CLASSIFICAZIONE CLINICO-PATOLOGICA DEI LINFOMI CUTANEI

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Table 1. WHO/EORTC classification of cutaneous lymphomas

WHO/EORTC classification of cutaneous lymphomas	WHO classification for lymphoid tissues† (ICD-O Code)
Cutaneous T-cell and NK-cell lymphomas	Mature T-cell and NK-cell neoplasms
Mycosis fungoides (MF)	Mycosis fungoides (MF) (9700/3)
MF variants and subtypes	MF variants and subtypes
Folliculotropic MF	Folliculotropic MF
Pagetoid reticulosis	Pagetoid reticulosis
Granulomatous slack skin	Granulomatous slack skin
Sézary syndrome	Sézary syndrome (9701/3)
Adult T-cell leukaemia/lymphoma	Adult T-cell leukaemia/lymphoma (9827/3)
Primary cutaneous CD30+ lymphoproliferative disorders	Primary cutaneous CD30+ T-cell lymphoproliferative disorders
Primary cutaneous anaplastic large cell lymphoma	Primary cutaneous anaplastic large cell lymphoma (9718/3)
Lymphomatoid papulosis	Lymphomatoid papulosis (9718/1)
Subcutaneous panniculitis-like T-cell lymphoma*	Subcutaneous panniculitis-like T-cell lymphoma* (9708/3)
Extranodal NK/T-cell lymphoma, nasal type	Extranodal NK/T-cell lymphoma, nasal type (9719/3)
Primary cutaneous peripheral T-cell lymphoma, unspecified	Primary cutaneous peripheral T-cell lymphoma, rare subtypes
Primary cutaneous aggressive epidermotropic CD8+ T-cell lymphoma (provisional)	Primary cutaneous CD8+ aggressive epidermotropic cytotoxic T-cell lymphoma (provisional) (9709/3)
Cutaneous γ/δ+ T-cell lymphoma (provisional)	Primary cutaneous γ/δ T-cell lymphoma (9726/3)‡
Primary cutaneous CD4+ small/medium-sized pleomorphic T-cell lymphoma (provisional)	Primary cutaneous CD4+ small/medium T-cell lymphoma (provisional) (9709/3)
Cutaneous B-cell lymphomas	Mature B-cell neoplasms
Primary cutaneous marginal zone B-cell lymphoma	Extranodal marginal zone lymphoma of mucosa-associated lymphoid tissue (MALT lymphoma) (9699/3)
Primary cutaneous immunocytoma	
Primary cutaneous plasmacytoma	
Follicular hyperplasia with monotypic plasma cells	
Primary cutaneous follicle centre lymphoma	Primary cutaneous follicle centre lymphoma (9597/3)
Growth patterns: follicular, follicular and diffuse, diffuse	Diffuse large B-cell lymphoma, NOS (9680/3)
Primary cutaneous diffuse large B-cell lymphoma, leg type	Primary cutaneous diffuse large B-cell lymphoma, leg type (9680/3)
Primary cutaneous diffuse large B-cell lymphoma, other	Intravascular large B-cell lymphoma (9712/3)
Primary cutaneous intravascular large B-cell lymphoma	
Precursor haematological neoplasm	Precursor neoplasms
CD4+/CD56+ haematodermic neoplasm (formerly blastic NK cell lymphoma)	Blastic plasmacytoid dendritic cell neoplasm (9727/3)

*Phenotype (by definition): T-cell receptor α/β chain positive.
†List restricted to cutaneous lymphomas in the WHO classification (4th edn).
‡Provisional International Classification of Diseases (ICD)-O Code.

Kempf & Sander, 2011

Review article

WHO-EORTC classification for cutaneous lymphomas

Rein Willemze, Elaine S. Jaffe, Günter Burg, Lorenzo Cerroni, Emilio Berti, Steven H. Swerdlow, Elisabeth Ralfkiaer, Sergio Chimenti, José L. Diaz-Perez, Lyn M. Duncan, Florent Grange, Nancy Lee Harris, Werner Kempf, Helmut Kerl, Michael Kurrer, Robert Knobler, Nicola Pimpinelli, Christian Sander, Marco Santucci, Wolfram Sterry, Maarten H. Vermeer, Janine Wechsler, Sean Whittaker, and Chris J. L. M. Meijer

Primary cutaneous lymphomas are currently classified by the European Organization for Research and Treatment of Cancer (EORTC) classification or the World Health Organization (WHO) classification, but both systems have shortcomings. In particular, differences in the classification of cutaneous T-cell lymphomas other than mycosis fungoides, Sézary syndrome, and the group of primary cutaneous CD30+ lymphoproliferative disorders and the classification and terminology of different types of cutaneous B-cell lymphomas have resulted in considerable debate and confusion. During recent consensus meetings representatives of both systems reached agreement on a new classification, which is now called the WHO-EORTC classification. In this paper we describe the characteristic features of the different primary cutaneous lymphomas and other hematologic neoplasms frequently presenting in the skin, and discuss differences with the previous classification schemes. In addition, the relative frequency and survival data of 1905 patients with primary cutaneous lymphomas derived from Dutch and Austrian registries for primary cutaneous lymphomas are presented to illustrate the clinical significance of this new classification. (Blood. 2005;105:3768-3785)

Table 2. Relative frequency and disease-specific 5-year survival of 1905 primary cutaneous lymphomas classified according to the WHO-EORTC classification

WHO-EORTC classification	No.	Frequency, %*	Disease-specific 5-year survival, %
Cutaneous T-cell lymphoma			
Indolent clinical behavior			
Mycosis fungoides	800	44	88
Folliculotropic MF	86	4	80
Pagetoid reticulosis	14	< 1	100
Granulomatous slack skin	4	< 1	100
Primary cutaneous anaplastic large cell lymphoma	146	8	95
Lymphomatoid papulosis	236	12	100
Subcutaneous panniculitis-like T-cell lymphoma	18	1	82
Primary cutaneous $CD4^+$ small/medium pleomorphic T-cell lymphoma†	39	2	75
Aggressive clinical behavior			
Sézary syndrome	52	3	24
Primary cutaneous NK/T-cell lymphoma, nasal-type	7	< 1	NR
Primary cutaneous aggressive CD8+ T-cell lymphoma†	14	< 1	18
Primary cutaneous γ/δ T-cell lymphoma†	13	< 1	NR
Primary cutaneous peripheral T-cell lymphoma, unspecified‡	47	2	16
Cutaneous B-cell lymphoma			
Indolent clinical behavior			
Primary cutaneous marginal zone B-cell lymphoma	127	7	99
Primary cutaneous follicle center lymphoma	207	11	95
Intermediate clinical behavior			
Primary cutaneous diffuse large B-cell lymphoma, leg type	85	4	55
Primary cutaneous diffuse large B-cell lymphoma, other	4	< 1	50
Primary cutaneous intravascular large B-cell lymphoma	6	< 1	65

NR indicates not reached.

*Data are based on 1905 patients with a primary cutaneous lymphoma registered at the Dutch and Austrian Cutaneous Lymphoma Group between 1986 and 2002.

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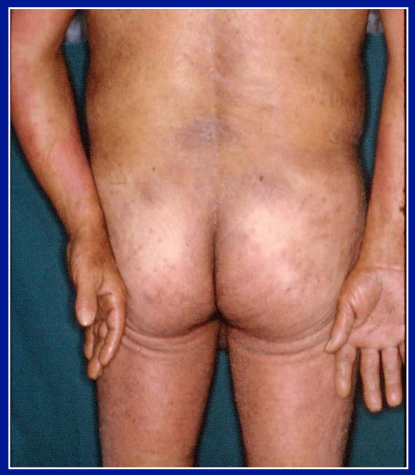

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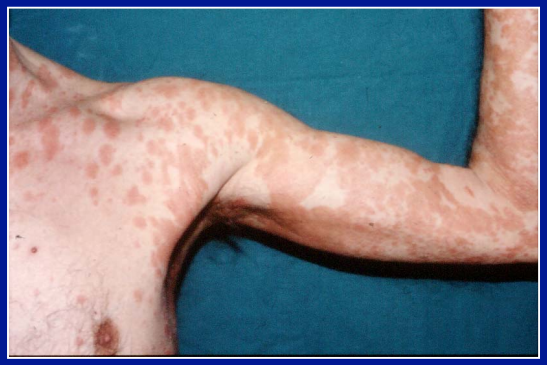

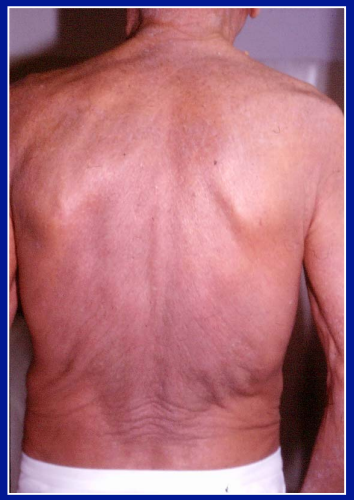

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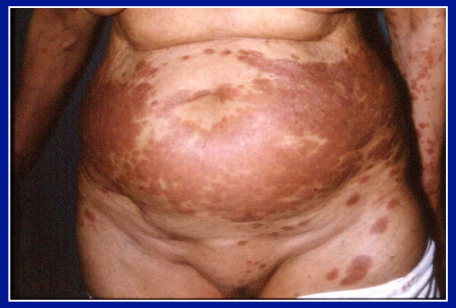

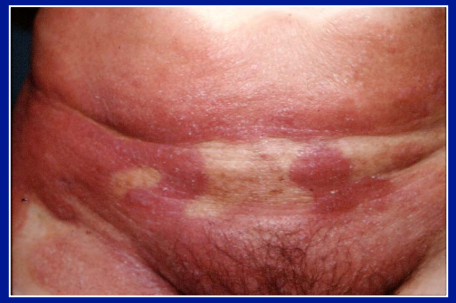

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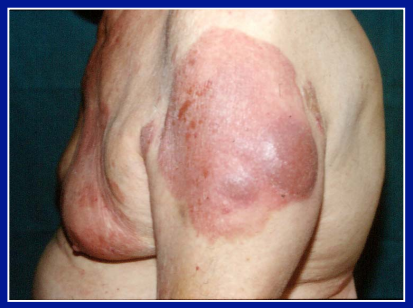
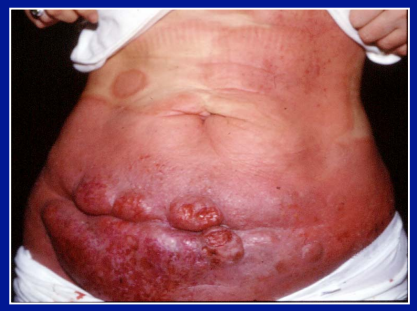




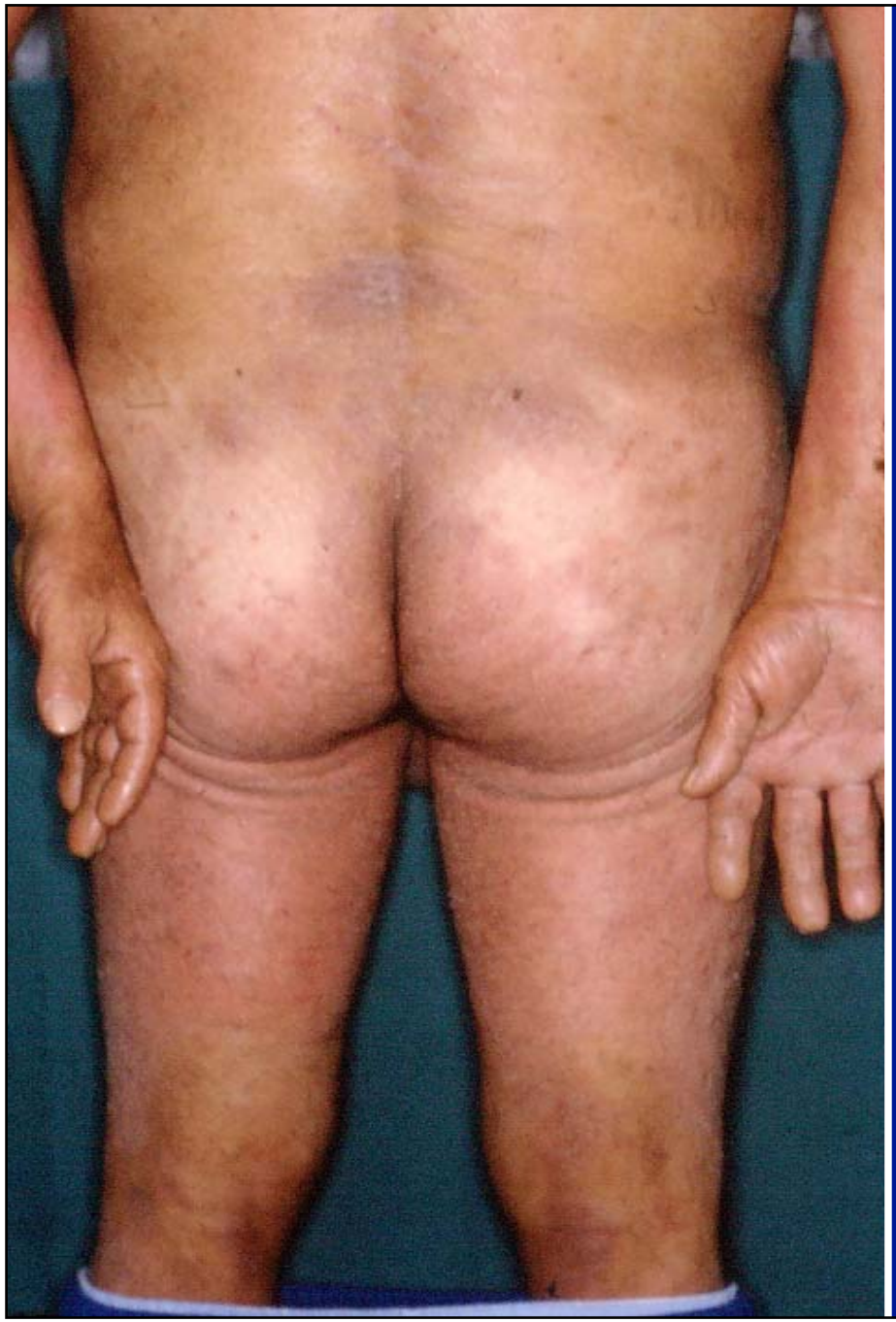
TUMOR STAGE

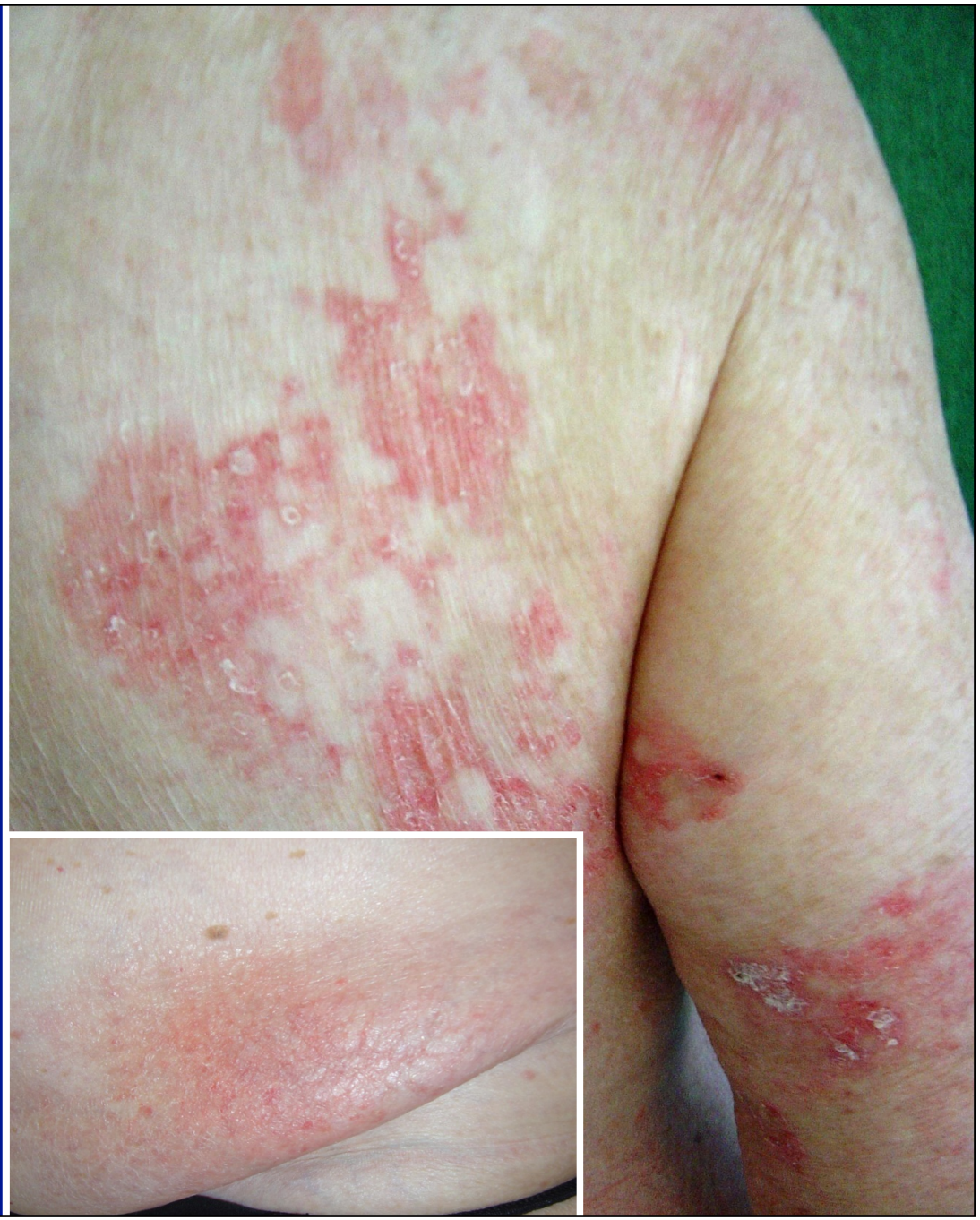
STAGING/CLASSIFICATION OF MF/SS

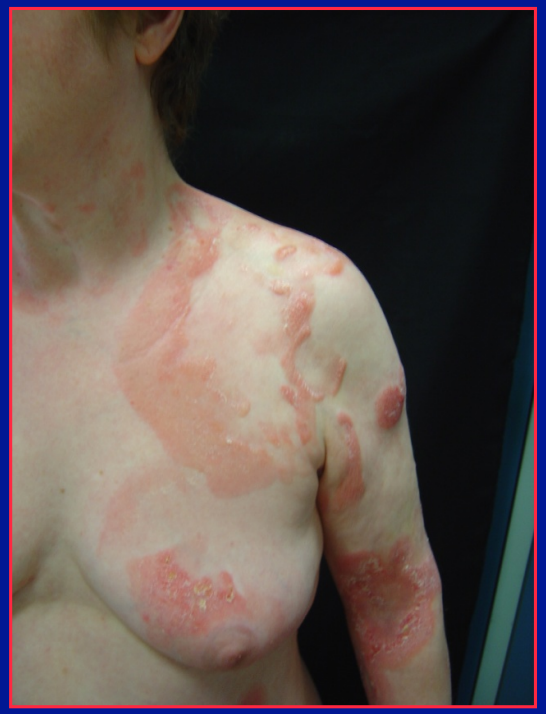
MF Cooperative Group 1979				ISCL- EORTC 2007					
	T	N	M		T	N	M	B	
IA	1	0	0	IA_1 IA_2	1_a 1_b	0 0	0 0	0 0	
1B	2	0	0	IB_1 IB_2	2_a 2_b	0 0	0 0	0 0	
IIA	1-2	1	0	IIA IIA_1 IIA_2	1-2 1_a ,2_a 1_b, 2_b	1 1 1	0 0 0	0 0 0	
IIB	3	0, 1	0	**IIB**	**3**	**0,1**	**0**	**0**	
III	**4**	**0, 1**	**0**	**III**	**4**	**0,1**	**0**	**0, 1**	
IVA	**1-4**	**2-3**	**0**	**IVA_1** **IVA_2**	**1-4** **1-4**	**3** **0-3**	**0** **0**	**0,1** **2**	
IVB	1-4	0-3	1	IVB	1-4	0-3	1	0-2	

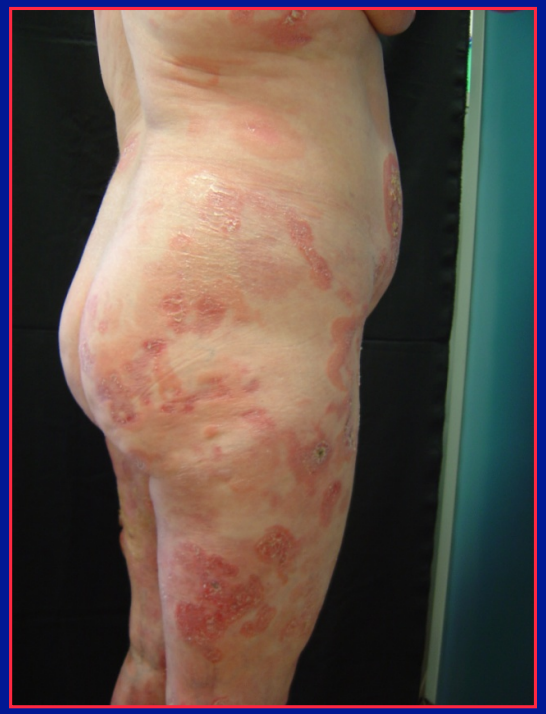
a=patch only
B=patch and plaque

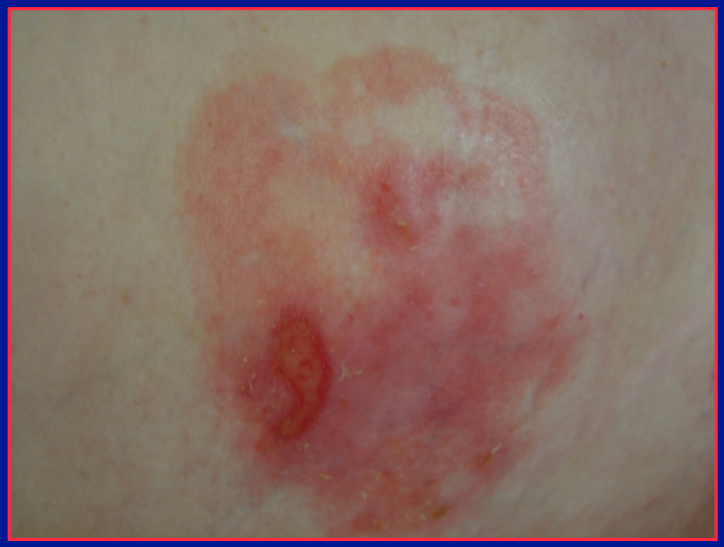
Olsen E, Vonderheid E, Pimpinelli N, et al for ISCL/EORTC. BLOOD, 2007

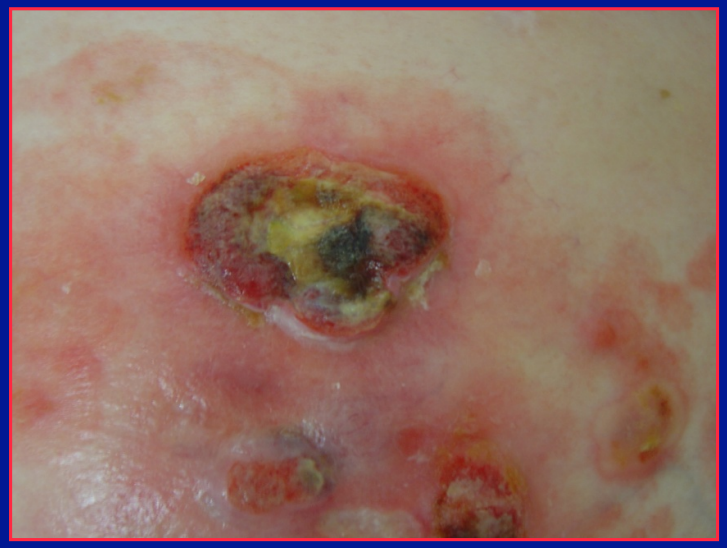












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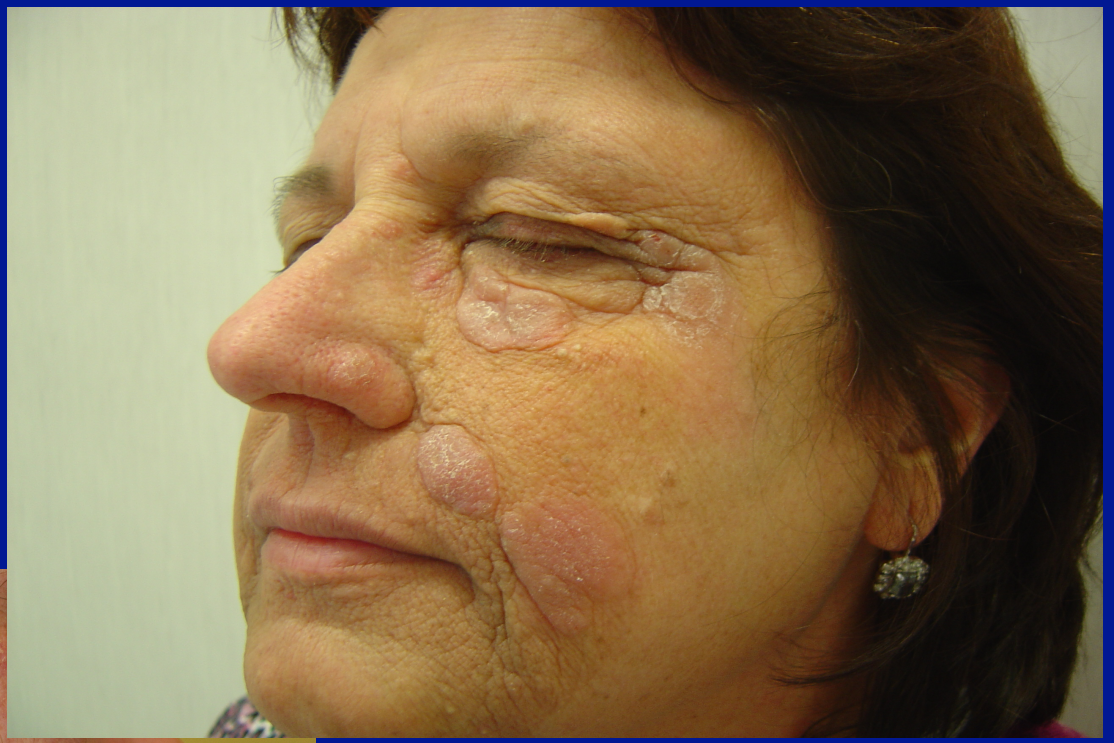

