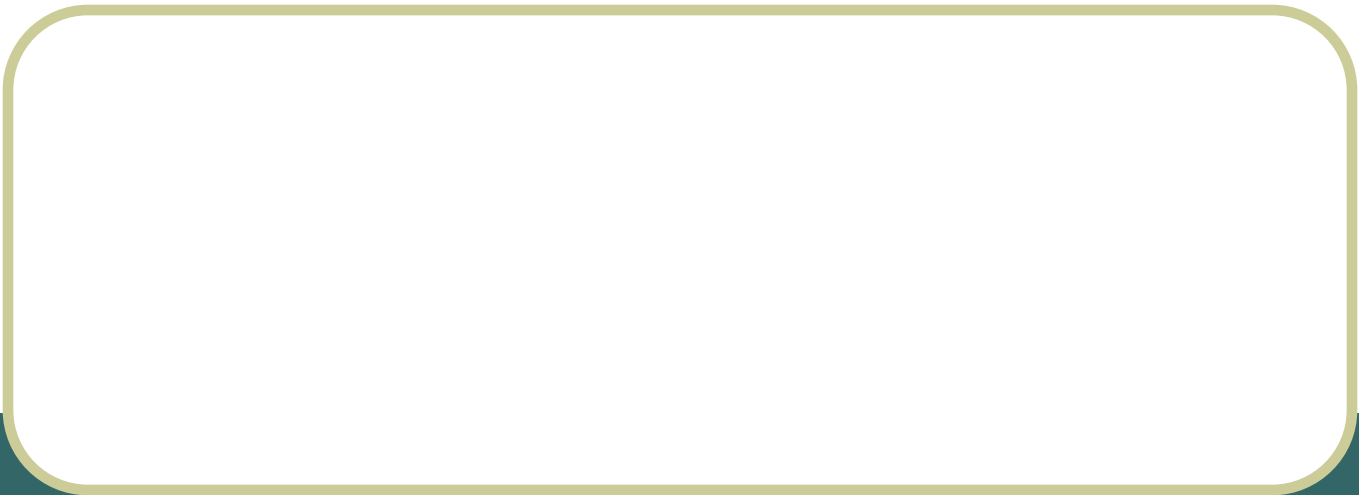


***WHO/EORTC
Classification of
Cutaneous Lymphomas
2005***



Introduction

- Lymphoma in skin, in the past were considered to be manifestations of systemic processes.
- A variety of T- and B-cell neoplasms can involve the skin, either primarily or secondarily.
- After GI, skin is the 2nd most common site of extranodal non-Hodgkin lymphoma, annual incidence of 1:100,000.
- Primary cutaneous lymphomas, completely different clinical behavior and prognosis from histologically similar systemic lymphomas, which may involve the skin secondarily
- Treatment for primary vs secondary is usually different.

Introduction 2

- EORTC – Classification 1997
- WHO – Classification 2001
- Consensus meetings on 2003 and 2004 lead to the 2005 WHO/EORTC classification.

CLASSIFICATION FOR PRIMARY CUTANEOUS LYMPHOMAS

Table 1. EORTC Classification for Primary Cutaneous Lymphomas

Primary CTCL	Primary CBCL
Indolent	Indolent
MF	Follicle center cell lymphoma
MF + follicular mucinosis	
Pagetoid reticulosis	Immunocytoma (marginal zone B-cell lymphoma)
Large cell CTCL, CD30 ⁺	
Anaplastic,	
Immunoblastic	
Pleomorphic	Intermediate
Lymphomatoid papulosis	Large B-cell lymphoma of the leg
Aggressive	
SS	
Large cell CTCL, CD30 ⁻	
Immunoblastic,	
Pleomorphic	
Provisional	Provisional
Granulomatous slack skin	Intravascular large B-cell lymphoma
	Plasmacytoma
CTCL, pleomorphic small/medium-sized	
Subcutaneous panniculitis-like T-cell lymphoma	

Abbreviations: CTCL, cutaneous T-cell lymphoma; CBCL, cutaneous B-cell lymphoma; MF, mycosis fungoides; SS, Sezary syndrome.

Table 1.2: WHO Classification of NHL

B-cell	
Mature B-cell lymphomas	
Precursor B-cell lymphoma	Small lymphocytic lymphoma
	Lymphoplasmacytic lymphoma
	Splenic marginal zone lymphoma
	Hairy cell leukaemia
	Plasma cell neoplasms
	Extranodal marginal zone B-cell lymphoma of mucosa-associated lymphoid tissue (MALT lymphoma)
	Nodal marginal zone B-cell lymphoma
	Follicular lymphoma (grades 1, 2, 3a and 3b)
	Diffuse follicle centre lymphoma
	Mantle cell lymphoma
	Diffuse large B-cell lymphoma
	Mediastinal (thymic) large B-cell lymphoma
	Intravascular large B-cell lymphoma
	Primary effusion lymphoma
	Burkitt lymphoma
B-cell proliferations of uncertain malignant potential	Lymphomatoid granulomatosis
	Post-transplant lymphoproliferative disorder, polymorphic
T-cell and NK-cell	
Precursor T- and NK-cell lymphomas	Precursor T lymphoblastic lymphoma
	Blastic NK-cell lymphoma
Mature T-cell and NK-cell lymphomas	T-cell prolymphocytic leukaemia
	T-cell large granular lymphocytic leukaemia
	Aggressive NK-cell leukaemia
	Adult T-cell lymphoma/leukaemia
	Extranodal NK-/T-cell lymphoma, nasal type
	Enteropathy-type T-cell lymphoma
	Hepatosplenic T-cell lymphoma
	Subcutaneous panniculitis-like T-cell lymphoma
	Mycosis fungoides
	Peripheral T-cell lymphoma unspecified
	Angioimmunoblastic T-cell lymphoma
Anaplastic large cell lymphoma	

WHO/EORTC 2005 Classification

- Combines features of both classifications.
- Takes into account morphologic, molecular, and clinical features of each entity.
- Comprises primary cutaneous B, T, and NK lymphomas.
- Terminology is compatible with that of systemic lymphomas (WHO), but reflects the organ specific peculiarities of cutaneous lymphomas.

Table 1. The WHO/EORTC classification for cutaneous lymphomas¹⁻³

Mature T-cell and NK-cell neoplasms

Mycosis fungoides (MF)

Variants of MF

 Pagetoid reticulosis (localized disease)

 Folliculotropic, syringotropic, granulomatous variants

Subtype of MF

 Granulomatous slack skin

Sézary syndrome

CD30⁺ T-cell lymphoproliferative disorders of the skin

 Lymphomatoid papulosis

 Primary cutaneous anaplastic large cell lymphoma

*Subcutaneous panniculitis-like T-cell lymphoma**

Primary cutaneous peripheral T-Cell lymphoma (PTL), unspecified

Subtypes of PTL

 Primary cutaneous aggressive epidermotropic CD8⁺ T-cell lymphoma (provisional)

 Cutaneous gamma/delta-positive T-cell lymphoma (provisional)

 Primary cutaneous CD4⁺ small/medium-sized pleomorphic T-cell lymphoma (provisional)

Extranodal NK/T-cell lymphoma, nasal type[†]

 Hydroa vacciniforme-like lymphoma (variant)

Adult T-cell leukemia/lymphoma[†]

Angioimmunoblastic T-cell lymphoma[†]

Mature B-cell neoplasms

Cutaneous marginal zone B-cell lymphoma (MALT-type)

Primary cutaneous follicle center lymphoma

Growth patterns

Follicular

Follicular and diffuse

Diffuse

Cutaneous diffuse large B-cell lymphoma, leg type

Cutaneous diffuse large B-cell lymphoma, others

Intravascular large B-cell lymphoma[†]

Lymphomatoid granulomatosis[†]

Chronic lymphocytic leukemia[†]

Mantle cell lymphoma[†]

Burkitt lymphoma[†]

Immature hematopoietic malignancies

Blastic NK-cell lymphoma[†] CD4⁺/CD56⁺ hematodermic neoplasm

Precursor lymphoblastic leukemia/lymphoma[†]

T-lymphoblastic lymphoma[†]

B-lymphoblastic lymphoma[†]

Myeloid and monocytic leukemias[†]

Hodgkin lymphoma

*Definition is restricted to lymphomas of alpha/beta T-cell origin.

[†]This table also contains entities of extracutaneous lymphomas frequently involving the skin as a secondary site.

[‡]Recent evidence suggests an origin from a dendritic cell precursor. In recognition of uncertain histogenesis the term CD4⁺/CD56⁺ hematodermic neoplasm is preferred.

Cutaneous T-cell lymphomas

Mycosis Fungoides

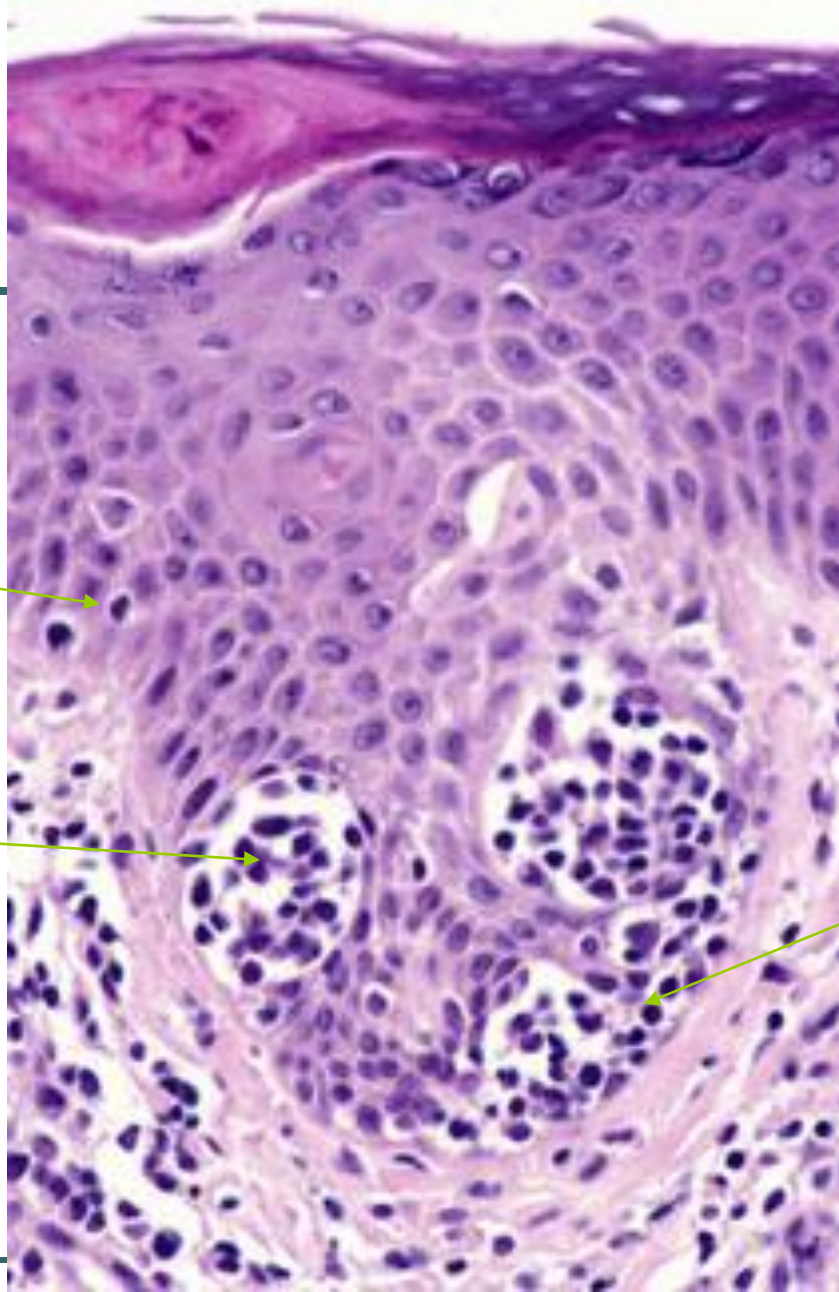
- *Mycosis fungoides* - 44% CL
- MF initially presents in the skin and shows a characteristic stepwise clinical progression with potential extracutaneous involvement.
- Skin lesions do not resolve.
- Patches → thin plaques → thick plaques, minority progress to systemic disease.

Patch Stage





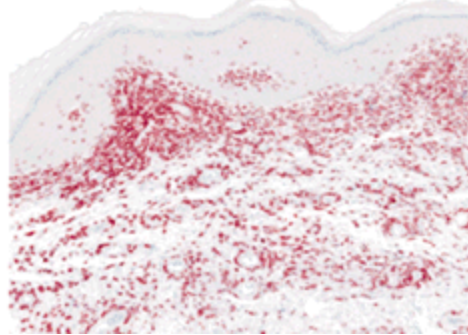
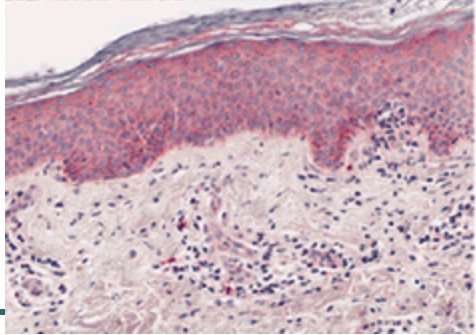
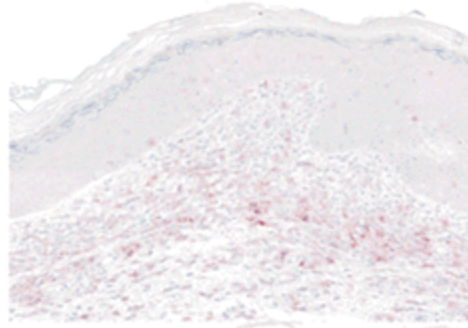
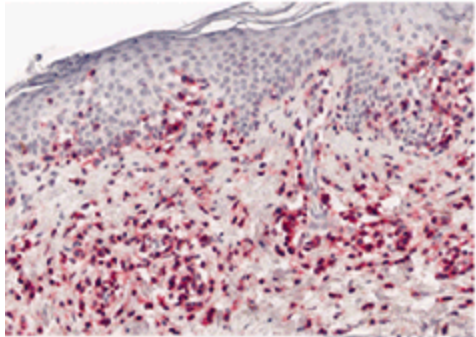
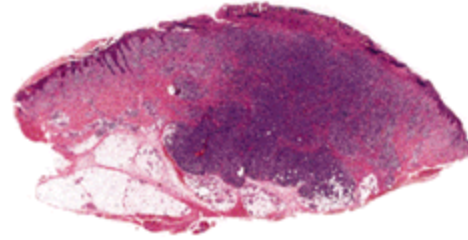
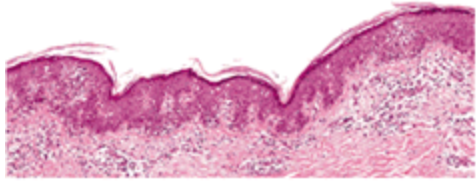
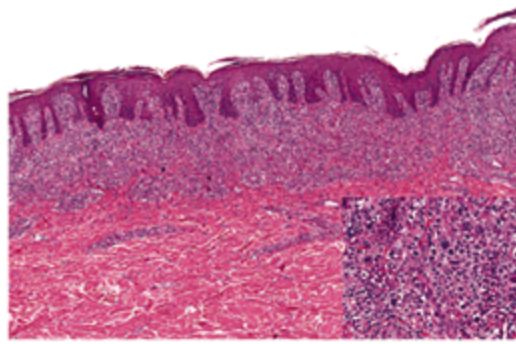
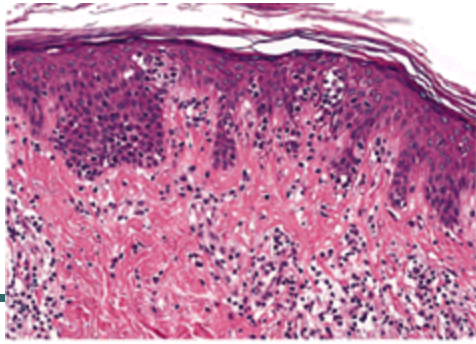
Plague Stage



