Complexo Hospitalario Universitario de Santiago de Compostela

Parapsoriasis

Un viejo problema de actualidad

Congreso SEAP

Zaragoza 18-21 Mayo 2011

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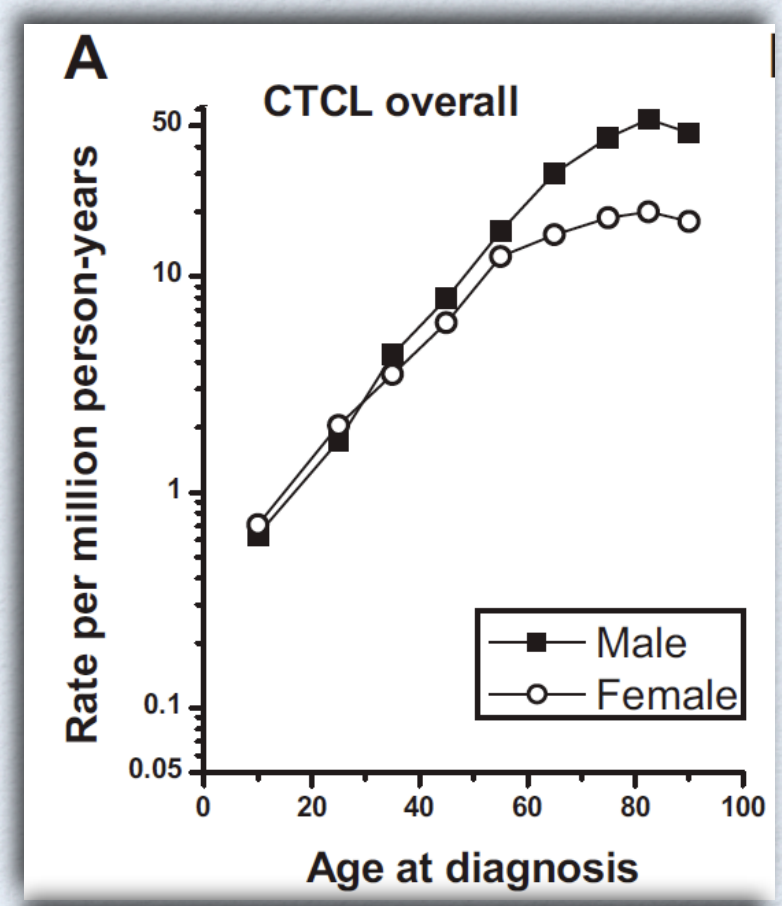
Linfomas cutáneos

- Piel: 2º localización de linfomas extranodales
- Primarios: comportamiento indolente
- Más comunes derivados de células T que B
  - Células T: 7.7/1 000 000 / año
  - Células B: 3.1/1 000 000 / año



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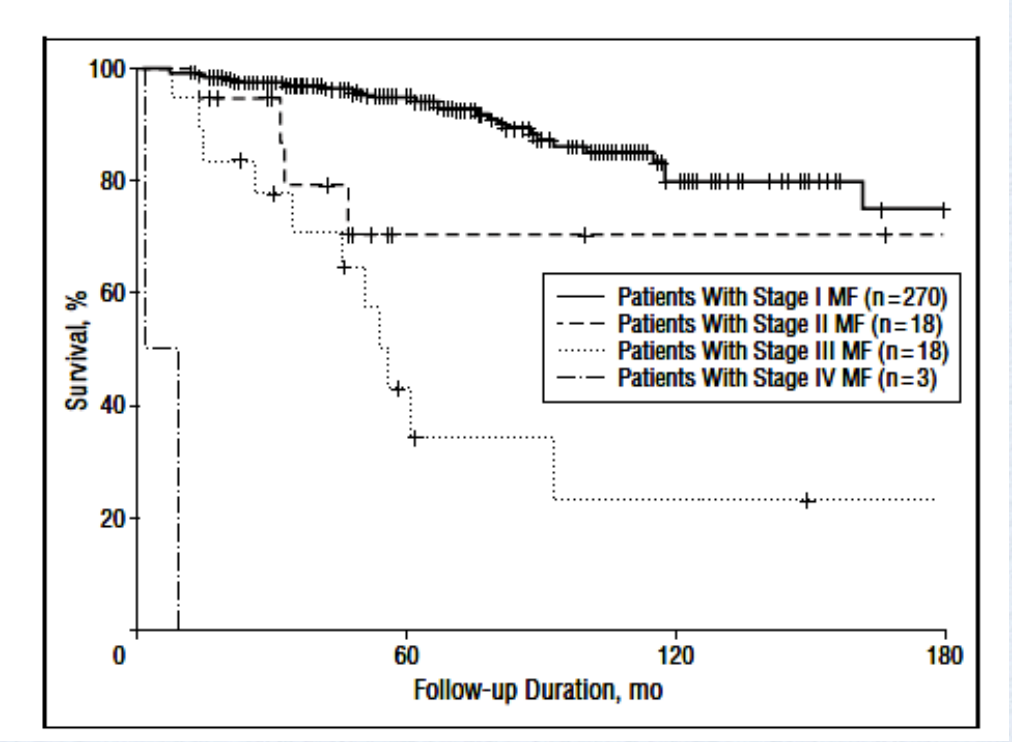
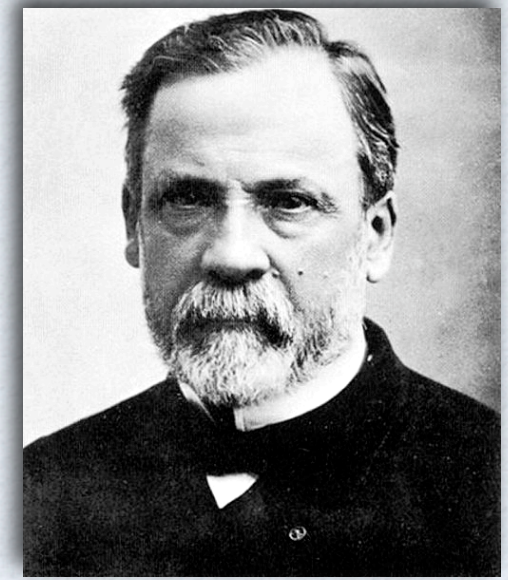