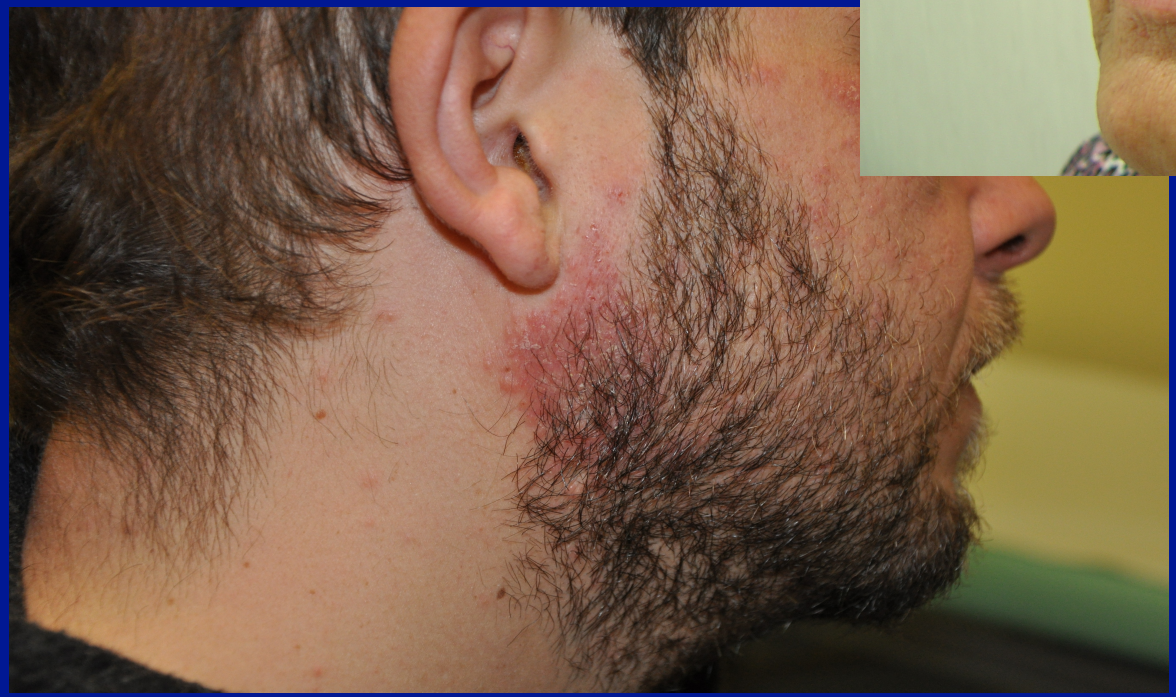

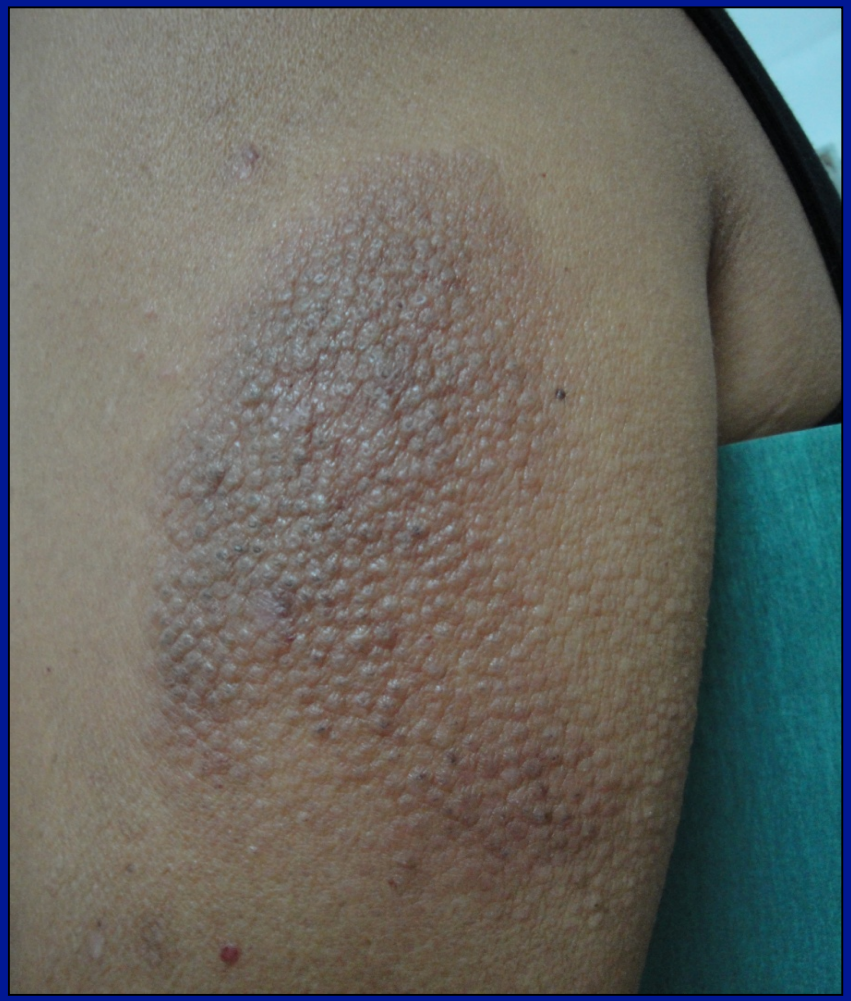

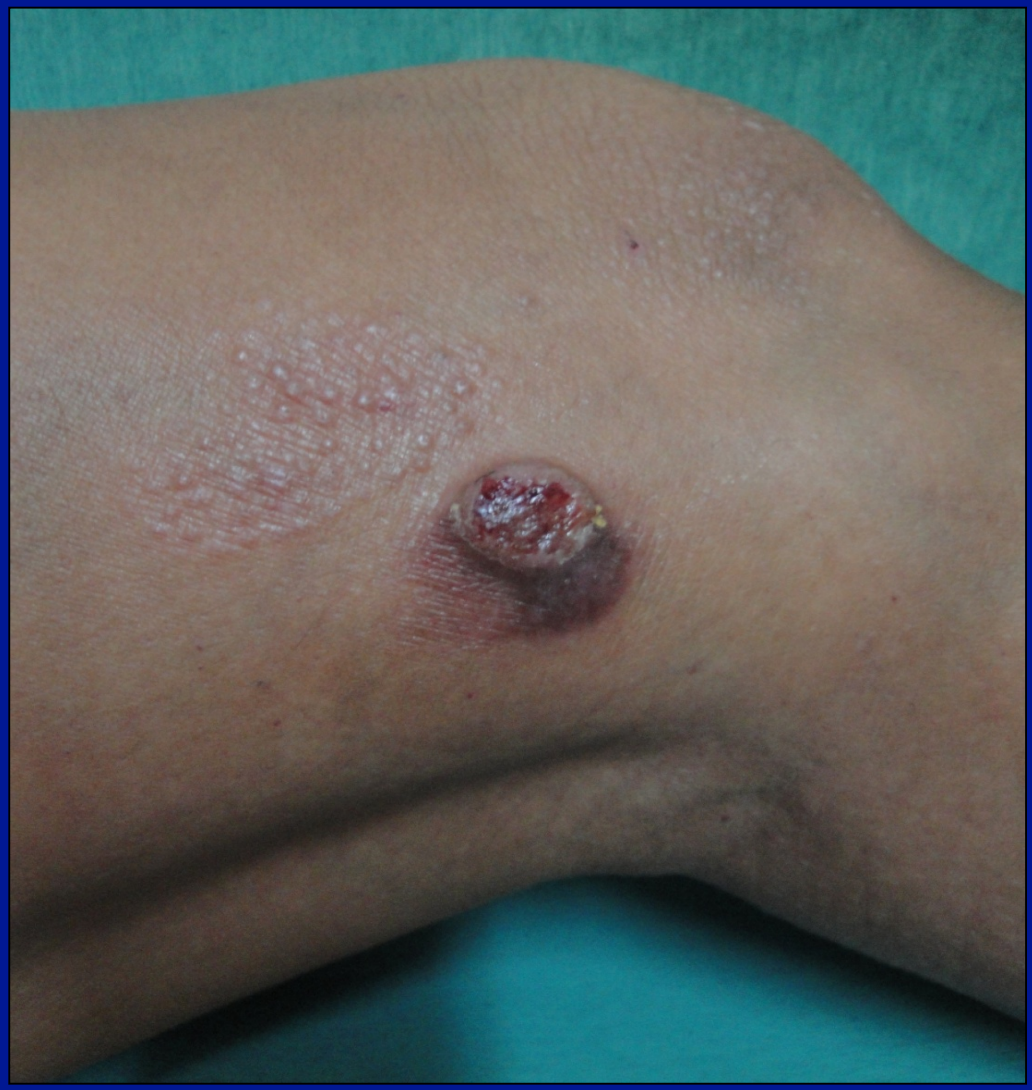

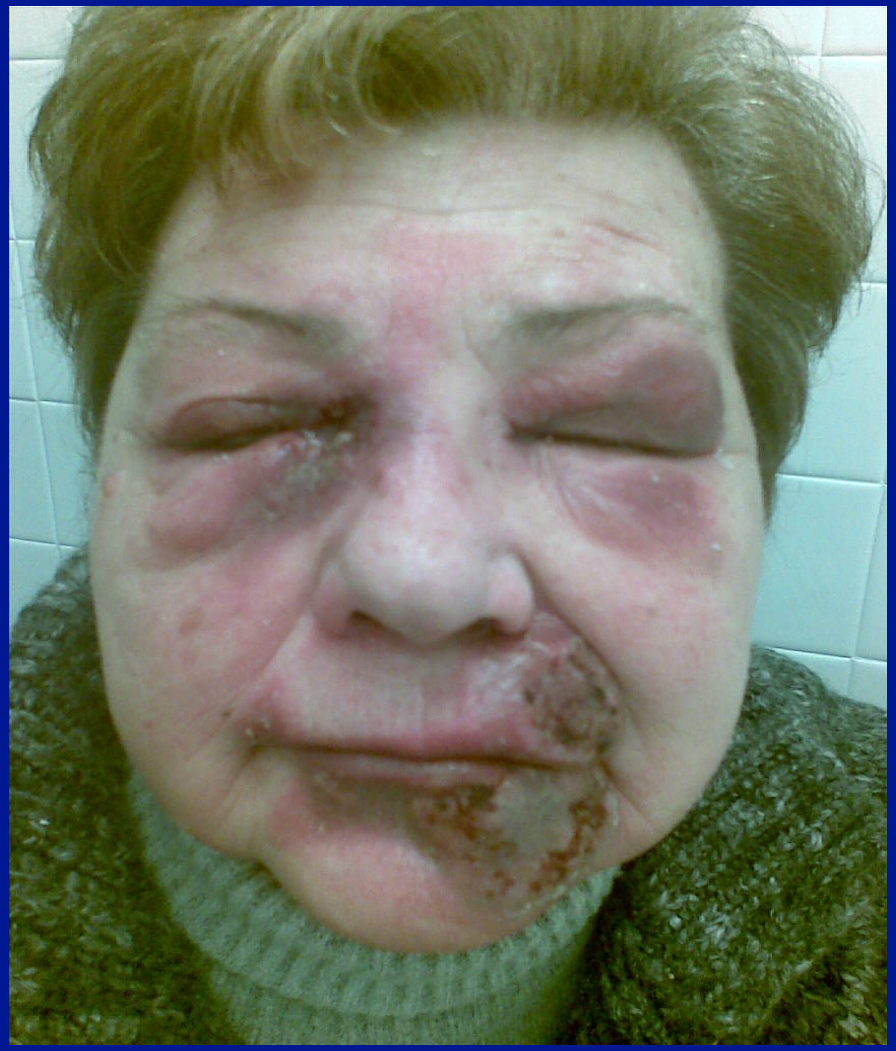

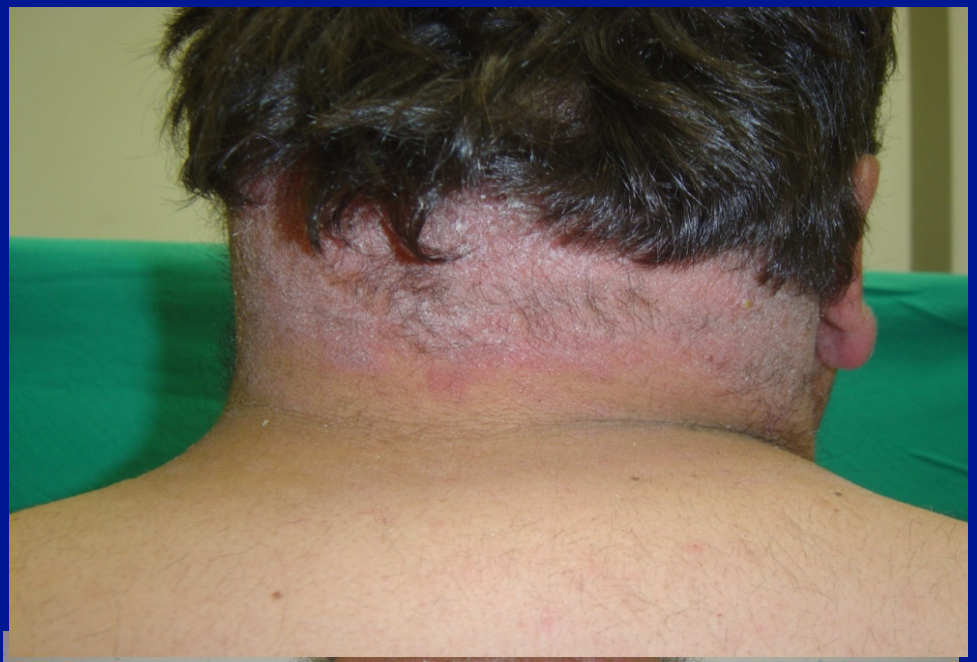

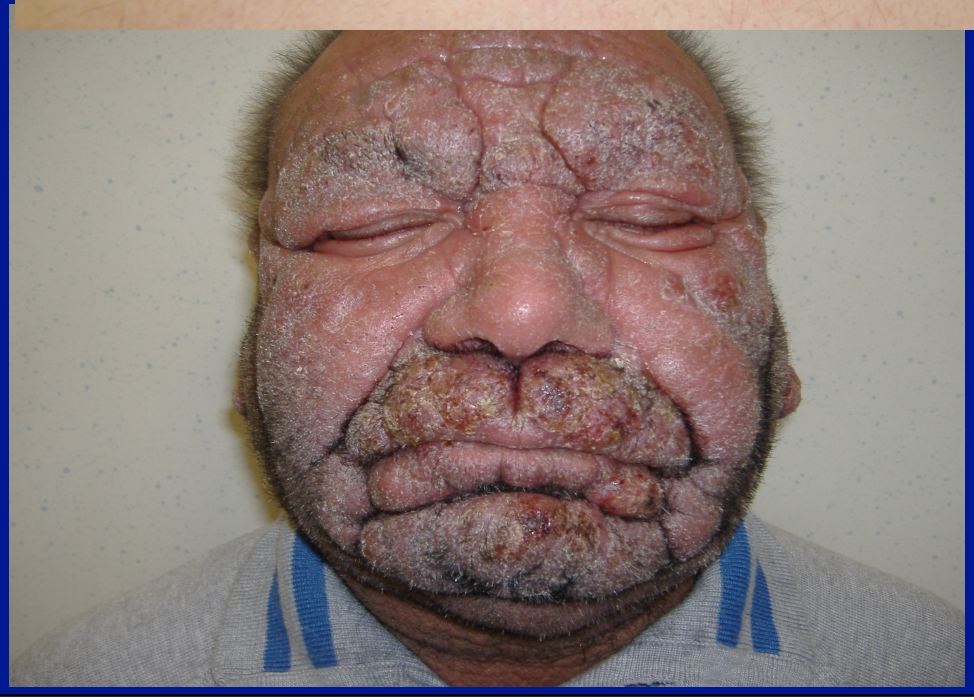

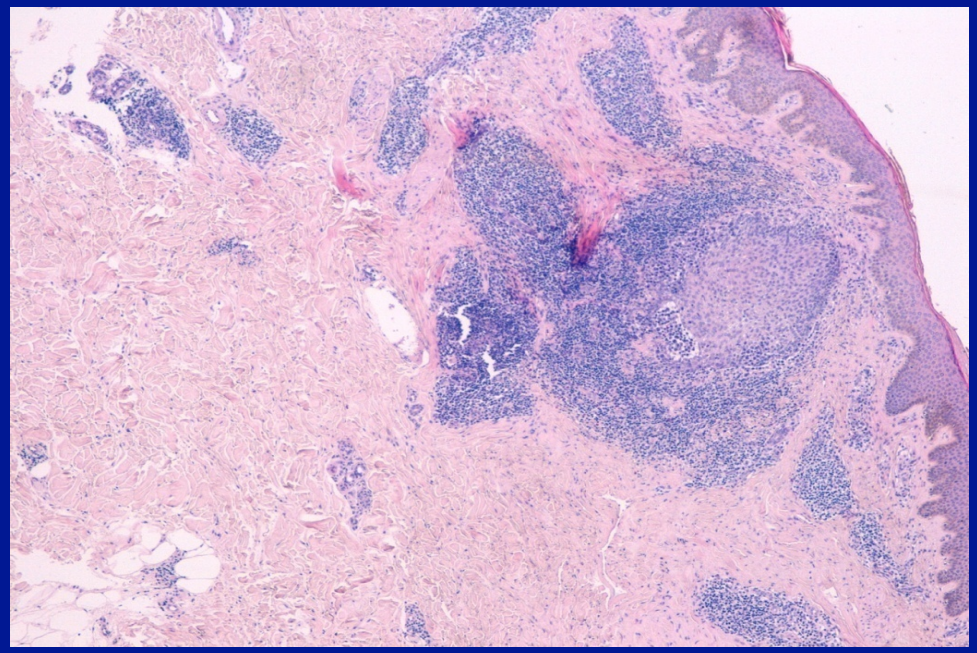

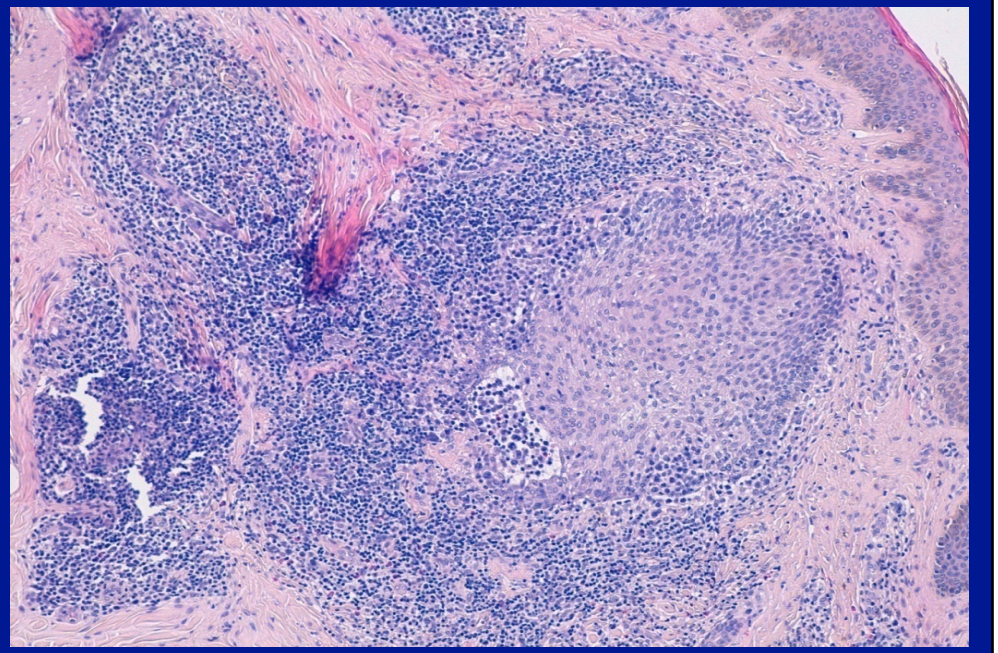

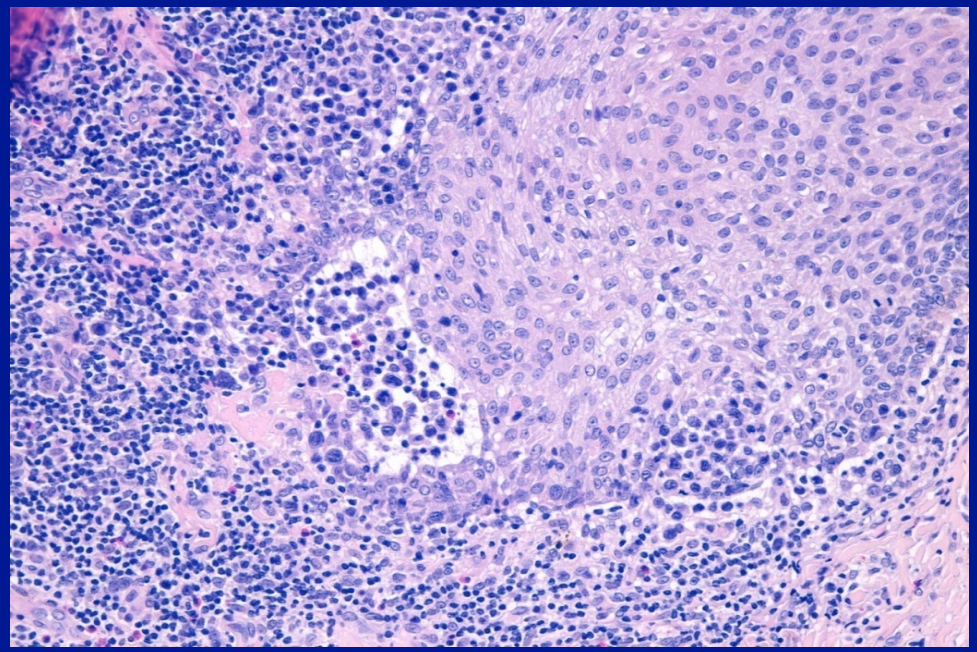

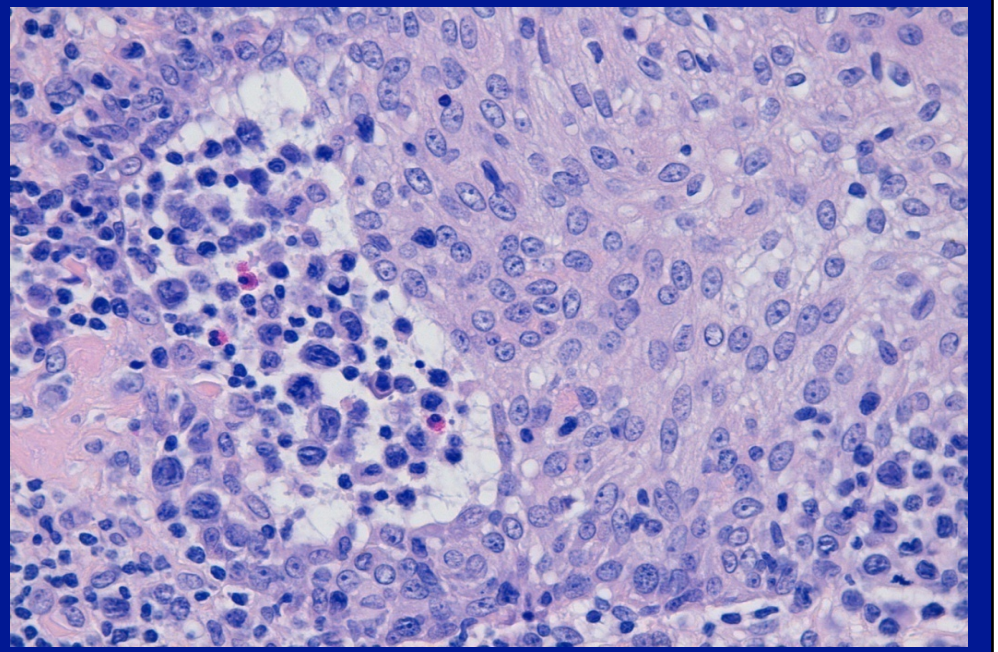

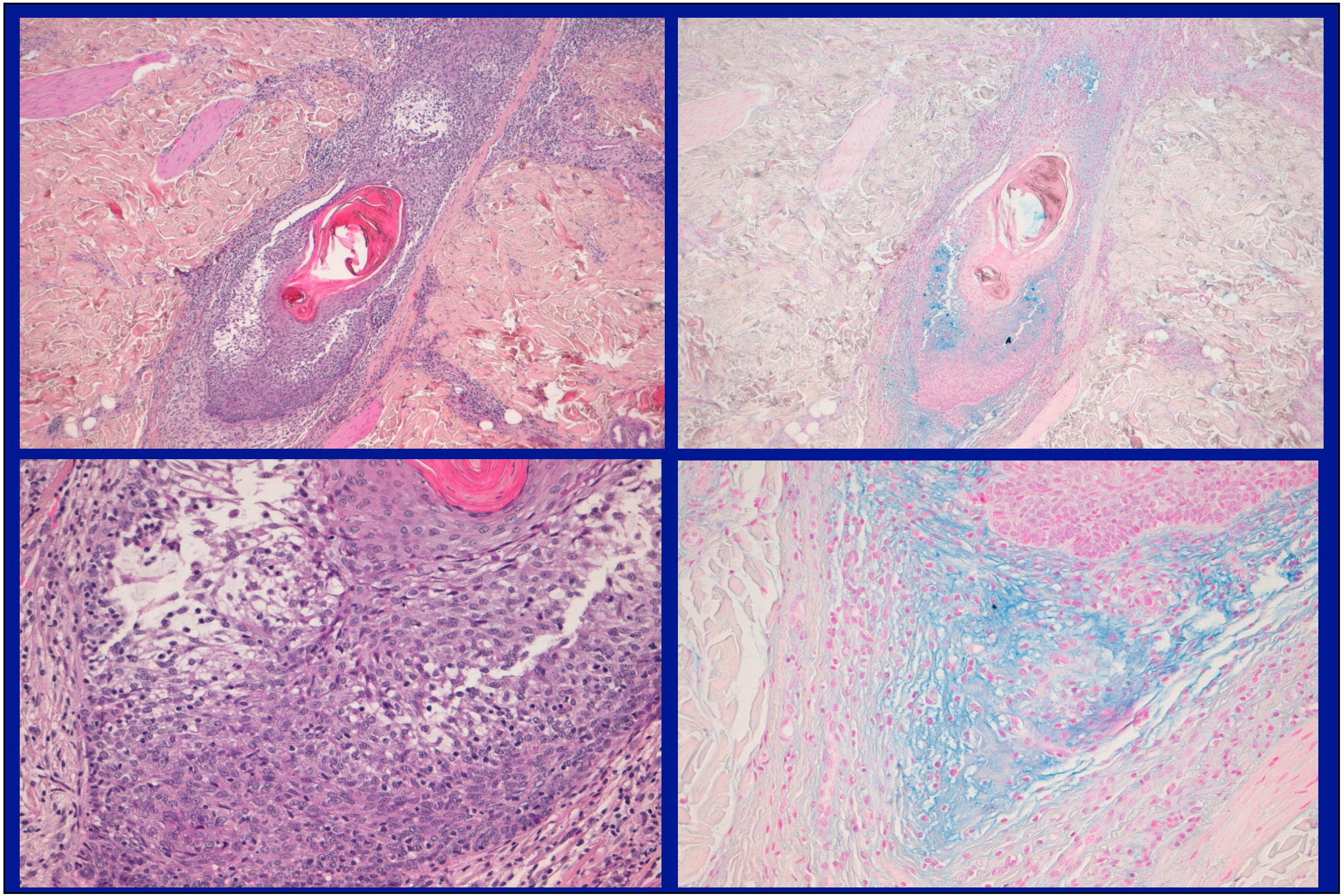

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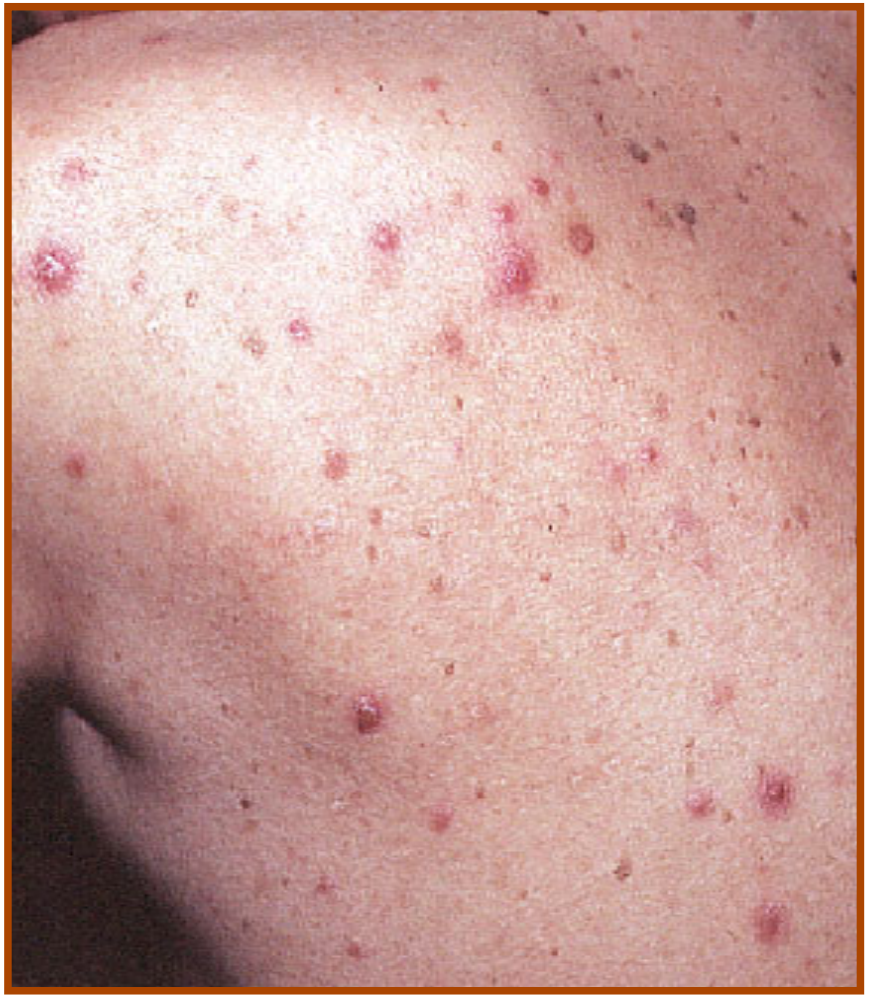
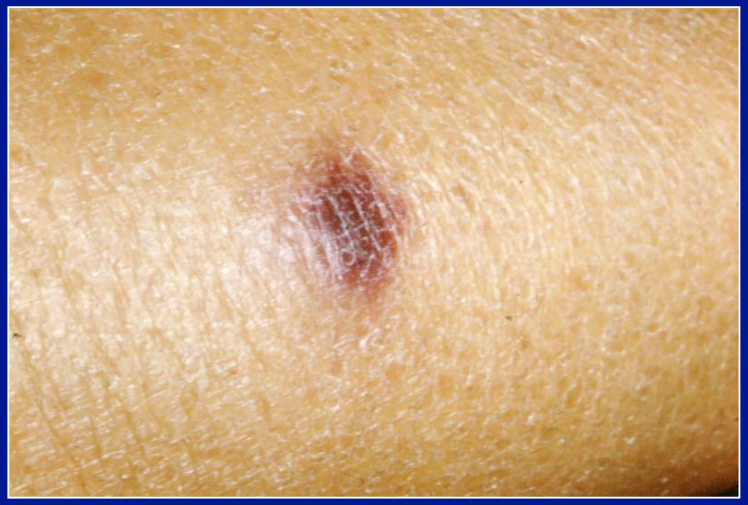
SPECTRUM OF CD30+ LYMPHOPROLIFERATIVE DISORDERS