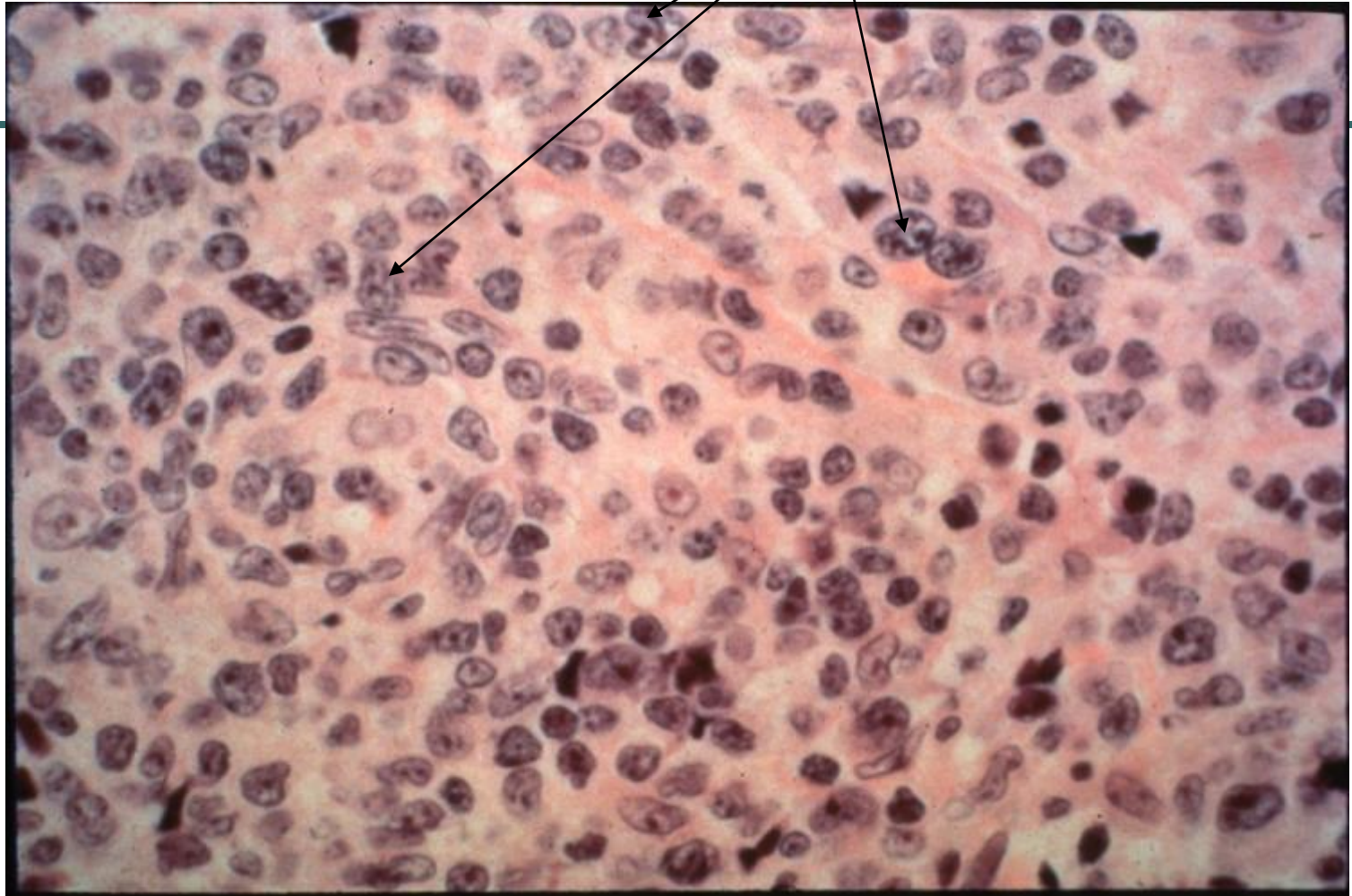
Epidermotrophism

Pautrier Microabscess

Lymphocytes
aligned on
basal layer



Cerebriform cells



Histologic features of MF

- Pautrier microabscesses – 10%
- Psoriasiform – lichenoid pattern
- Lymphocytes aligned in the basal layer (at tips of rete ridges)
- Epidermotropism 95%
- Cerebriiform cells seen in 50% of cases
- Eosinophils, plasma cells, and macrophages may be admixed.

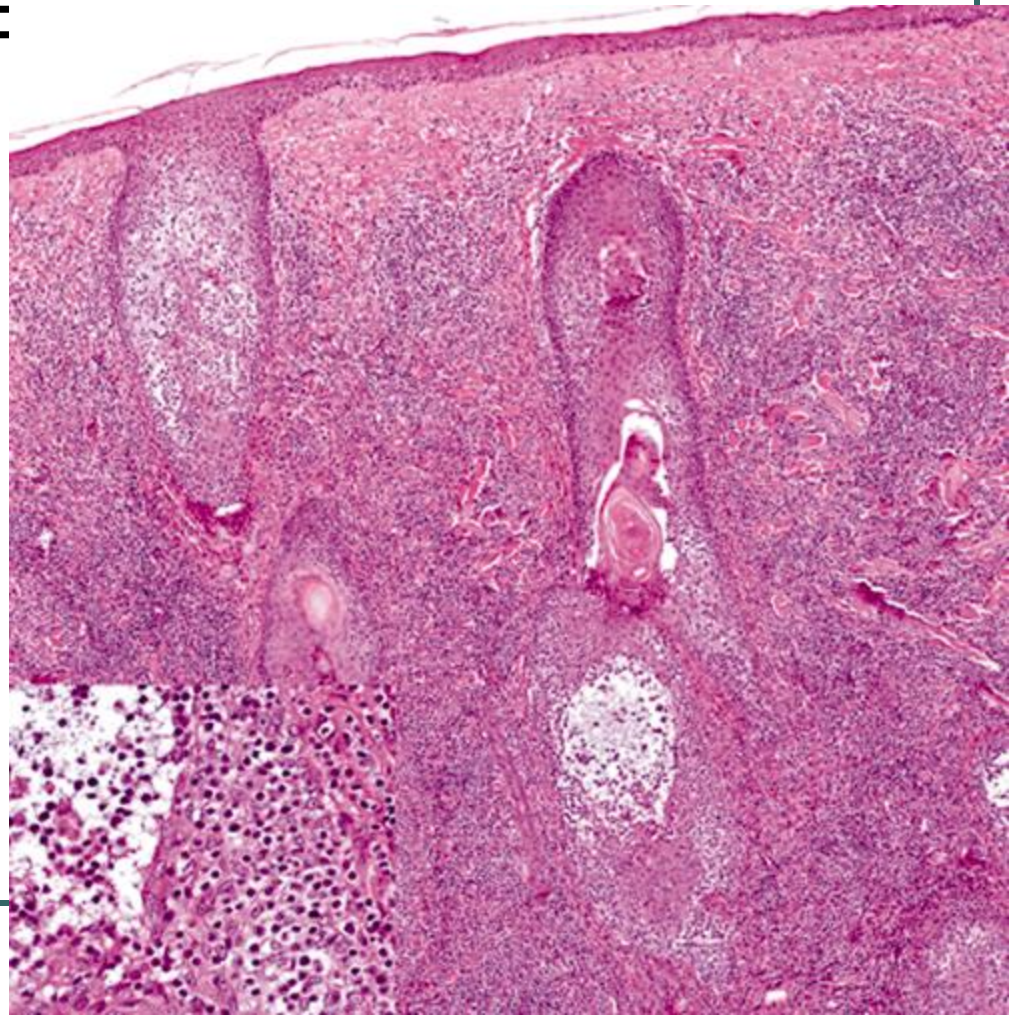
Mycosis Fungoides

- Immunohistochemistry is helpful but not necessary for dx.
- CD2 +, CD3 +, CD4+, CD5+ TCRbeta+
- CD8 -, Cd 30 –
- Rare cases may be CD8 + or CD30+
- 5yr survival 90%
- Tx Skin-targeted therapies as photo (chemo)–therapy PUVA, topical nitrogen mustard or chlormustine (BCNU), or radiotherapy, including total skin electron beam irradiation.

MF variants

- Folliculotropic MF

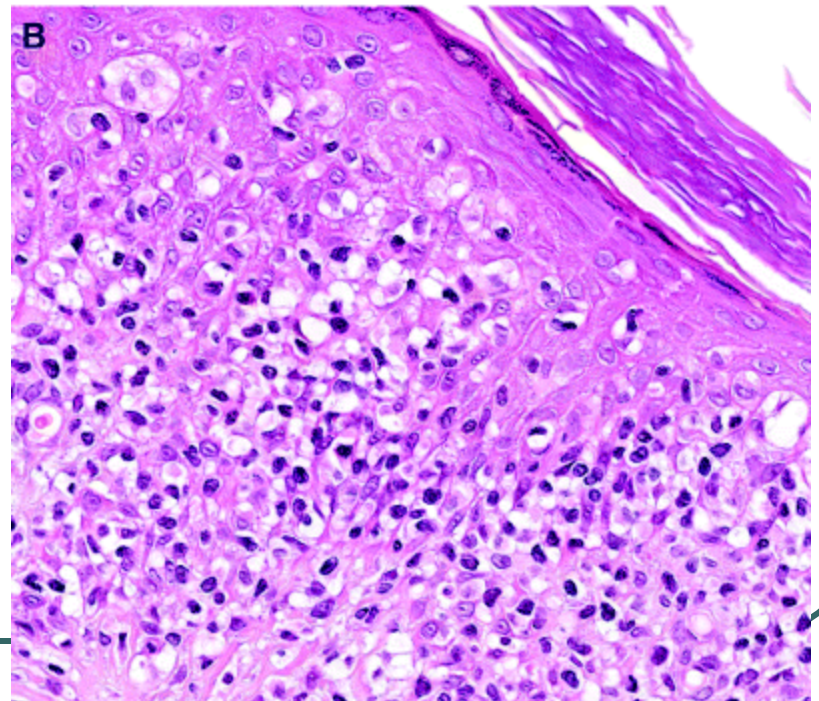
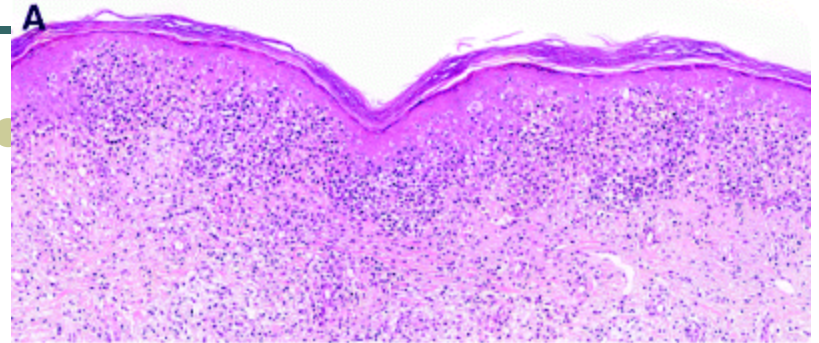
Worse px
5yr 80%



MF variants

Pagetoid Reticulosis

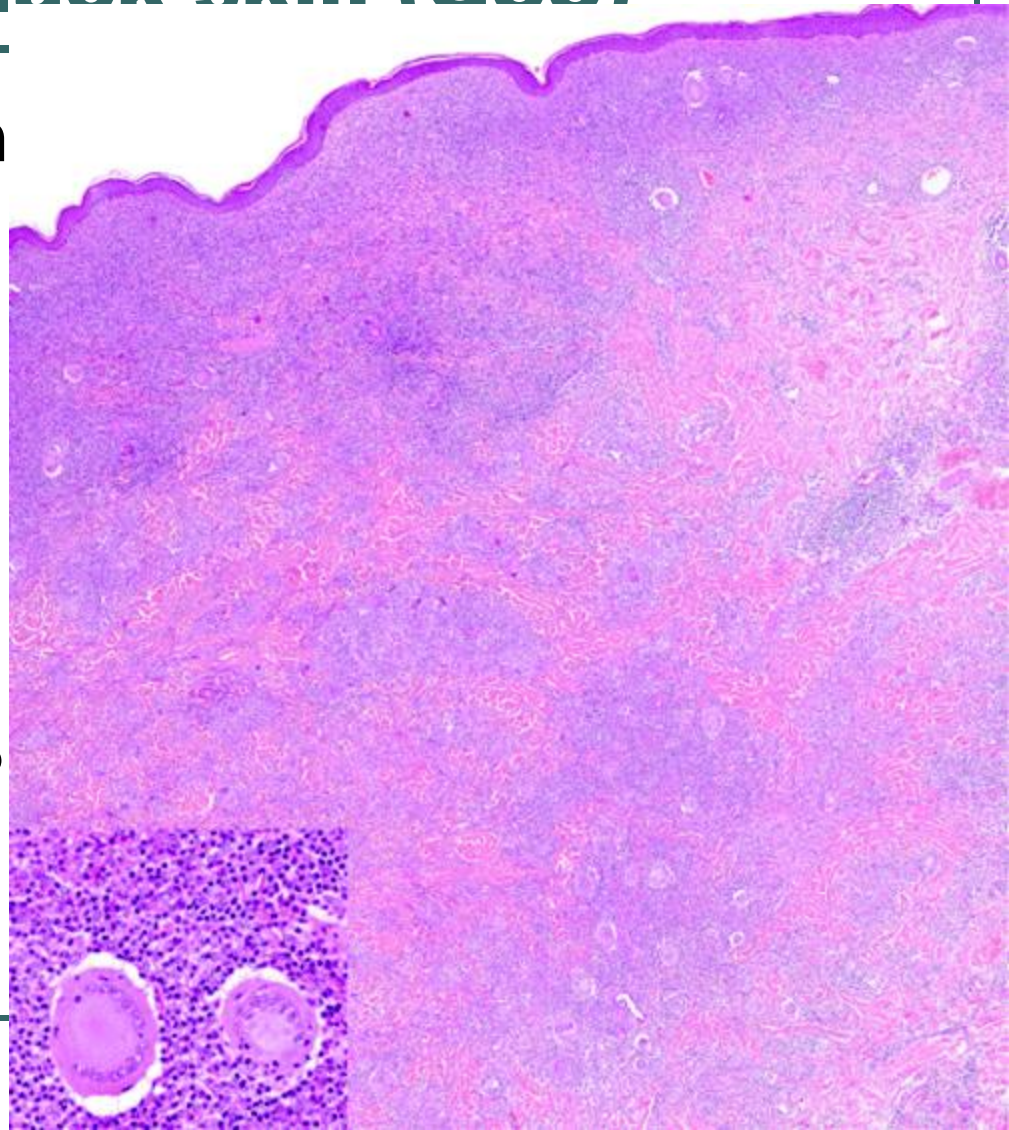
- Solitary or multiple
- T(H) phenotype
 - CD4,CD5,CD3 +
 - CD8-
- Strong expression of CLA, and alpha E beta 7
- May mimic superficial melanoma or paget's disease
- 100% 5 yr survival