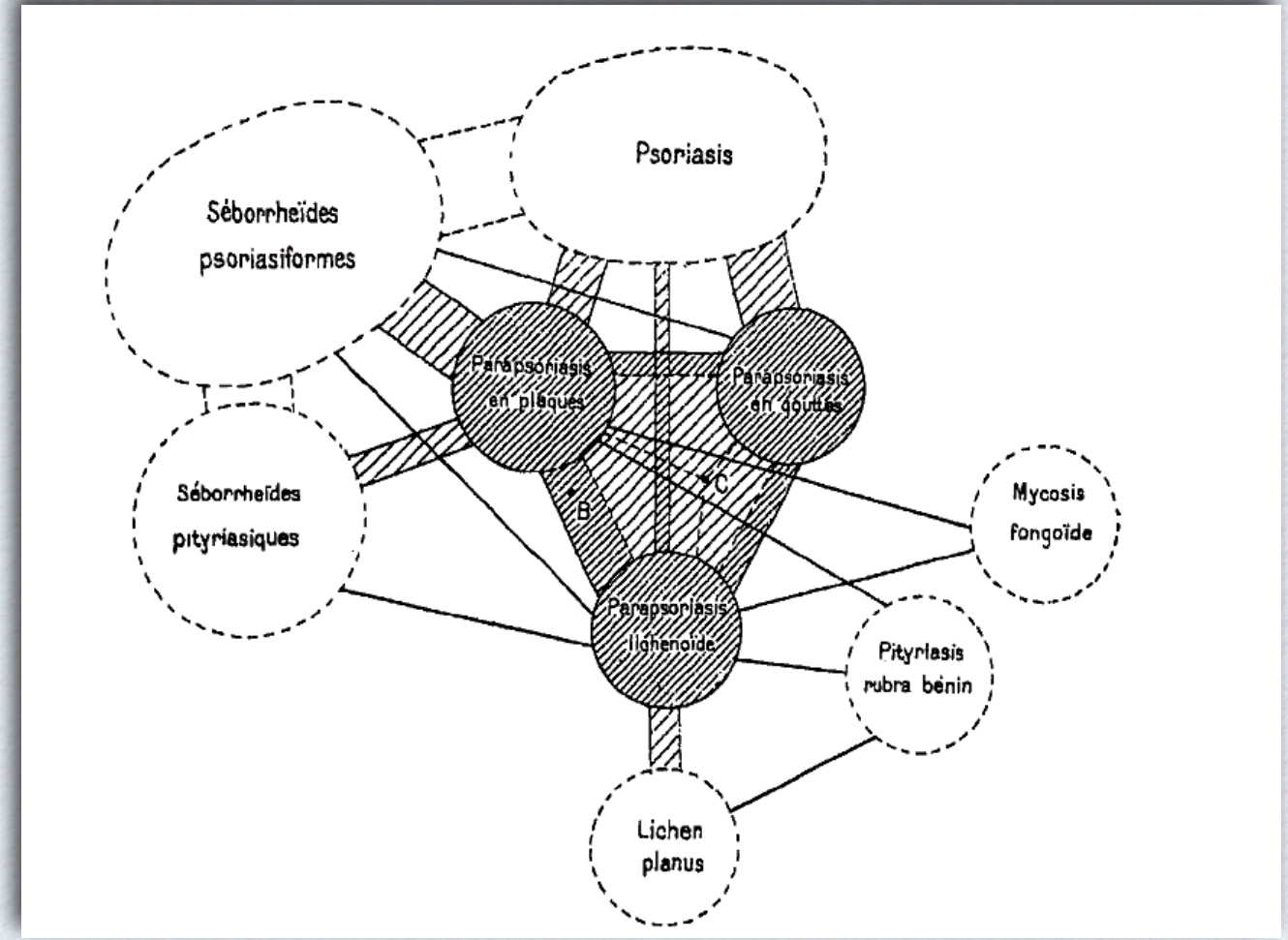
Clasificación (EORTC) y frecuencia relativa

WHO-EORTC	Frequency (%)	5-year Survival (%)
Cutaneous T-cell and NK-cell lymphoma		
Indolent		
Mycosis fungoides	44	88
Follicular MF	4	80
Pagetoid reticulosis	<1	100
Granulomatous slack skin	<1	100
$CD30^+$ lymphoproliferative diseases		
Anaplastic large cell lymphoma	8	95
Lymphomatoid papulosis	12	100
Subcutaneous panniculitis-like T-cell lymphoma	1	82
$CD4^+$ small/medium pleomorphic T-cell lymphoma	2	72
Agressive		
Sézary syndrome	3	24
Cutaneous aggressive $CD8^+$ T-cell lymphoma	<1	18
Cutaneous γ/δ T-cell lymphoma	<1	—
Cutaneous peripheral T-cell lymphoma unspecified	2	16
Cutaneous NK/T-cell lymphoma, nasal-type	<1	—

Micosis fungoides

- Linfoma no Hodgkin T periférico, epidermotropo, de bajo grado de malignidad de presentación cutánea y evolución progresiva
- Características histológicas (excepto en fases iniciales), fenotípicas y genotípicas bien definidas
  - CD2+, CD3+, CD4+, CD5+, CD44RO+, TCRβ+
  - CD8- (CD8+), CD30-
  - Pérdida de CD2, CD7, CD4, CD5