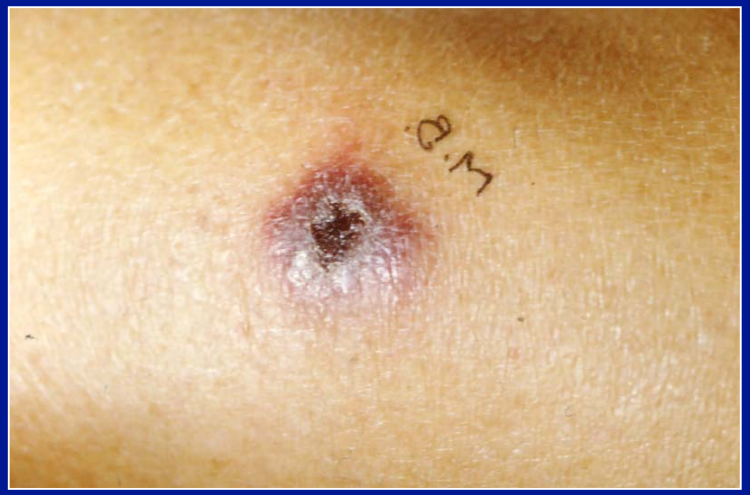
- **CD30 antigen expression** by neoplastic cells (variable numbers) AT PRESENTATION
- **Spontaneous regression of skin lesions** (partial to complete)
- **Favourable prognosis** (>90% 5-year survival)

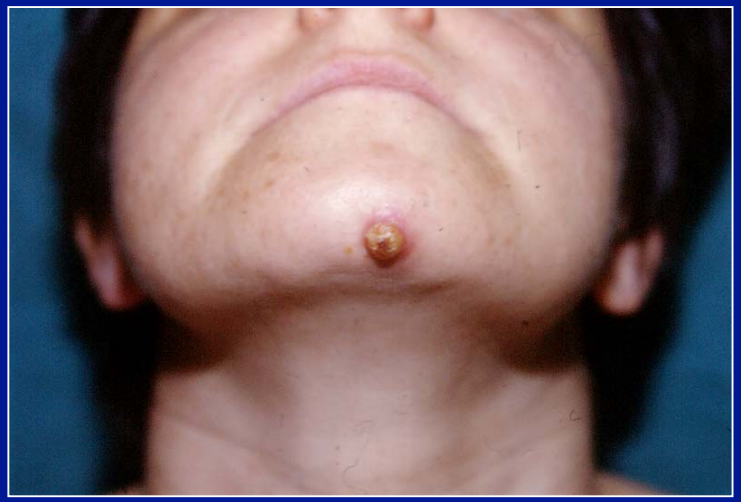
Lymphomatoid Papulosis
(Clinical Features)

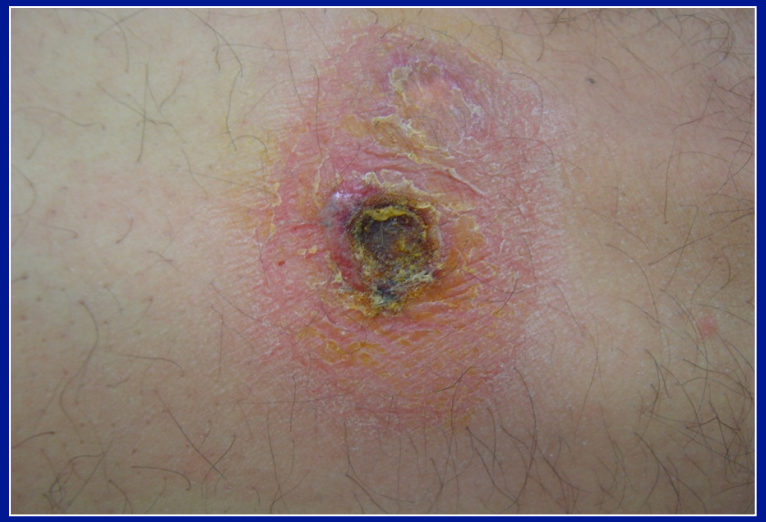
- **Age: 30-50 yrs. (rare in children)**
- **M:F=1.5:1**
- **Papular, papulo-necrotic, and/or nodular skin lesions at different stages of development**
- **Spontaneous regression of individual lesions within 3-6 weeks**
- **Trunk and limbs**



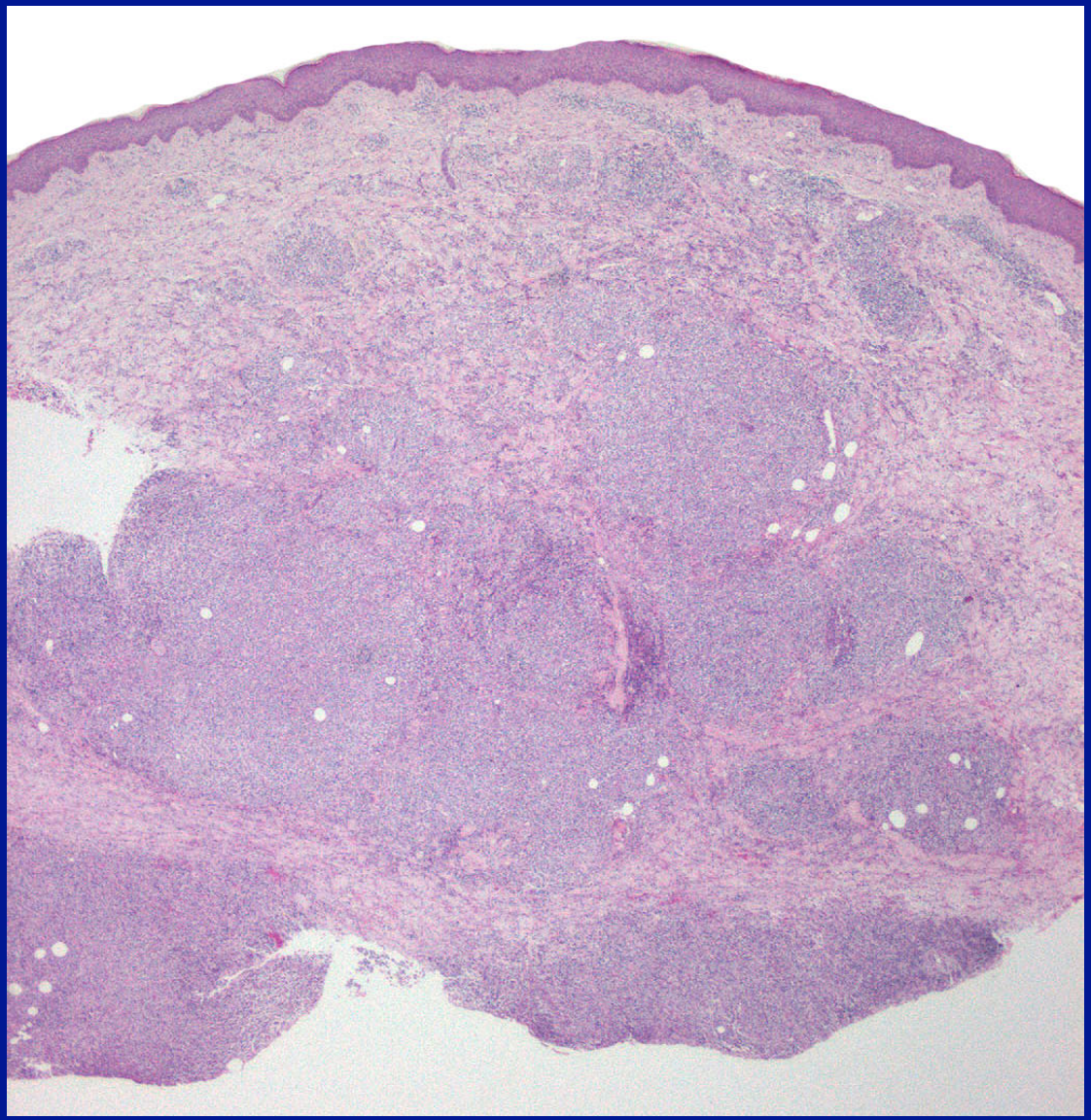


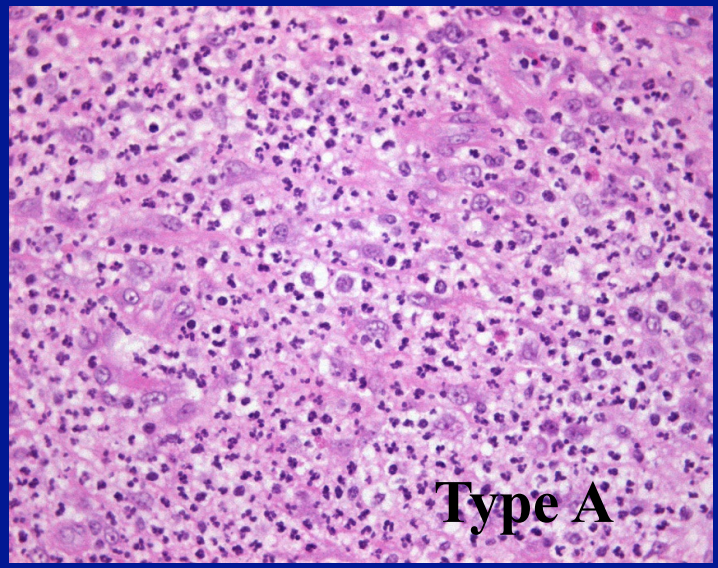


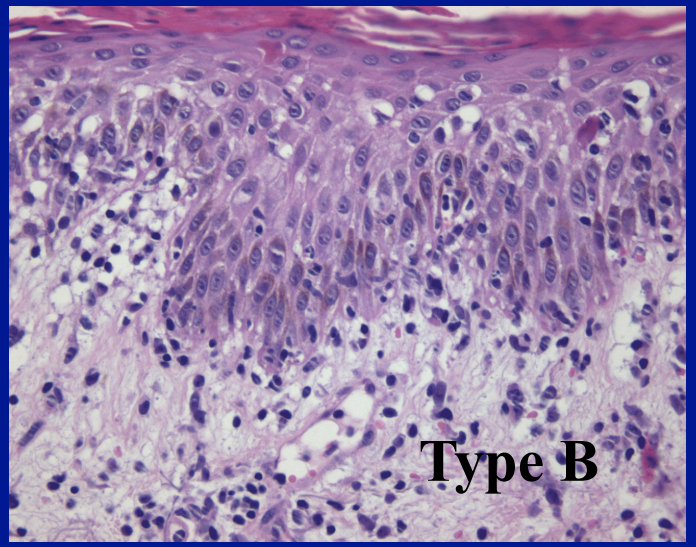




Lymphomatoid Papulosis



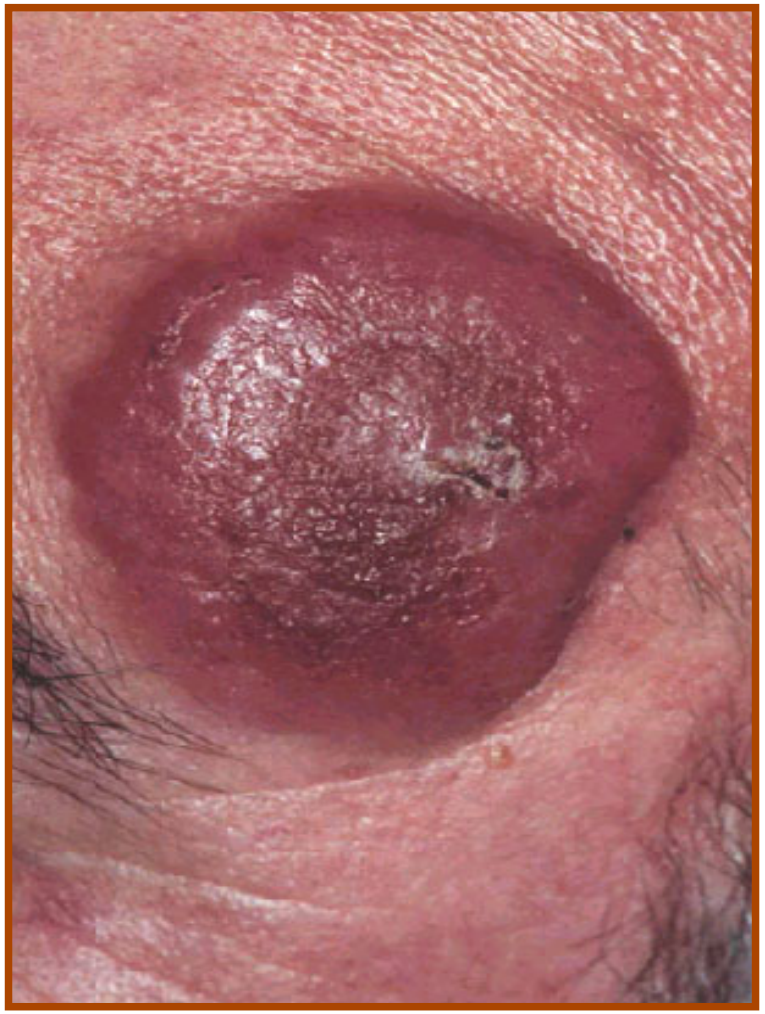


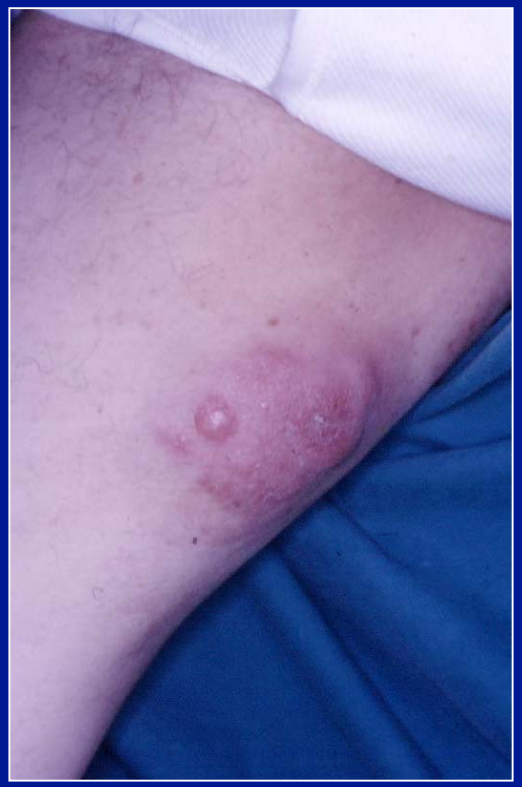


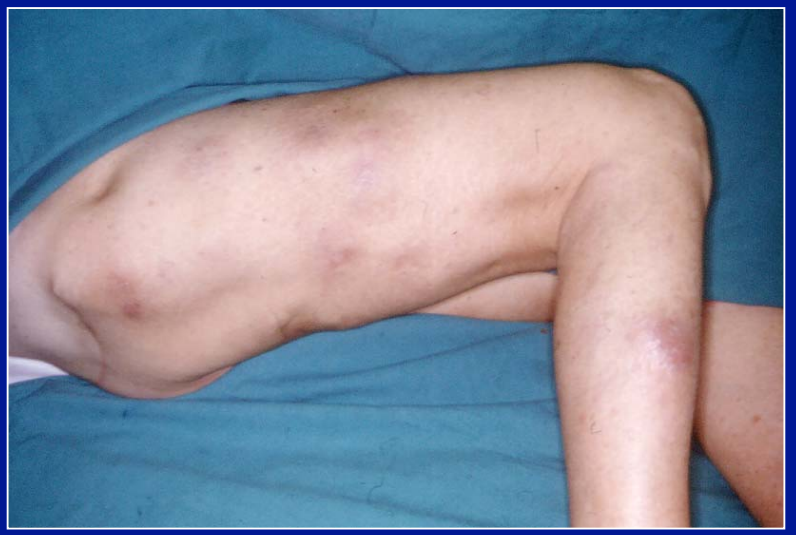
Type C (ALCL-like), Type D (epidermotropic aggressive CD8+ PTL-like), Type E (angiocentric) …

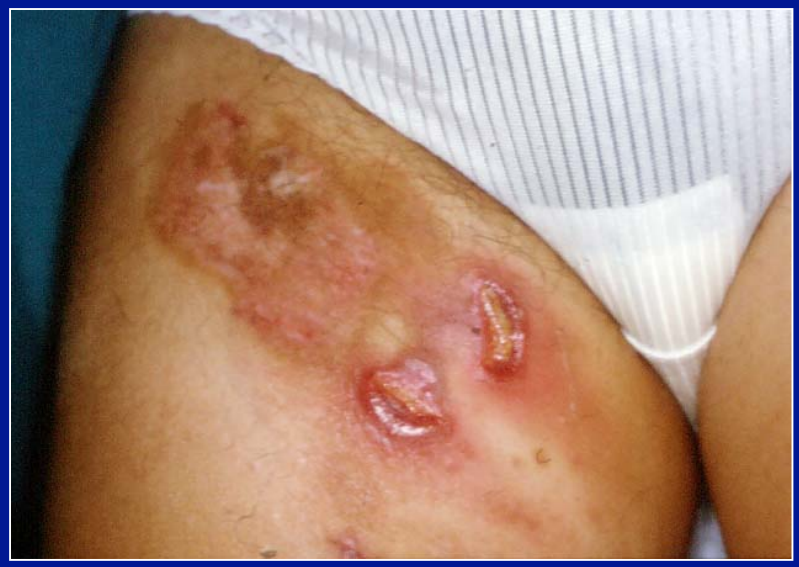
Primary Cutaneous ALCL (CD30+)

- Age: 16-89 yrs (mean 60 yrs); M:F=2:1
- Solitary or localized (ulcerating) nodules or tumors
- Complete or partial regression in up 25% of patients
- Regional lymph node involvement in up 20% of patients
- Rare involvement of internal organs

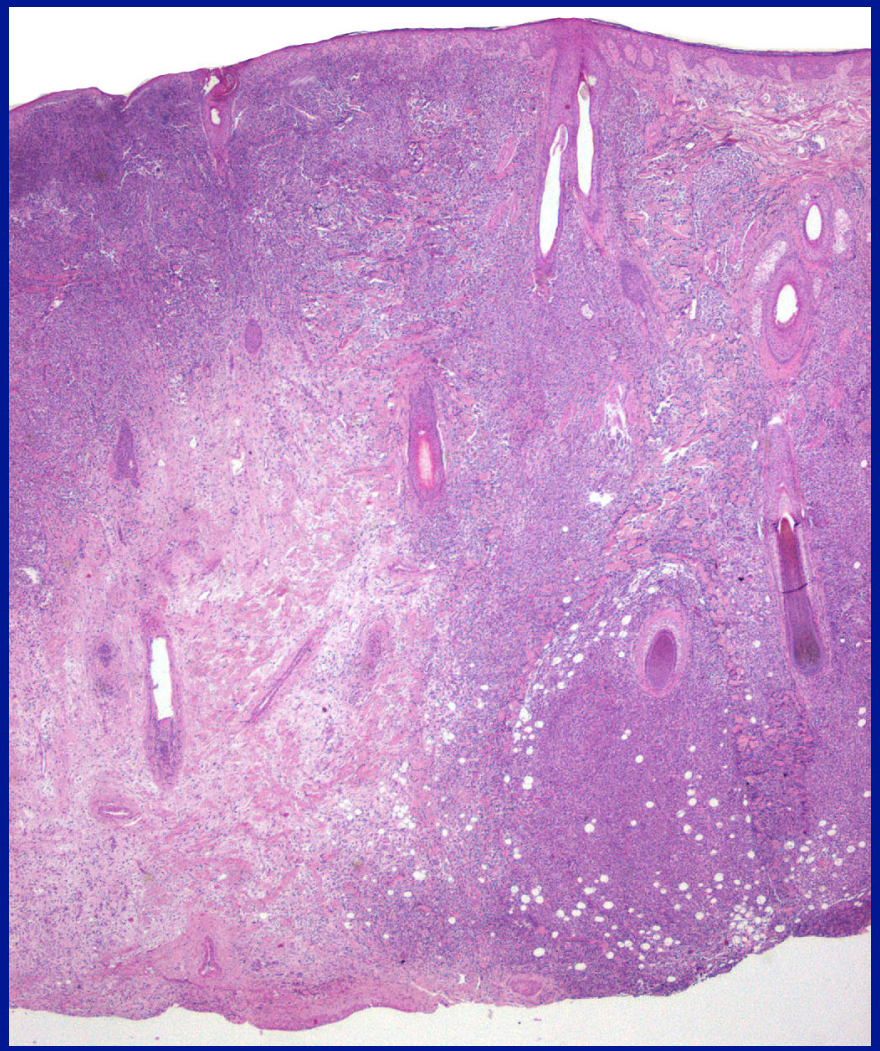


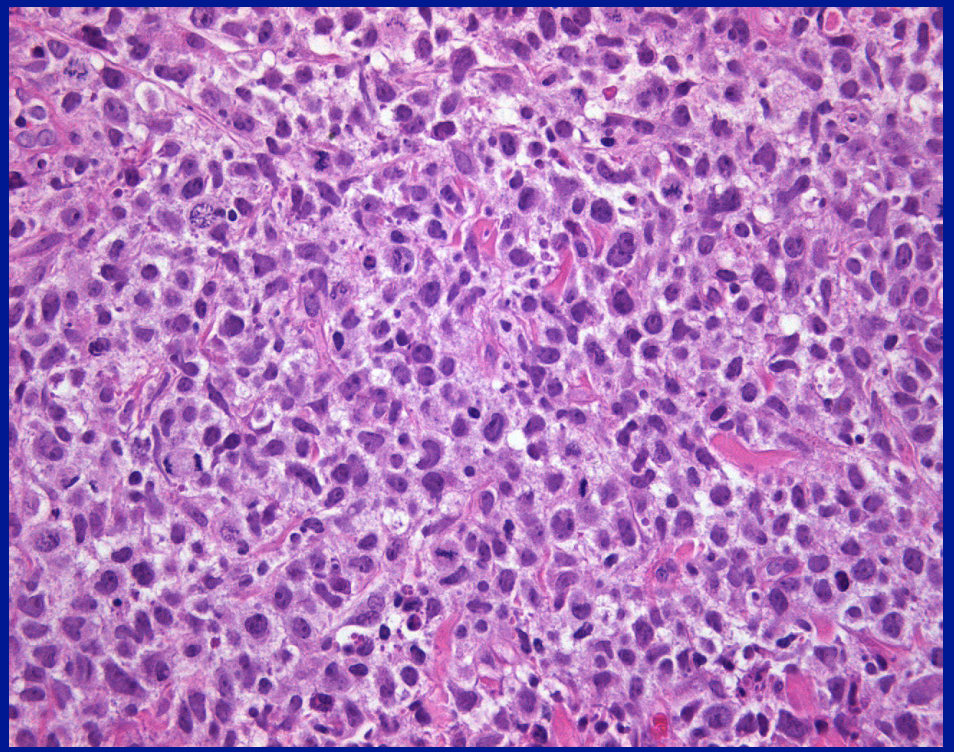






CD30+ anaplastic large cell CTCL





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Folliculotropic MF	86	4	80
Pagetoid reticulosis	14	< 1	100
Granulomatous slack skin	4	< 1	100
Primary cutaneous anaplastic large cell lymphoma	146	8	95
Lymphomatoid papulosis	236	12	100
Subcutaneous panniculitis-like T-cell lymphoma	18	1	82
Primary cutaneous CD4+ small/medium pleomorphic T-cell lymphoma†	39	2	75
Aggressive clinical behavior			
Sézary syndrome	52	3	24
Primary cutaneous NK/T-cell lymphoma, nasal-type	7	< 1	NR
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Primary cutaneous peripheral T-cell lymphoma, unspecified‡	47	2	16
Cutaneous B-cell lymphoma			
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Primary cutaneous marginal zone B-cell lymphoma	127	7	99
Primary cutaneous follicle center lymphoma	207	11	95
Intermediate clinical behavior			
Primary cutaneous diffuse large B-cell lymphoma, leg type	85	4	55
Primary cutaneous diffuse large B-cell lymphoma, other	4	< 1	50
Primary cutaneous intravascular large B-cell lymphoma	6	< 1	65

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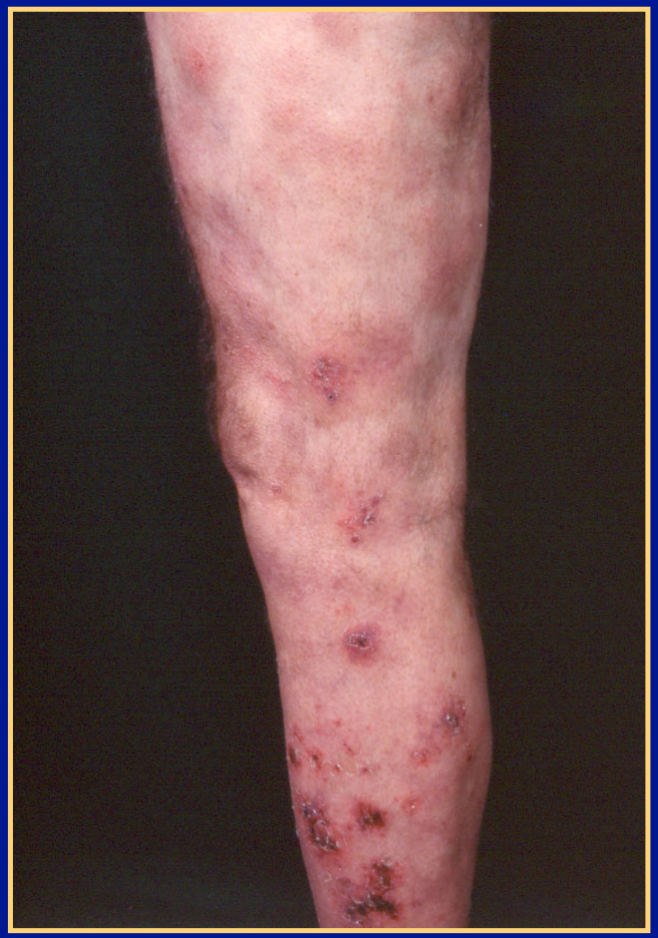
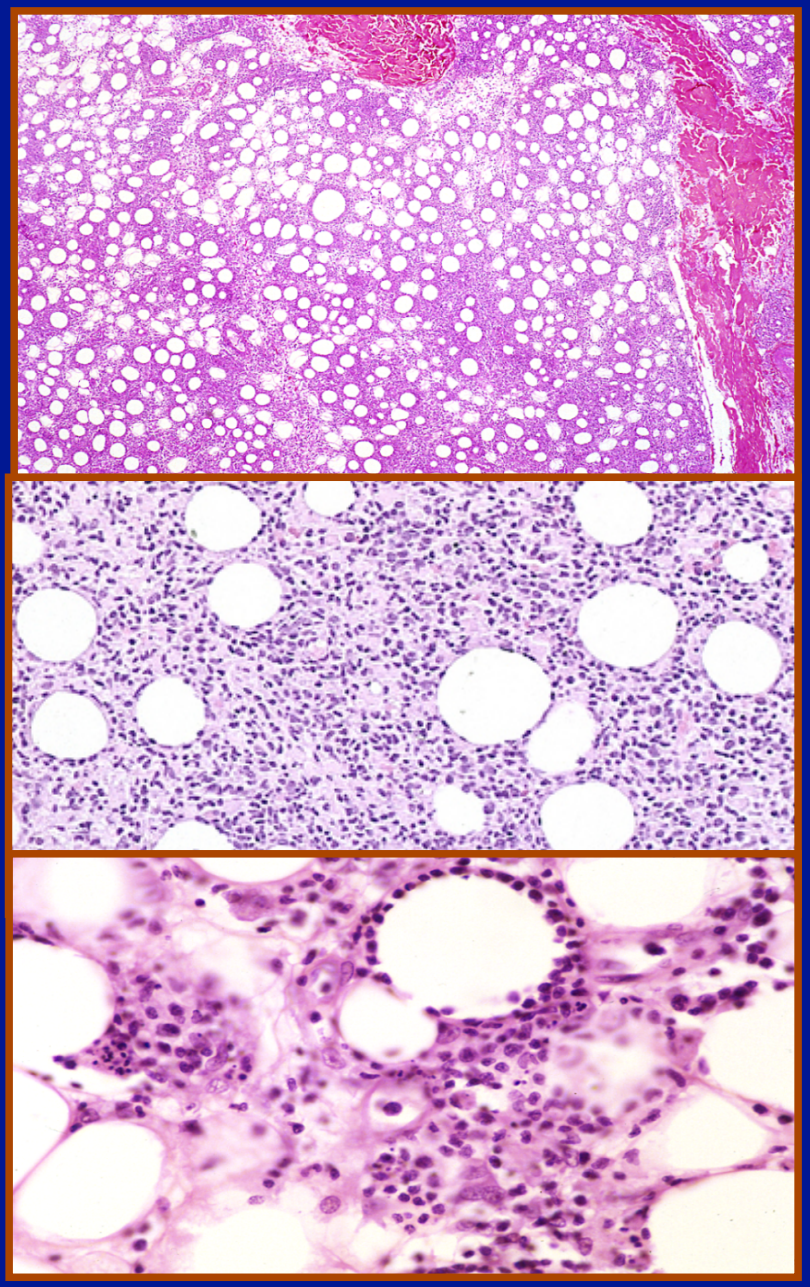
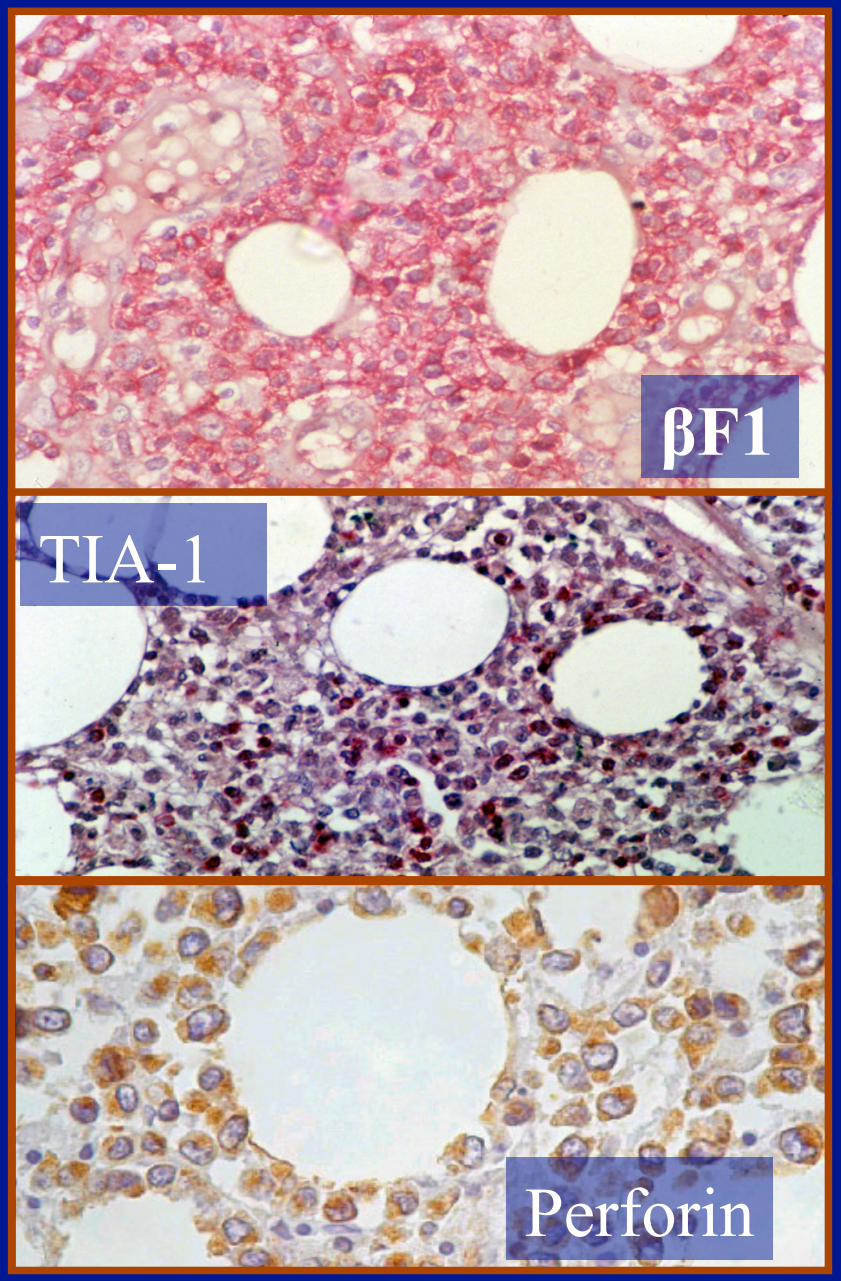
SUBCUTANEOUS PANNICULITIS-LIKE T-CELL LYMPHOMA

Clinical presentation and course: subcutaneous plaques and/or nodules mainly involving the lower extremities, rarely undergoing ulceration, seldom accompanied by fever and associated with hemophagocytic sy.