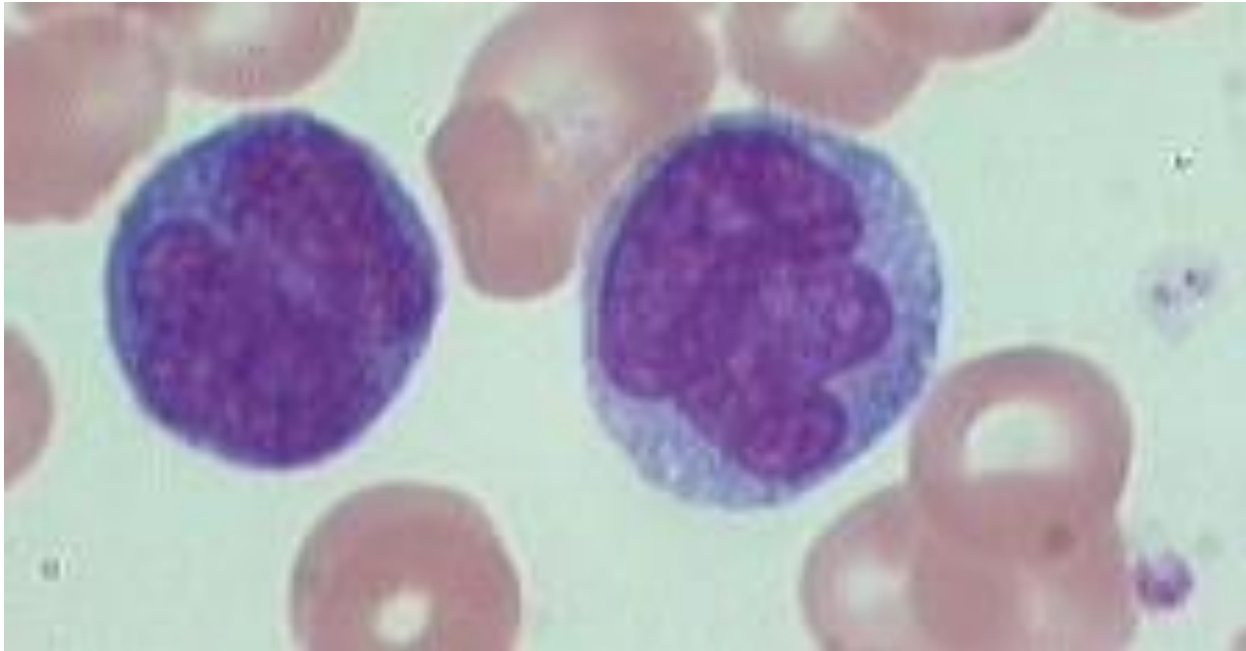
MF variants

Granulomatous slack skin (GSS)

- Bulky skin lesions on major skin folds
- Band like infiltrate with giant cells
- Elastophagocytosis and emperipolesis
- T(H) phenotype
- Giant cells are CD68 +
- 100 % 5 yr survival



Sezary Syndrome



Sezary Syndrome

- ISCL (International society of cutaneous lymphoma)
 - Absolute sezary cell count of 1000 cells/mm
 - Expanded CD4 population resulting in CD4/CD8 ratio of > 10
 - Loss of any of the T cell antigens (CD2, CD3, CD4, and CD5) or demonstration of T cell clonality by molecular studies.
- 20% 5 yr survival
- Tx Extracorporeal photopheresis (ECP), either alone or in combination with other treatment modalities (interferon alpha, methotrexate, campath)

CD 30 + T cell lymphoproliferative disorders of the skin

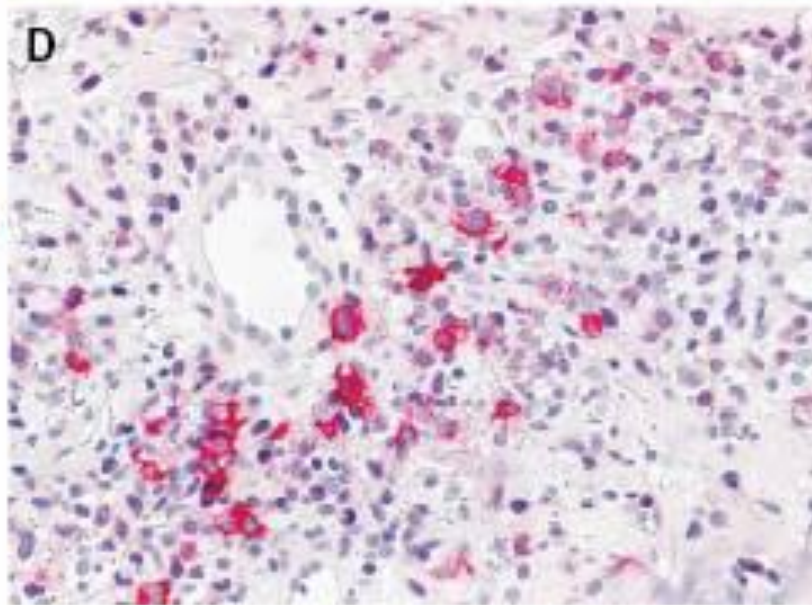
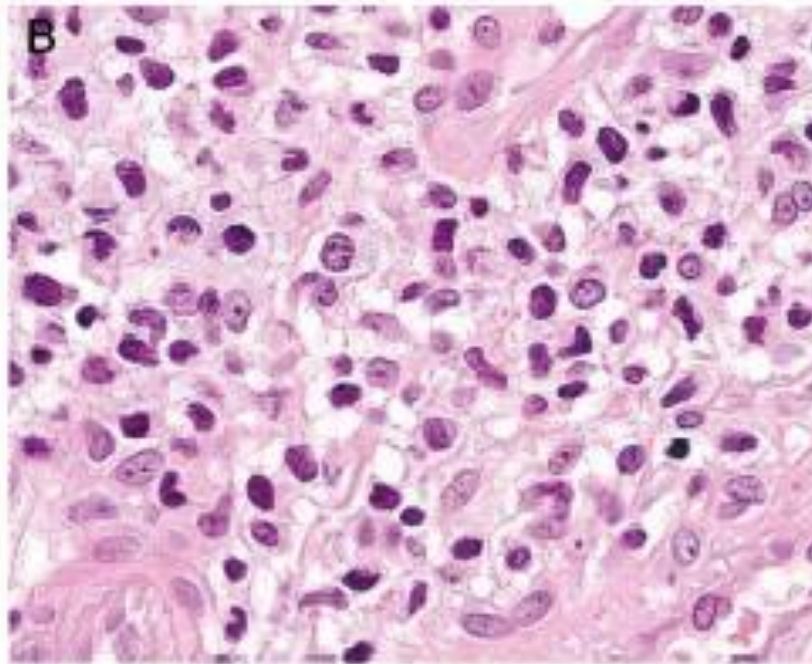
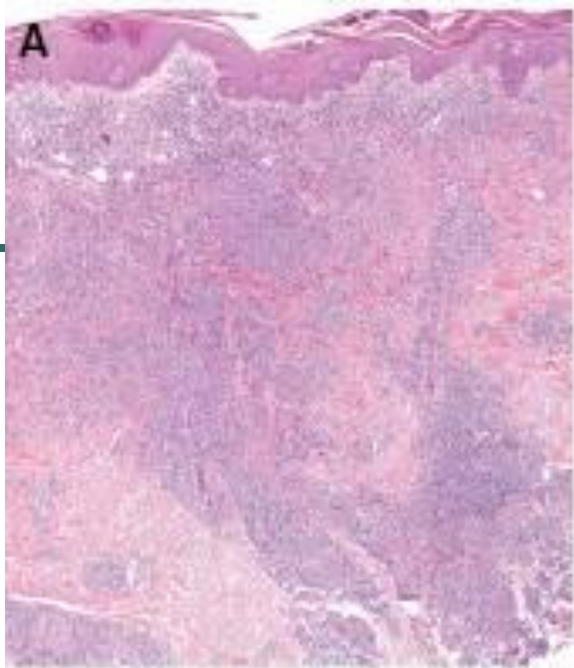
- Lymphomatoid papulosis
- Primary cutaneous anaplastic large cell lymphoma
- Hallmark is CD 30 +, however both entities differ in clinical and histologic presentations.
- Represent 30% of cutaneous lymphomas

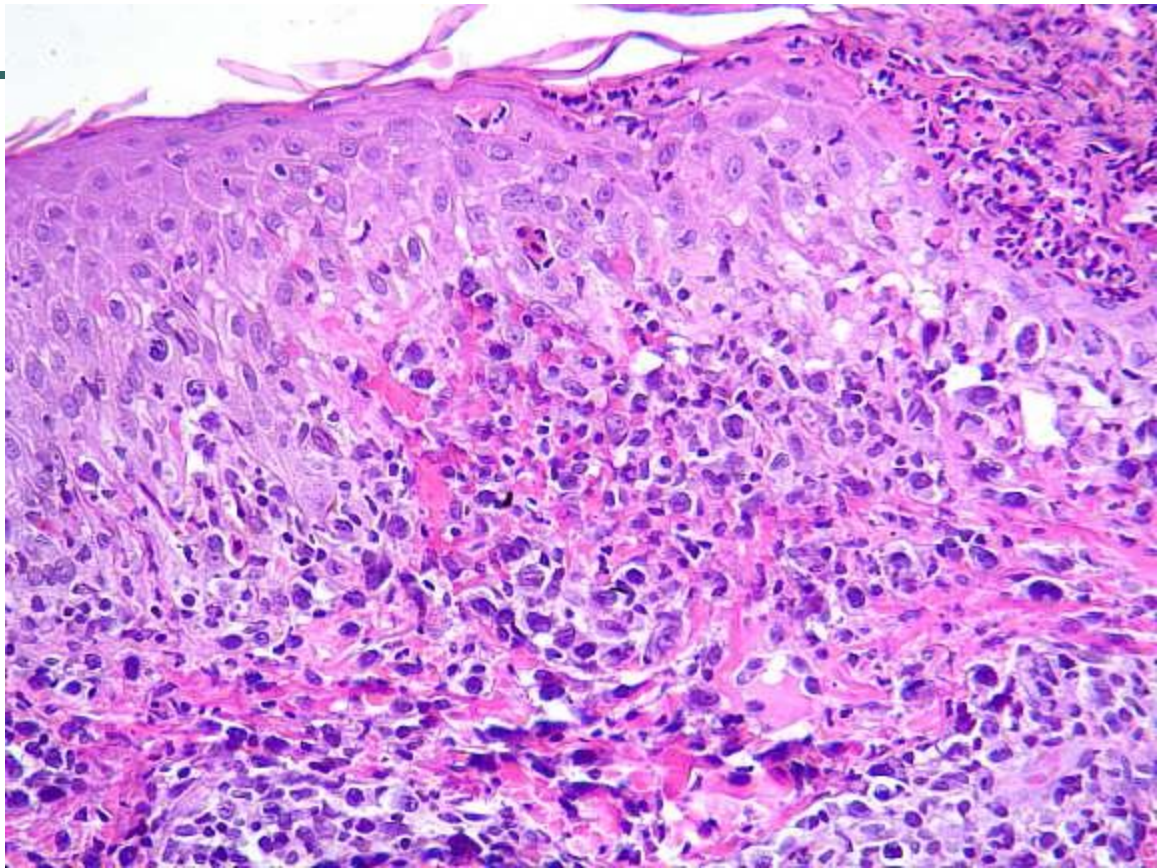
Lymphomatoid Papulosis

- Three histologic subtypes of LyP (types A, B, and C) have been described, which represent a spectrum with overlapping features
- LyP type A lesions, scattered or small clusters of large, sometimes multinucleated or Reed-Sternberg-like, CD30+ cells are intermingled with numerous inflammatory cells, such as histiocytes, small lymphocytes, neutrophils, and/or eosinophils.
- LyP type C lesions demonstrate a monotonous population or large clusters of large CD30+ T cells with relatively few admixed inflammatory cells.
- LyP type B is uncommon (less than 10%) and is characterized by an epidermotropic infiltrate of small atypical cells with cerebriform nuclei similar to that observed in MF.
- The large atypical cells in the LyP type A and type C lesions have the same phenotype as the tumor cells in C-ALCL.
- The atypical cells with cerebriform nuclei in the LyP type B lesions have a CD3+, CD4+, CD8- phenotype and do not express CD30 antigen.
- Clonally rearranged T-cell receptor genes have been detected in approximately 60%-70% of LyP lesions.



