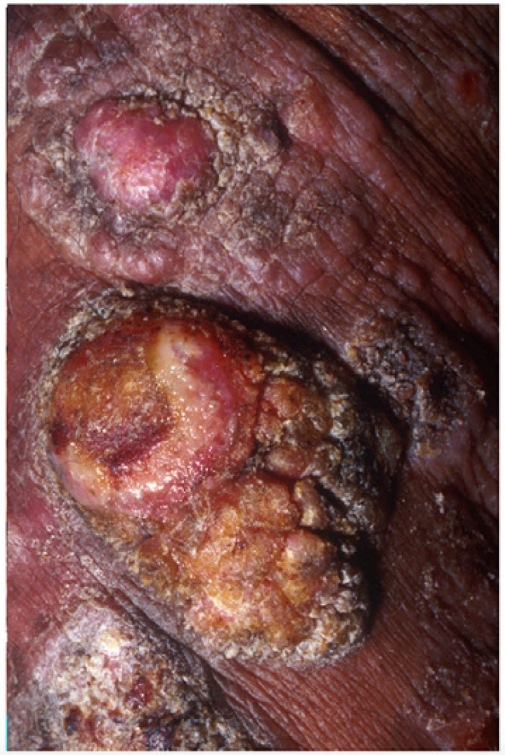

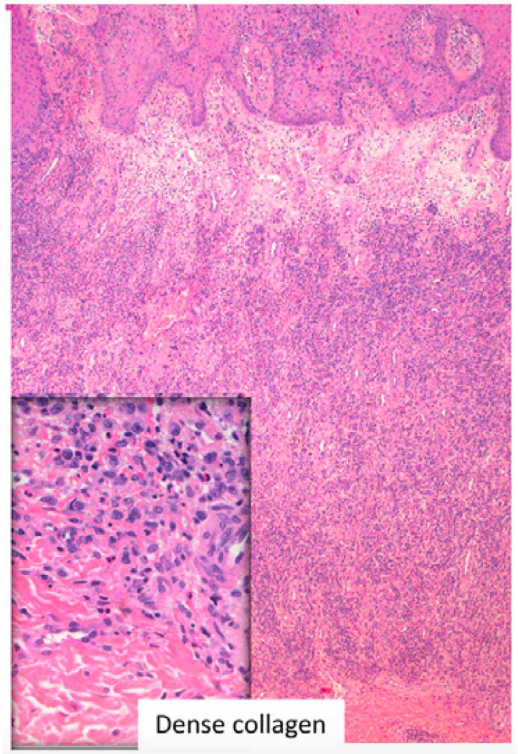

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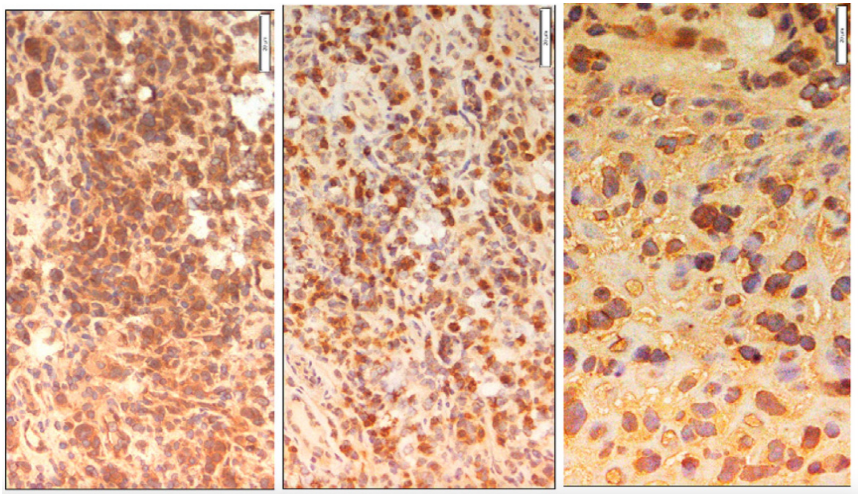


- Tumor-stage MF (TMF) poor prognosis
- Must be distinguished from less aggressive primary cutaneous ALCL (pcALCL).
- Similar presentation.
- There are no current reliable pathologic criteria to distinguish TMF from pcALCL.