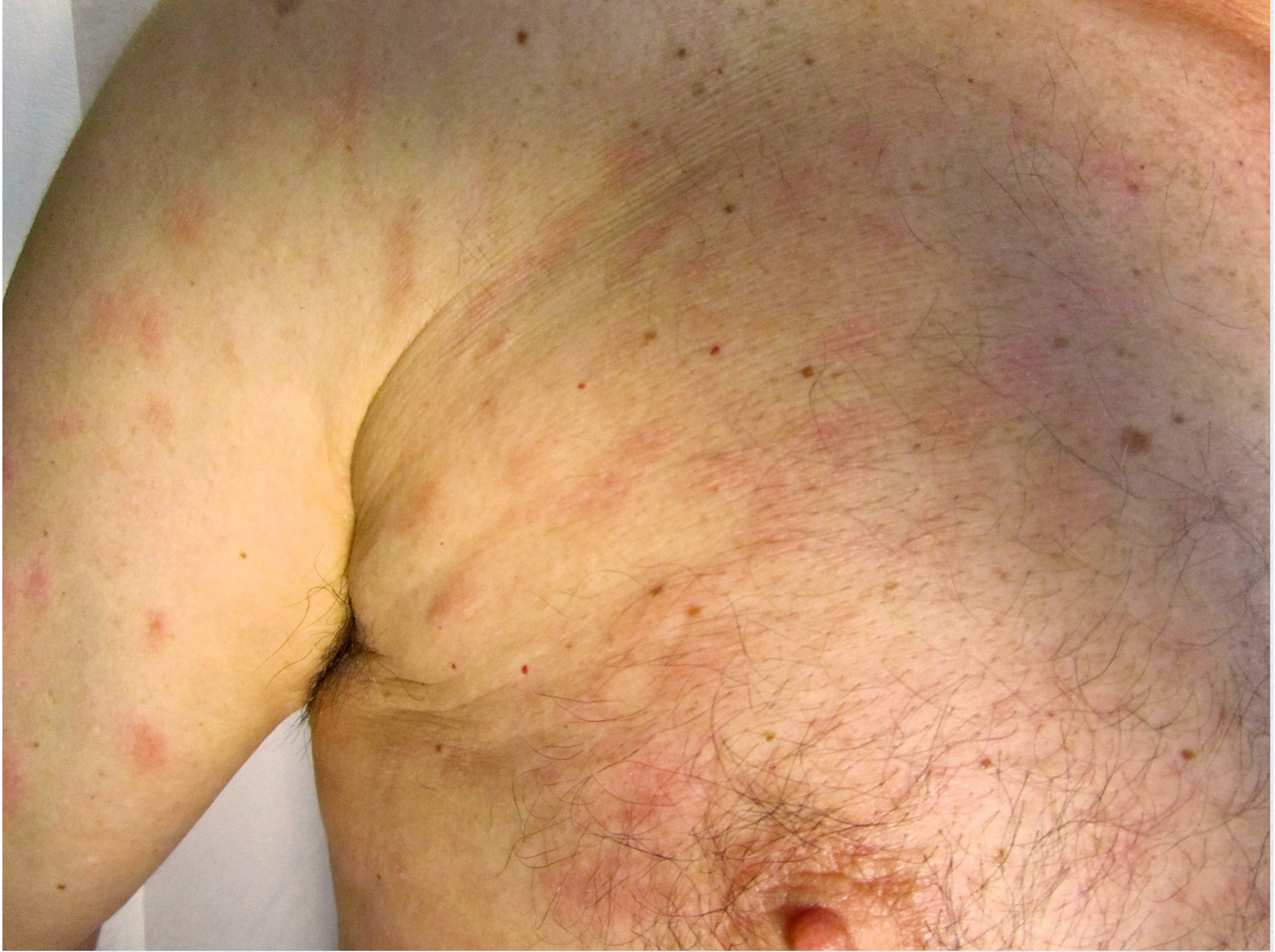


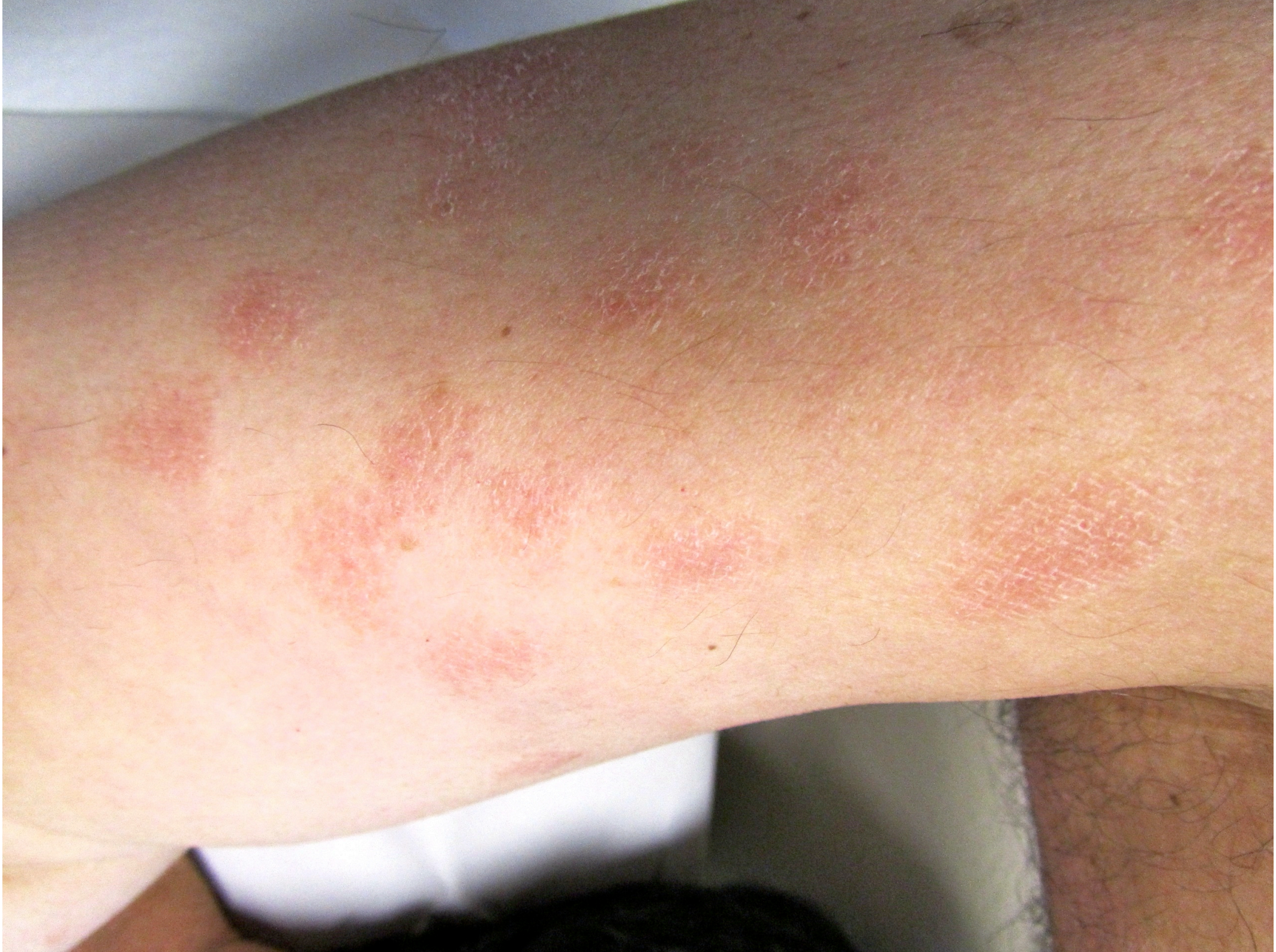
- Parapsoriasis en gotas → psoriasis *gutata*
- Parapsoriasis en placas → dermatitis seborreica
- Parapsoriasis liquenoide → liquen plano