<http://dermis.net>



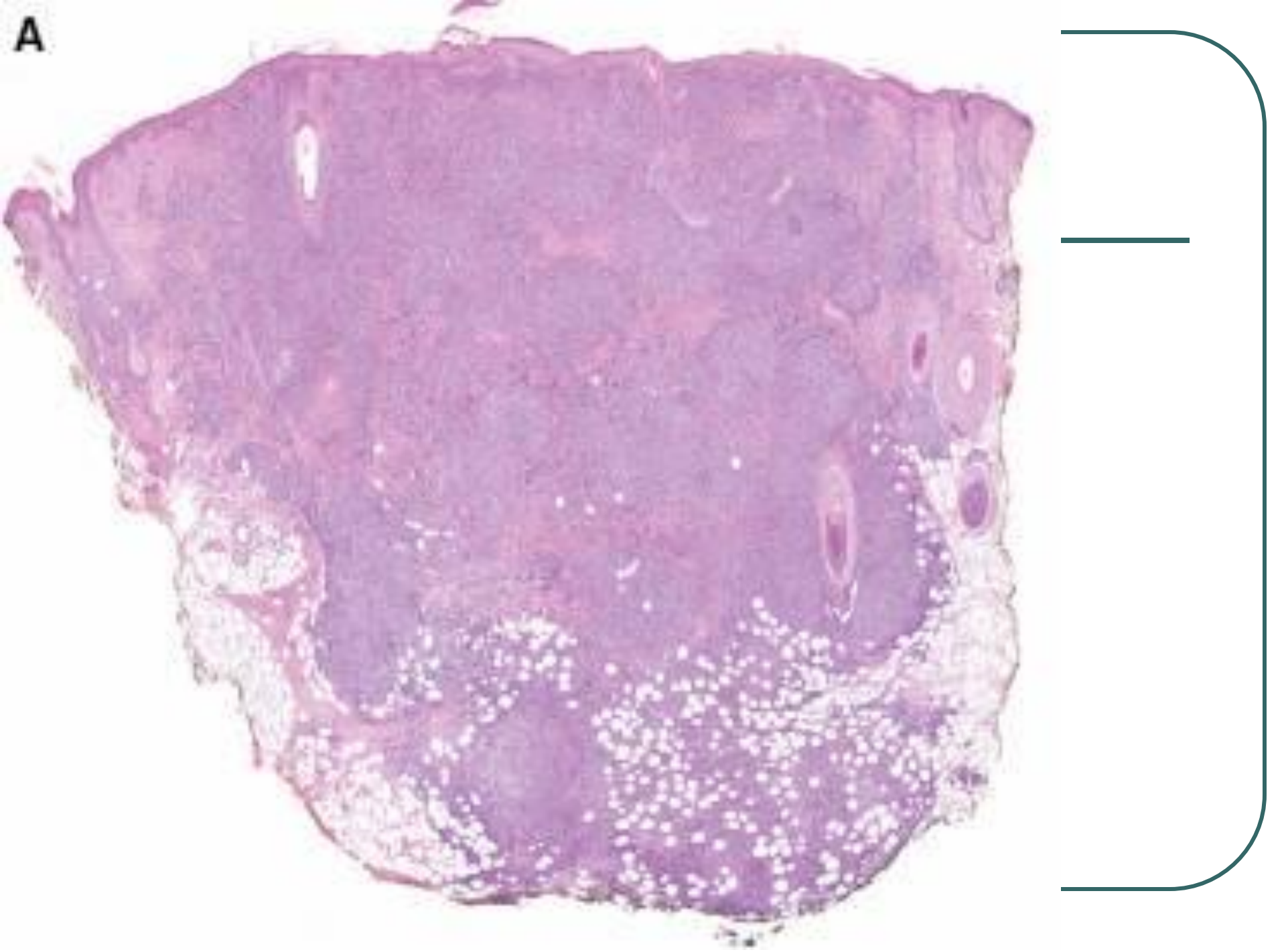


Lymphomatoid Papulosis

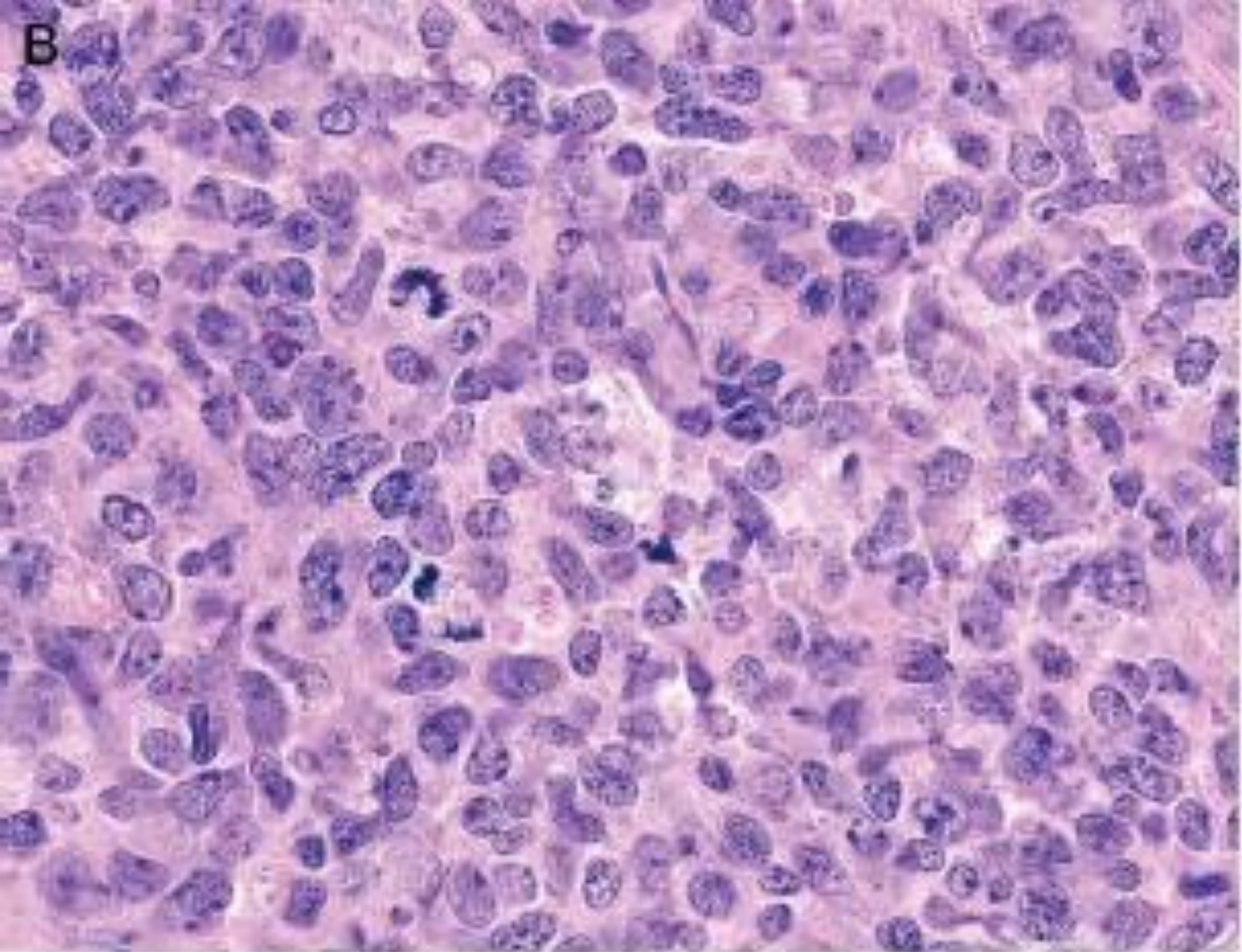
- Chronic recurrent, self-healing papulonecrotic or papulonodular skin eruption with histologic features of a malignant lymphoma.
- Histologically malignant but clinically benign
- Considered a low-grade cutaneous T-cell lymphoma
- Adults, median age 45, M:F 1.5, trunks and limbs
- Individual lesions disappear within 3 to 12 weeks
- 10-15% of the cases may be followed by a cutaneous lymphoma, usually MF
- 4% develop systemic lymphomas
- Px - 98% 5yr survival
- Tx – Long term follow up without active tx

Primary Cutaneous Anaplastic Large Cell Lymphoma

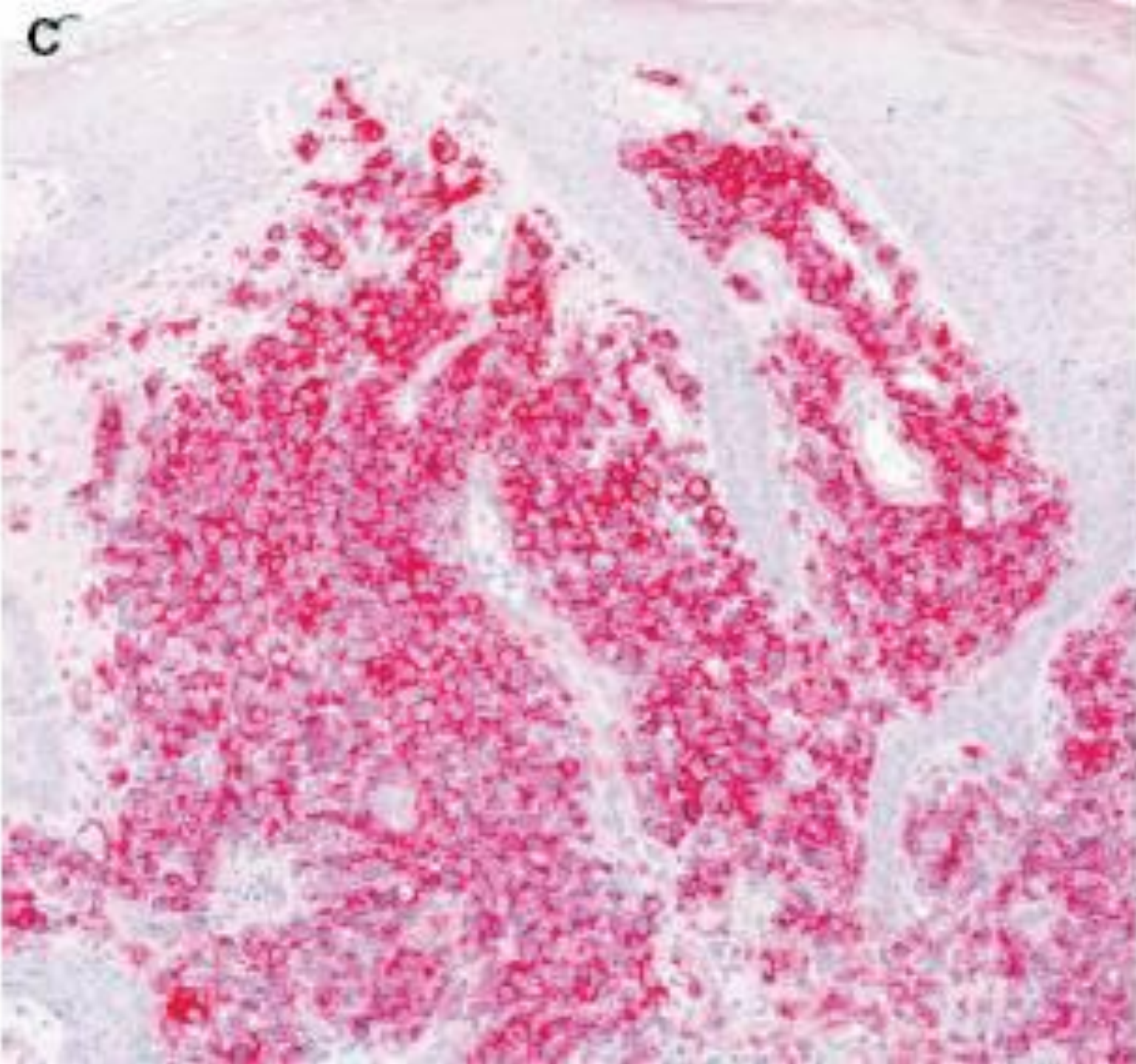
- Shares histologic and immunohistochemical features with nodal type.
- Differs in age of onset, genetic features, etiology, and prognosis.
- Presents as single or multiple grouped nodules or tumors confined to one extremity of body area.



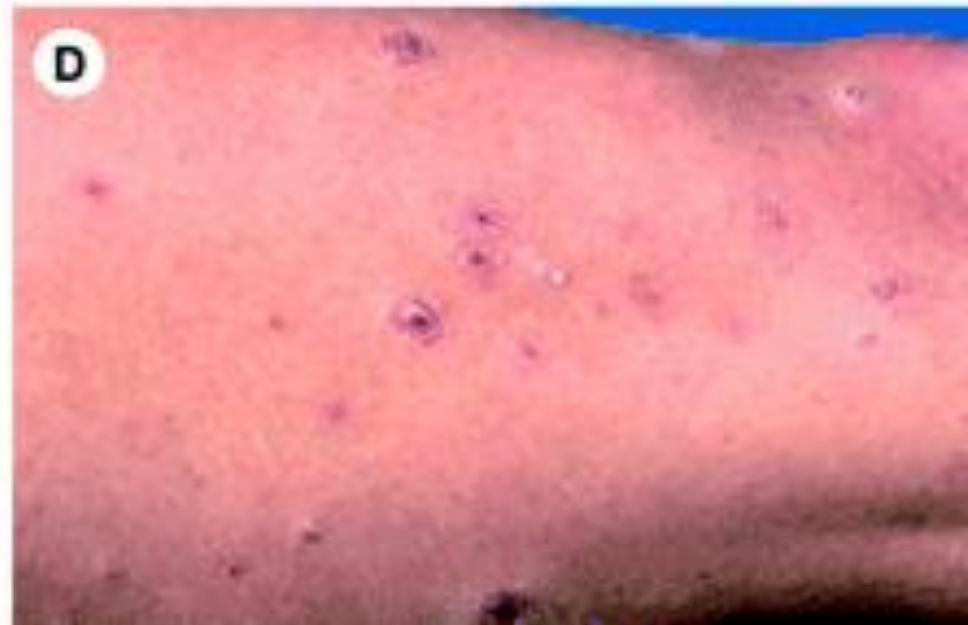
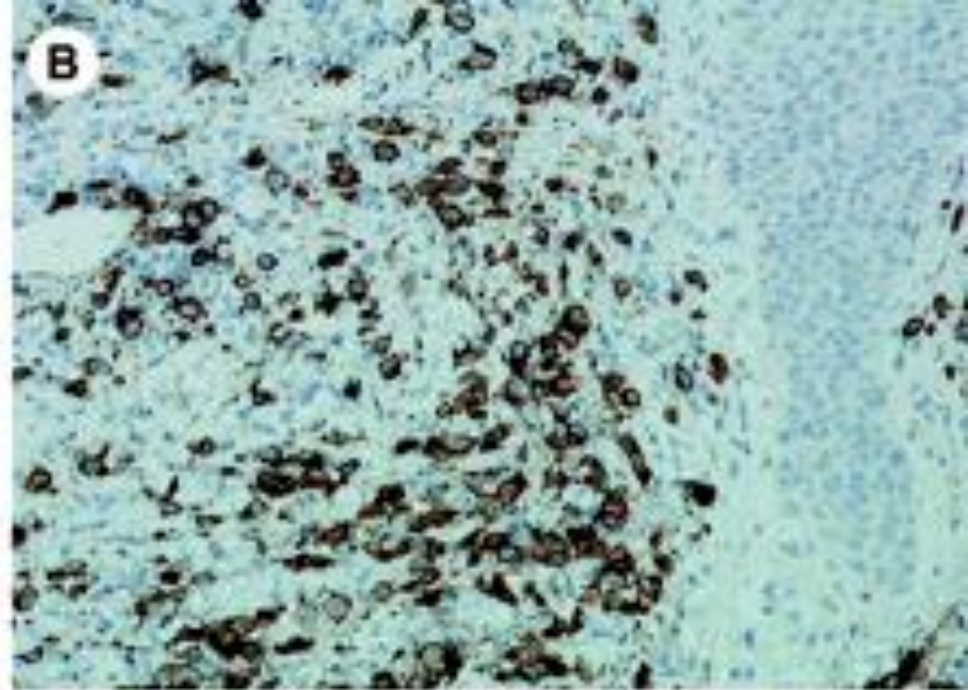
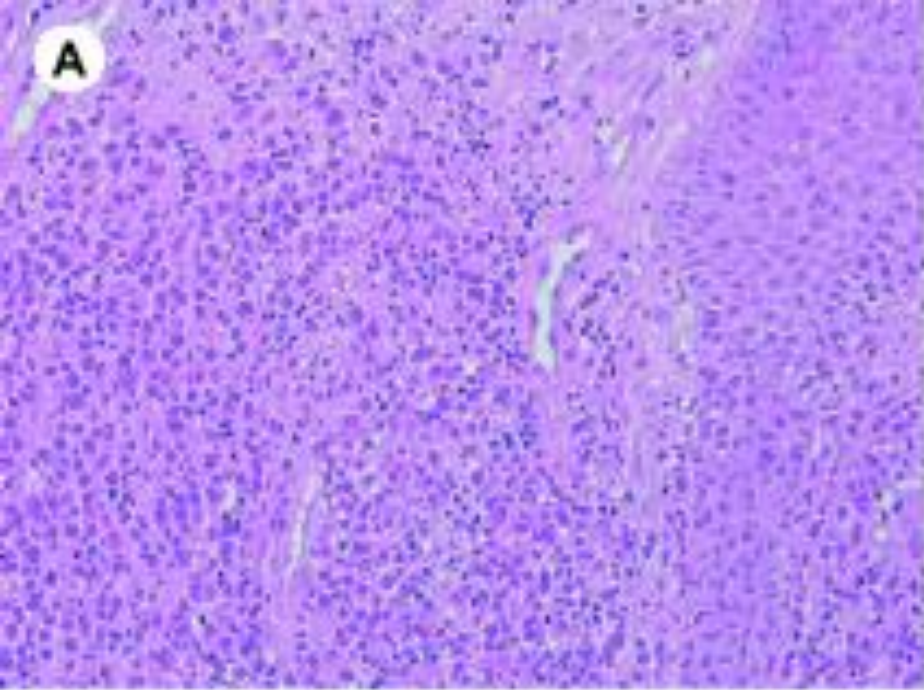
B