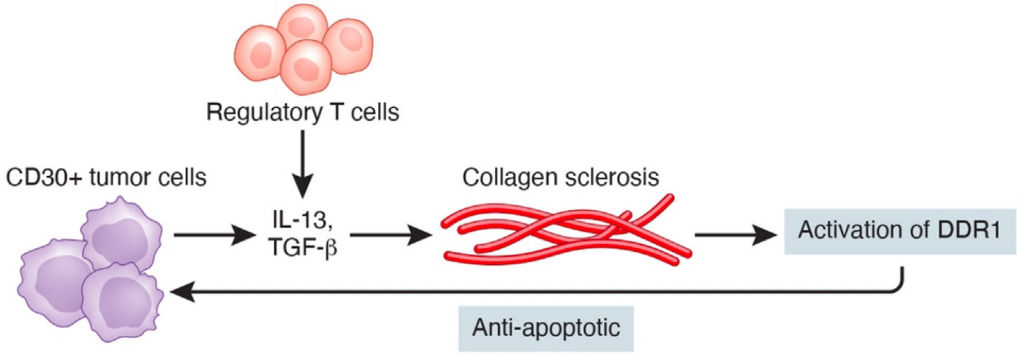
CTCL behavior by disease

Based on the WHO-EORTC guidelines

	Clinical behavior	Frequency (%)	Disease-specific 5-year survival (%)
Mycosis fungoides	Indolent	39	88
Mycosis fungoides variants			
Folliculotropic mycosis fungoides	Indolent	5	75
Pagetoid reticulosis	Indolent	<1	100
Granulomatous slack skin	Indolent	<1	100
Sézary syndrome	Aggressive	2	36
Primary Cutaneous CD30+ Lymphoproliferative Disorders			
Primary cutaneous anaplastic large T-cell lymphoma	Indolent	8	95
Lymphomatoid papulosis	Indolent	12	99
Adult T-cell lymphoma/leukemia	Indolent / Aggressive	<1	NR
Subcutaneous panniculitis-like T-cell lymphoma	Indolent	1	87
Extranodal NK/T-cell lymphoma, nasal type	Aggressive	<1	16
Chronic active EBV infection	Indolent	<1	NR
Primary cutaneous T-cell lymphoma γ/δ	Aggressive	<1	11
Primary cutaneous aggressive epidermotropic CD8+ T-cell lymphoma	Aggressive	<1	31
Primary cutaneous acral CD8+ T-cell lymphoma	Indolent	<1	100
Primary cutaneous CD4+ small/medium T-cell lymphoproliferative disorder	Indolent	6	100
Primary Cutaneous Peripheral T-Cell Lymphoma, NOS	Aggressive	2	15

Sanches et al., *An Bras Dermatol*, 2021;

Fisiopatología



- MF:
  - Origen en células T CD4+ residentes en la piel.
  - Cambio microambiental Th1 → Th2 promueve progresión.
  - Mutaciones: STAT3, JAK2, NF-κB.
- SS:
  - Variante leucémica con células Sézary circulantes (CCR4+, CD26-, CD7-).

Stuver & Geller, *Front Immunol*, 2023; Kim et al., *Lancet Oncol*, 2018
Image *Marshall E. Kadin1 Journal of Investigative Dermatology (2024)*

Clasificación TNMB

Parámetro	MF	SS
Piel (T)	Parches/placas/tumores	Eritrodermia (T4)
Ganglios (N)	Dermatopáticos (N1-N3)	Infiltración linfoide (N3)
Sangre (B)	B0-B1 (<1000 células/μL)	B2 (>1000 células/μL)
Metástasis (M)	Raro en etapas tempranas	Visceral (M1)

Olsen et al., *J Clin Oncol*, 2011; Willemze et al., *Blood*, 2019.

Table 2: ISCL/EORTC revision to the staging of mycosis fungoides and Sezary syndrome with five-year disease-specific survival (DSS)

Stages of MF	T	N	M	B	Prognosis (DSS at five years)
IA	1	0	0	0-1	100%
IB	2	0	0	0-1	95%
IIA	1-2	1-2	0	0-1	84%
IIB	3	0-2	0	0-1	56%
III	4	0-2	0	0-1	65%
IIIA	4	0-2	0	0	65%
IIIB	4	0-2	0	1	65%
IVA_1	1-4	0-2	0	2	30%
IVA_2	1-4	3	0	0-2	30%
IVB	1-4	0-3	1	0-2	30%

T- Tumour, N- Node, M- Metastasis, B-Blood involvement.

Singh GK et al. Indian J Derm Ven Lepr 2025

Table 2 Revised TNMB classification for mycosis fungoides and Sézary syndrome.[2,5]

Skin (T)	
T1	Limited patches/plaques (covering <10% of the total skin surface)
T1a	Patches only
T1b	Plaques ± patches
T2	Generalized patches/plaques (covering ≥10% of the total skin surface)
T2a	Patches only
T2b	Plaques ± patches
T3	Tumor(s)
T4	Erythroderma
Lymph node (N)	
N0	No clinically abnormal peripheral lymph nodes;
N1	Clinically abnormal peripheral lymph nodes; dermatopathic lymphadenopathy or histopathological involvement by isolated atypical lymphocytes, without alteration of the lymph node architecture
N1a	Negative clone
N1b	Positive clone
N2	Clinically abnormal peripheral lymph nodes; histopathological involvement by aggregates of atypical lymphocytes, without alteration of the lymph node architecture
N2a	Negative clone
N2b	Positive clone
N3	Clinically abnormal peripheral lymph nodes; frank histopathological involvement and partial/complete effacement of the lymph node architecture
NX	Clinically abnormal peripheral lymph nodes; without histopathological confirmation
Viscera (M)	
M0	Without visceral involvement
M1	With visceral involvement
Blood (B)	
B0	There are no circulating atypical cells (Sézary cells; or <5% of atypical lymphocytes)
B0a	Negative clone
B0b	Positive clone
B1	Low tumor load in the blood (≥5% of lymphocytes are Sézary cells, but not B2 cells)
B1a	Negative clone
B1b	Positive clone
B2	Elevated tumor load in the blood and clonal rearrangement of the TCR (≥1000 Sézary cells/microL. and/or CD4:CD8 ≥ 10, and/or CD4 + CD7 - ≥40%, and/or CD4 + CD26 - ≥30%)