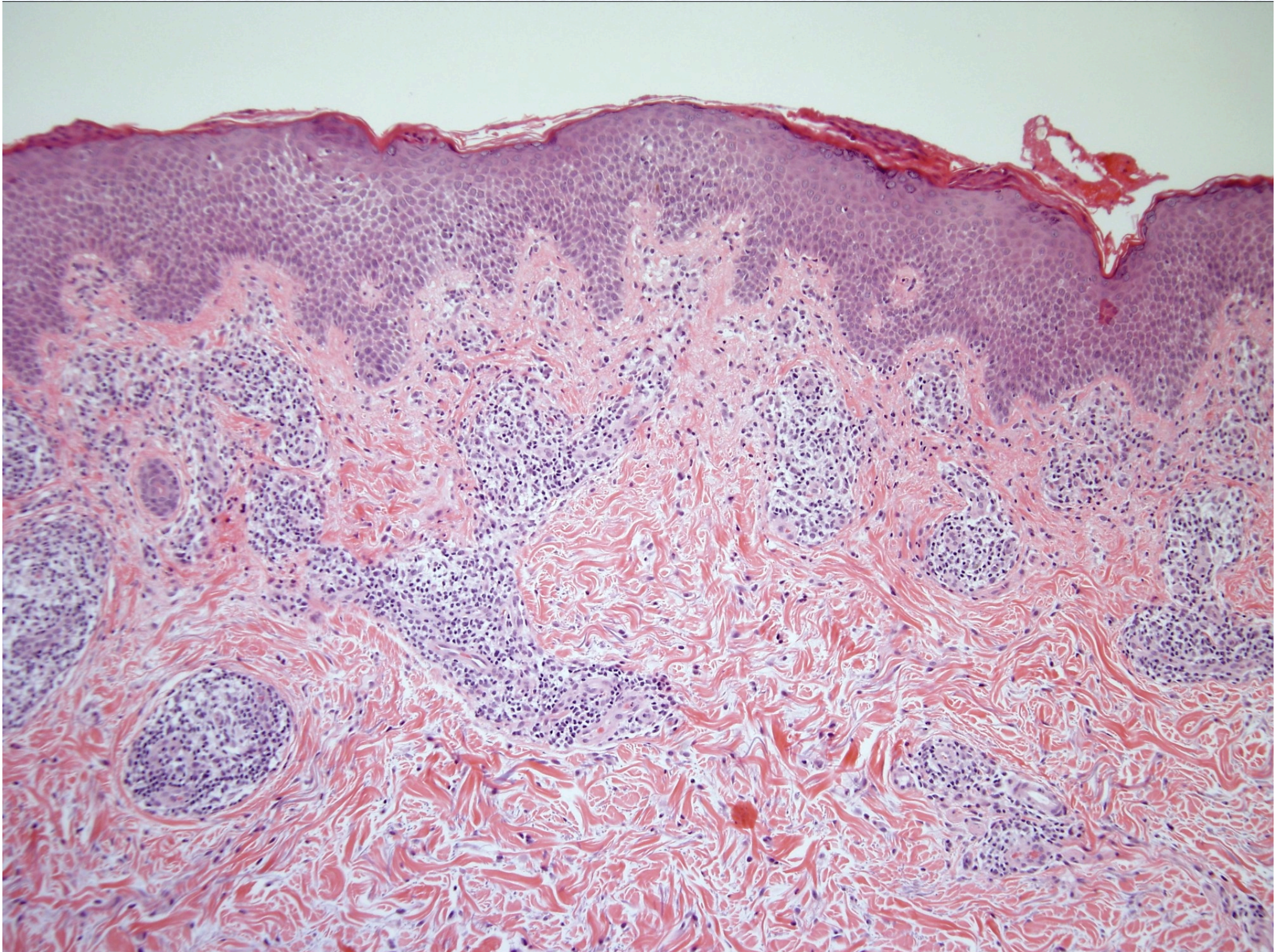
Parapsoriasis

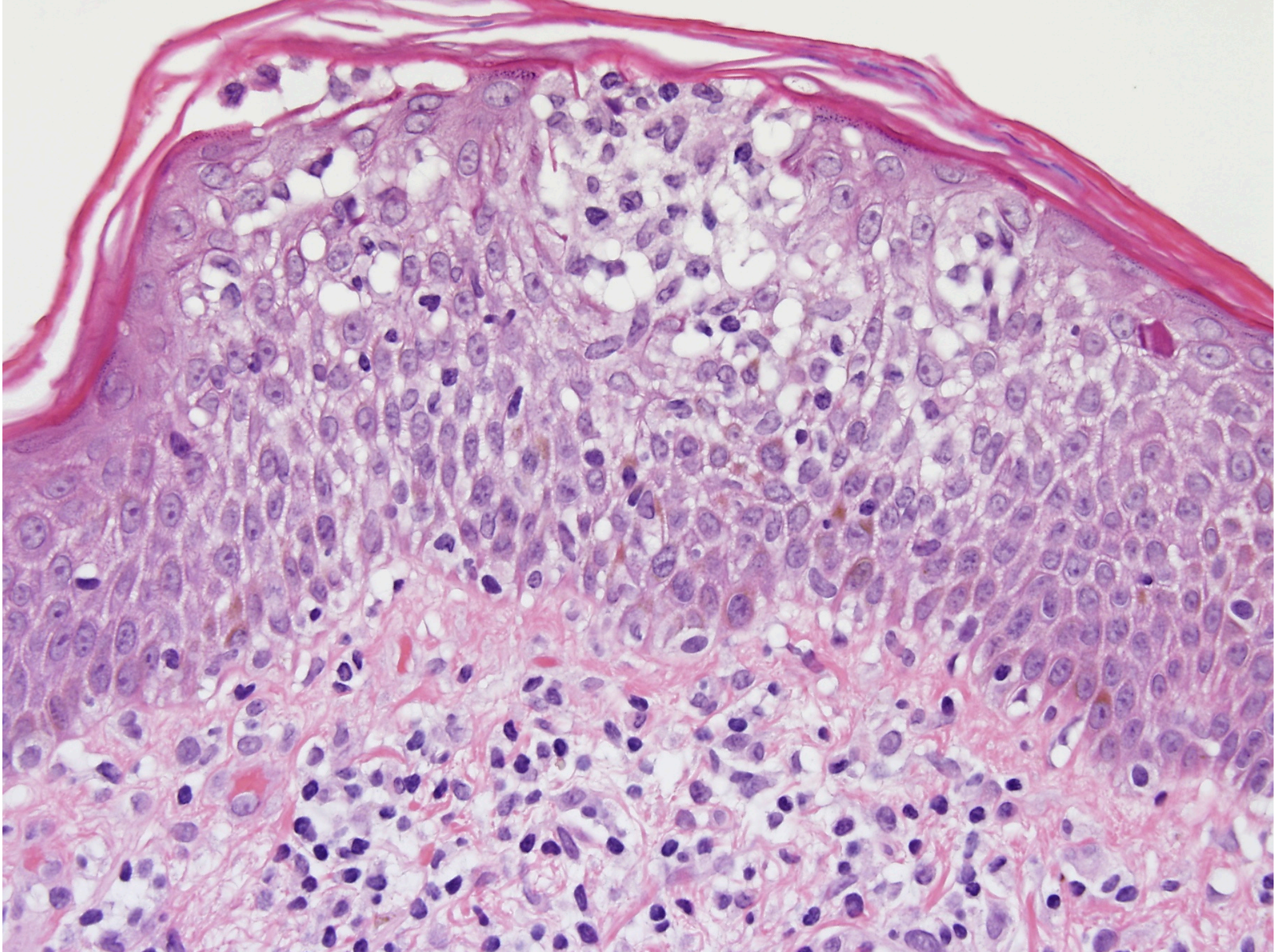
- Conjunto de enfermedades previamente descritas
- Idiopáticas, crónicas, asintomáticas y resistentes al tratamiento
- Características compartidas por la psoriasis: **"parapsoriasis"**
- División placas pequeñas y placas grandes:
  - Degos 1952
  - Palmer 1962

	Placas pequeñas	*Placas grandes*
Edad	Adultos	Todas las edades
Género (h:m)	5:1	2:1
Clínica	Múltiples placas 2-5 cm, ovales o digitiformes Eritematosas y con descamación	Pocas placas (>5 cm) Descamativas, con o sin telangiectasia e hiperpigmentación
Localización	Tronco y extremidades superiores	Mama y nalgas
Histopatología	Infiltrado linfoide perivascular sin epidermotropismo Paraqueratosis	Infiltrado en banda sin epidermotropismo significativo Atrofia epidérmica
Pronóstico	No progresión	Puede evolucionar a MF









Acta Derm Venereol 2005; 85: 318–323

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INVESTIGATIVE REPORT

A Retrospective Study of the Probability of the Evolution of Parapsoriasis en Plaques into Mycosis Fungoides

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Parapsoriasis en plaque has been suggested to be an early manifestation of mycosis fungoides (cutaneous T-cell lymphoma). We explored the disease course of patients with small plaque or large plaque parapsoriasis in a 26-year retrospective cohort analysis of 105 parapsoriasis patients, who were clinically and histopathologically followed up in Helsinki and Tampere University Hospitals. Eventual later cancers of these patients were verified from the Finnish Cancer Registry. In the small plaque parapsoriasis group, 7 patients (10%) and in the large plaque parapsoriasis group 12 patients (35%), developed histologically confirmed mycosis fungoides during a median of 10 and 6 years, respectively. No significant differences were found regarding the risk of developing mycosis fungoides or the tendency to remission in patients treated with or without phototherapy. Our results show that not only large plaque parapsoriasis, but also small plaque parapsoriasis, as currently defined in textbooks, can progress to mycosis fungoides. The benefits of phototherapy are equivocal in parapsoriasis treatment as far as progression to cancer is concerned.
Key words: parapsoriasis; mycosis fungoides; cutaneous T-cell lymphoma; phototherapy; PUVA.

(Accepted December 22, 2004.)

Acta Derm Venereol 2005; 85: 318–323.

A patient with clinicopathologic features of small plaque parapsoriasis presenting later with plaque-stage mycosis fungoides: Report of a case and comparative retrospective study of 27 cases of "nonprogressive" small plaque parapsoriasis