c



CD 30 +

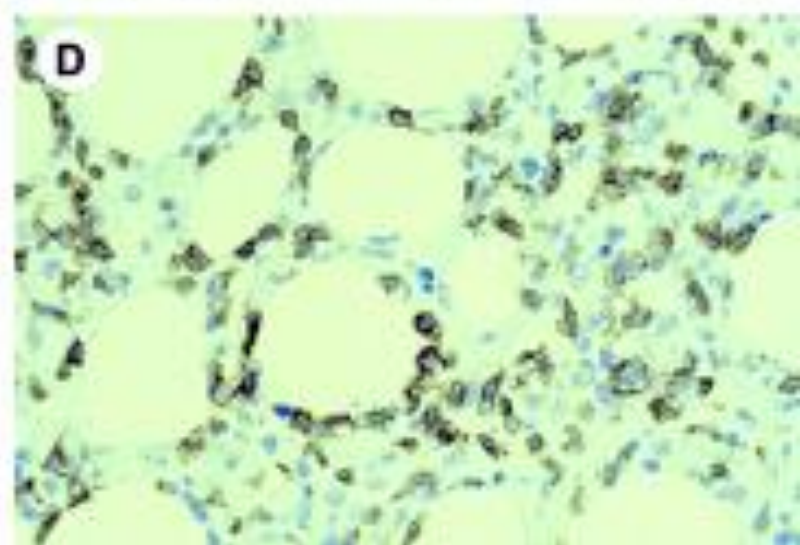
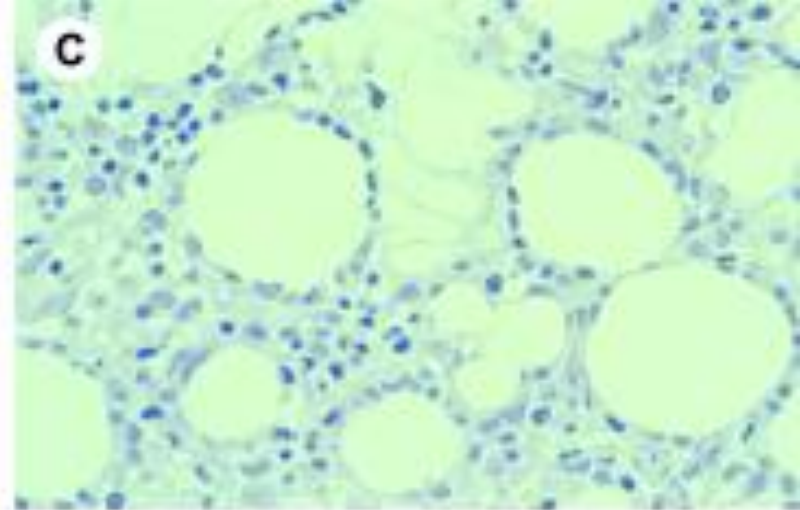
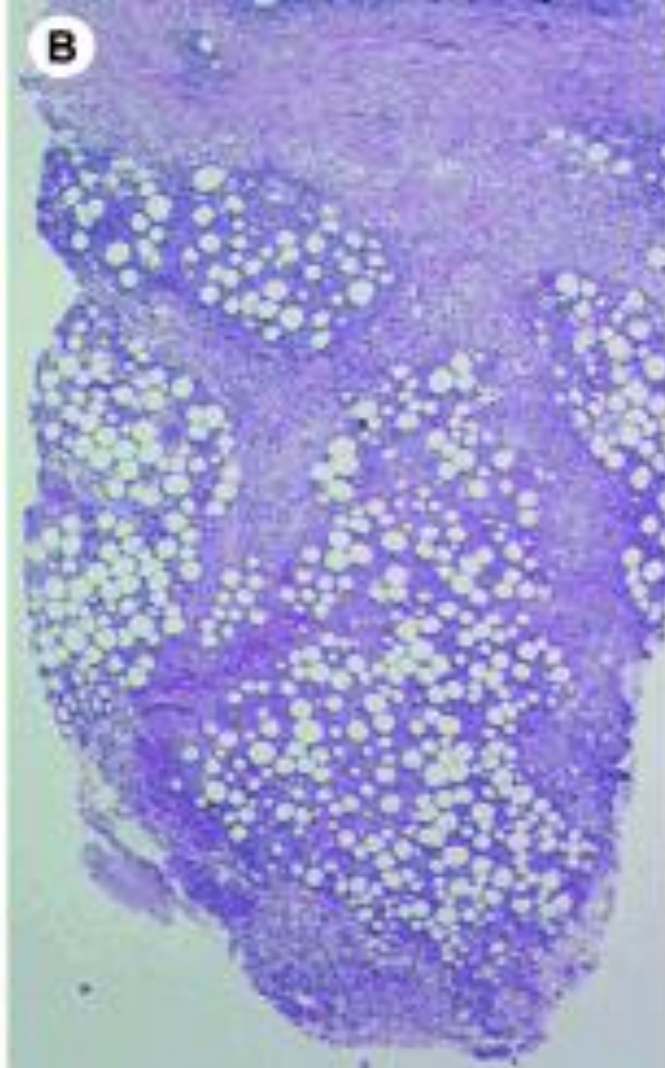


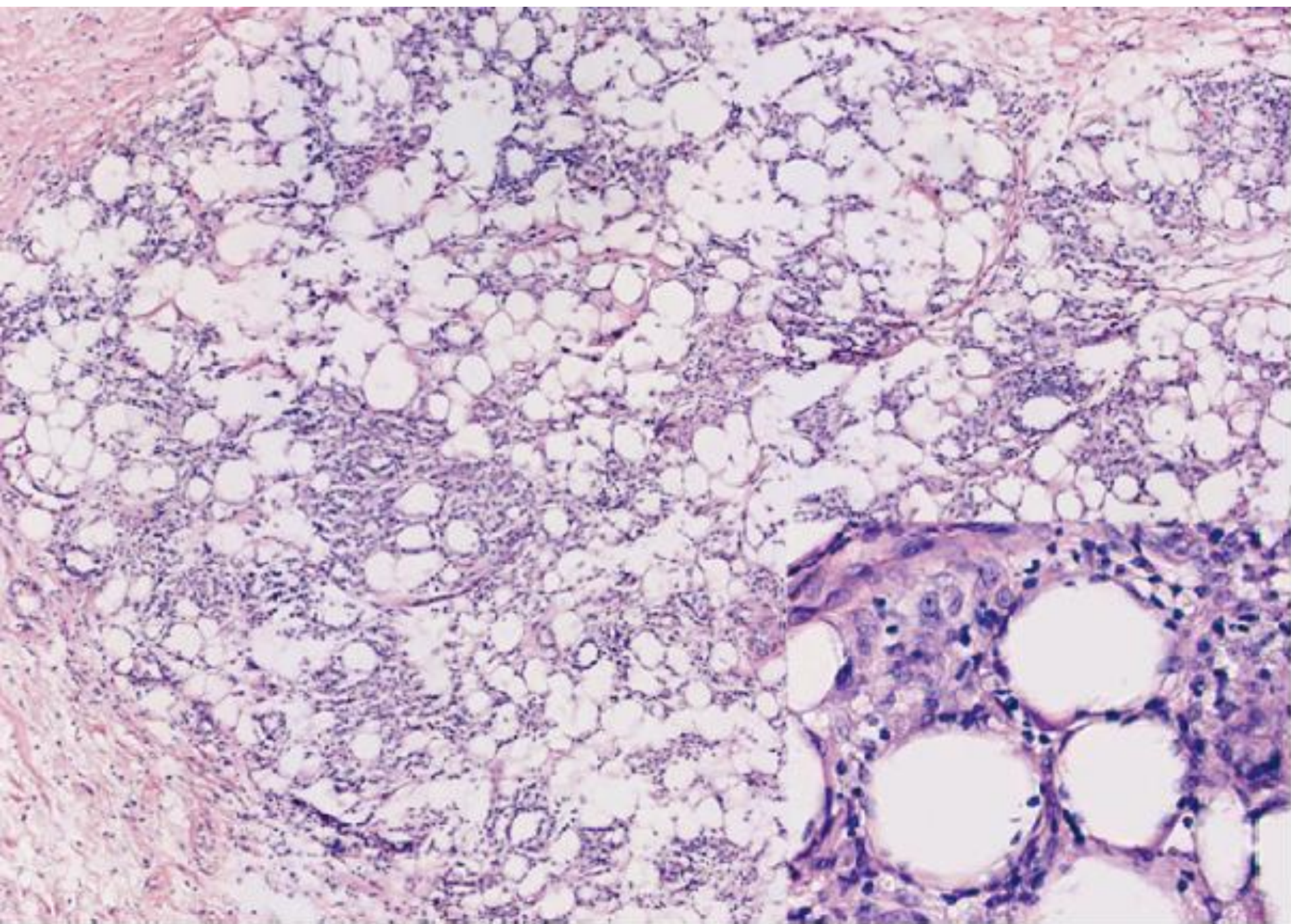
Primary Cutaneous Anaplastic Large Cell Lymphoma

- Large cells with an anaplastic, pleomorphic, or immunoblastic cytomorphology
- There is no clinical evidence or history of LyP, MF, or another type of CTCL
- CD30 antigen by more than 75% of tumor cells.
- Activated T cell phenotype, CD4+ , CD25, CD71, HLADR.
- Variable loss of CD2, CD3, and CD5
- CLA +, **EMA -**, **ALK-**
- **Lack translocation 2:5**, 90% have clonal rearrangement of TCR.
- **Px is excellent, 10 yr survival of over 90%**
- Tx radiotherapy or surgery

Subcutaneous panniculitis like T-cell lymphoma

- Adults and children, no sex predilection
- Most often on legs
- Systemic symptoms such as fever, fatigue, and weight loss may be present.
- Can be associated with hemophagocytic syndrome, rapidly progressive course.





Subcutaneous panniculitis like T-cell lymphoma

- Panniculitis like growth of T cells with hyperchromatic nuclei and often many macrophages.
- Overlying epidermis and dermis are typically uninvolved.
- Rimming of individual fat cells by neoplastic T cells
- Necrosis, karyorrhexis, and cytophagocytosis are common findings
- Plasma cells and reactive lymphoid follicles are generally absent, in contrast to lupus profundus, and other forms of lobular panniculitis.
- Cytotoxic T-cell lymphoma, subcutaneous infiltrates of pleomorphic T cells.

Subcutaneous panniculitis like T-cell lymphoma

- **α/β T cells**, CD 2+, CD3+, CD5+, CD4-, **CD8 +**, CD43 +, TIA-1 + granzyme B +, perforin +
- Neoplastic T cells show clonal T-cell receptor (TCR) gene rearrangements
- 5-year survival of 80%
- Tx Doxorubicin-based chemotherapy and radiotherapy

Primary cutaneous peripheral T-cell lymphoma, unspecified

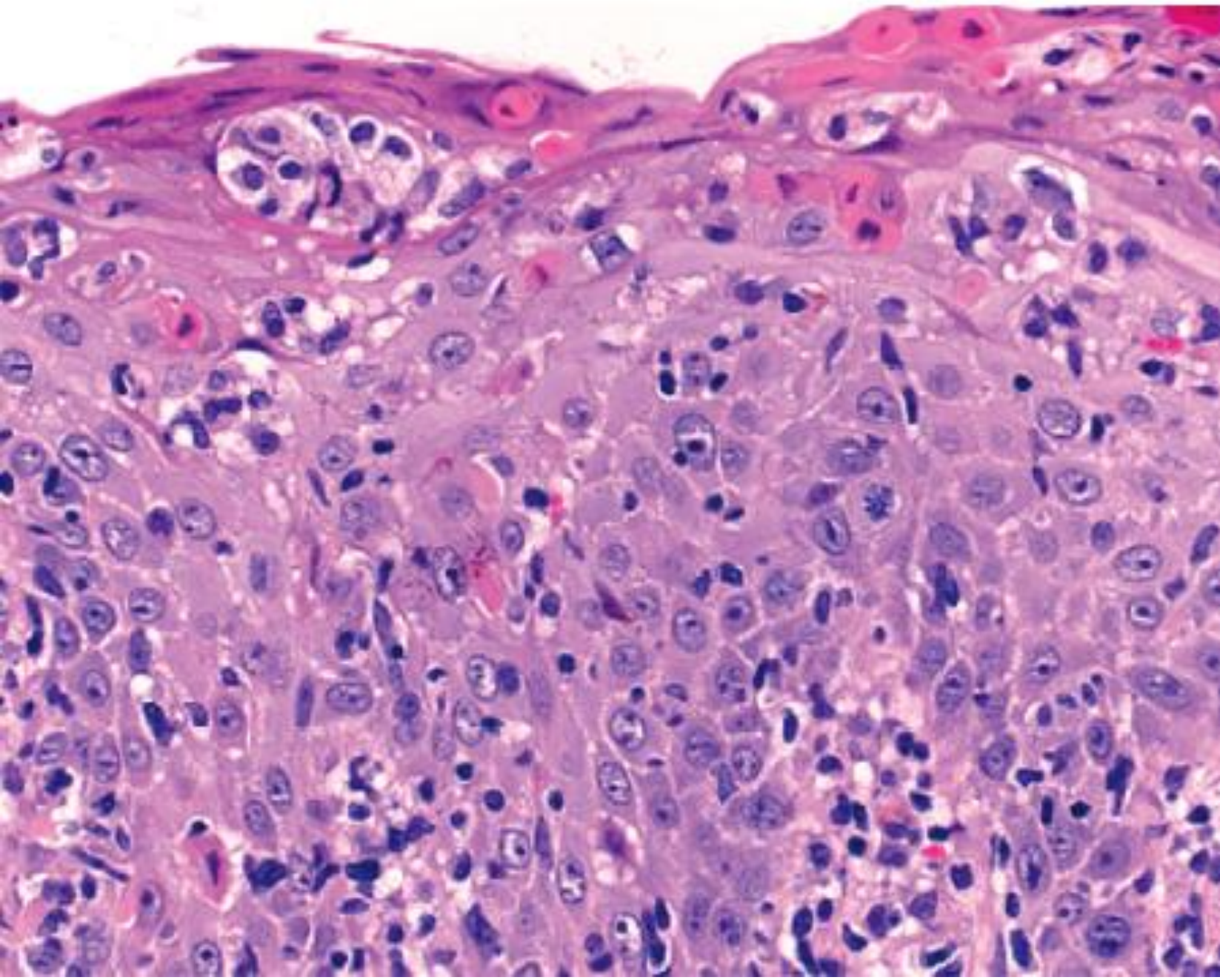
- T-cell neoplasms that do not fit into any of the better defined subtypes of T-cell lymphoma/leukemia .
- 10% of CTLs
- Cutaneous gamma/delta T-cell lymphoma
- Primary cutaneous aggressive epidermotropic CD8+ cytotoxic T-cell lymphoma
- Primary cutaneous CD4+ small/medium-sized pleomorphic T-cell lymphoma

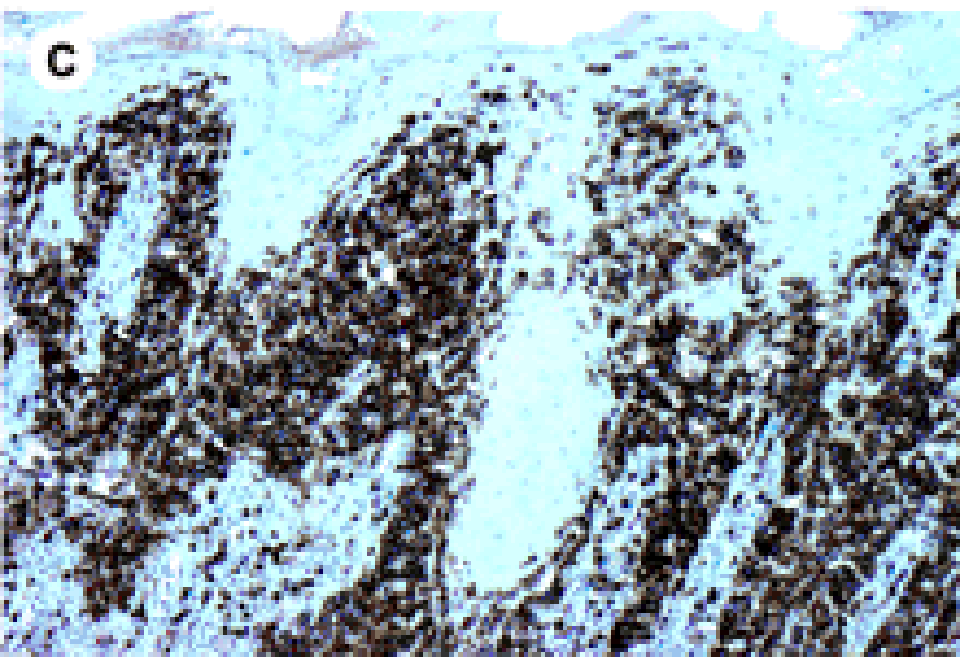
Cutaneous gamma/delta T-cell lymphoma

- Clonal proliferation of mature, activated γ/δ T cells expressing a cytotoxic phenotype.
- Epidermotropic, dermal, and subcutaneous.
- Dermal and epidermal involvement often coexists with subcutaneous disease, in contrast to SPTCL of α/β origin.
- Apoptosis and necrosis are common, often with vascular invasion.
- CD3+, CD2+, CD43+, CD45RO+,
- CD15-, CD30-, CD20-, CD25-
- CD4-, CD8 –
- Positive for TIA-1 and the cytotoxic proteins granzyme B, granzyme M, and perforin.