Mostly indolent course; if not, consider transplantation procedures.






βF1
TIA-1
Perforin

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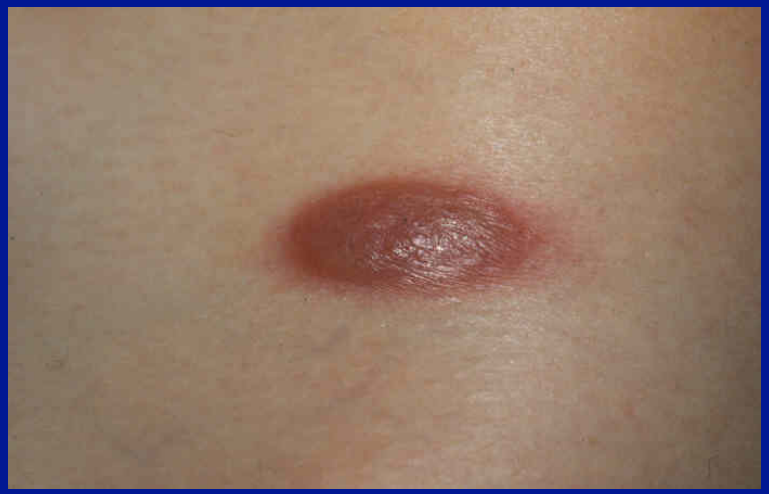
CD4+ pleomorphic small/medium –sized

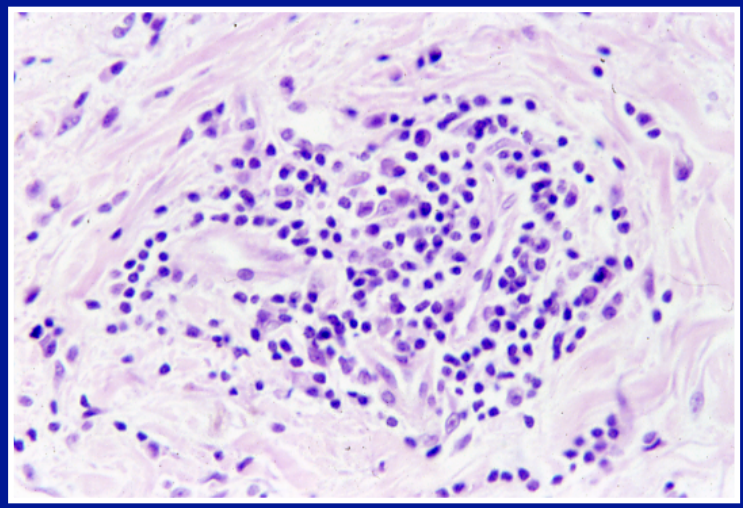
Clinical presentation : plaques or nodules more often isolated

Histology: small/medium sized, pelomorphic, CD4+ T-cells

Course and treatment: indolent course, RT or surgery

D.D. pseudoCTCL





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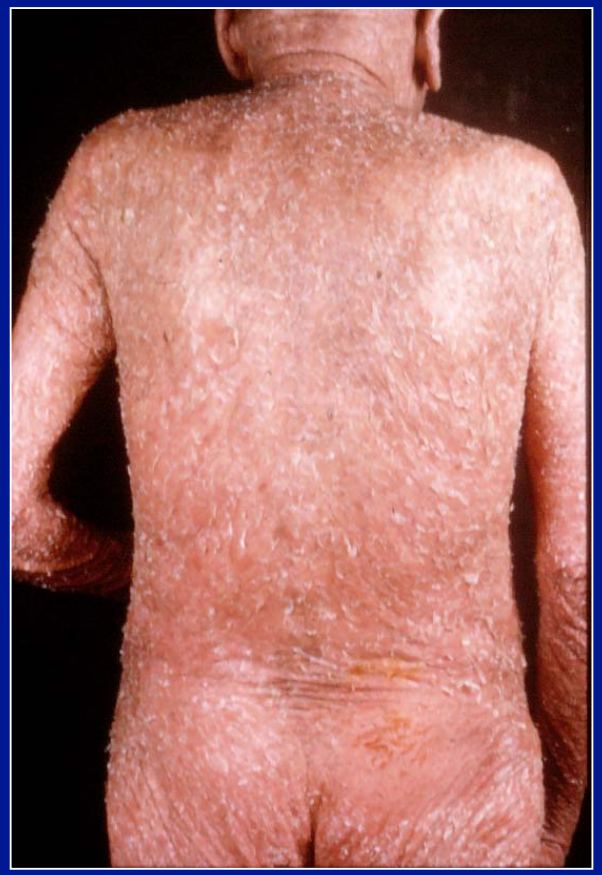

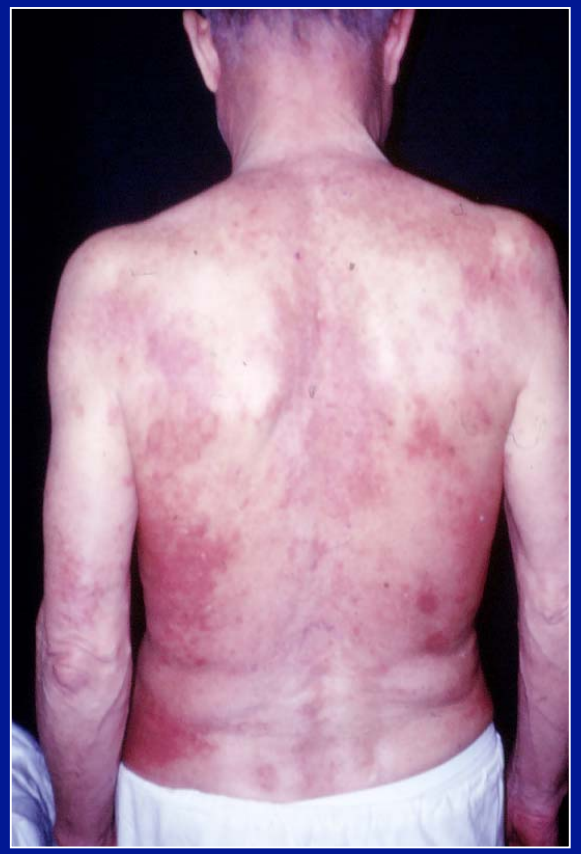

Sézary Syndrome

SEZARY SYNDROME (stage IVA) vs. MF (stage I-III)
ISCL criteria

Hematologic criteria:

- **Sézary cells count >1000/mm^3**
- **CD4/CD8 ratio > 10**
- **deficit pan-T cell markers (CD2,CD3,CD4,CD5)**
- **CD4+CD7- and/or CD4+/CD26- cells >33%**
- **Sézary cells >20% total lymph. count + clonality (SB, PCR)**
- **T-cell clone with chromosomal aberrancies**

Vonderheid EC et al. (ISCL) 2002; Olsen E et al. (ISCL) 2007

TCR Vbeta restriction ?

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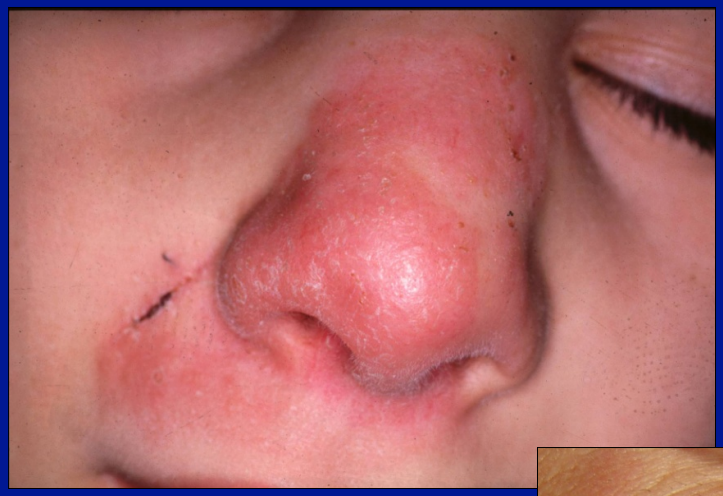
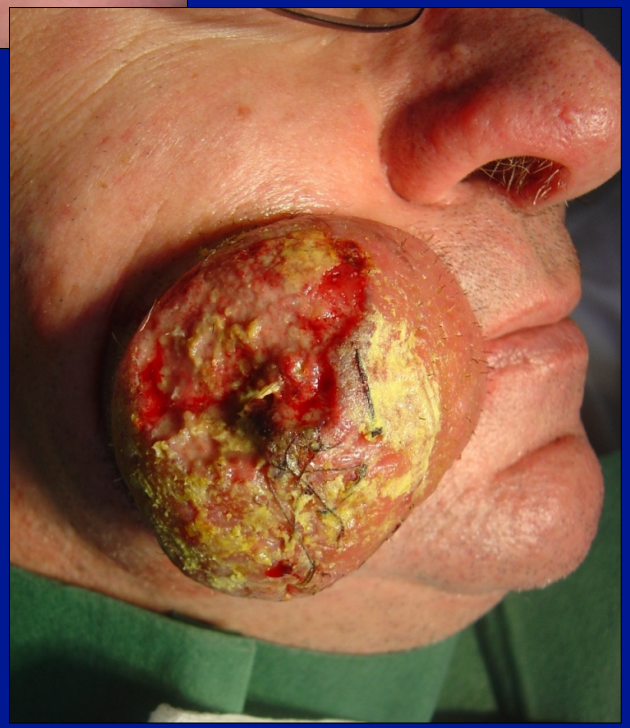
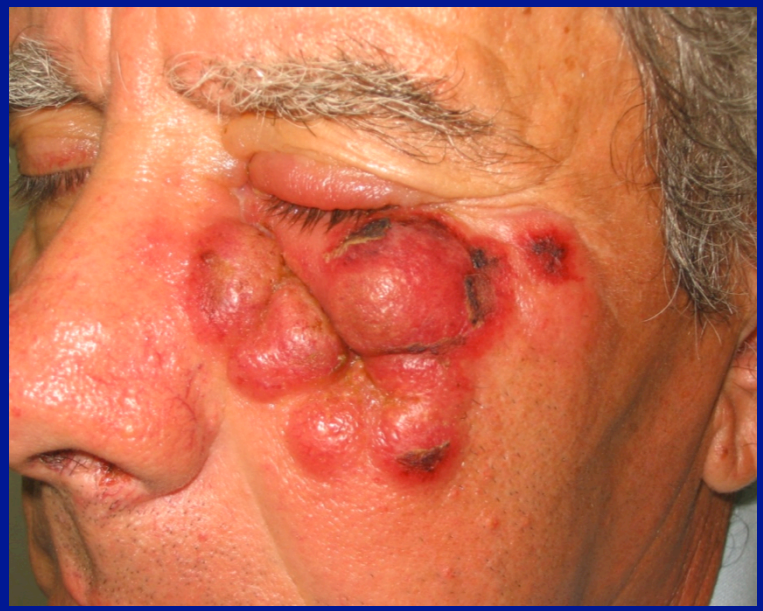
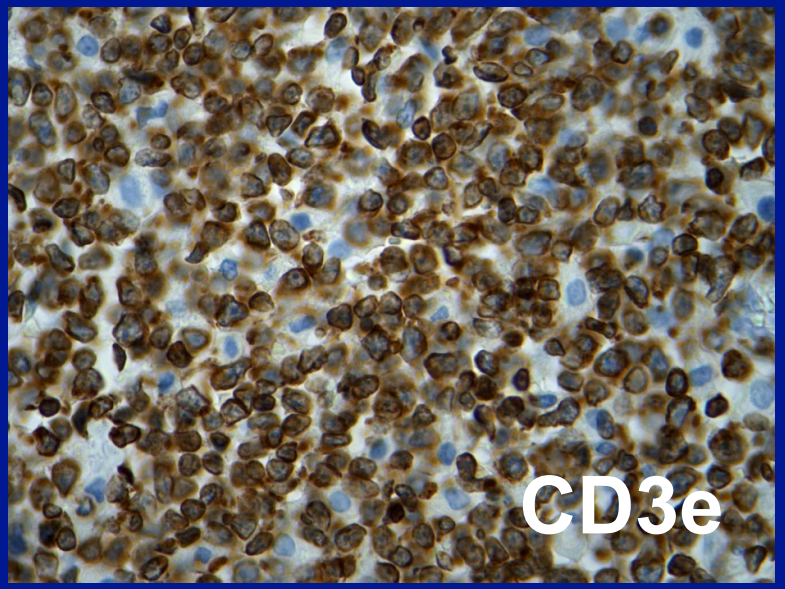
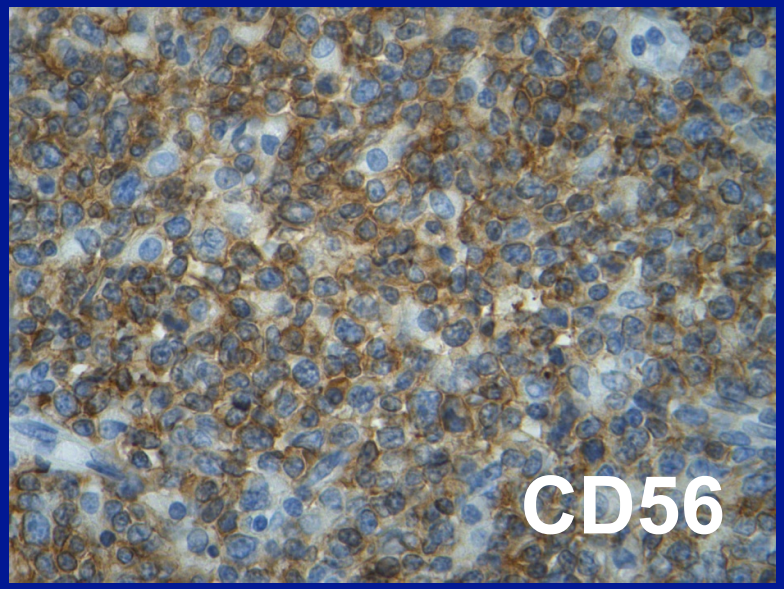
Extranodal NK/T-cell lymphoma, nasal type

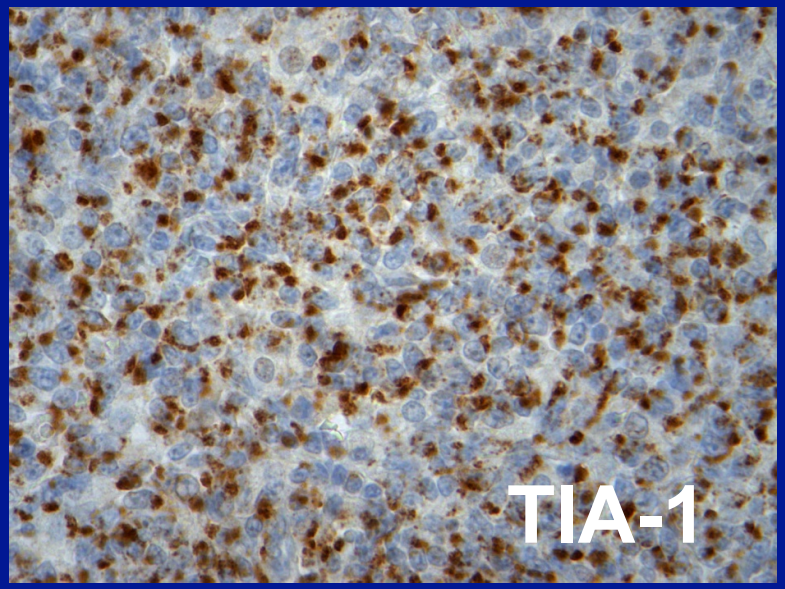







CD3e


CD56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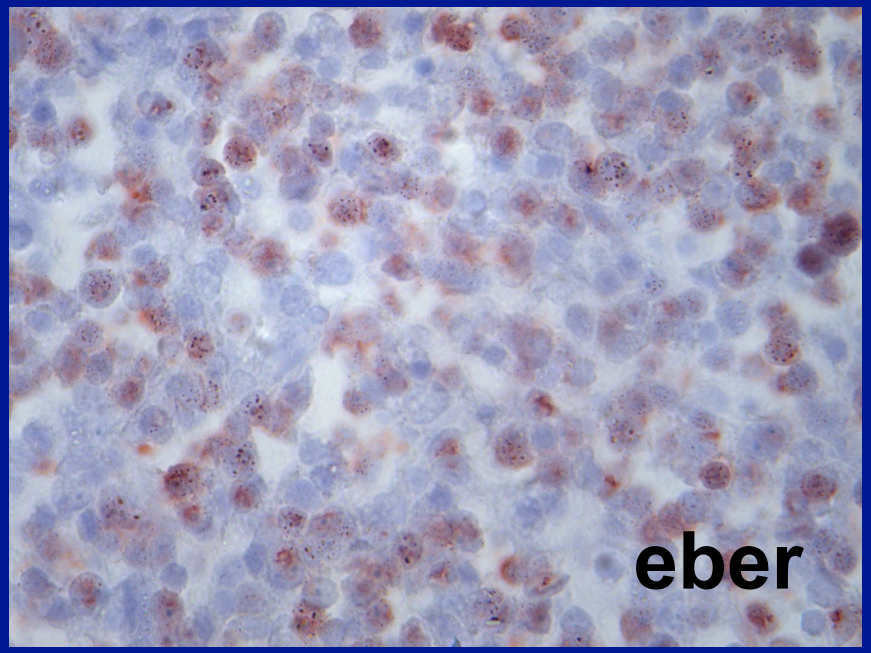
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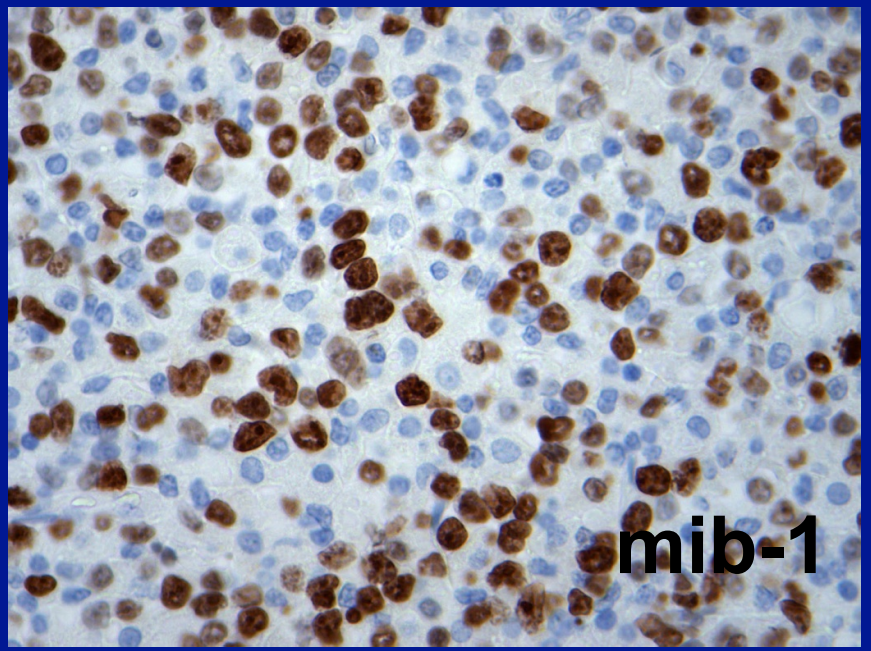
mib-1

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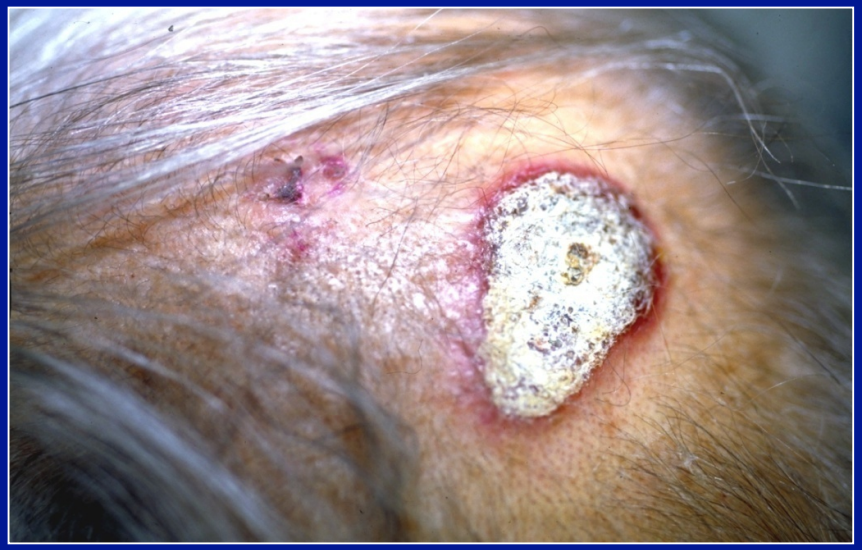
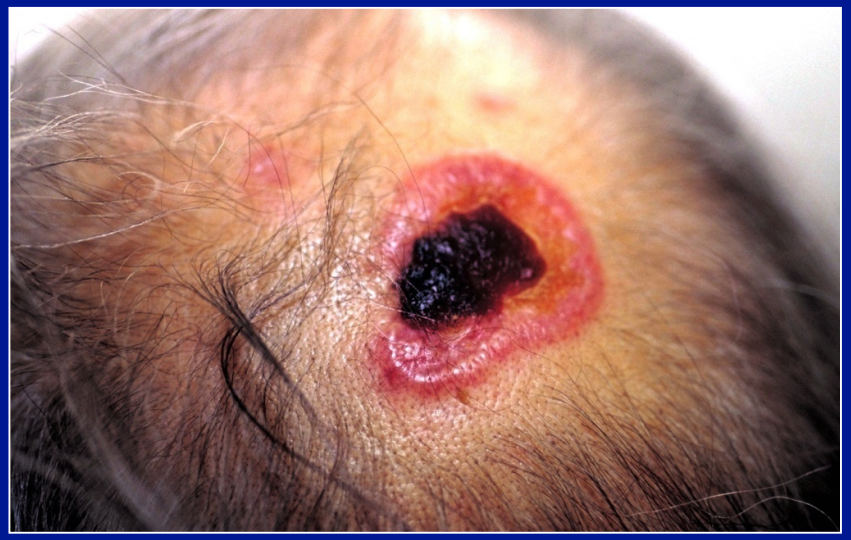
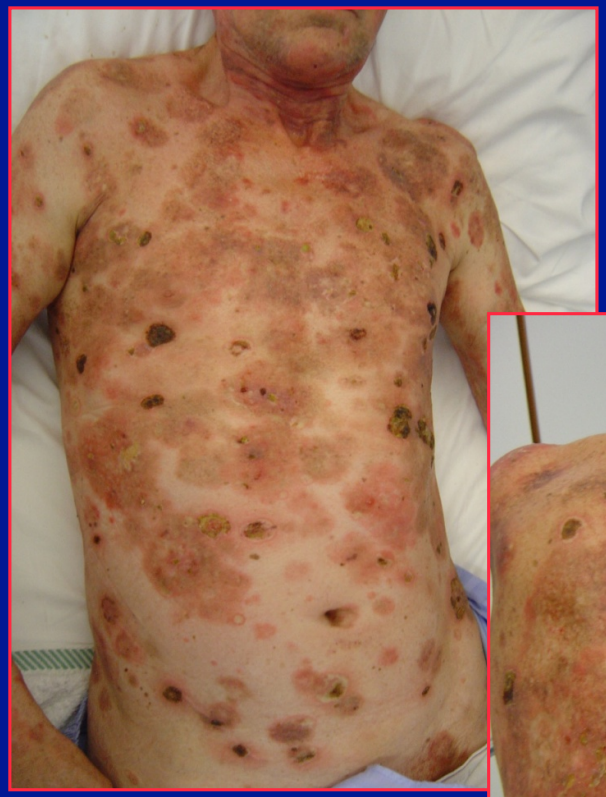
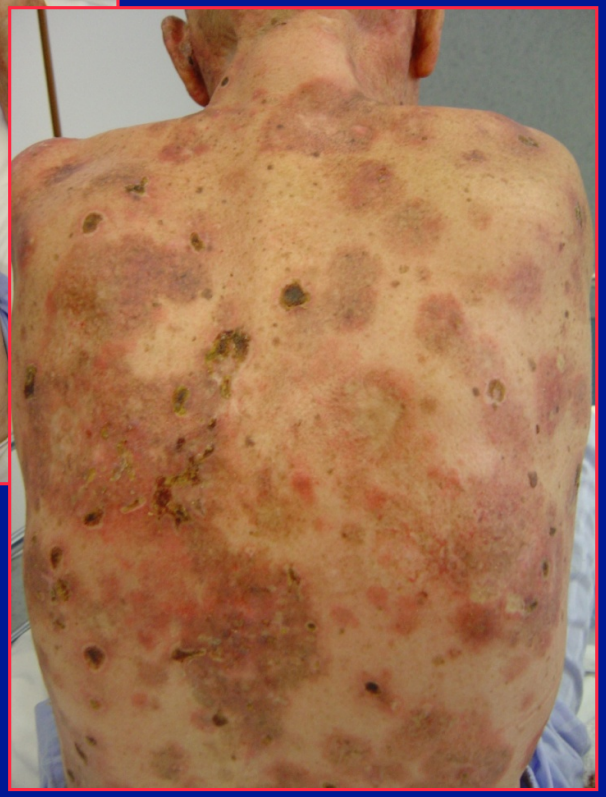