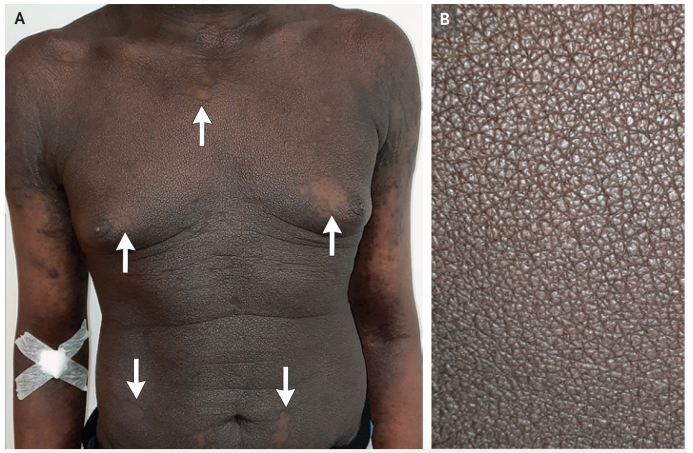
Sanches et al., *An Bras Dermatol*, 2021

Estadificación Clínica

Estadio Clínico	TNMB	Supervivencia a 5 años (%)
IA	T1 N0 M0 B0/B1	~98%
IIB	T3 N0-N2 M0 B0/B1	~50%
IVB	T1-T4 N0-N3 M1 B0-B2	~18%

Olsen et al., *J Clin Oncol*, 2011; Sanches et al., *An Bras Dermatol*, 2021

CASO CLÍNICO: IIB



Heloise Mazoyer & Bachmeyer C. Sorbonne Université, Paris.
N Engl J Med 392;12 March 27, 2025

PROGNOSIS FACTORS

Table 4: Poor prognostic factors in MF and SS

Variable	Poor prognosis
Age*	More than 60 years
Gender*	Male
Body surface area (BSA) involvement	More the BSA involved, worse is the prognosis (T2b > T2a)
Type of skin lesions*	Thick plaques, nodules
Folliculotropism*	Folliculotropism present
Course	New onset nodules, ulceration
Cell morphology	Large cell transformation (presence of CD30− or CD30+ large cells (at least four times larger than a small lymphocyte) exceeding 25% of the infiltrate or forming microscopic nodules)
Lymph node*	Lymph node involvement
Histopathology of lymph nodes	Higher the grade, worse the prognosis
Metastasis*	Early metastasis
Hematopoietic system (blood)/Sezary syndrome*	Absolute Sezary cell count of >1000/μL or an expanded $CD4^+$ T-cell population resulting in a CD4/CD8 ratio ≥10, $CD4^+/CD7^-$ cells ≥40% or $CD4^+/CD26^-$ cells ≥30%)
Serum lactate dehydrogenase (LDH)	High Lactate dehydrogenase
T-cell clonality	Present

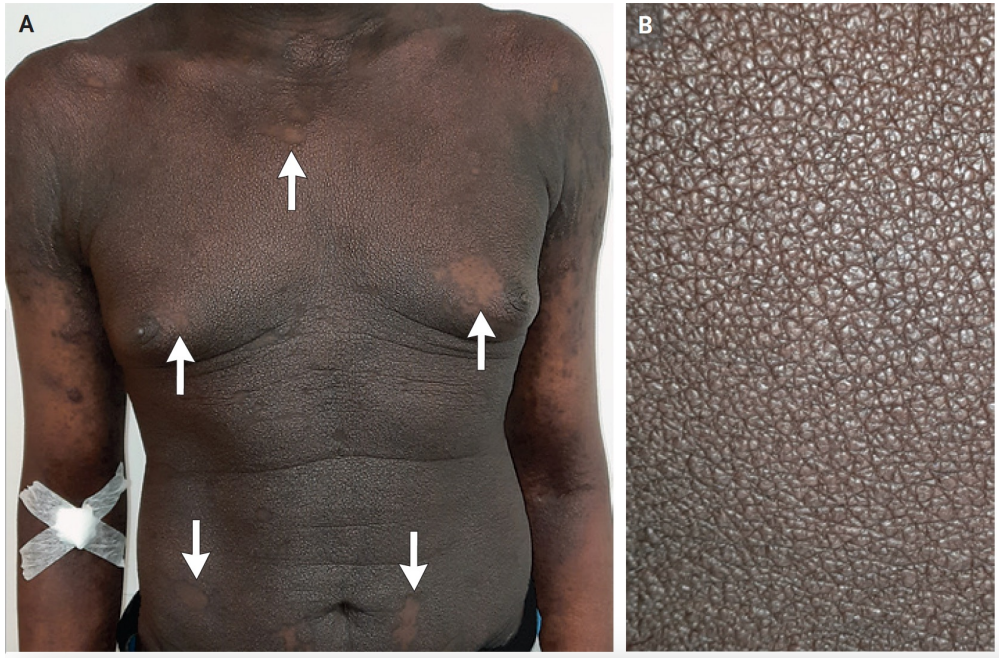
The variables marked (*) are included in CLIPi (Cutaneous Lymphoma International Prognostic index).