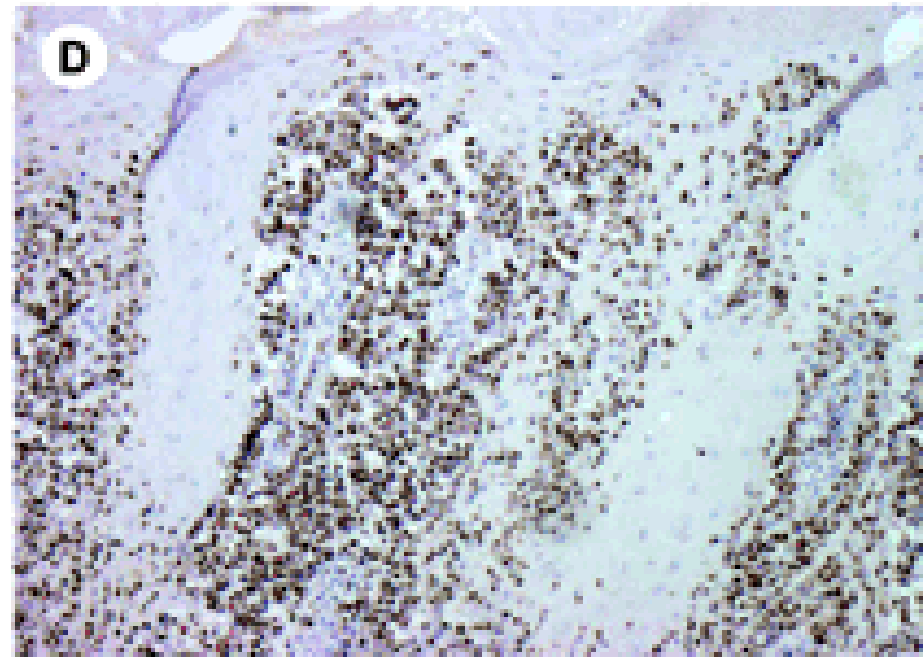
Primary cutaneous aggressive epidermotropic CD8+ cytotoxic T-cell lymphoma

- Clinically, this form of CD8+ cutaneous lymphoma differs from the slowly progressive CD8+ form similar to classic MF. It presents with erosive plaques rather than patches. It exhibits an unfavorable prognosis with rapid course .
- The tumor cells have a CD3+, CD8+, granzyme B+, perforin+, TIA-1+, CD45RA+, CD45RO-, CD2-, CD4-, CD5-
- The neoplastic T cells show clonal TCR gene rearrangements. Specific genetic abnormalities have not been described.
- Median survival of 32 months
- Patients are generally treated with doxorubicin-based multiagent chemotherapy.





CD 8+



TIA-1+

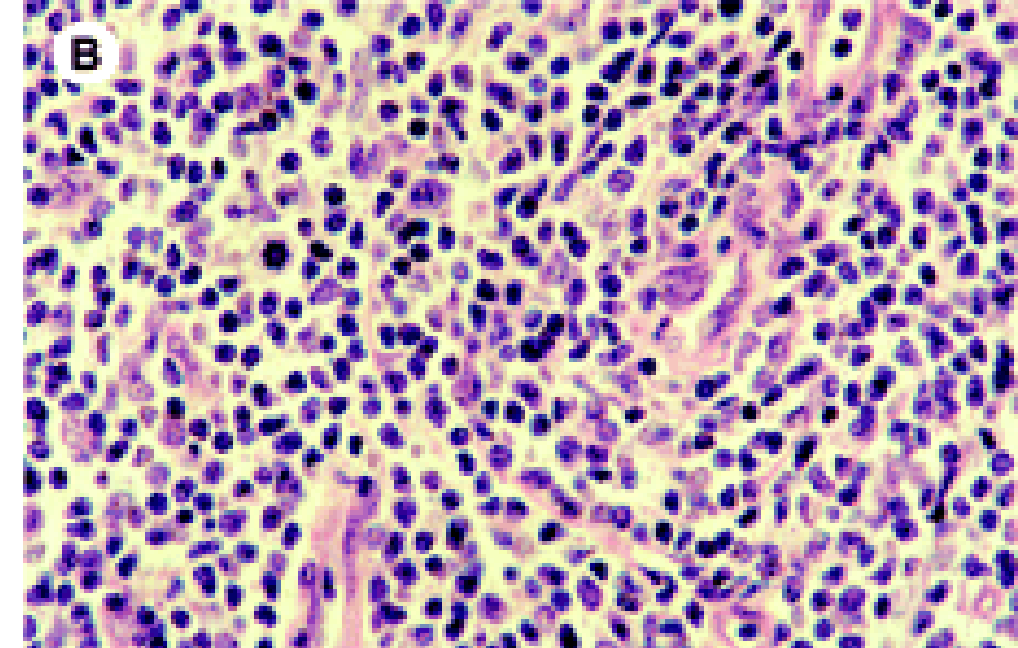
Primary cutaneous CD4+ small/medium-sized pleomorphic T-cell lymphoma

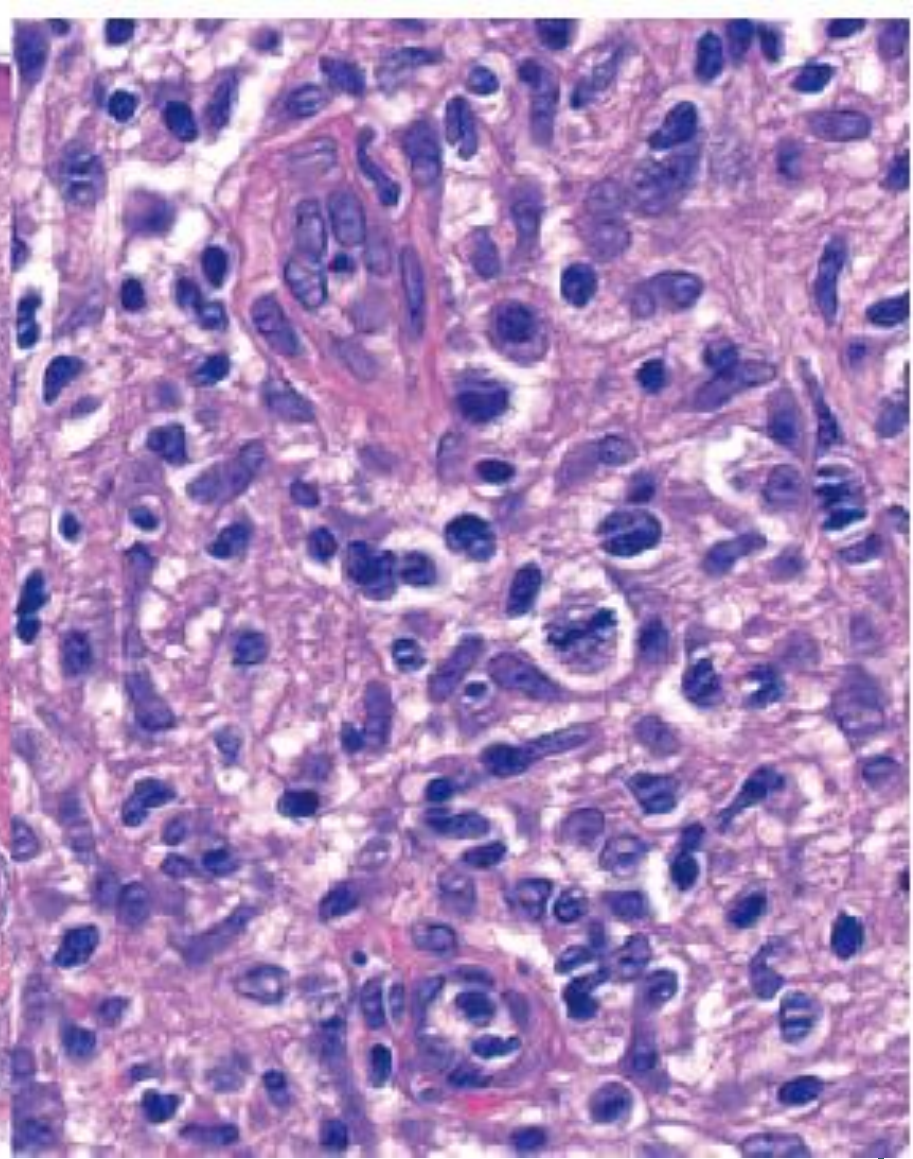
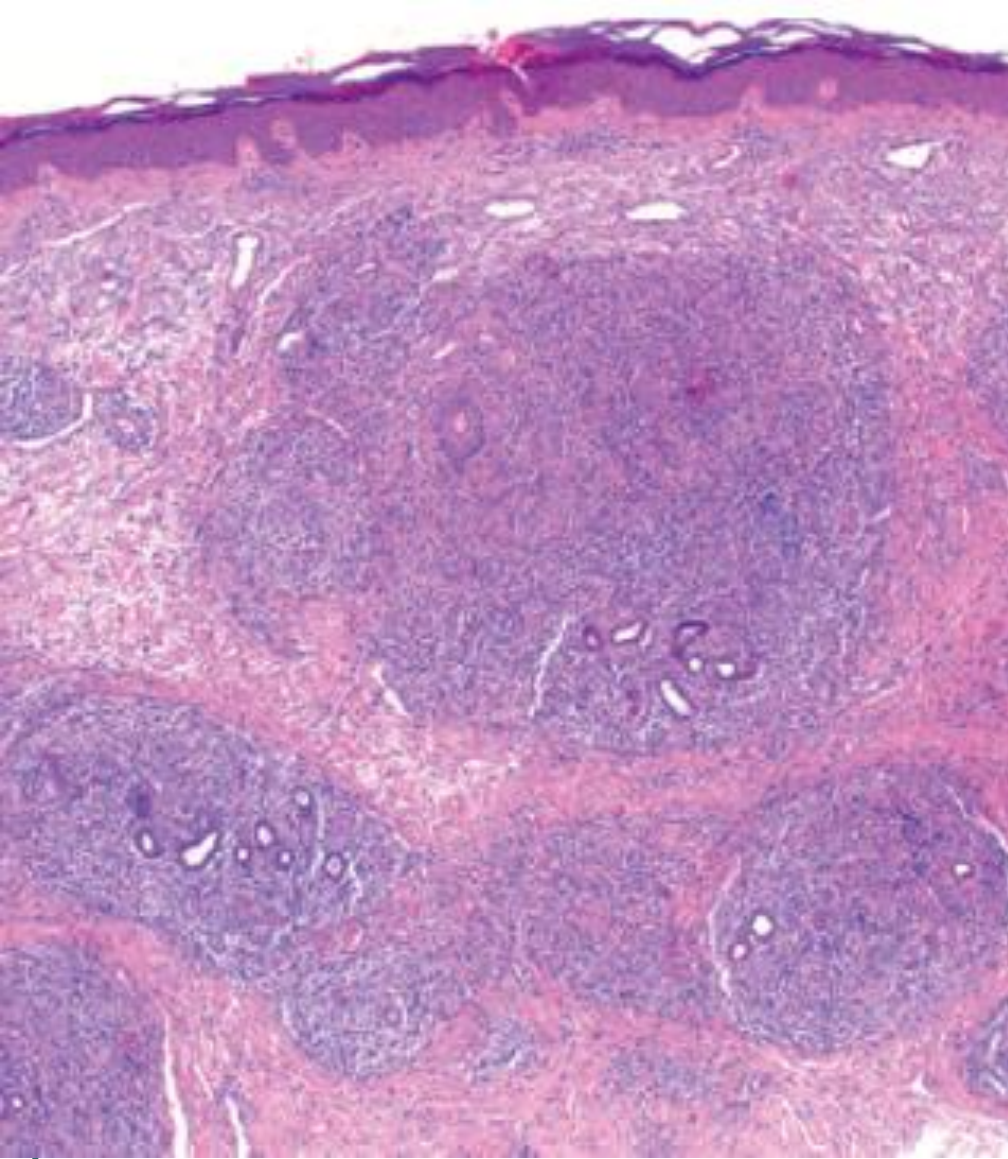
- This is a non-cytotoxic CTCL characterized by a predominance of small to medium-sized CD4+ pleomorphic T cells with clinical features not compatible with MF.
- Diffuse or nodular lymphoid infiltrate, predominantly perivascular and periadnexal and shows a tendency to extend to the subcutaneous tissue.
- It consists of small-to-medium-sized pleomorphic lymphoid cells with irregular hyperchromatic nuclei and a pale scanty cytoplasm
- Patients are commonly adults, who present with solitary, localized, or more frequently generalized nodules or tumors. No sites of predilection have been recorded

A



B





Cutaneous B-cell lymphomas

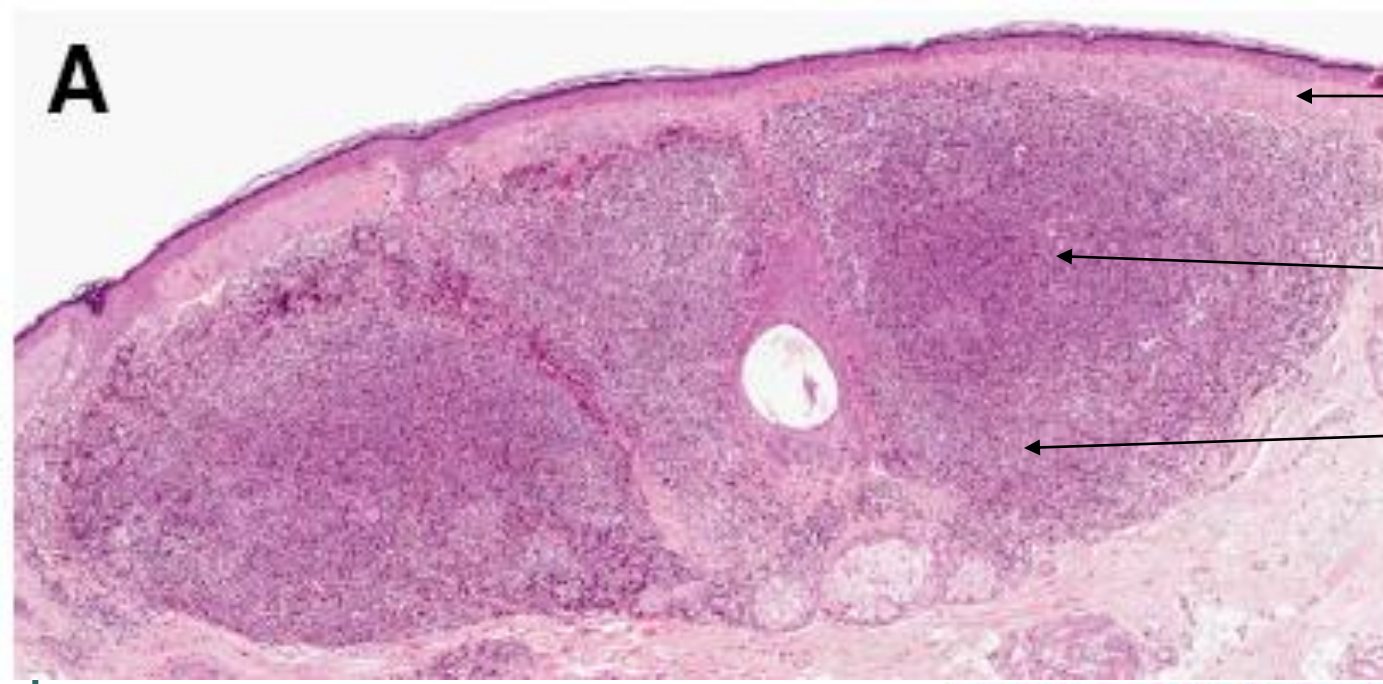
- Cutaneous marginal zone B cell lymphoma
- Primary cutaneous follicle center lymphoma
- Cutaneous large B cell lymphoma, leg type
- Cutaneous large B cell lymphoma, other
- Cutaneous intravascular large B cell lymphoma