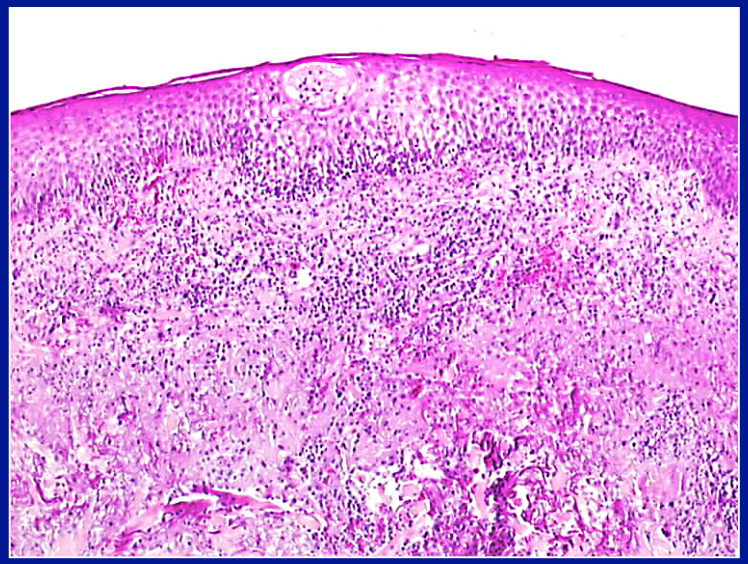


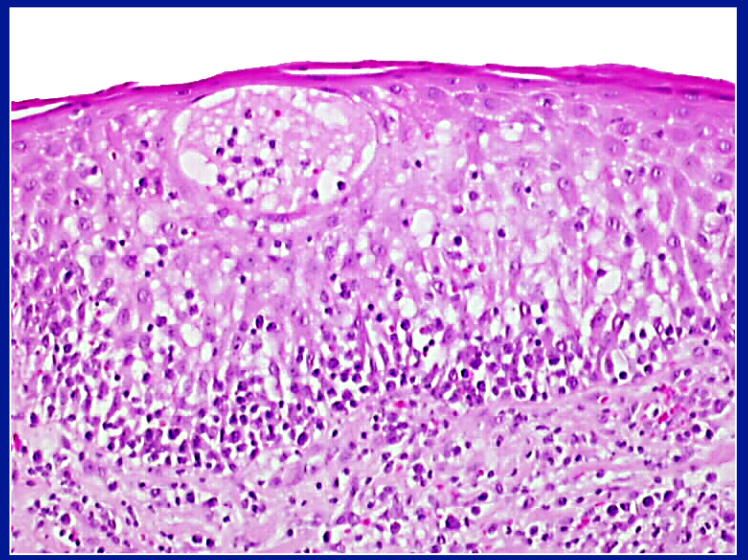


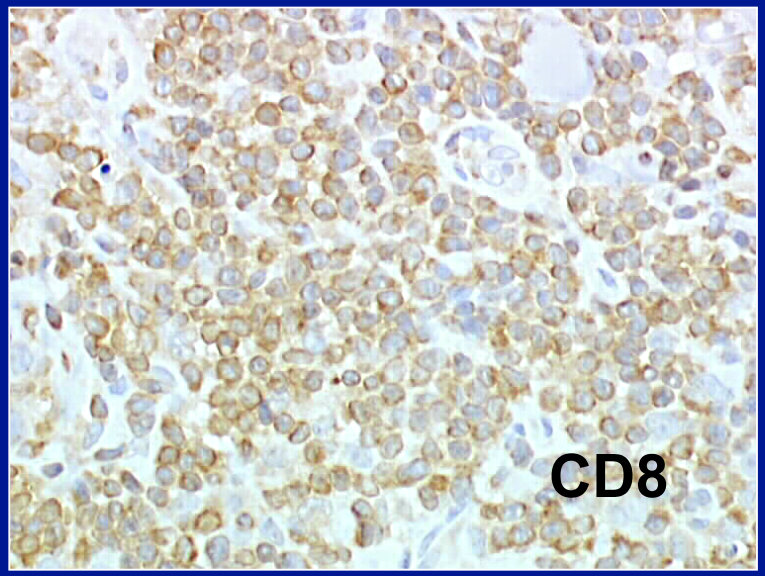


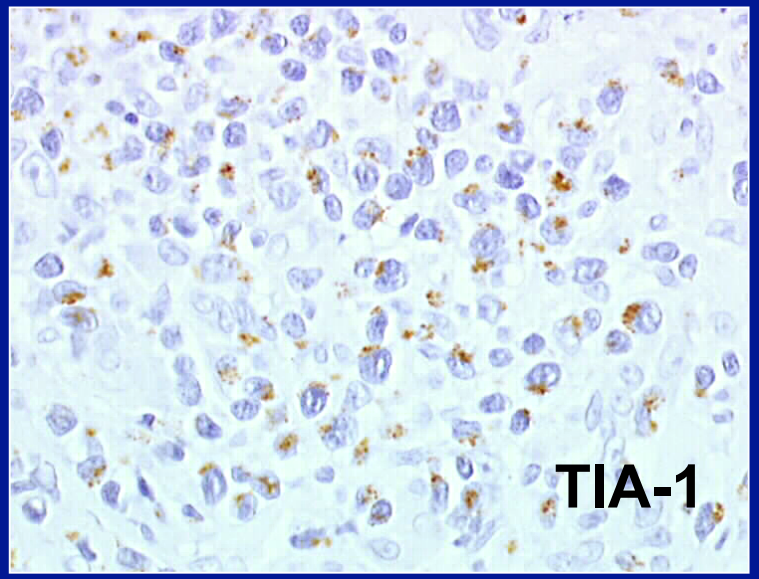
PTL-U (CD8+ epidermotropic aggressive CTCL)

EPIDERMOTROPIC CD8+ CYTOTOXIC CTCL









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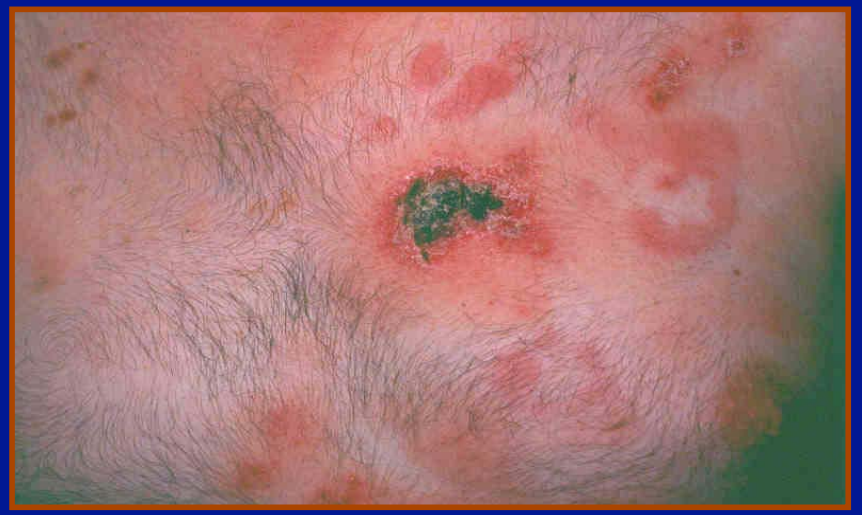
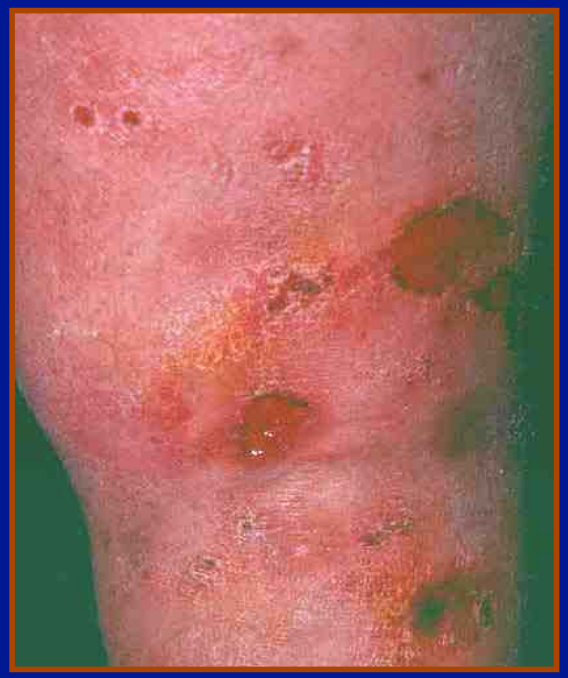
Cutaneous γ/δ T-cell Lymphoma

- Disseminated plaques and/or ulceronecrotic nodules or tumors, particularly on the extremities
- Involvement of mucosal and other extranodal sites frequent; involvement of lymph nodes, spleen or bone marrow uncommon
- Median survival: ≈ 15 months





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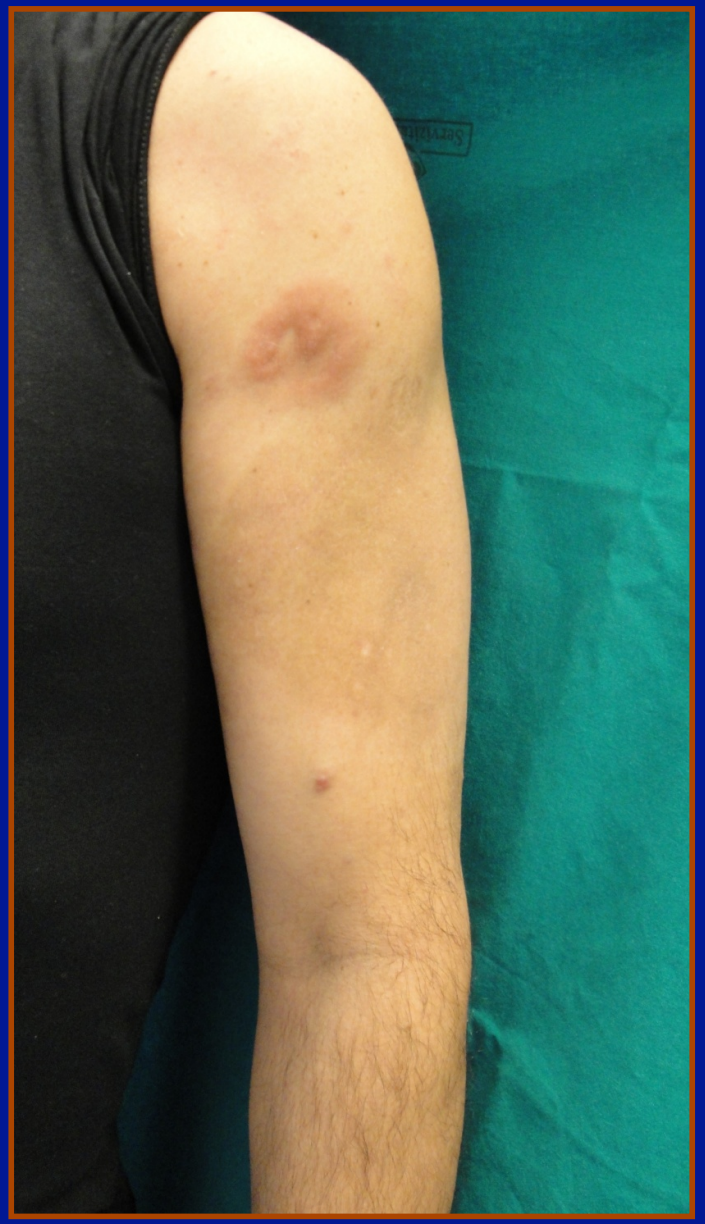
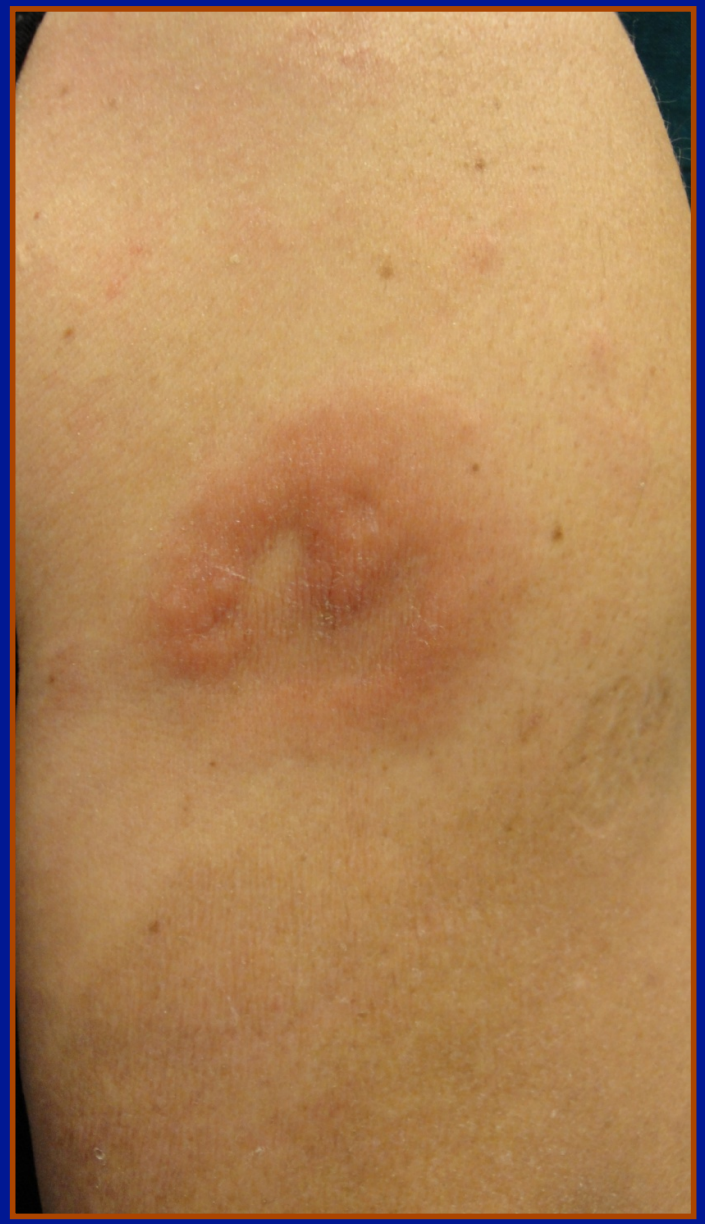
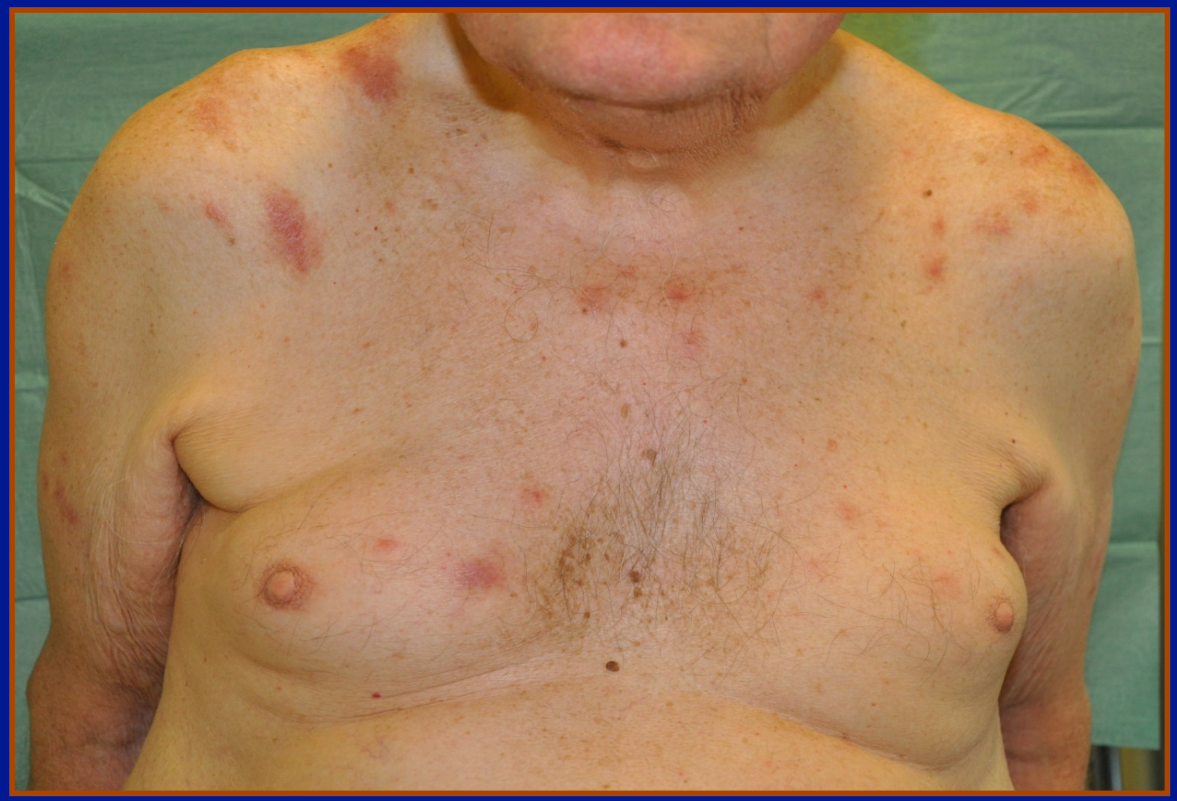
Marginal Zone B-cell Lymphoma

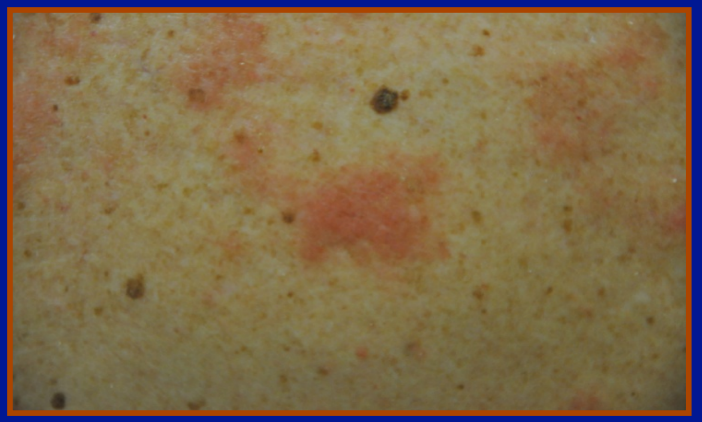
Epidemiology, Clinical Features, and Prognosis

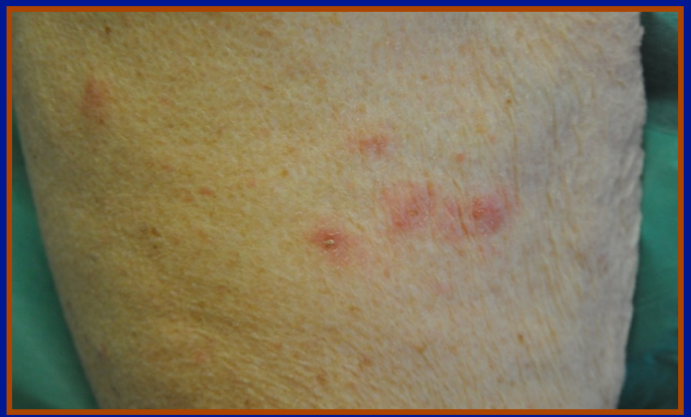
- Most commonly affects adults aged >40 years, with a slight male gender prevalence
- Predominantly localized on the extremities and trunk, less often on the head & neck; multifocal skin lesions frequently observed (≈ 70%)
- MZL presents with erythematous to cianotic papules, plaques or nodules; subcutaneous involvement uncommon
- Ulceration and extracutaneous dissemination uncommon
- Very good prognosis (99% 5-year survival)

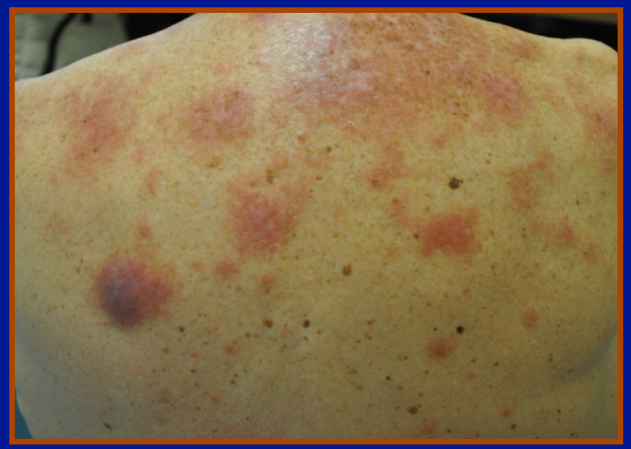


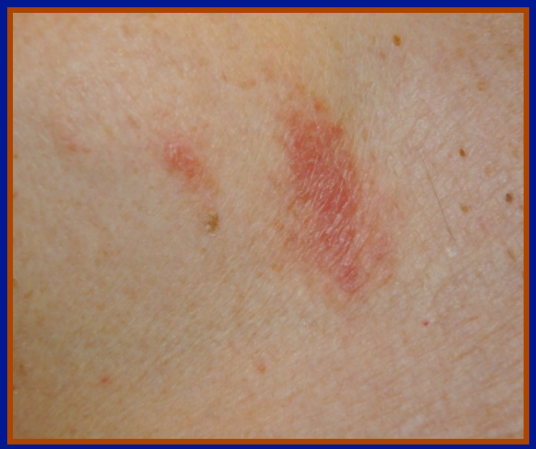


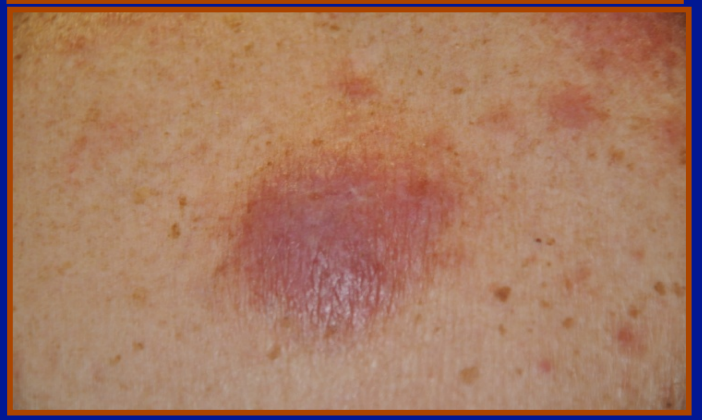




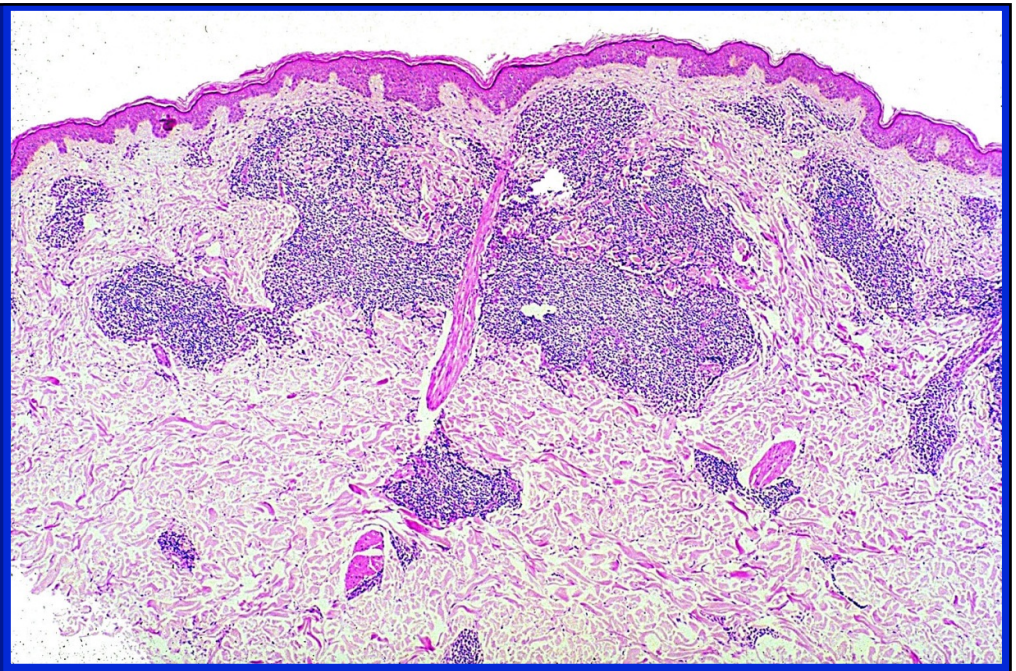


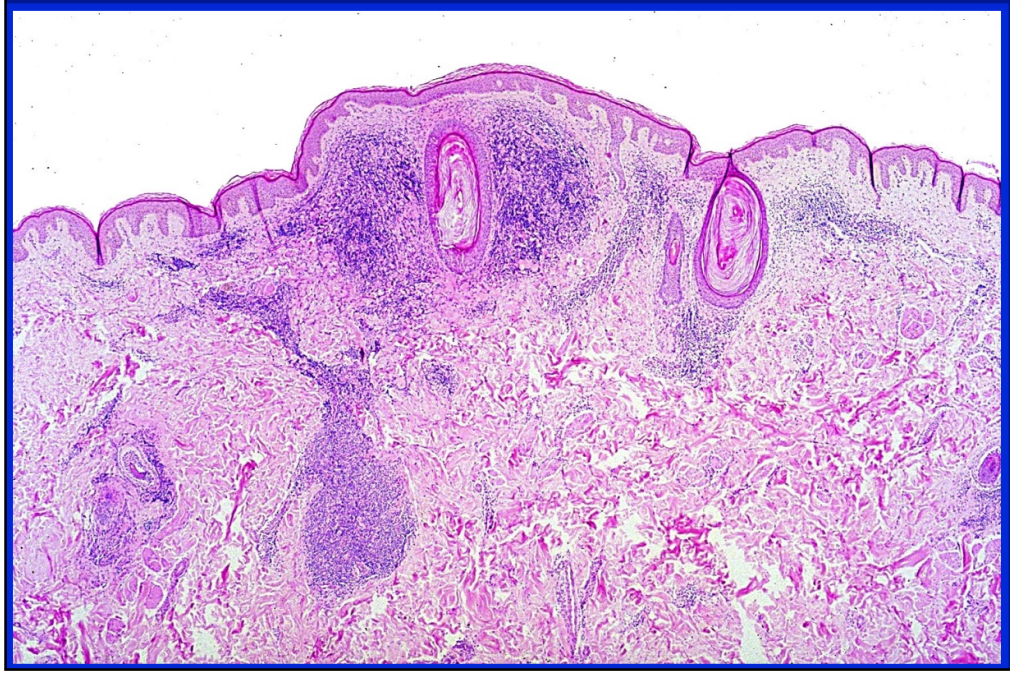


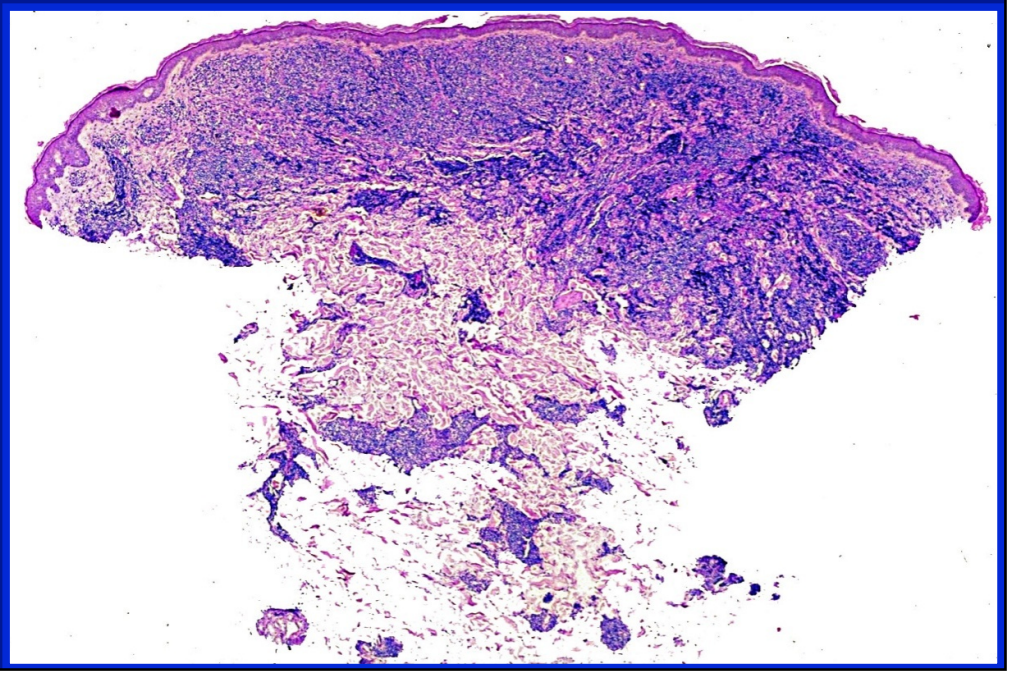




- ✓ ***MZL*** **shows nodular or diffuse infiltrates, with the almost constant sparing of the epidermis**
- ✓ **The histologic picture is variable, and primarily relates to the age and growth rate of skin lesions**