Singh GK et al. Indian J Derm Ven Lepr 2025

The NEW ENGLAND JOURNAL of MEDICINE

IMAGES IN CLINICAL MEDICINE

Sézary Syndrome



- *Sézary syndrome (stage B2)*
- *Treatment with*
- *Systemic therapy was initiated,*
- *CR within 4 months.*

- *Relapse 1 year later*
- *Currently undergoing evaluation for bone marrow transplantation.*

The NEW ENGLAND JOURNAL of MEDICINE

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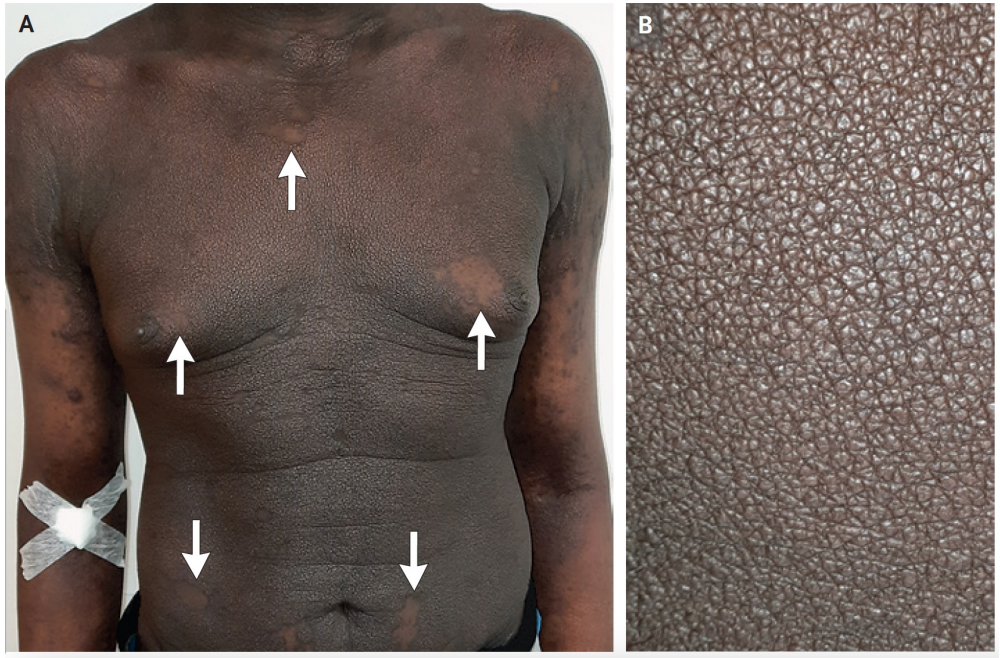
Sézary Syndrome



- *Sézary syndrome (stage B2)*
- *Treatment with*
- *Systemic therapy was initiated,*
- *CR within 4 months.*

- *Relapse 1 year later*
- *Currently undergoing evaluation for bone marrow transplantation.*

Stage	First-Line Treatment	Second-Line Treatment	Third-Line Treatment
Early MF (IA-IIA)	Topical corticosteroids, mechlorethamine gel	PUVA/NB-UVB, localized radiotherapy	Low-dose TSEBT
Advanced MF/SS	Brentuximab vedotin, mogamulizumab	ECP + interferon alpha, oral bexarotene	HDAC inhibitors, methotrexate
Refractory MF/SS	Romidepsin/vorinostat, pembrolizumab	Lacutamab, systemic chemotherapy	Allo-HSCT

SANGHES
OLSEN

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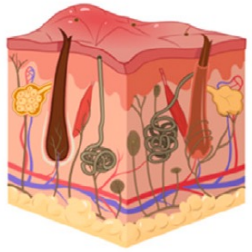
WHEN AND HOW TO TREAT??

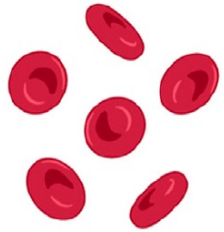
Treatment By Compartment

Epidermal Skin
Bexarotene
Brentuximab vedotin
Liposomal doxorubicin
Methotrexate
Mogamulizumab
Romidepsin
Vorinostat



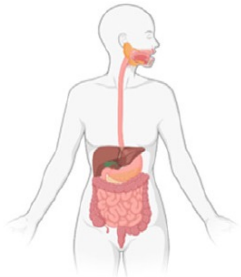
Dermal Skin
Bexarotene
Liposomal doxorubicin
Pralatrexate
Romidepsin

Blood
Brentuximab vedotin
Liposomal doxorubicin
Mogamulizumab
Romidepsin



Nodes
Brentuximab vedotin
Pralatrexate
Romidepsin

Viscera
Brentuximab vedotin
Liposomal doxorubicin
Romidepsin



- CRITERIA
- Based on disease stage, symptom burden, and patient-specific factors

→ “Really”?

Brazel D & Pinter-Brown L. Clinical Lymphoma, Myeloma and Leukemia June 2023

Recommendations:
Dermatology and Hematology Divisions of HC-FMUSP for the treatment of mycosis fungoides and Sézary syndrome.

CS	First-line	Second line	Third line
Ia	Topical corticosteroids Phototherapy with NB-UVB Topical mechlorethamine[a] Topical carmustine Bexarotene gel[a] Imiquimod / Resiquimod	Localized topic PUVA PUVA	Localized radiotherapy (if regionalized disease)
Ib, IIa	Phototherapy with NB-UVB PUVA phototherapy	Systemic retinoids[b] ± phototherapy with NB-UVB or PUVA Alpha interferon ± phototherapy with NB-UVB or PUVA	Total skin electron beam radiotherapy Low-dose methotrexate
IIb	Systemic retinoids ± phototherapy with NB-UVB or PUVA ± localized RT Alpha-interferon ± phototherapy with NB-UVB or PUVA ± localized RT	Total skin electron beam radiotherapy Low-dose methotrexate Brentuximab vedotin (if CD30-positive)	Monochemotherapy (gemcitabine, pegylated liposomal doxorubicin) HDACi[a] Mogamulizumab Polychemotherapy (CHOP type) Allogeneic hematopoietic stem