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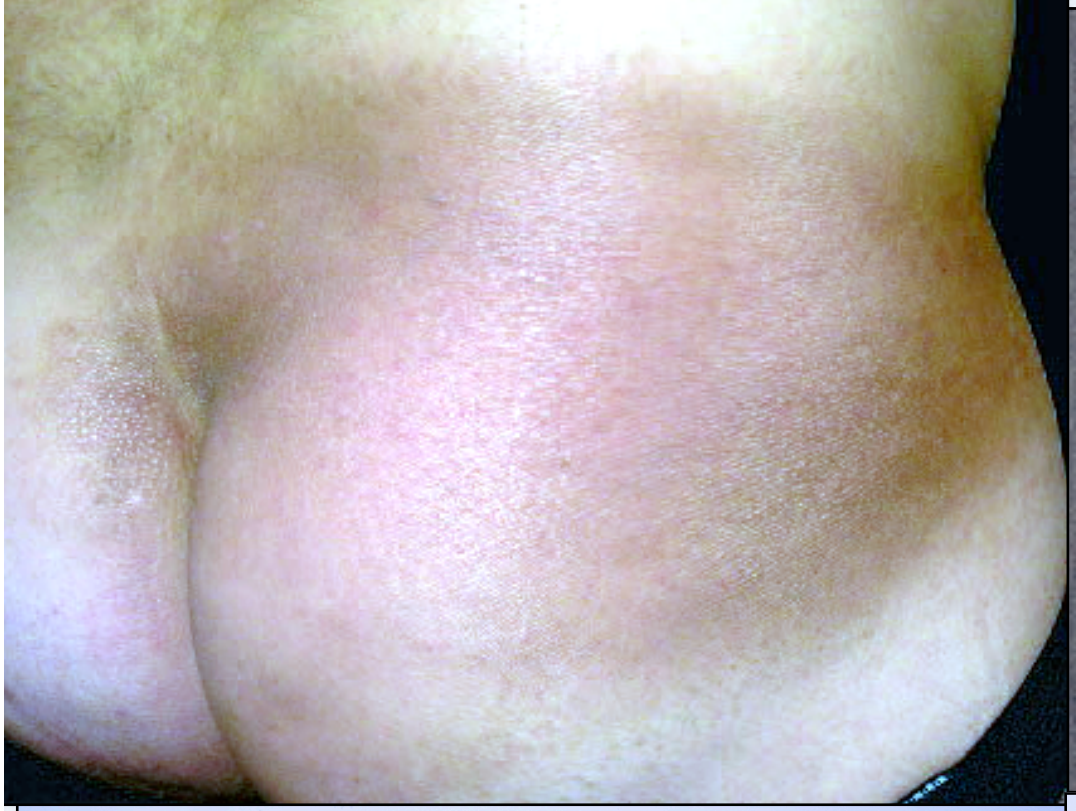
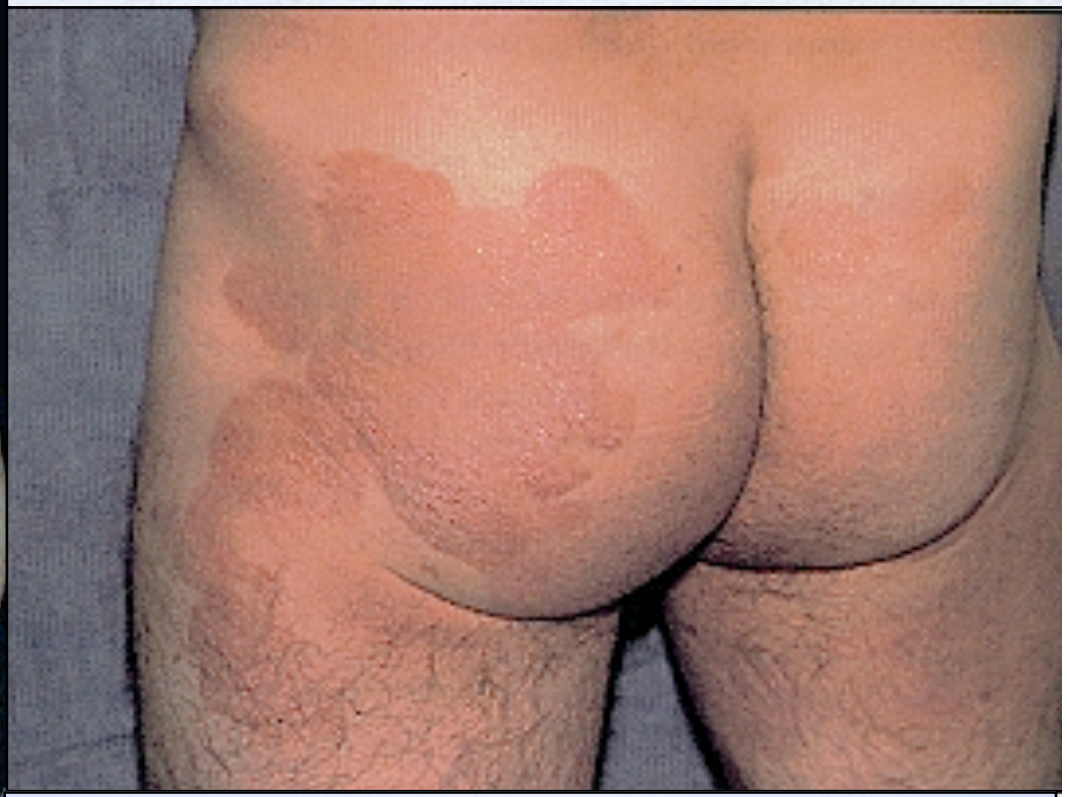
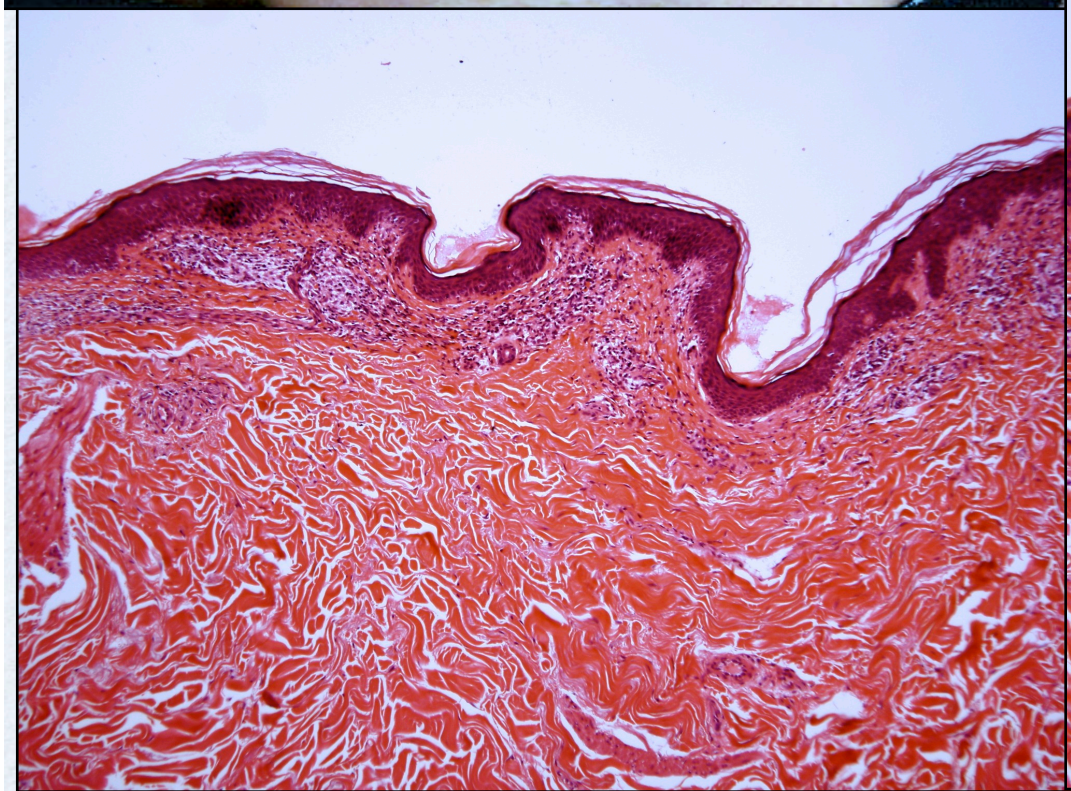
Conclusions: Occasionally patients with the clinical and pathologic presentation of SPP may develop typical features for MF. This event seems to be extremely rare; at present there appears to be no means to predict such a course. The vast majority of SPP patients will never have disease progression to MF. (J Am Acad Dermatol 2008;59:474-82.)