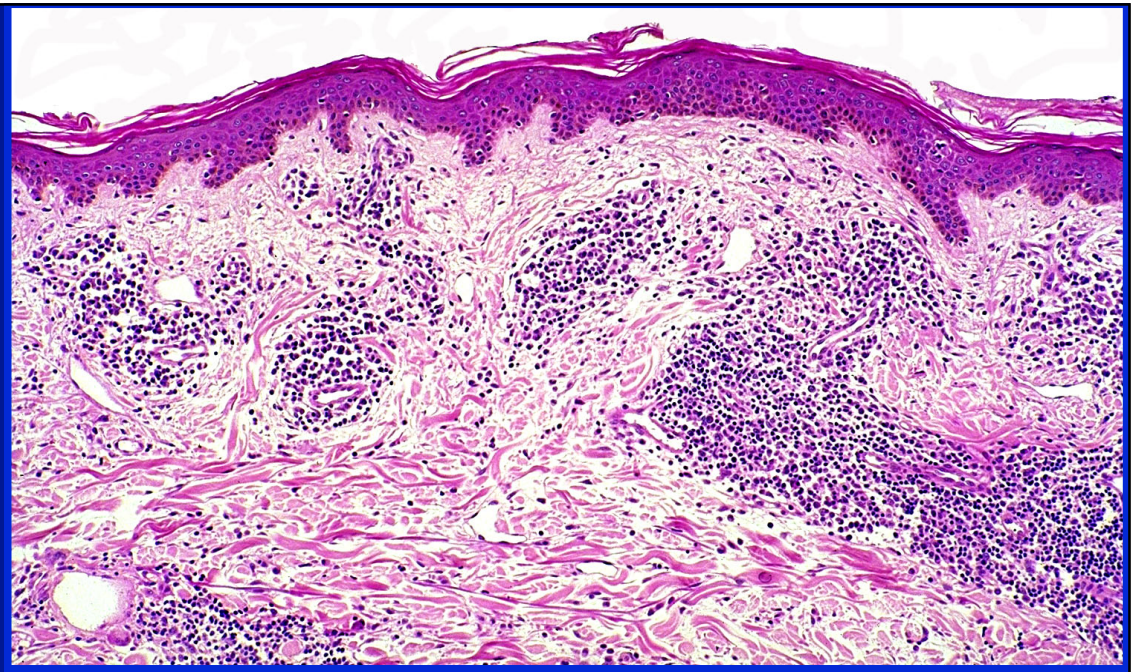


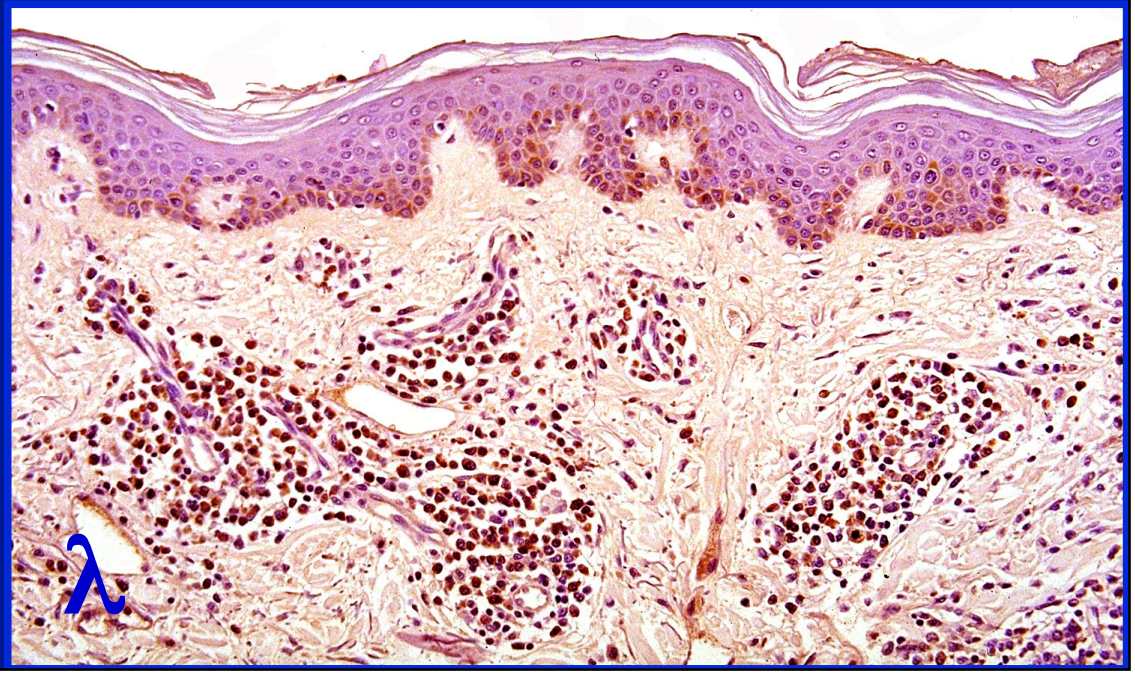


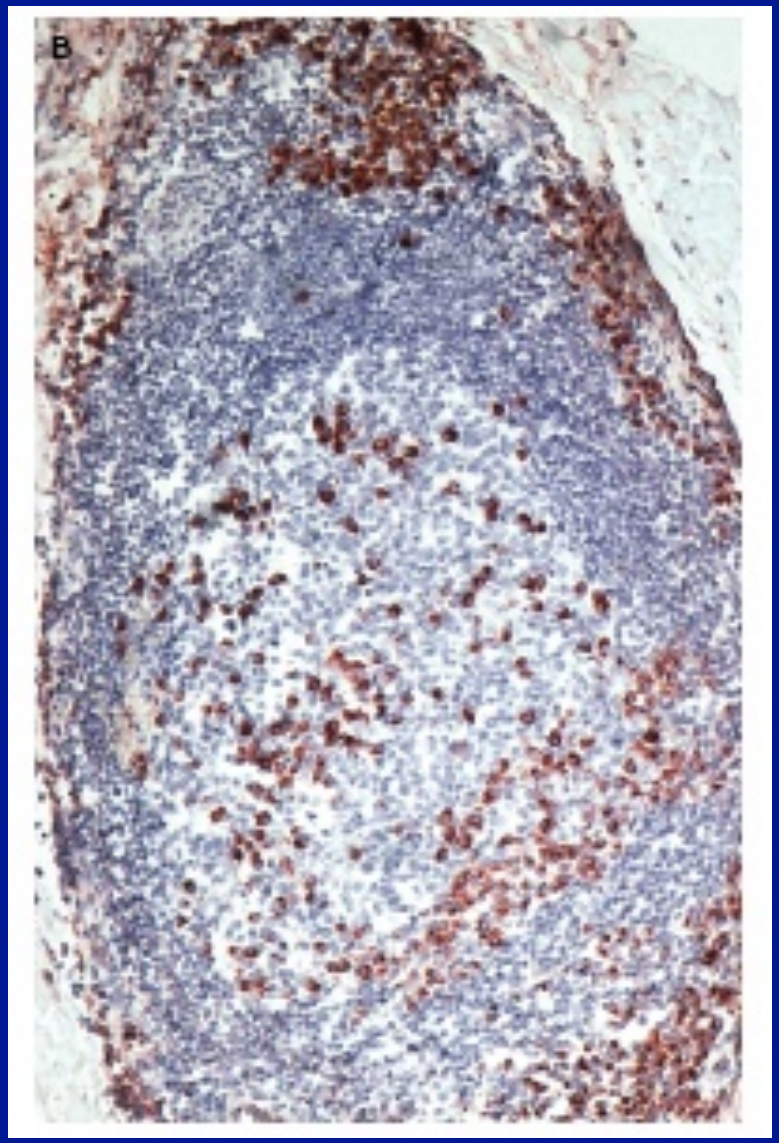


Marginal Zone B-cell Lymphoma

- ✓ **Infiltrates are composed of small lymphocytes, centrocyte-like cells, and plasma cells admixed with variable numbers of centroblast-, and immunoblast-like cells**
- ✓ **The plasma cells are located at the periphery of the infiltrates**
- ✓ **Tumor cells are monotypic cIg^{+}, CD79a^{+}, CD5^{-}, CD10^{-}**







“colonization” of lymphoid follicles

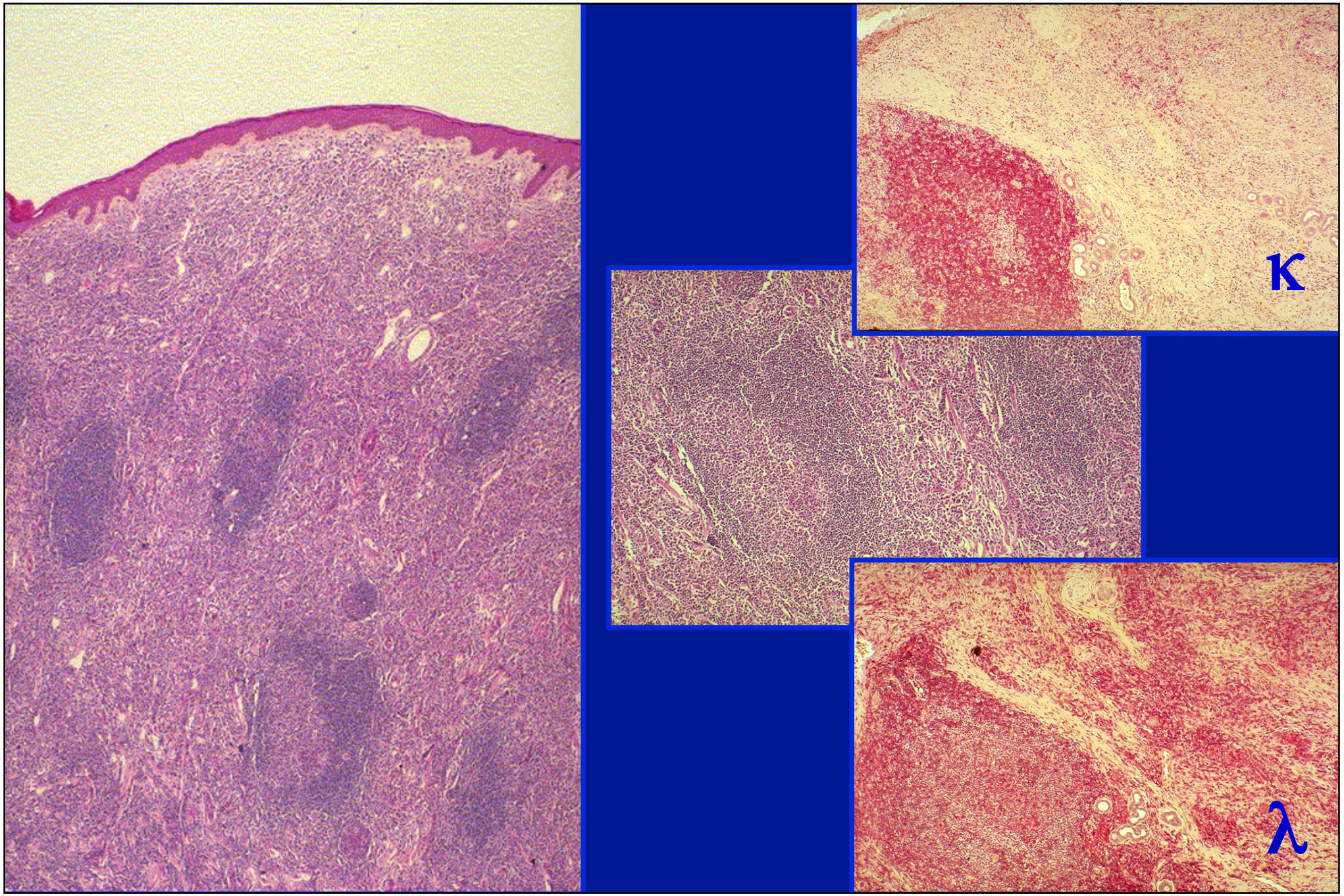

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Table 2. Relative frequency and disease-specific 5-year survival of 1905 primary cutaneous lymphomas classified according to the WHO-EORTC classification

WHO-EORTC classification	No.	Frequency, %*	Disease-specific 5-year survival, %
Cutaneous T-cell lymphoma			
Indolent clinical behavior			
Mycosis fungoides	800	44	88
Folliculotropic MF	86	4	80
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Primary cutaneous $CD4^+$ small/medium pleomorphic T-cell lymphoma†	39	2	75
Aggressive clinical behavior			
Sézary syndrome	52	3	24
Primary cutaneous NK/T-cell lymphoma, nasal-type	7	< 1	NR
Primary cutaneous aggressive CD8+ T-cell lymphoma†	14	< 1	18
Primary cutaneous γ/δ T-cell lymphoma†	13	< 1	NR
Primary cutaneous peripheral T-cell lymphoma, unspecified‡	47	2	16
Cutaneous B-cell lymphoma			
Indolent clinical behavior			
Primary cutaneous marginal zone B-cell lymphoma	127	7	99
Primary cutaneous follicle center lymphoma	207	11	95
Intermediate clinical behavior			
Primary cutaneous diffuse large B-cell lymphoma, leg type	85	4	55
Primary cutaneous diffuse large B-cell lymphoma, other	4	< 1	50
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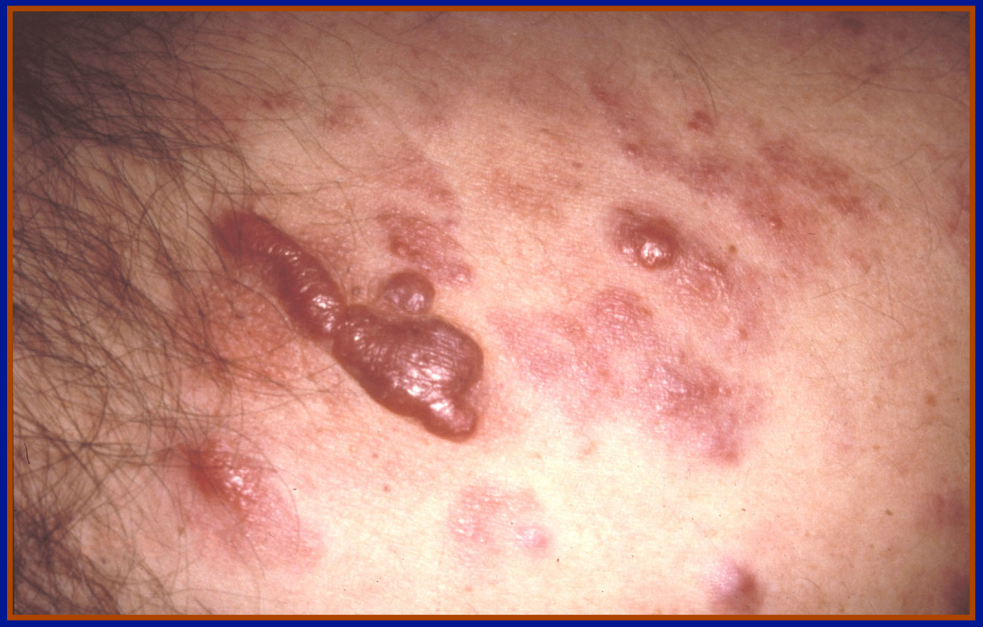
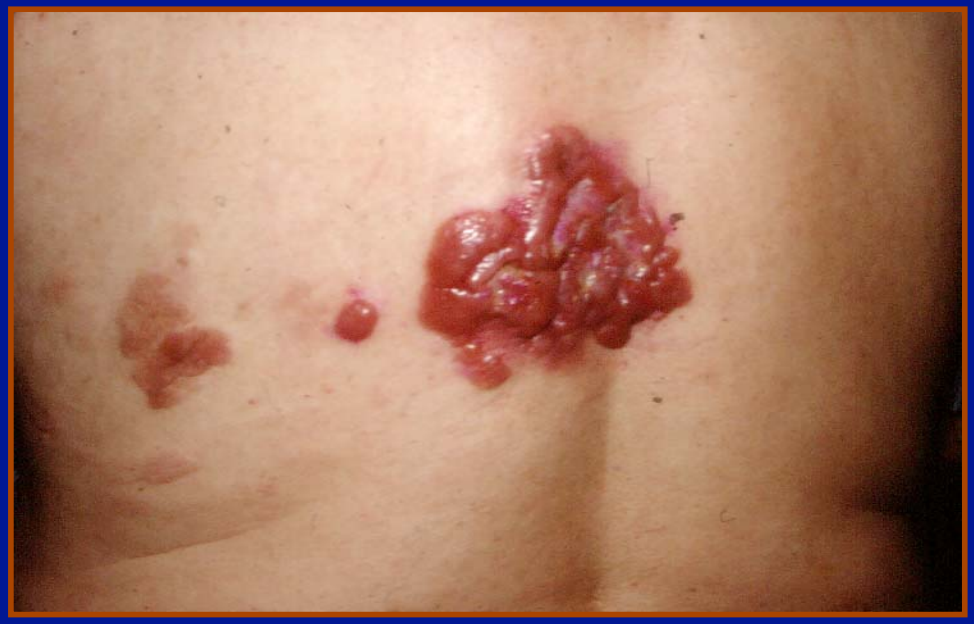
NR indicates not reached.
*Data are based on 1905 patients with a primary cutaneous lymphoma registered at the Dutch and Austrian Cutaneous Lymphoma Group between 1986 and 2002.
†Primary cutaneous peripheral T-cell lymphoma, unspecified excluding the three provisional entities indicated with a double dagger (‡).

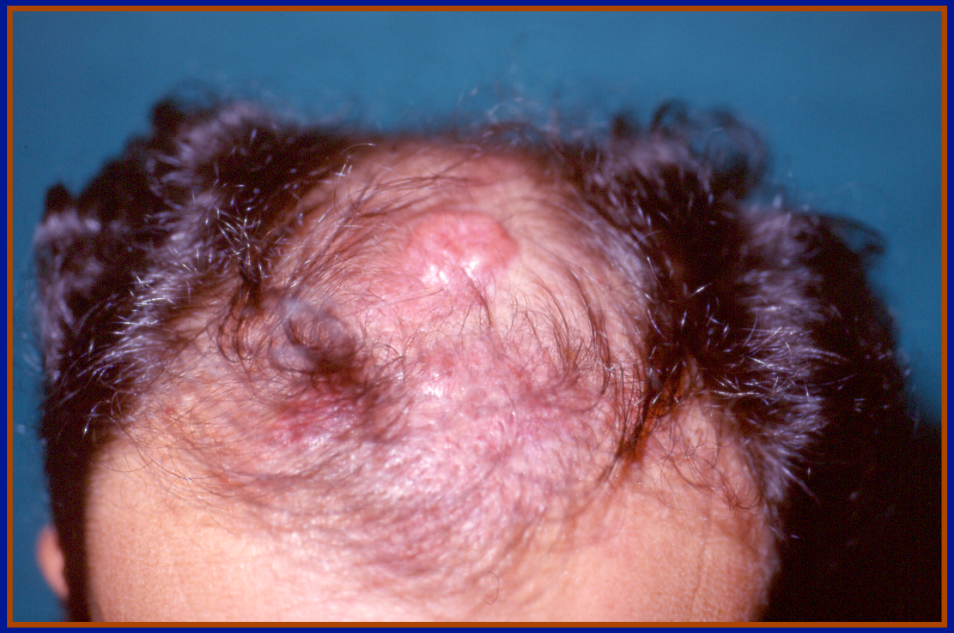
FOLLICLE CENTRE B-CELL LYMPHOMA

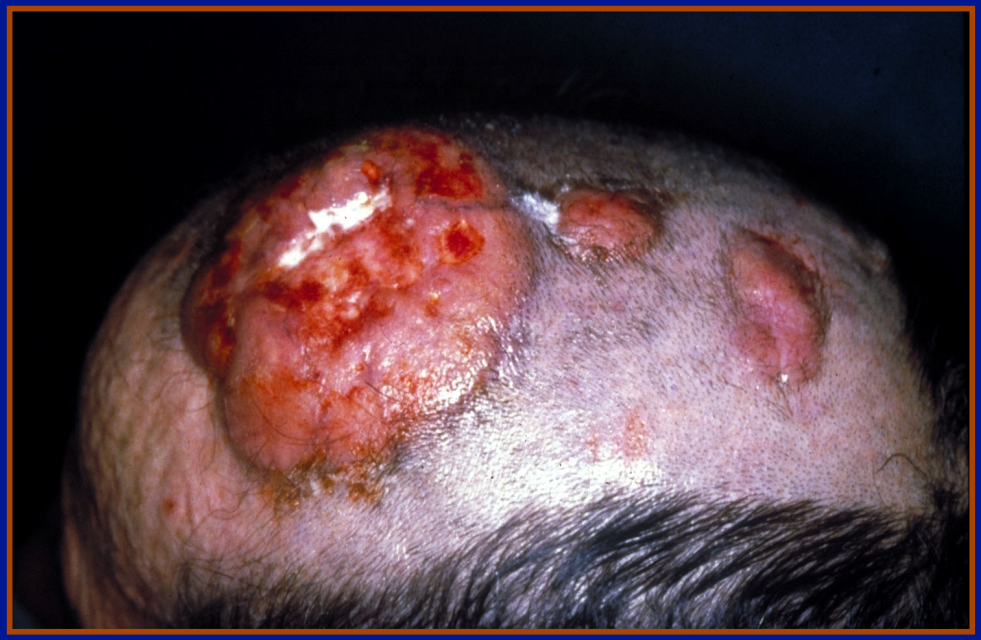
Epidemiology, Clinical Features, and Prognosis

- Most commonly affects adults aged >50 years, with a slight male gender prevalence
- Predominantly localized on the trunk and head & neck; multifocal skin lesions unfrequently observed (<20%)
- FCL presents with erythematous to cianotic papules, plaques or nodules; typical, multiphasic evolution: possible, rapid growth of large tumours - extenting into subcutaneous fat - after months to years
- Ulceration possible, extracutaneous dissemination uncommon
- Very good prognosis

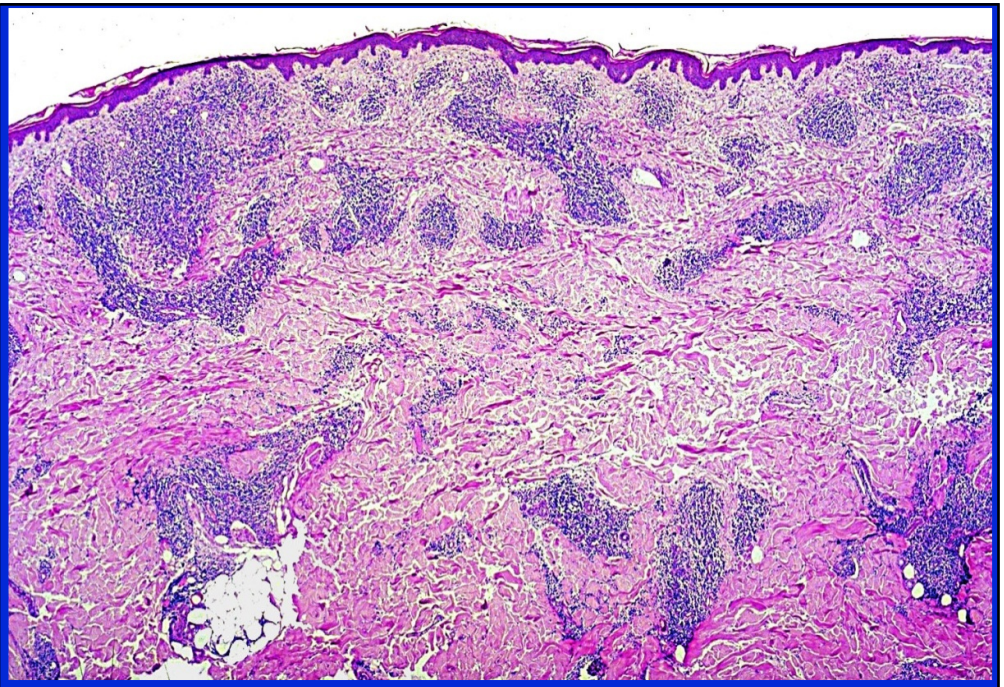


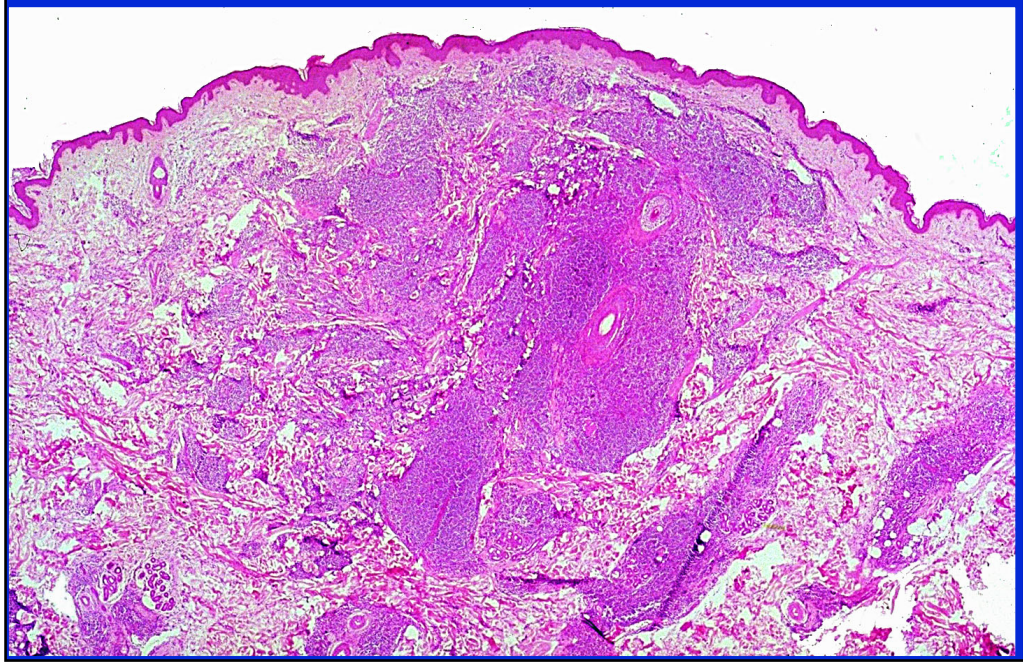


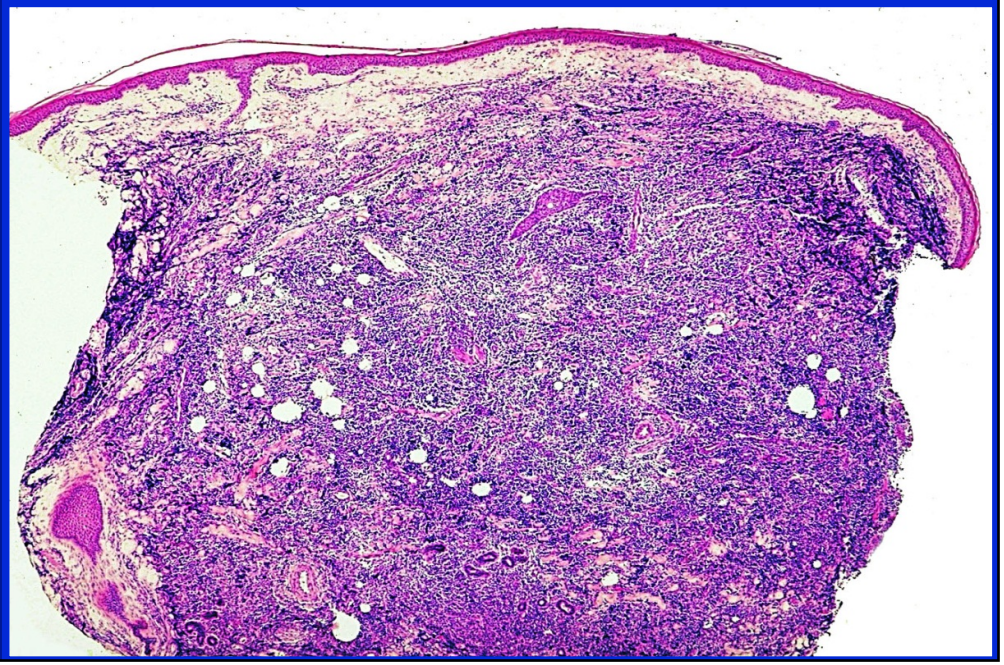


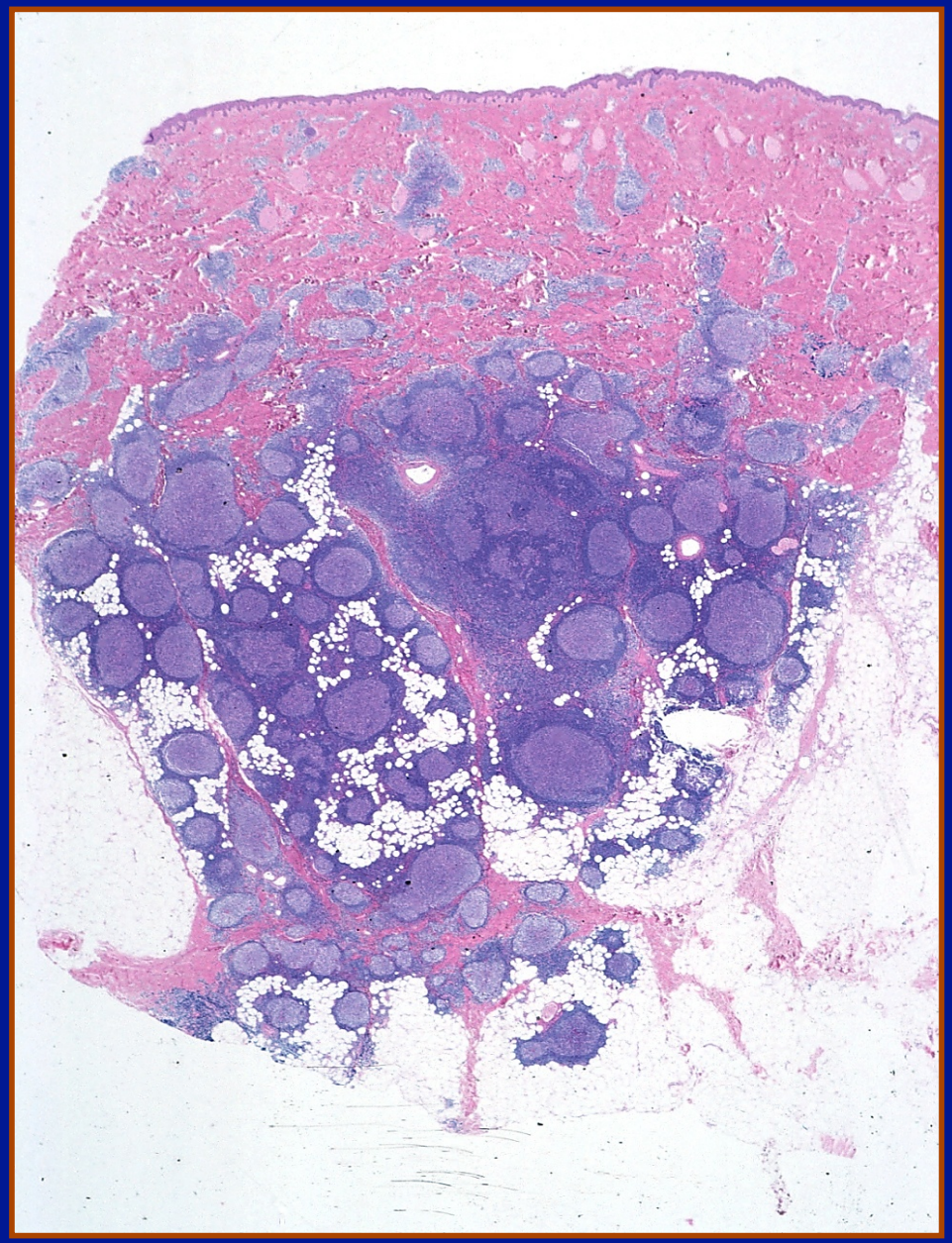


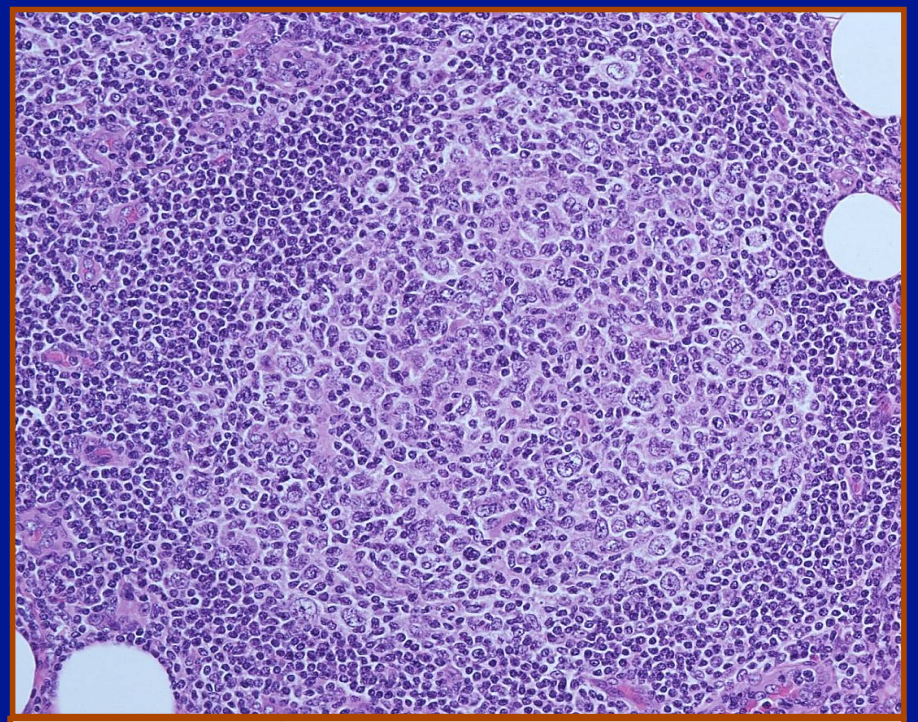
- ✓ *FCCL* shows nodular or diffuse infiltrates, with the almost constant sparing of the epidermis
- ✓ The histologic picture is variable, and primarily relates to the age and growth rate of skin lesions