Cutaneous marginal zone B-cell lymphoma (MZL)

- Adults trunk or extremities, especially the arms
- The nodular or diffuse infiltrate
- Small to medium-sized lymphocytes
- Irregular nuclei, dispersed chromatin, inconspicuous nucleoli and an abundant pale cytoplasm
- Monocytoid appearance (reniform nuclei) or plasma cell differentiation.
- Typified by darker centers surrounded by brighter zones of pale-staining cells.
- Dutcher bodies and intracytoplasmic PAS+ globular inclusions may be seen



A



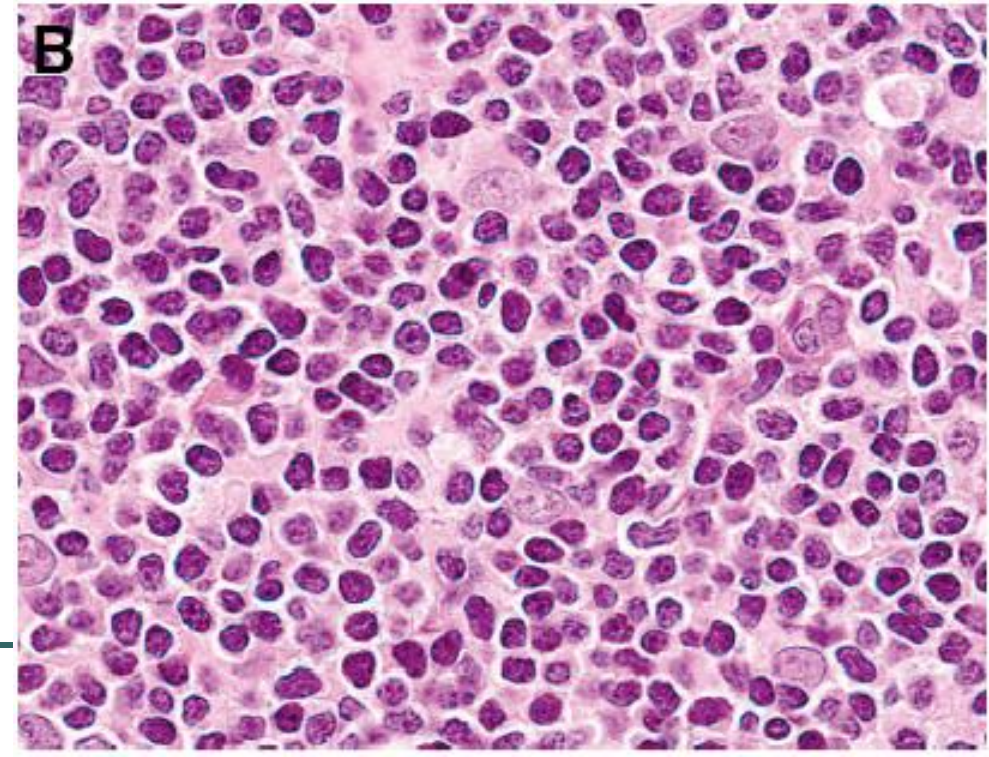
Grenz zone

Darker center

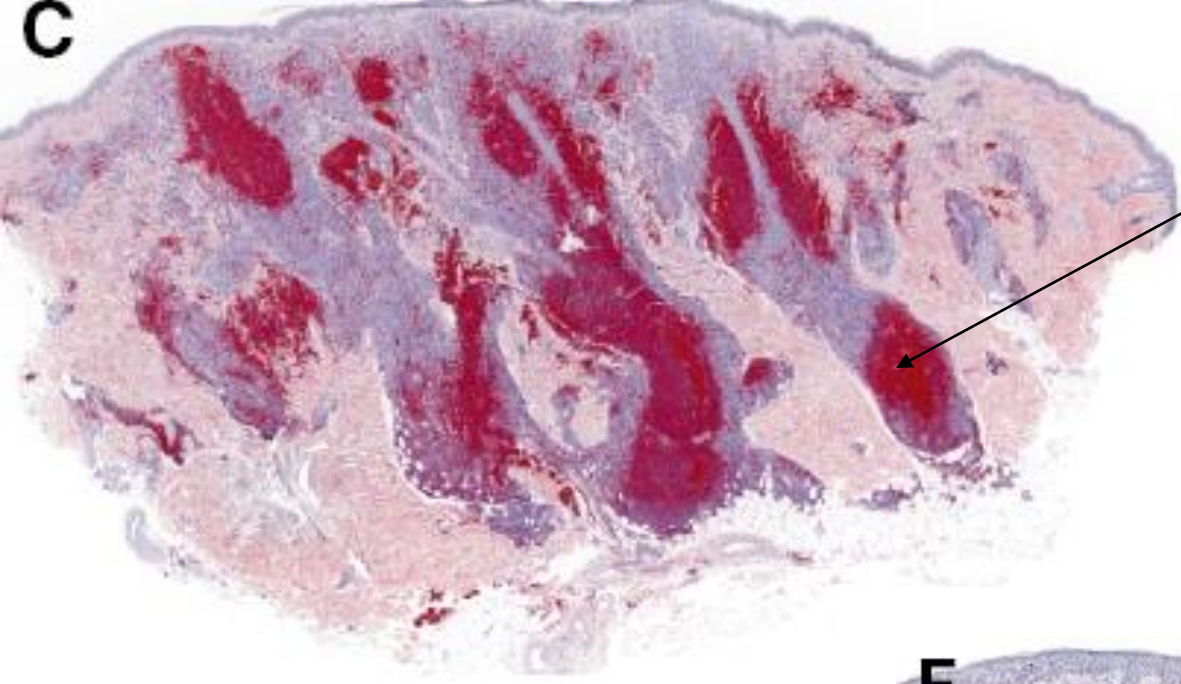
Lighter Periphery

Monocytoid lymphocytes

B

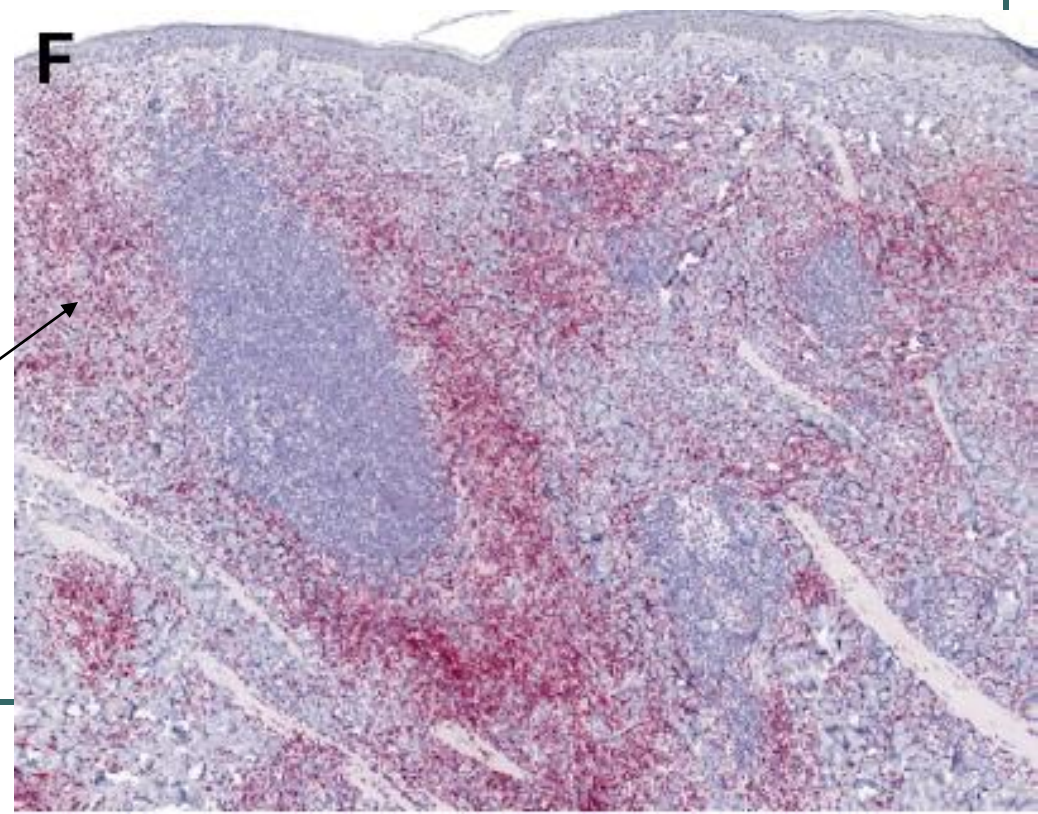


C



CD20 + Neoplastic cells

F



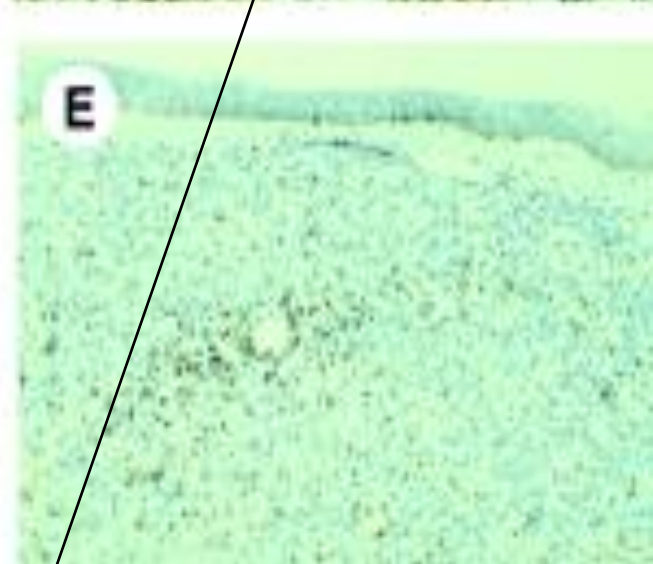
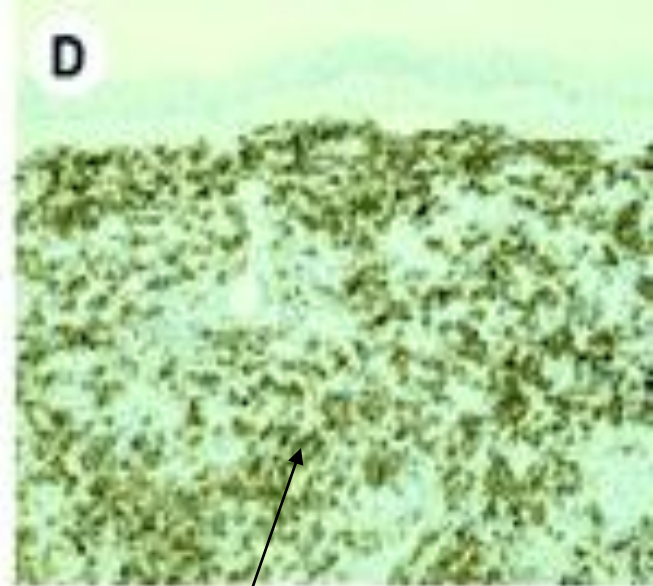
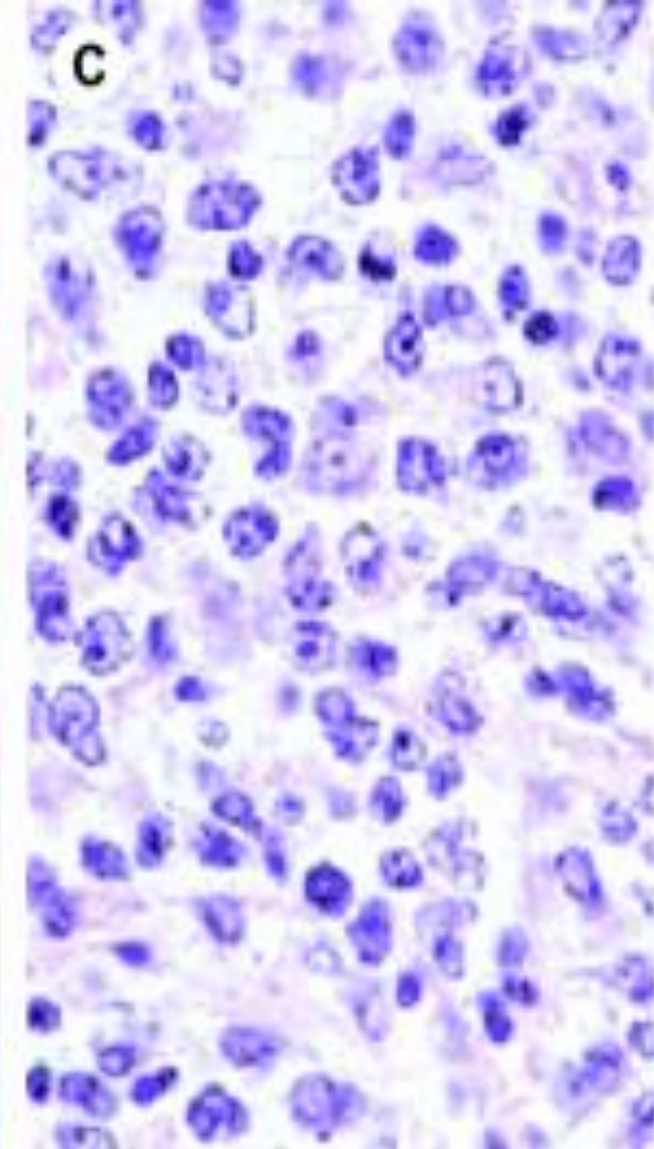
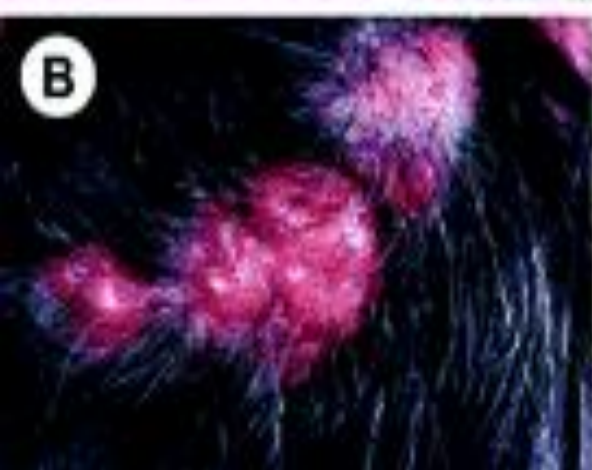
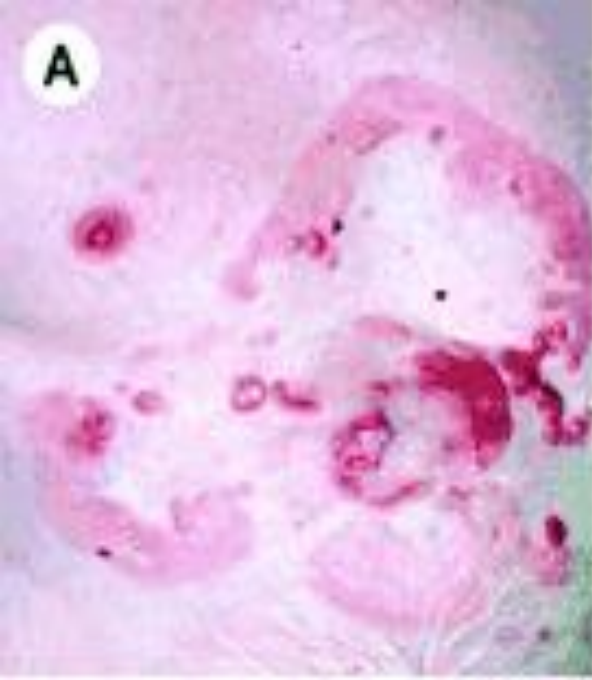
CD 3 + Reactive T cells