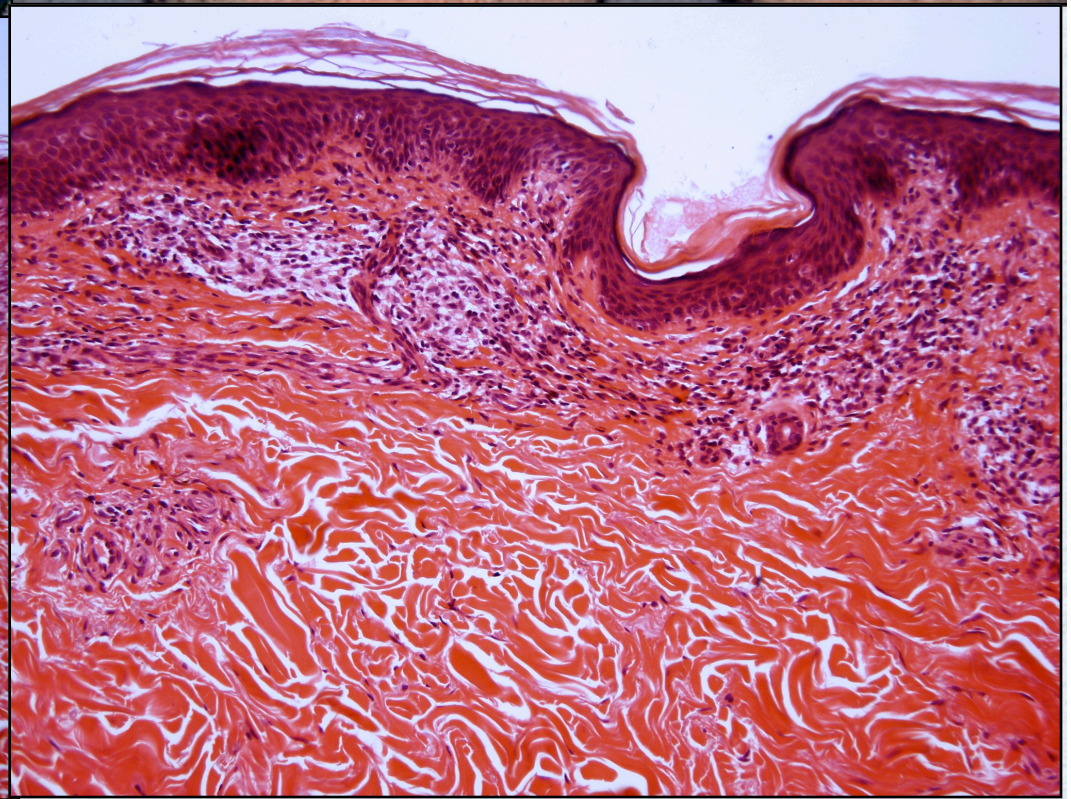
Parapsoriasis en placas grandes

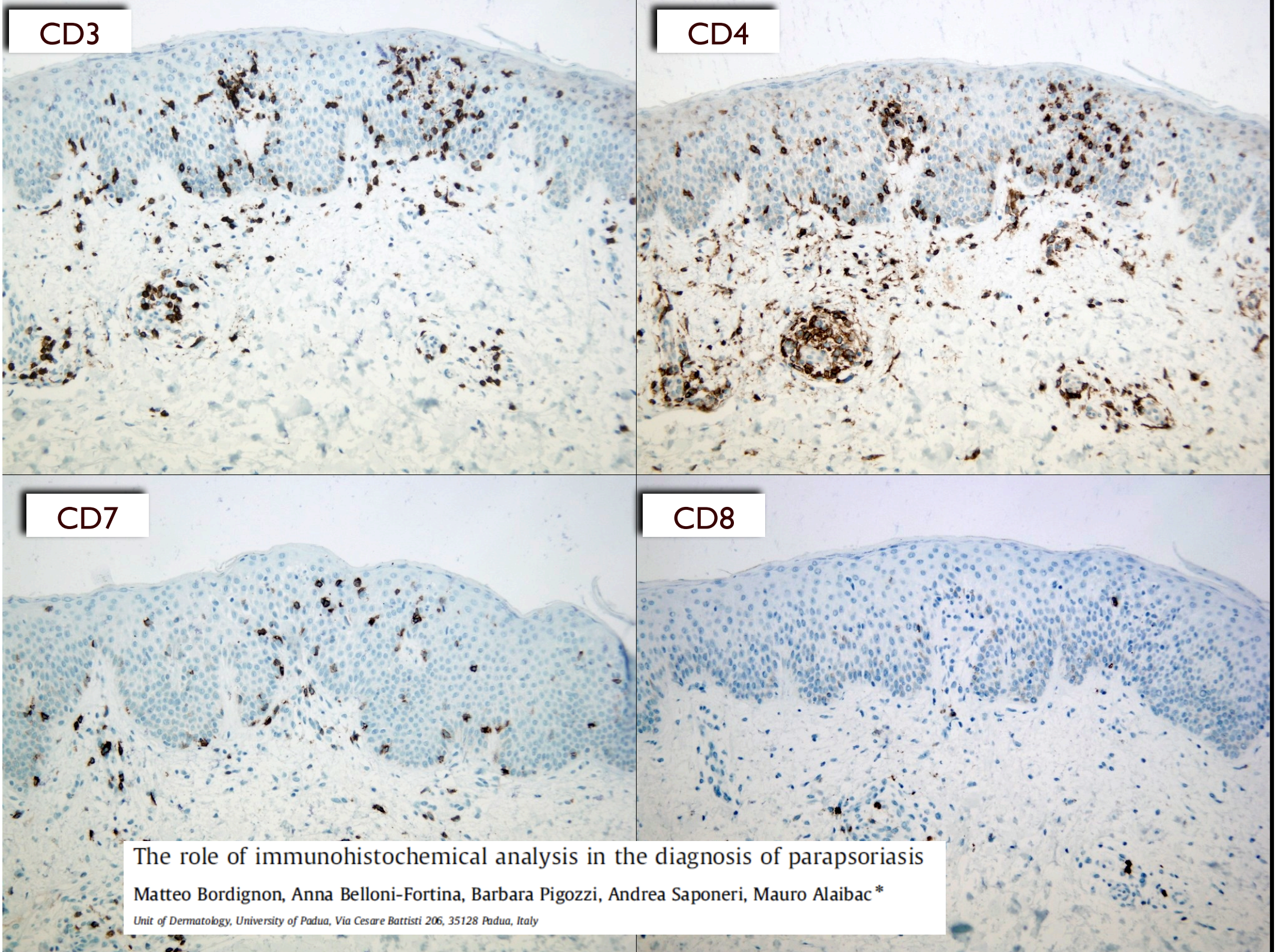
- Lesión premaligna tendente a la evolución a MF vs. MF inicial
  - Tasa evolución 7,5-30%
  - Casos comprobados (NCI): 16%
  - Casos de LCCT con dermatosis previas 77%
- Esperanza de vida no alterada ¿linfoma?











The role of immunohistochemical analysis in the diagnosis of parapsoriasis

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Parapsoriasis en placas grandes

- Kikucki 1993:
  - 4/20 (Southern blot)
- Mielke 1994:
  - 11/22 (TCRγ)
- Simon 2000:
  - 12 casos: 6 clonales (TCRγ)
  - 1➟ MF tras 2-21 (9) años de seguimiento

Table 1. Summary of clonality results

	No. of cases	Monoclonal	Oligoclonal	Polyclonal
CTCL				
Mycosis fungoides	15	13	0	2
Pleomorphic T cell lymphoma	2	2	0	0
Angioimmunoblastic lymphadenopathy	1	1	0	0
Peripheral T cell lymphoma	4	4	0	0
LyP	1	1	0	0
Idiopathic lymphocytic proliferations associated with CTCL				
LPP	5	5	0	0
Pigmentary purpuric dermatosis	11	3	1	7
Pytiriasis lichenoides chronica	9	2	1	6
Pityriasis lichenoides et varioliformis acuta	4	2	0	2
Idiopathic follicular mucinosis	4	0	2	2
Drug-induced lymphomatoid hypersensitivity				
Reversible T-cell dyscrasias	16	5	2	8
Reactive non-lymphoid dermatitis				
Lymphomatoid lupus erythematosus	2	1	0	1
Morphea	3	0	1	3
T-cell rich pseudolymphoma	3	0	1	2

CTCL, cutaneous T-cell lymphoma, LyP, lymphomatoid papulosis, LPP, large plaque parapsoriasis.

Plaza JA, Morrison C, Magro CM. Assessment of TCR-beta clonality in a diverse group of cutaneous T-cell infiltrates. J Cutan Pathol 2008; 35: 358–365. © Blackwell Munksgaard 2007.

Parapsoriasis en placas grandes

Klemke (*J Pathol 2002*)

- Clonalidad
  - Lesiones cutáneas: parapsoriasis 19,6% vs. MF 66,6%