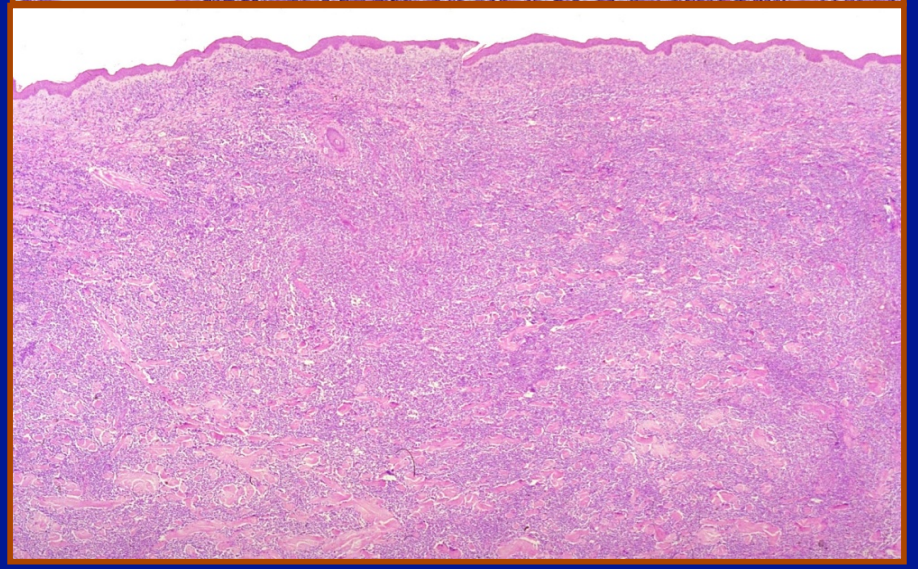






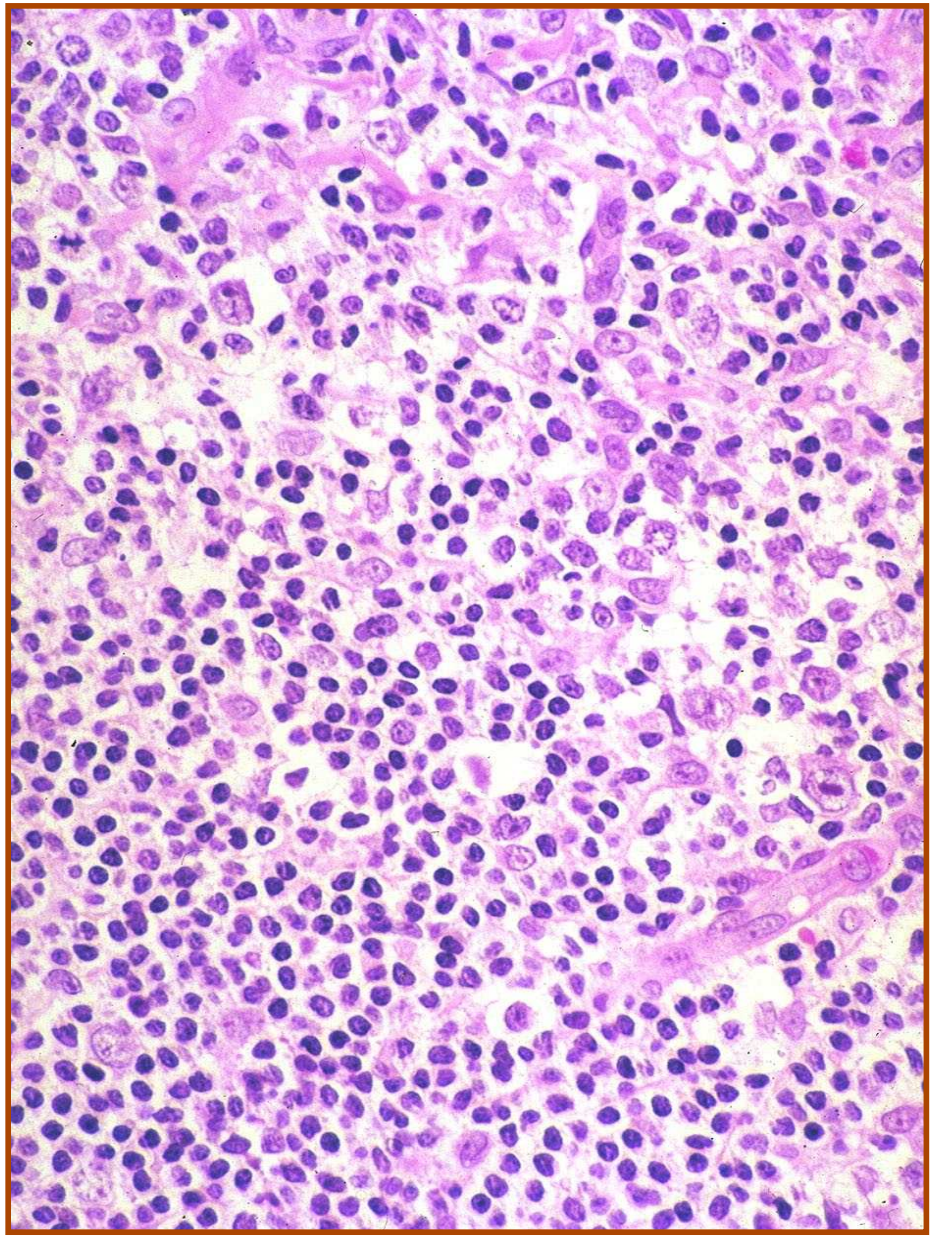






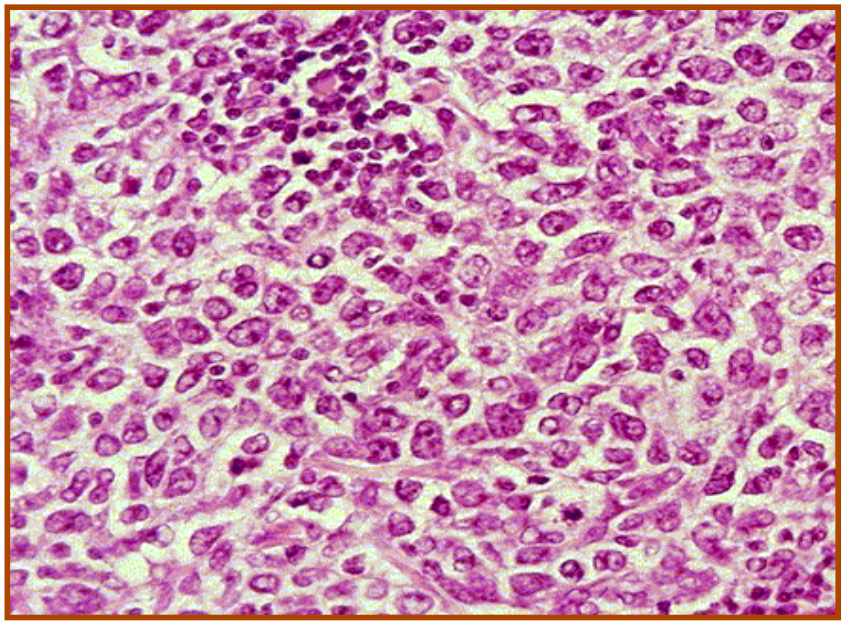
Follicle Centre Lymphoma

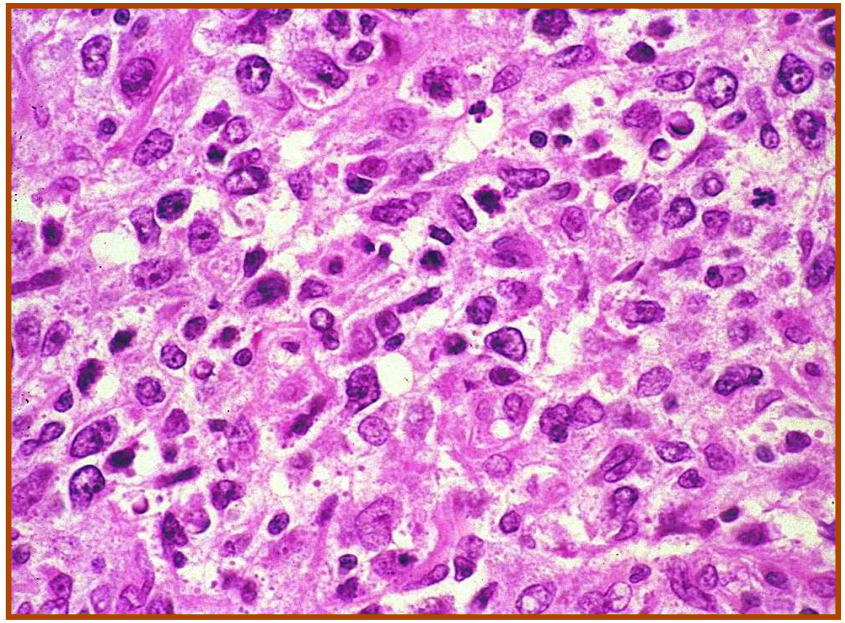
- Small and early lesions contain a mixture of small cleaved cells (centrocytes), relatively few noncleaved cells (centroblasts), and many reactive T cells
- Large cleaved cells (centrocytes), often multilobated, are a common feature



Follicle Centre Lymphoma

- With progression to tumorous lesions, neoplastic cells increase both in number and size, whereas the number of tumor infiltrating T cells decreases
- Tumorous skin lesions generally show a fairly monotonous infiltrate of large cleaved/multilobated cells, with variable admixture of centroblasts and immunoblasts





Follicle Centre Lymphoma

Immunoprofile

- Neoplastic B cells express CD19, CD20, CD22, CD79a and show monotypic sIg (the lack of detectable sIg is common in tumorous lesions with large cell histology)
- Follicles are associated with follicular dendritic cells (positive for **CD21**, CD23, CD35)
- FCLs are generally **bcl6+**, variably express CD10, **rarely (and faintly) express the bcl-2 protein**, and are CD5, CD43, IgM, Fox-P1, and **MUM-1/ IRF4 negative**

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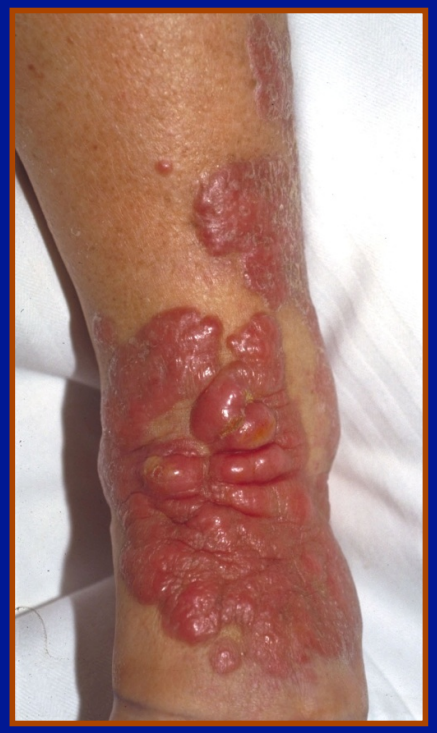
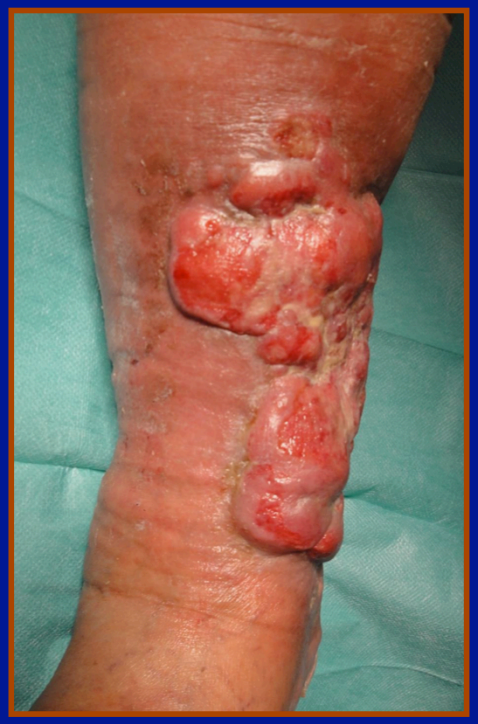
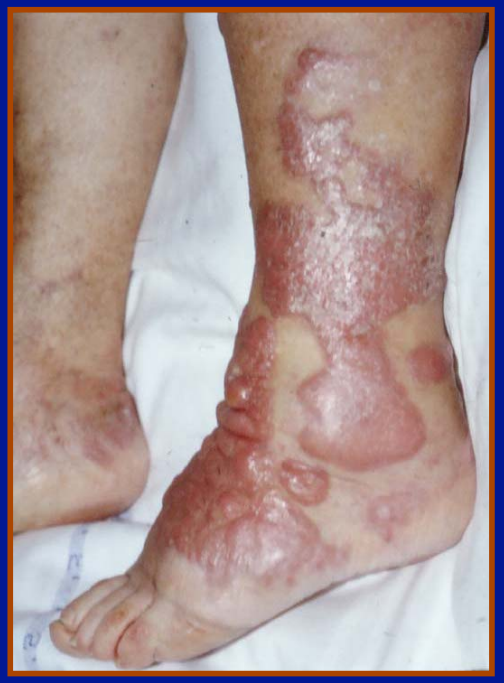
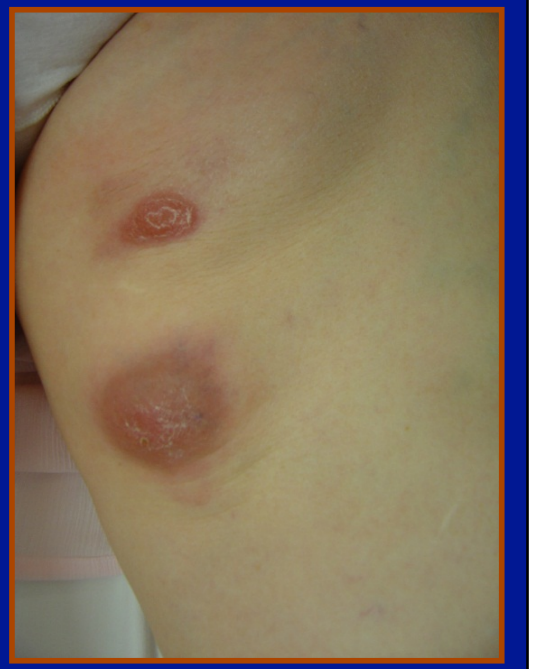
DIFFUSE LARGE B-CELL LYMPHOMA, LEG-TYPE
Epidemiology, Clinical Features, and Prognosis

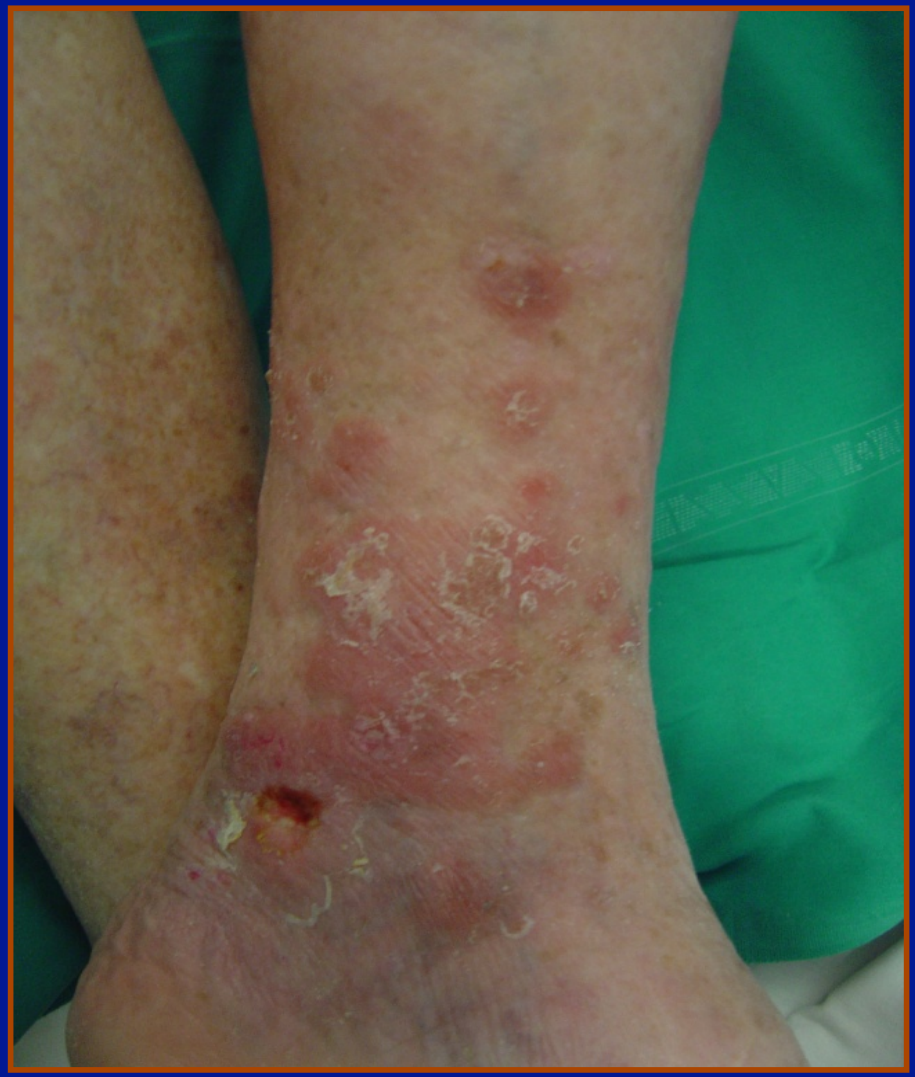
- Most commonly affects adults aged >70 years, with a clearcut female gender prevalence
- Predominantly localized on the legs (one or both, most often below the knee); rare lesions elsewhere (≈10%)
- DLBCL, LT presents with erythematous to cianotic plaques, nodules or tumors; rapid growth from the beginning
- Ulceration frequent, extracutaneous dissemination common
- Intermediate prognosis (55% 5-year survival; 40% vs > 90% when stratified multiple vs single lesion)

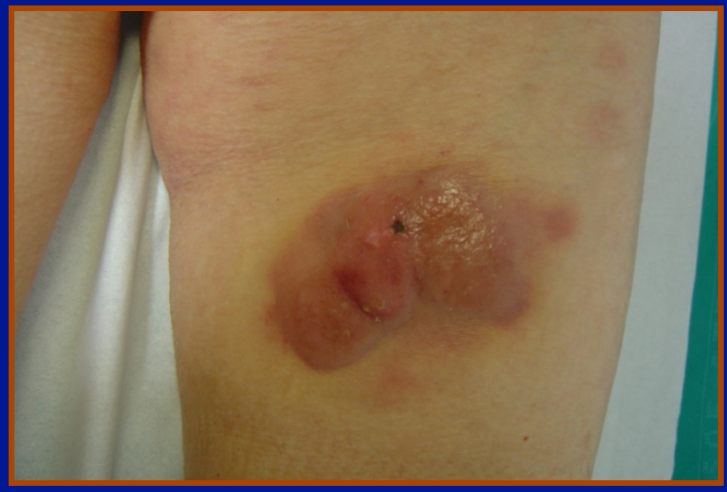


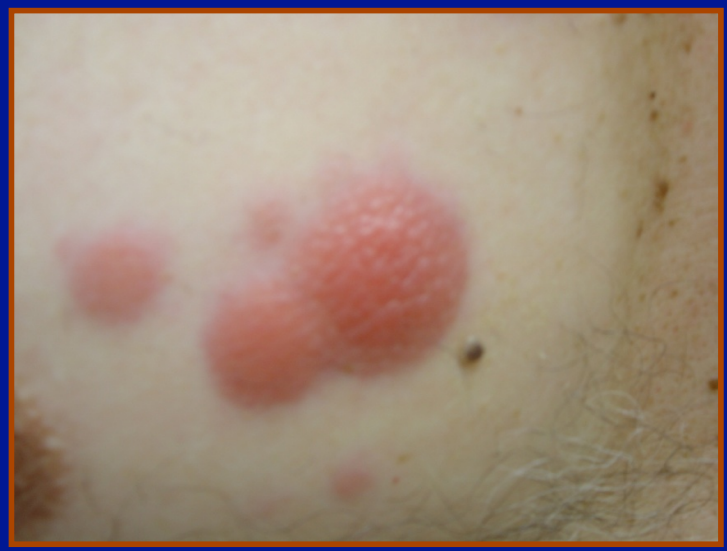






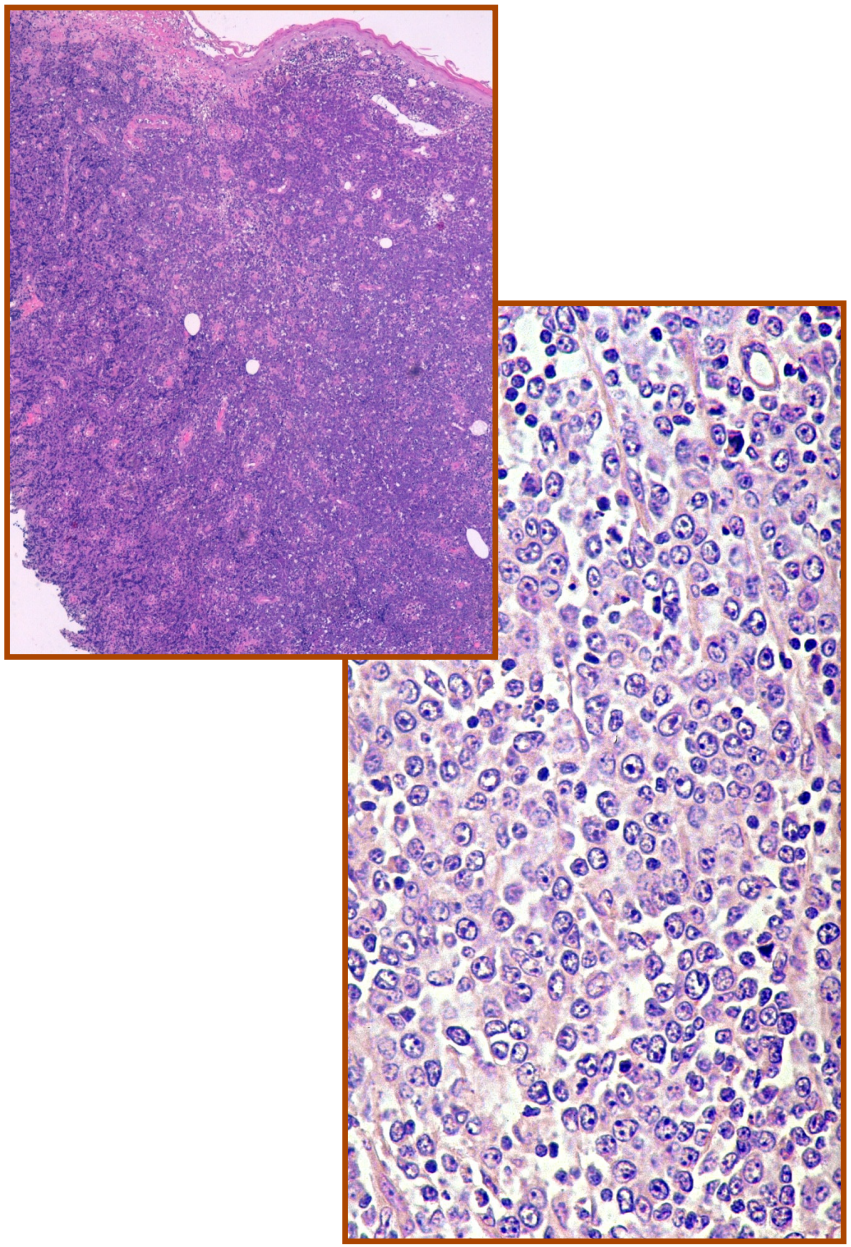






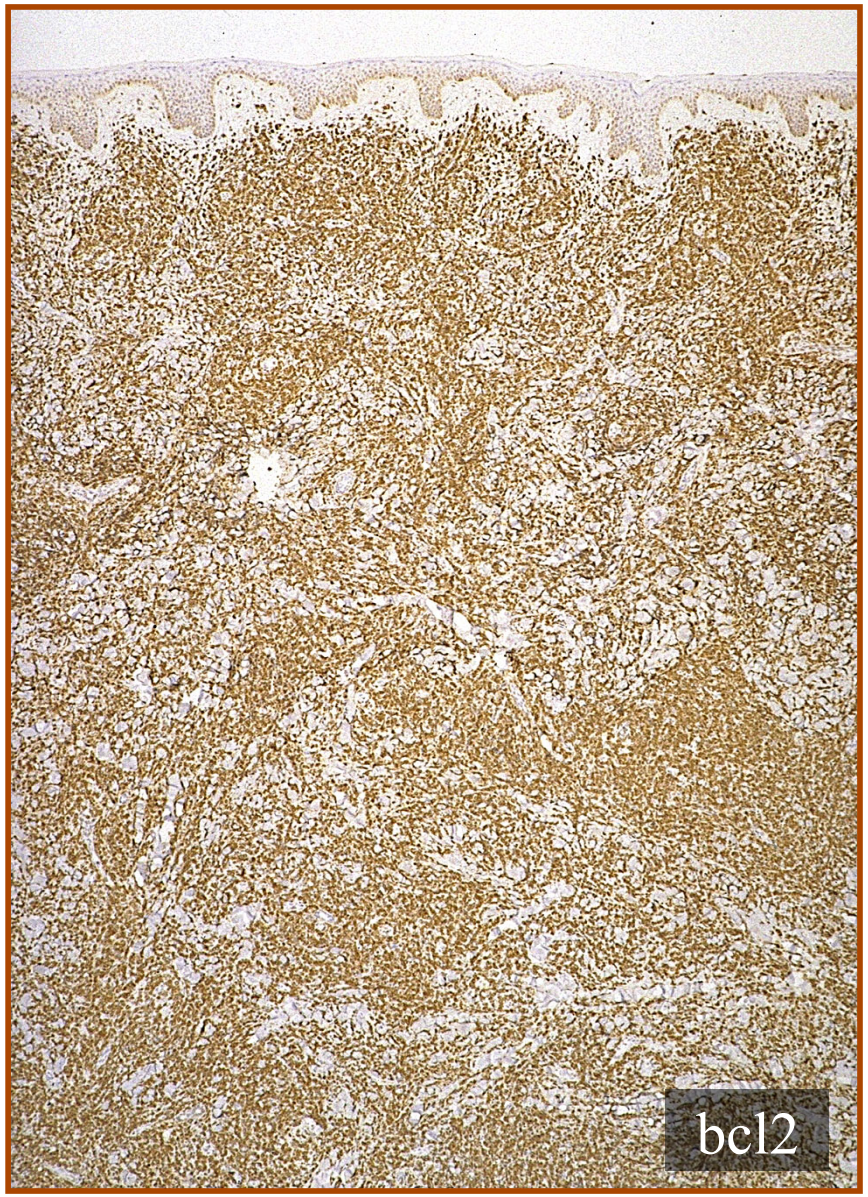
Diffuse Large B-cell Lymphoma, leg type

- Diffuse monotonous nonepidermotropic infiltrates composed of large B cells (confluent sheets of centroblasts and immunoblasts
- Frequent mitotic figures
- Reactive T cells few and confined to perivascular areas