Sanches et al., *An Bras Dermatol*, 2021

Recommendations:
Dermatology and Hematology Divisions of HC-FMUSP for the treatment of mycosis fungoides and Sézary syndrome.

			cell transplantation
IIIa, IIIb	Systemic retinoids[b] ± PUVA Alpha interferon ± PUVA ECP ± systemic retinoids[b] ± alpha-interferon	Chlorambucil + prednisone Low-dose methotrexate Monochemotherapy (gemcitabine, pegylated liposomal doxorubicin)	HDACi[a] Polychemotherapy (CHOP type) Allogeneic hematopoietic stem cell transplantation
IVa1, SS	ECP ± alpha-interferon ± systemic retinoids[b] Alpha interferon ± PUVA	Chlorambucil + prednisone Low-dose methotrexate Alemtuzumab Mogamulizumab[a]	Monochemotherapy Polychemotherapy (CHOP type) Allogeneic hematopoietic stem cell transplantation
IVa2	Monochemotherapy (gemcitabine, pegylated liposomal doxorubicin) Radiotherapy if localized nodal disease	Brentuximab vedotin (if CD30-positive)	HDACi[a] Polychemotherapy (CHOP type) Allogeneic hematopoietic stem cell transplantation
IVb	Monochemotherapy (gemcitabine, pegylated liposomal doxorubicin)	Brentuximab vedotin (if CD30-positive)	Polychemotherapy (CHOP type) Allogeneic hematopoietic stem cell transplantation

Sanches et al., *An Bras Dermatol*, 2021

Tratamiento por Etapas Tempranas

Terapias dirigidas a la piel:

- Corticoides tópicos: Clobetasol propionato (63%63% respuesta completa en T1).
- Mecloretamina gel (58%−93%58%−93% respuesta global).
- Fototerapia PUVA/NB-UVB (65%−85%65%−85% respuesta completa).

Zackheim, *Dermatol Ther*, 2003; Lessin et al., *JAMA Dermatol*, 2013.

Response rates and duration of responses for skin treatments for MF/SS.

Table 4 Response rates and duration of responses for skin treatments for MF/SS.

Treatment	Staging	Overall response (%)	Complete response (%)	Response duration	References
Corticosteroid	T1–T2	82–94	25–63	RD: 9 months	9
Nitrogen mustard	T1–T2	58.5–93	13.8–80	–	10, 11
Carmustine	T1–T2	84–98	47–86	–	12
Bexarotene	T1–T2	54–63	10–21	RD: 25 months	13
Imiquimod	T1–T2	80	45	–	14
Resiquimod	T1–T2	75	33	–	15
NB-UVB	T1–T2	–	54–91	RD: 22 months	16
PUVA	T1–T2	–	65–85	RD: 23 months	16
Localized RT	un. MF, 30Gy	100	–	DFS, 5 years: 86%	19
	Ia–III, 9–45Gy	94	88	PFS, 5 years: 85%	17
Total skin irradiation with electrons	T1–T2, 36Gy	95	85–88	PFS, 5 years: 50%	20
	Ia–III, 12Gy	88	27	RD: 70.7 weeks	21

RD, Response Duration; PFS, Progression-Free Survival; DFS, Disease-Free Survival; NB-UVB, narrow-band UVB; RT, Radiotherapy; Gy, Gray; un. MF, unilesional mycosis fungoides.

Sanches et al., *An Bras Dermatol*, 2021

PEG-IFNα-2a

- IFN-α estándar - 3–9 millones de unidades - veces/semana
- PEG-IFNα-2a : 1.5 µg/kg - 1 vez/semana
- Duración: Mínimo 6 meses; ajuste según respuesta y toxicidad

- Terapia inmunomoduladora : +++ respuesta Th1, activa células NK y apoptosis de linfocitos T malignos.
- Antiangiogénesis
- Regulación epigenética (genes asociados a la proliferación celular)

Estadios Tempranos (IA-IIA):

- Estudios prospectivos: CR 84% (IA-IIA)
- - Monoterapia: ORR: 29-80%,
- - + PUVA: ORR del 75-98%
- PFS a 5 años: 27-75%

- Estadios Avanzados (IIB-IVB) y SS:

- +retinoides (bexaroteno) o quimioterapia: ORR:45-58%
- + PUVA: ORR 50-83%.

→ SS eritrodérmico: + fotoféresis extracorpórea: ORR 45-88%

- -Ventajas: administración semanal) y mejor perfil de seguridad
- Supervivencia global a 5 años del 75% (IB-IIIA_