  - Sangre periférica 26,7 % vs. 12,5%
- Conclusiones:
  - LCCT está asociado a clonalidad desde inicio
  - Clonalidad no es un requisito para lesiones precursoras
  - Céls. T circulantes clonales se asocian a progresión
- Dermatitis clonal

Técnicas auxiliares

- Emplear en conjunción con la histología
- Variabilidad según tipo de técnica y fijación del material
- Estudio de clonalidad:
  - PCR > Southern blot (60-90% vs. 59%)
  - Preferible material congelado
  - TCRγ mediante PCR/DGCE
  - Cuidadosa selección de controles

Algoritmo para el diagnóstico de MF inicial

Defining early micosis fungoides. J Am Acad Dermatol 2005

Table 3. Algorithm for the diagnosis of early MF[15]

Criteria	Major (2 points)	Minor (1 point)
Clinical		
Persistent and/or progressive patches and plaques plus	Any 2	Any 1
(1) Non–sun-exposed location		
(2) Size/shape variation		
(3) Poikiloderma		
Histopathologic		
Superficial lymphoid infiltrate plus	Both	Either
(1) Epidermotropism without spongiosis		
(2) Lymphoid atypia*		
Molecular/biologic: clonal TCR gene rearrangement	NA†	Present
Immunopathologic		
(1) CD2,3,5 less than 50% of T cells	NA†	Any 1
(2) CD7 less than 10% of T cells		
(3) Epidermal discordance from expression of CD2,3,5 or CD7 on dermal T cells		

— indicates not applicable.

*Lymphoid atypia is defined as cells with enlarged hyperchromatic nuclei and irregular or cerebriform nuclear contours.

†Not applicable since it cannot fulfill any major criteria.

Conclusiones

- Parapsoriasis en placas pequeñas (dermatitis crónica superficial, dermatitis superficial persistente):
  - No relacionada con linfoma
  - Evitar sobretratamiento
- Parapsoriasis en placas grandes:
  - Superposición con fases iniciales de LCCT