Diffuse Large B-cell Lymphoma, leg type

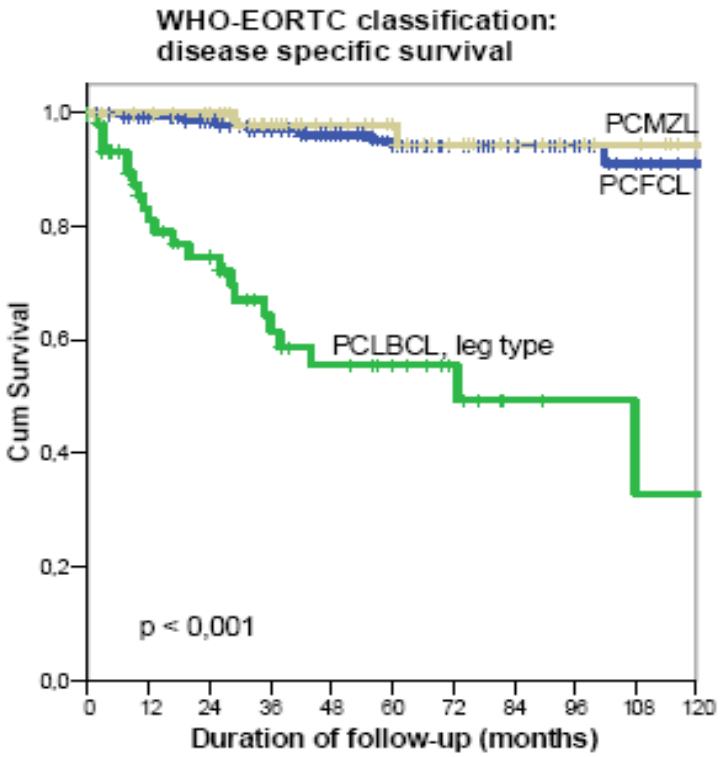
- The tumor cells express monotypic sIg/cIg, CD19, CD20, CD22, and CD79a
- Tumor cells are: bcl-2++ (strong and in most neoplastic cells), bcl6+/−, CD10−, MUM1/IRF4+ (activated B-cell gene expression profile), FOXP1+, and IgM+
- Absence of the t(14;18) translocation



Primary cutaneous marginal zone B-cell lymphoma (PCMZL)

Primary cutaneous follicle center lymphoma (PCFCL)

Primary cutaneous large B-cell lymphoma, leg type (PCLBCL-LT)



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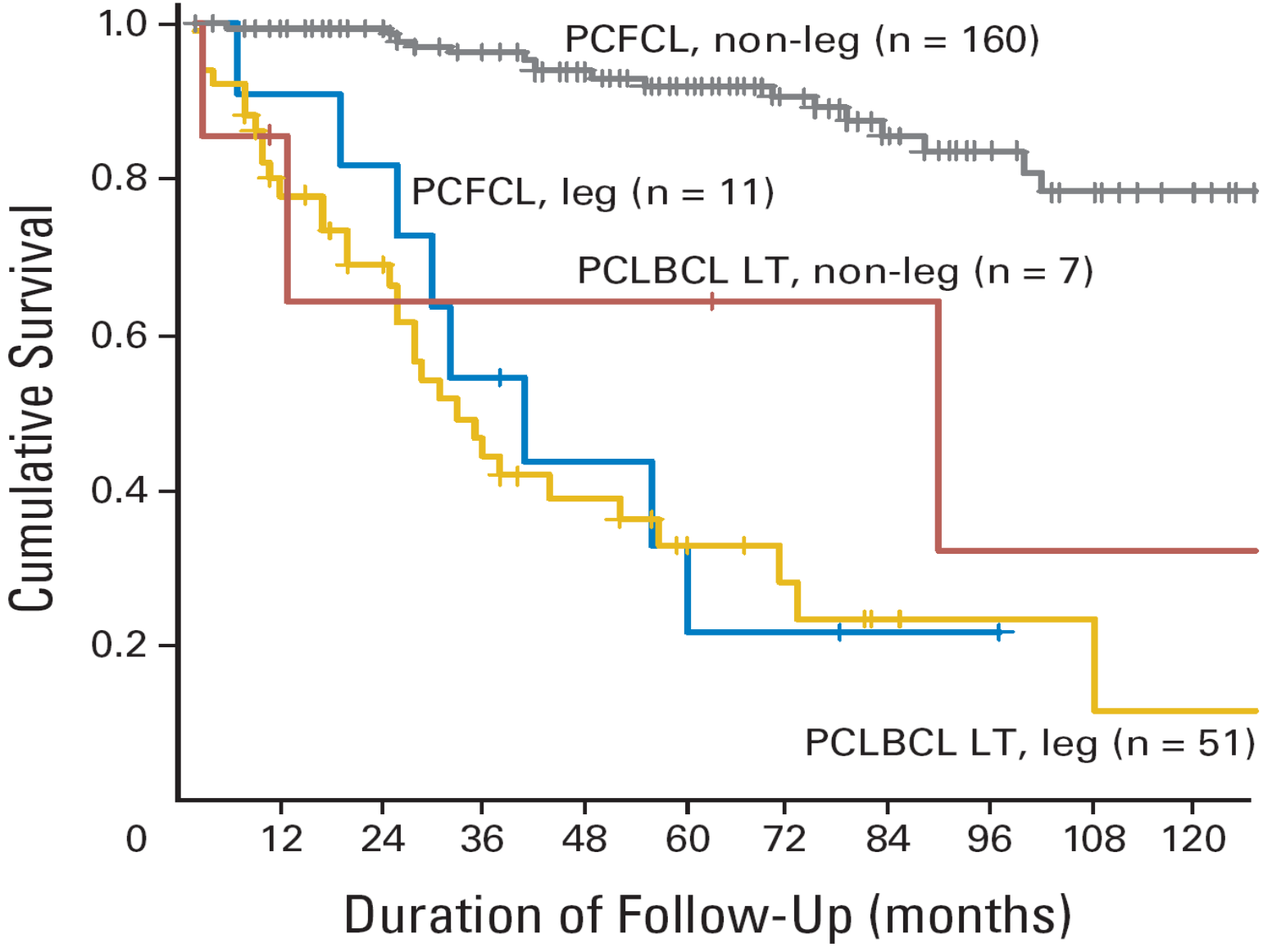

Senff et al. J Clin Oncol 25:1581-1587,2007

Cancer Medicine

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ORIGINAL RESEARCH

Primary cutaneous B-cell lymphoma other than marginal zone: clinicopathologic analysis of 161 cases: Comparison with current classification and definition of prognostic markers

Marco Lucioni[1], Emilio Berti[2], Luca Arcaini[3], Giorgio A. Croci[1], Aldo Maffi[1], Catherine Klersy[4], Gaia Goteri[5], Carlo Tomasini[6], Pietro Quaglino[7], Roberta Riboni[1], Mariarosa Arra[1], Elena Dallera[1], Vieri Grandi[8], Mauro Alaibac[9], Antonio Ramponi[10], Sara Rattotti[11], Maria Giuseppina Cabras[12], Silvia Franceschetti[13], Giulio Fraternali-Orcioni[14], Nicola Zerbinati[15], Francesco Onida[16], Stefano Ascani[17], Maria Teresa Fierro[7], Serena Rupoli[18], Marcello Gambacorta[19], Pier Luigi Zinzani[20], Nicola Pimpinelli[8], Marco Santucci[21] & Marco Paulli[1]

Table 1. Histologic features.

Histopathologic features	PCFCCL	PCDLBCL-NOS	PCDLBCL-LT	*P*
Cytology	Prevalence of small to large, cleaved cells (centrocytes)	Prevalence of round, nucleolated cells (centroblasts, rarely immunoblasts)	Almost exclusively round, nucleolated cells (centroblasts and immunoblasts)	—
Reactive T cells	Present	Present	Very scanty	—
Growth pattern (%)	Nodular to diffuse	Typically diffuse	Diffuse	NA
Nodular	33/96 (34)	0/40 (0)	0/25 (0)	
Nodular/diffuse	39/96 (41)	15/40 (38)	0/25 (0)	
Diffuse	24/96 (25)	25/40 (62)	25/25 (100)	
Dendritic meshwork, present (%)	80/96 (83)	11/40 (27)[1]	1/25 (4)[1]	<0.001
Infiltrate extension (%)				<0.001
Dermic	44/96 (46)	22/40 (55)	0/25 (0)	
Dermic/hypodermic	52/96 (54)	18/40 (45)	25/25 (100)	
Skin ulceration (%)	0/96 (0)	4/40 (10)	4/25 (16)	NA
Adnexal effacement, present (%)	3/96 (3)	6/40 (15)	10/25 (40)	NA
Necrosis	0/96 (0)	2/40 (5)	4/25 (16)	NA
Nuclear debris	0/96 (0)	4/40 (10)	15/25 (60)	NA
Starry sky appearance	0/96 (0)	0/40 (0)	11/25 (44)	NA
BCL2, +/total (%)	29/96 (30)	16/40 (40)	19/25 (76)	<0.001
CD10, +/total (%)	57/96 (59)	11/40 (27)	0/25 (0)	<0.001
BCL6, +/total (%)	84/96 (87)	33/40 (82)	14/25 (56)	0.001
MUM1, +/total (%)	0/96 (0)	14/40 (40)	20/25 (80)	0.004
HGAL, +/total (%)	50/54 (93)	9/40 (22)	1/25 (4)	<0.001
MYC, +/total (%)	0/40 (0)	10/21 (48)	11/13 (85)	<0.001
Ki67 median % (range)	30 (10–90)	50 (10–90)	70 (50–90)	—
Histogenetic profile, GC/total (%)	96/96 (100)	26/40 (65)	5/25 (20)	<0.001
DHS (%)				All: <0.001
0–1	NA	16/24	5/18	NOS vs. LT: 0.28
2	NA	8/24	13/18	
BCL2 translocation +/total (%)	15/75 (20)	3/27 (11)	1/20 (5)	0.234
BCL2 status (p)	(<0.001)	(0.273)	(1)	—
FISH+/IHC+ (%)	11/23 (48)	3/17 (17)	1/17 (6)	
FISH+/IHC− (%)	4/52 (8)	0/10 (0)	0/3 (0)	
EBV, +/total (%)	NA	0/15 (0)	0/20 (0)	NA

Table 2. Clinical features.

Clinical presentation	PCFCCL	PCDLBCL-NOS	PCDLBCL-LT (%)	*P*
Male/female (ratio)	53/43 (1.23)	27/13 (2.08)	17/8 (2.12)	0.432
Mean age (range)	54 (27–86)	63 (26–90)	76 (54–92)	<0.001
Number of lesions (%)				
Single lesion	67/96 (70)	27/40 (68)	18/25 (72)	0.889
Multiple lesions	29/96 (30)	13/40 (32)	6/25 (24)	
Diffuse	0/96 (0)	0/40 (0)	1/25 (4)	
Site involved (%)				
Head and neck	38/96 (40)	7/40 (17)	0/25 (0)	<0.001
Trunk	47/96 (49)	20/40 (50)	3/25 (12)	0.002
Upper limbs	8/96 (8)	8/40 (20)	1/25 (4)	NA
Lower limbs	7/96 (7)	9/40 (22)	21/25 (84)	<0.001
Type of lesion (%)				
Nodule/tumor	64/96 (67)	27/40 (67)	18/25 (72)	0.878
Plaque	17/96 (18)	10/40 (25)	5/25 (20)	0.625
Patch	4/96 (4)	1/40 (3)	2/25 (8)	NA
Papule	5/96 (5)	0/40 (0)	0/25 (0)	NA
Variable	6/96 (6)	2/40 (5)	0/25 (0)	NA
Therapy and follow-up				
First-line therapy				
Surgical only	20/96 (21)	2/40 (5)	0/25 (0)	0.004
Radiotherapy	47/96 (49)	15/40 (37)	9/25 (36)	<0.001
Chemotherapy (±radio)	26/96 (27)	22/40 (55)	15/25 (60)	0.006
Wait and see	3/96 (3)	1/40 (3)	1/25 (4)	NA
Response to therapy (%)				
CR	81/96 (84)	32/40 (80)	13/25 (52)	0.002
PR	15/96 (16)	8/40 (20)	12/25 (48)	
Relapse, /CR (%)	35/81 (43)	13/32 (41)	11/13 (85)	0.015
Extracutaneous relapse, /CR (%)	5/81 (6)	2/32 (6)	1/25 (4)	
Median time to relapse, months (range)	24 (6–156)	26 (5–159)	11 (5–28)	0.156
Follow-up				
ADF	76/96 (79)	25/40 (62)	4/25 (16)	<0.001
AWD	15/96 (16)	10/40 (25)	8/25 (32)	0.140
DOD	2/96 (2)	4/40 (10)	11/25 (44)	NA
DUC	3/96 (3)	1/40 (3)	2/25 (8)	NA
Median follow-up, months (range)	47 (12–237)	53 (8–210)	19 (6–126)	0.007

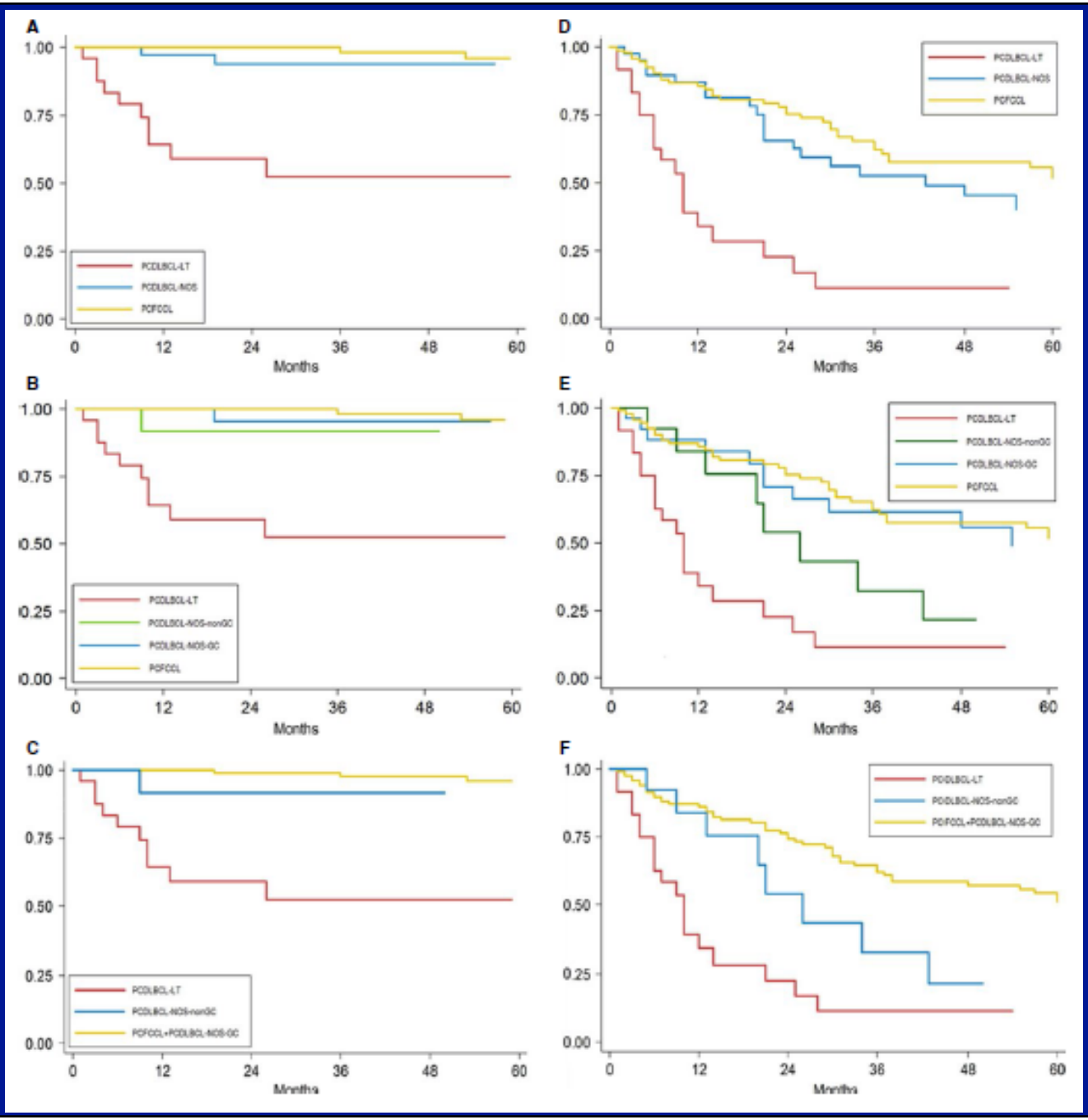
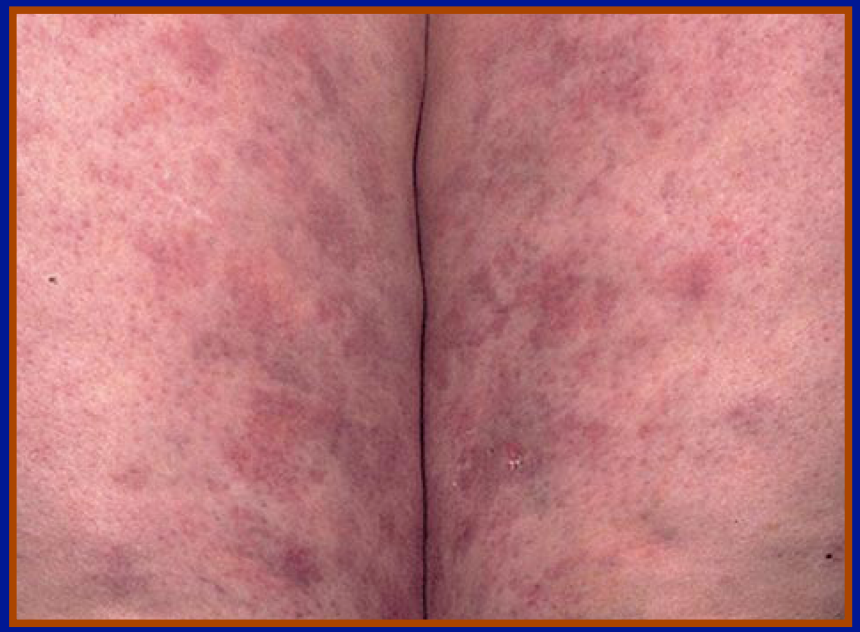
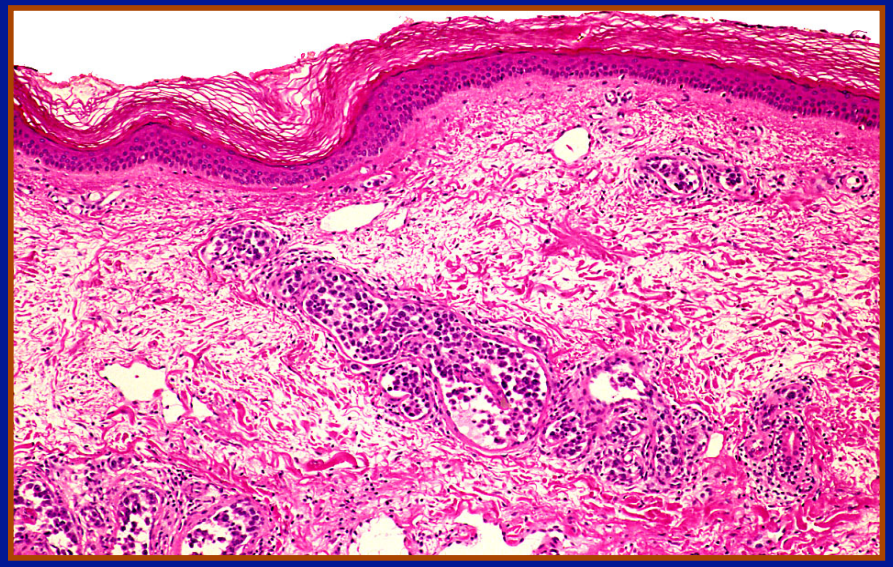

A
1.00
0.75
0.50
0.25
0.00
0
12
24
36
48
60
Months
PCDLBCL-LT
PCDLBCL-NOS
PCFCCL
B
1.00
0.75
0.50
0.25
0.00
0
12
24
36
48
60
Months
PCDLBCL-LT
PCDLBCL-NOS-nonGC
PCDLBCL-NOS-GC
PCFCCL
C
1.00
0.75
0.50
0.25
0.00
0
12
24
36
48
60
PCDLBCL-LT
PCDLBCL-NOS-nonGC
PCFCCL+PCDLBCL-NOS-GC
D
1.00
0.75
0.50
0.25
0.00
0
12
24
36
48
60
Months
PCDLBCL-LT
PCDLBCL-NOS
PCFCCL
E
1.00
0.75
0.50
0.25
0.00
0
12
24
36
48
60
Months
PCDLBCL-LT
PCDLBCL-NOS-nonGC
PCDLBCL-NOS-GC
PCFCCL
F
1.00
0.75
0.50
0.25
0.00
0
12
24
36
48
60
PCDLBCL-LT
PCDLBCL-NOS-nonGC
PCFCCL+PCDLBCL-NOS-GC

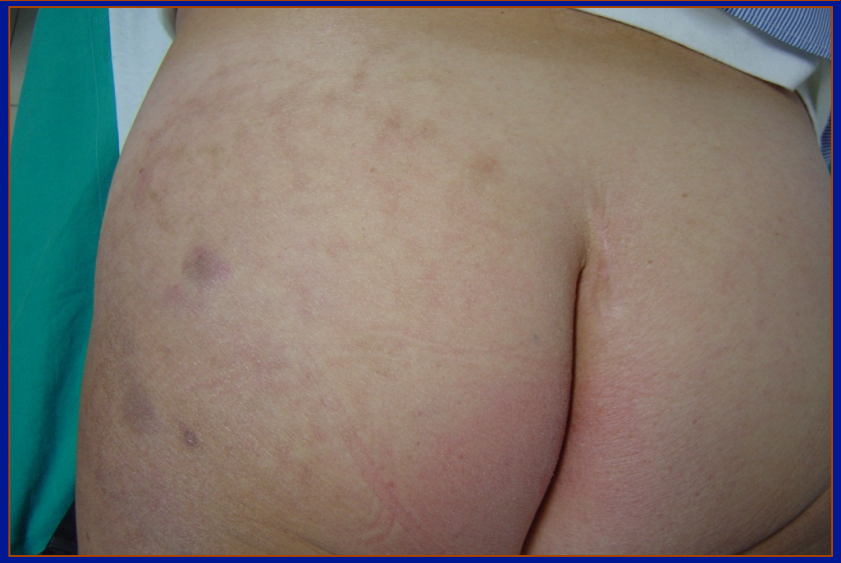
Table 2. Relative frequency and disease-specific 5-year survival of 1905 primary cutaneous lymphomas classified according to the WHO-EORTC classification

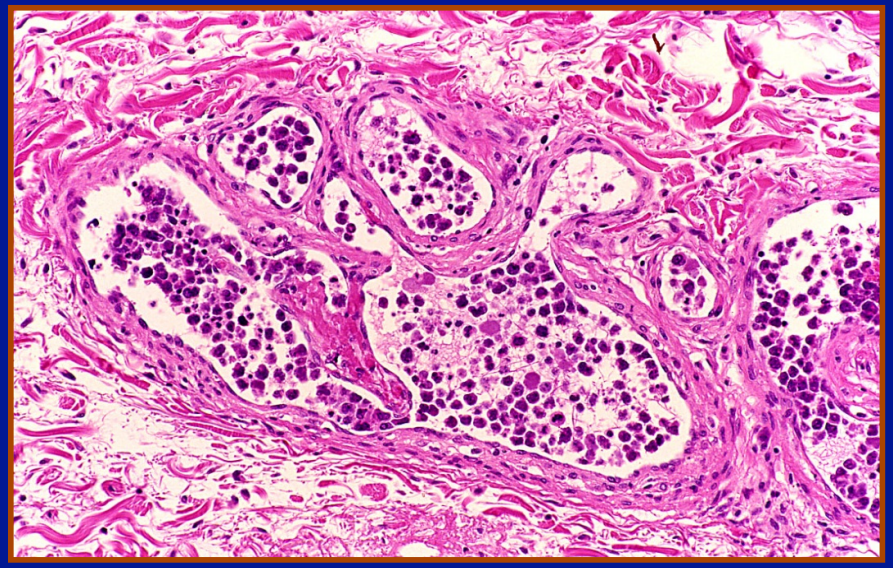
WHO-EORTC classification	No.	Frequency, %*	Disease-specific 5-year survival, %
Cutaneous T-cell lymphoma			
Indolent clinical behavior			
Mycosis fungoides	800	44	88
Folliculotropic MF	86	4	80
Pagetoid reticulosis	14	< 1	100
Granulomatous slack skin	4	< 1	100
Primary cutaneous anaplastic large cell lymphoma	146	8	95
Lymphomatoid papulosis	236	12	100
Subcutaneous panniculitis-like T-cell lymphoma	18	1	82
Primary cutaneous CD4+ small/medium pleomorphic T-cell lymphoma†	39	2	75
Aggressive clinical behavior			
Sézary syndrome	52	3	24
Primary cutaneous NK/T-cell lymphoma, nasal-type	7	< 1	NR
Primary cutaneous aggressive CD8+ T-cell lymphoma†	14	< 1	18
Primary cutaneous γ/δ T-cell lymphoma†	13	< 1	NR
Primary cutaneous peripheral T-cell lymphoma, unspecified‡	47	2	16
Cutaneous B-cell lymphoma			
Indolent clinical behavior			
Primary cutaneous marginal zone B-cell lymphoma	127	7	99
Primary cutaneous follicle center lymphoma	207	11	95
Intermediate clinical behavior			
Primary cutaneous diffuse large B-cell lymphoma, leg type	85	4	55
Primary cutaneous diffuse large B-cell lymphoma, other	4	< 1	50
Primary cutaneous intravascular large B-cell lymphoma	6	< 1	65

NR indicates not reached.
*Data are based on 1905 patients with a primary cutaneous lymphoma registered at the Dutch and Austrian Cutaneous Lymphoma Group between 1986 and 2002.
†Primary cutaneous peripheral T-cell lymphoma, unspecified excluding the three provisional entities indicated with a double dagger (‡).



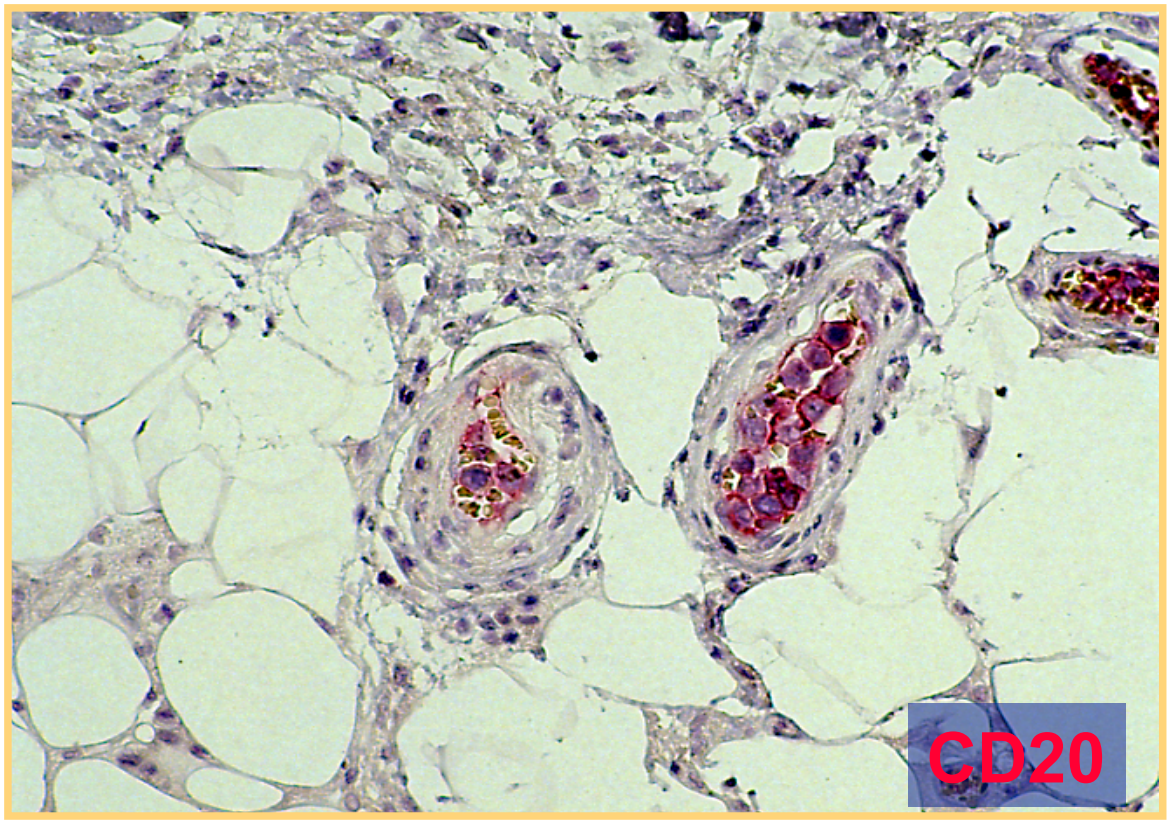






Intravascular Large B-cell Lymphoma

- Neoplastic cells are CDl9+, CD20+, CD22+, CD79a +, and monotypic sIg+



INTERNATIONAL SOCIETY FOR CUTANEOUS LYMPHOMAS

EORTC European Organisation for Research and Treatment of Cancer

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