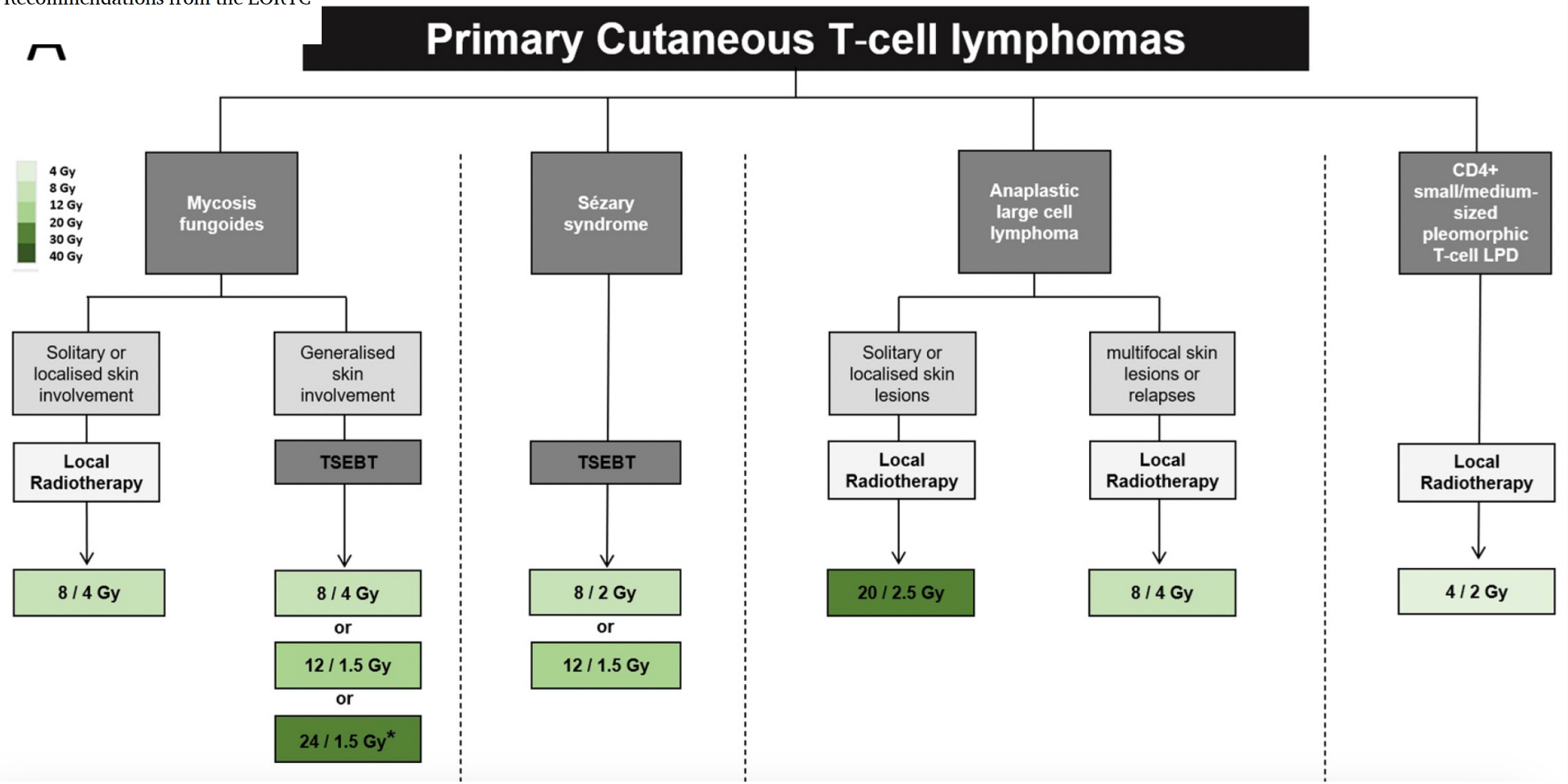
Gosman et al. Acta Derm Venereol 2023 ; Sanches et al., An Bras Dermatol, 2021;Stuver & Geller, Front Immunol, 2023

Contents lists available at ScienceDirect

European Journal of Cancer

journal homepage: www.ejcancer.com

Radiotherapy in cutaneous lymphomas: Recommendations from the EORTC cutaneous lymphoma tumour group



2. Radiotherapy dose recommendations for primary cutaneous lymphomas with the total radiation dose/fraction dose. * Before stem cell transplant ** Repeat if no complete response at 4 months *** Residual lesions or stable disease at 4 months or progressive disease at any time

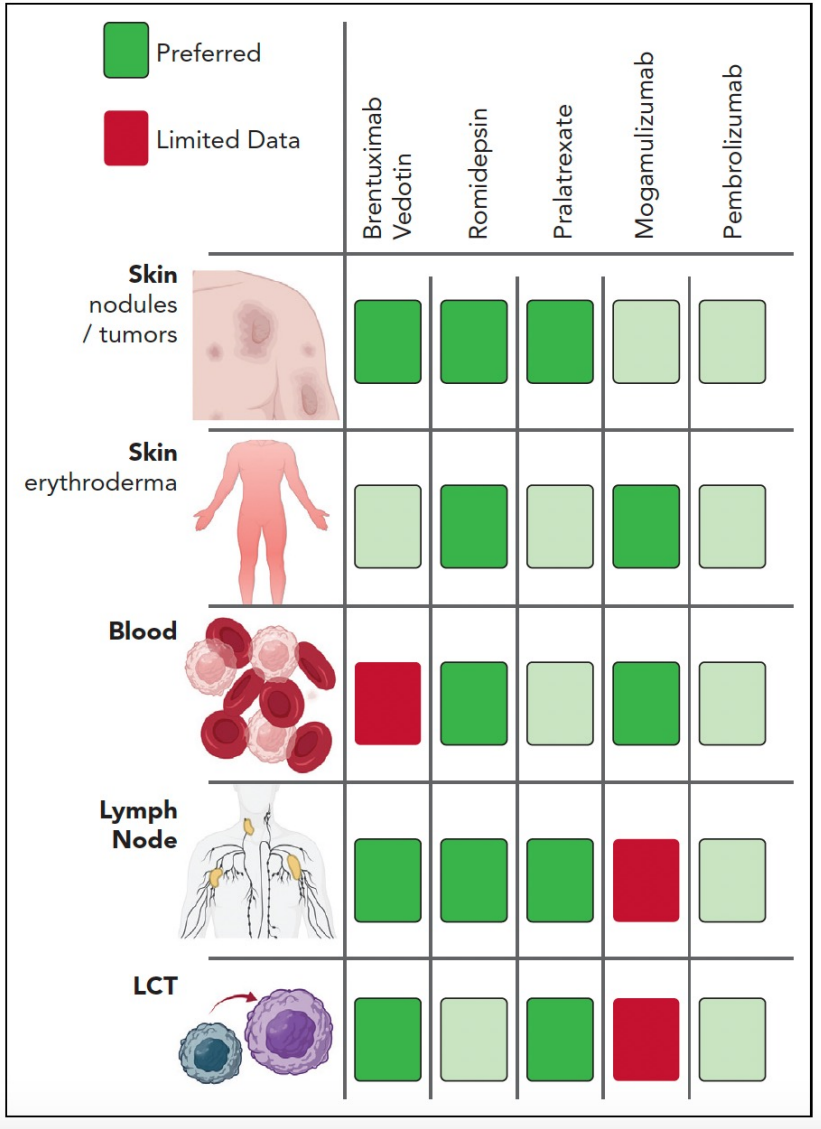
K. Elsayad et al. European Journal of Cancer 212 (2024) 115064

Integrating novel agents into the treatment of advanced mycosis fungoides and Sézary syndrome

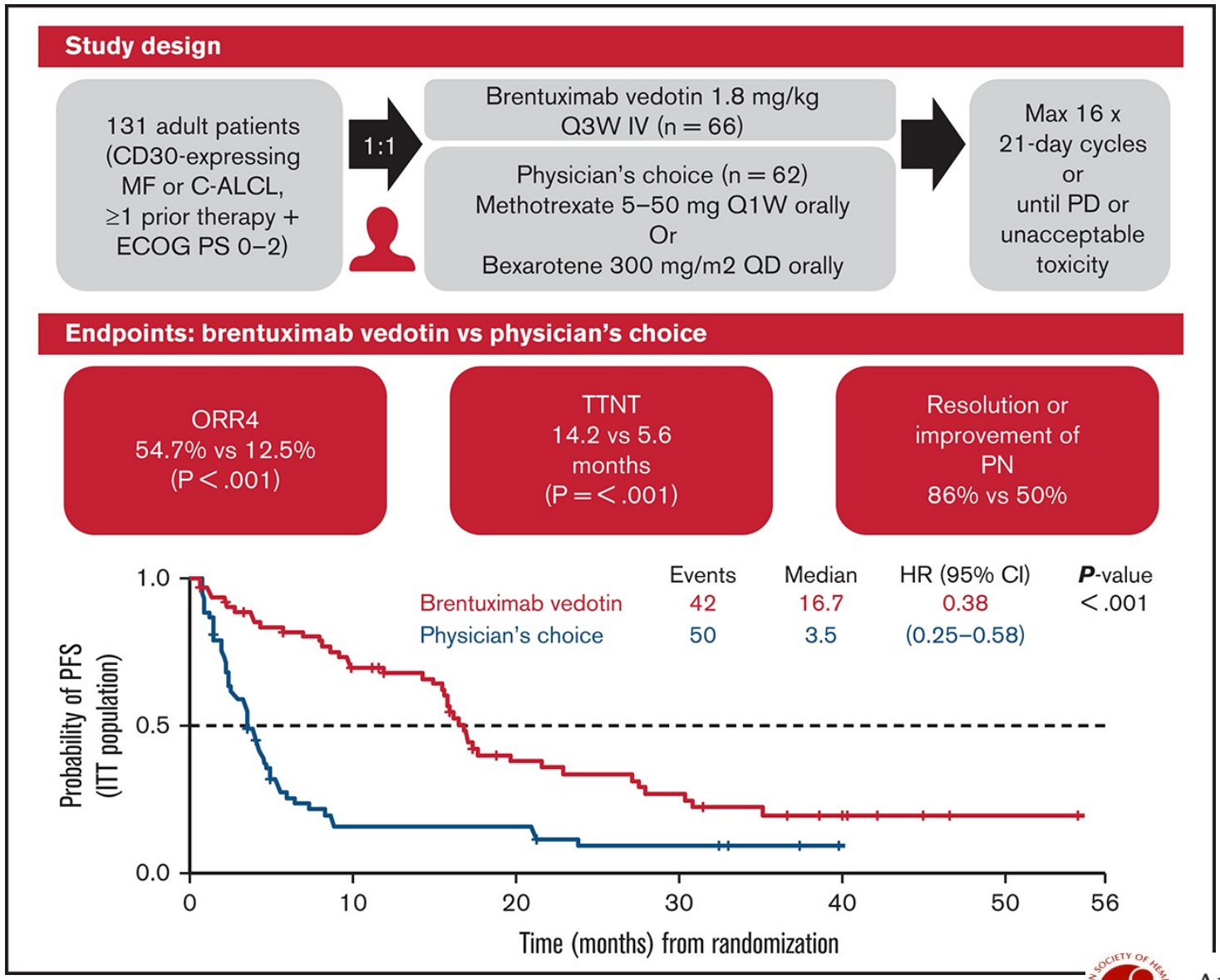
Michael S. Khodadoust,[1,2] Eric Mou,[3] and Youn H. Kim[1,2]

16 FEBRUARY 2023 I VOLUME 141, NUMBER 7

- *Disease compartment driven drug selection.*
- *Relative activity of single agent infusional therapies in various disease compartments*



Randomized phase 3 ALCANZA study of brentuximab vedotin vs physician's choice in RR cutaneous T-cell lymphoma: final data



Steven M et a. Blood Adv, 2021,

American Society of Hematology
Helping hematologists conquer blood diseases worldwide

SOME DRUGS FOR CTCL (NOW AND IN THE FUTURE…)

 # Monoclonal antibodies

- *Alemtuzumab (anti-CD52)*

Denileukin diftitox (diphtheria toxin and IL-2).

Mogamulizumab (anti CCR4)

Brentuximab vedotin (anti-CD30 + monomethyl auristatin E)

#Immune checkpoint Inhibitors

(anti-PD1)

Pembrolizumab

Nivolumab

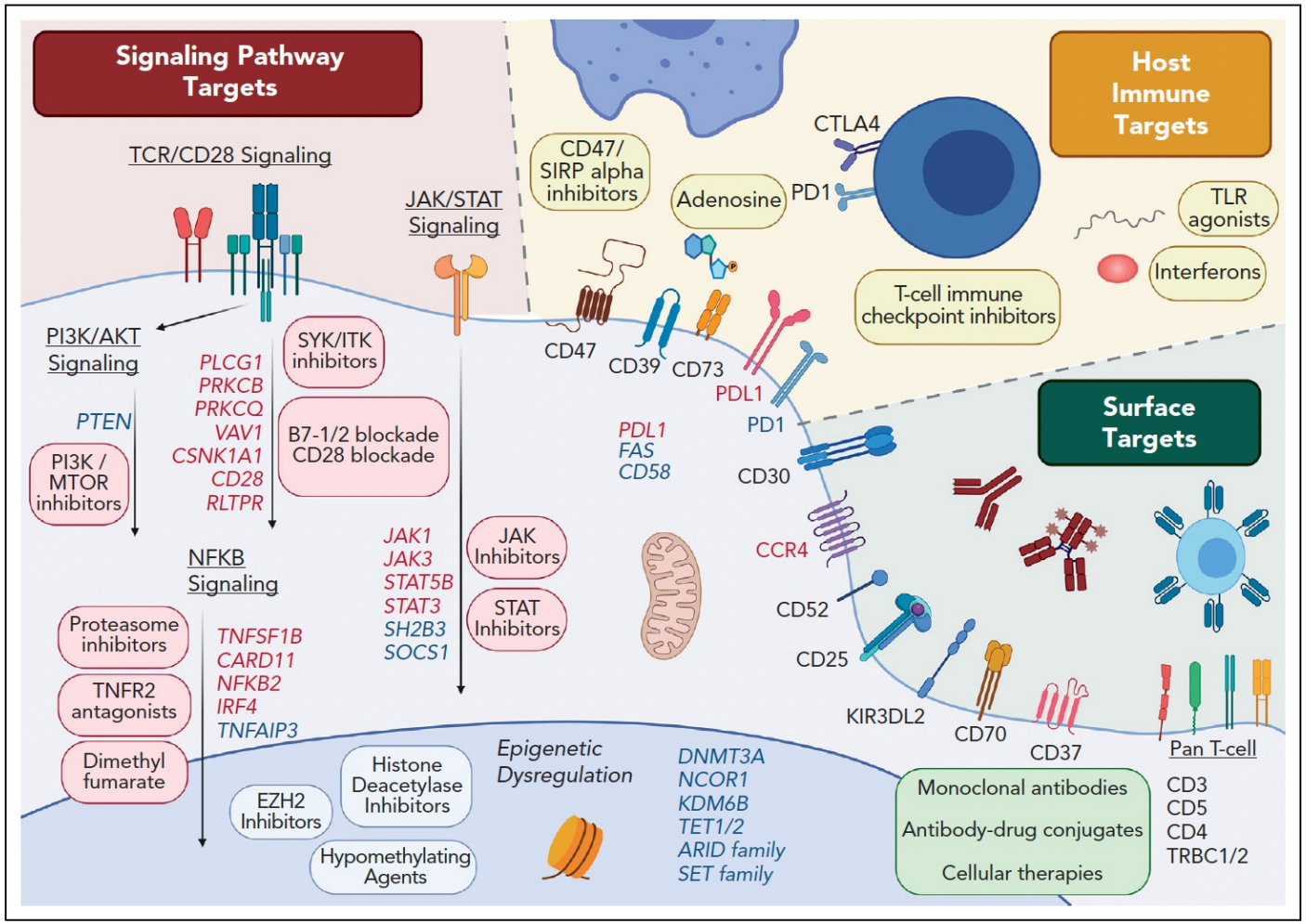
#JAK-STAT inhibitors

Ruxolitinib

Upadacitinib

Quadri I, Reneau JC, Hanel W andChung CG (2023) Front. Immunol.

Therapeutic targets in cutaneous T-cell lymphoma.



Genes recurrently affected by gain-of-function genomic variants (red) or loss-of-function variants (blue) are shown with each altered pathway.

Quadri I, Reneau JC, Hanel W and Chung CG,2023. Front. Immunol.

Conclusiones

1. MF y SS requieren un enfoque multidisciplinario para optimizar el manejo.
2. Terapias adaptadas a cada caso
3. Las terapias dirigidas a biomarcadores como CD30 y CCR4 están transformando el panorama terapéutico.
4. Los avances en inmunoterapia ofrecen nuevas esperanzas para pacientes refractarios.

GRACIAS!

OBRIGADA!

THANK YOU!