Cutaneous marginal zone B-cell lymphoma (CMZL)

- immunophenotype: CD19+, CD20+, CD22+, CD79a+, bcl-2+, CD5-, CD10-, CD23-, bcl-6-
- IgH genes are clonally rearranged in >70% of cases
- MZL, the t(11;18) **not seen** in primary cutaneous MZL
- t(14;18)(q32;q21) translocation involving IGH and MALT1 seen in 1/3 of cases.
- The prognosis of CMZL is excellent with a 5-year survival close to 100%.
- Tx radiotherapy or surgical excision

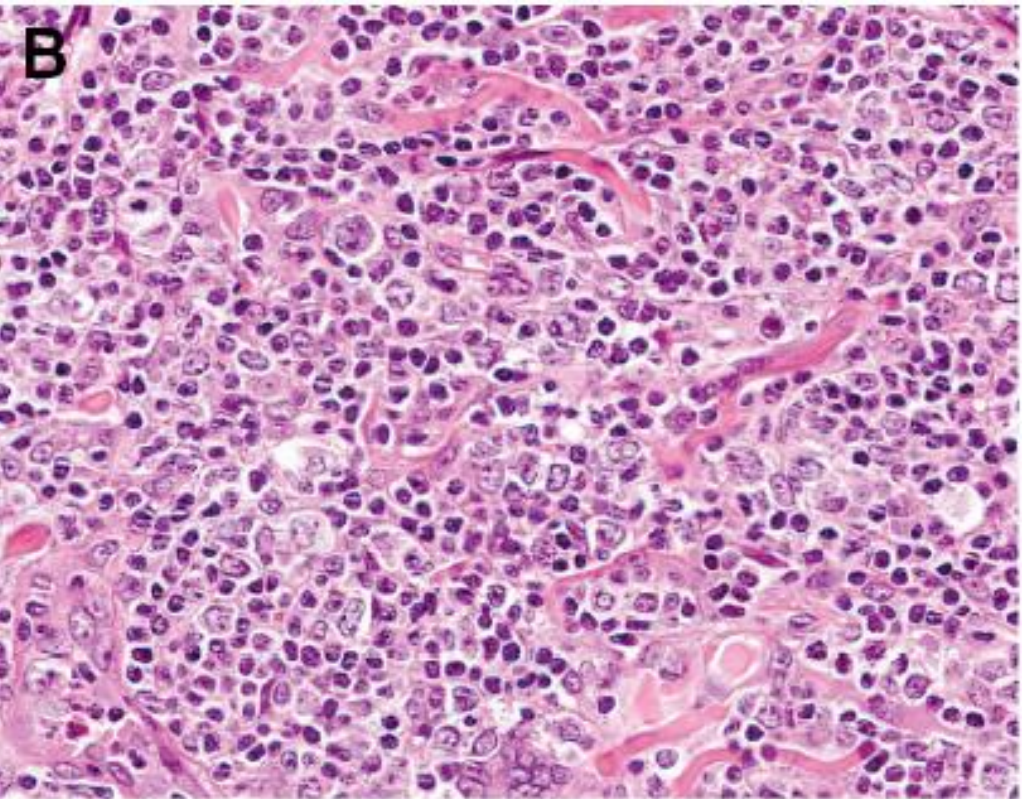
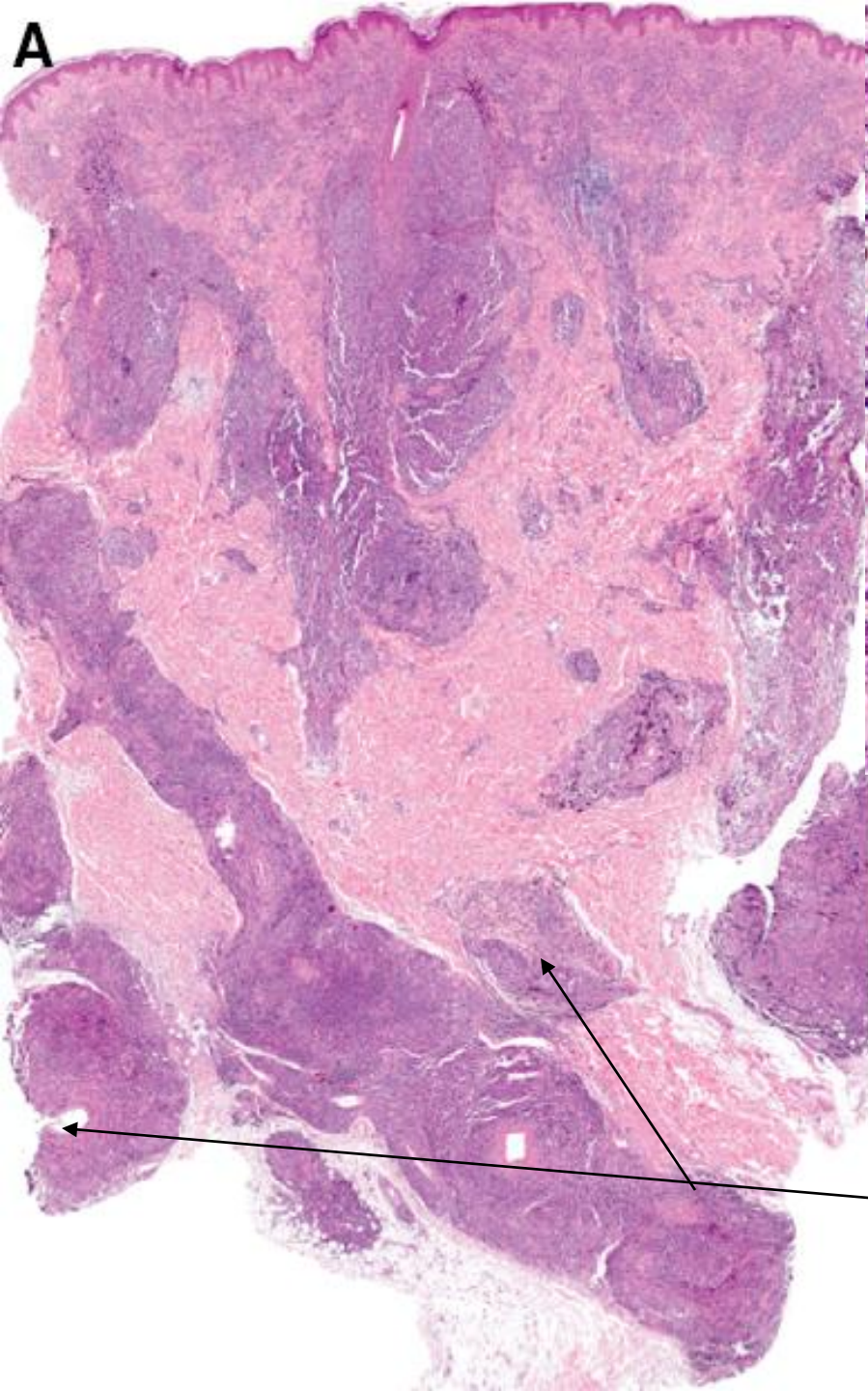
Primary Cutaneous Follicle Center Lymphoma

- Solitary or grouped plaques and tumors
- Scalp, forehead, and trunk
- Synonyms, reticulohistiocytoma of back, or Crosti lymphoma
- The skin lesions gradually increase in size over years, but dissemination to extracutaneous sites is uncommon.



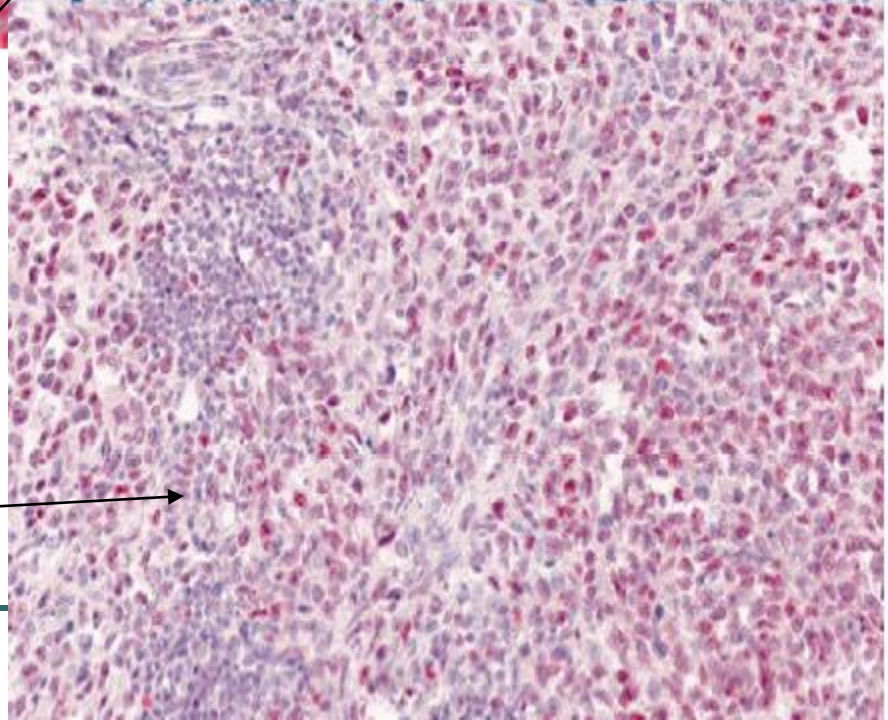
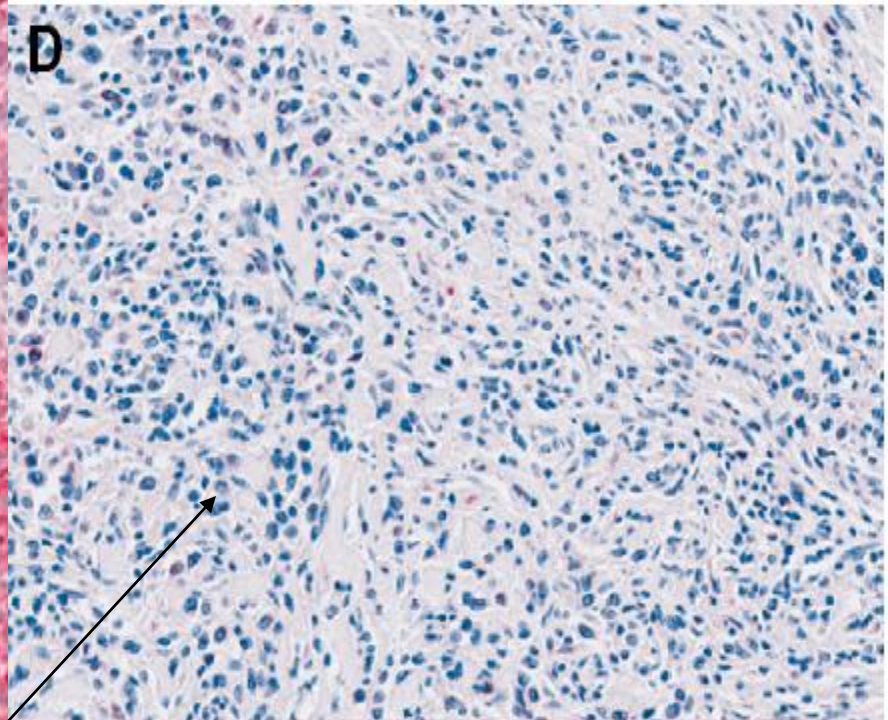
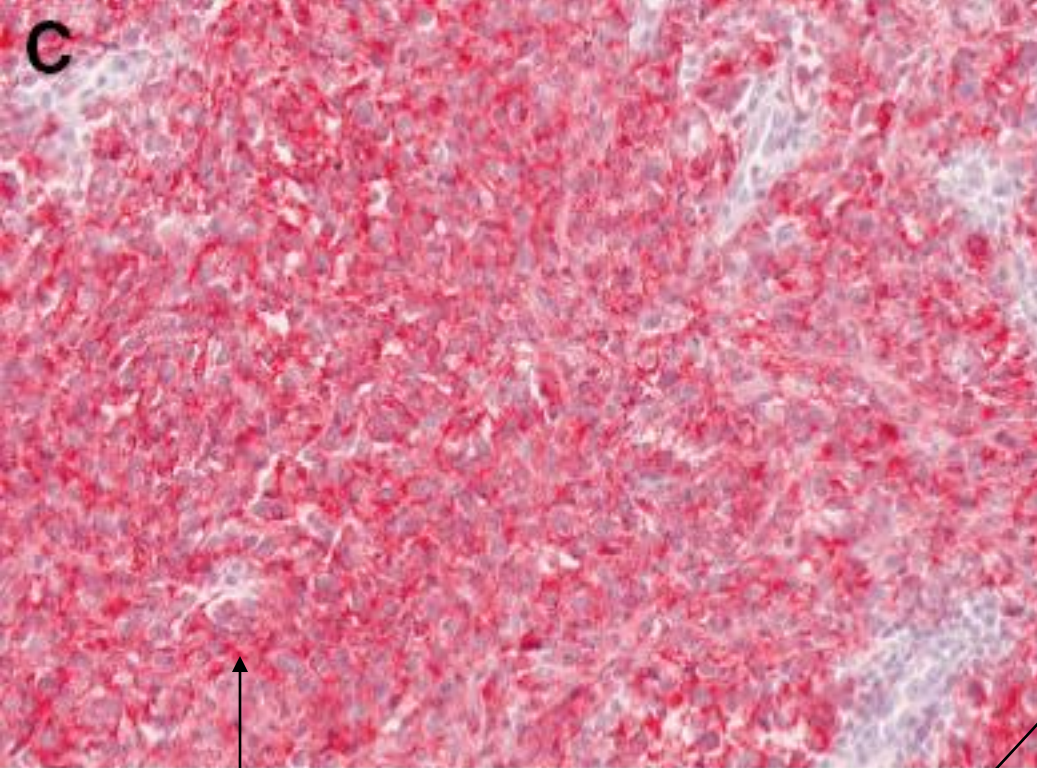
CD 20 +

Bcl-2 -



Mixed population of centrocyte like and centroblast like cells

Nodular infiltrate



CD 20 +

Bcl-2 negative

Bcl-6 positive

Primary Cutaneous Follicle Center Lymphoma

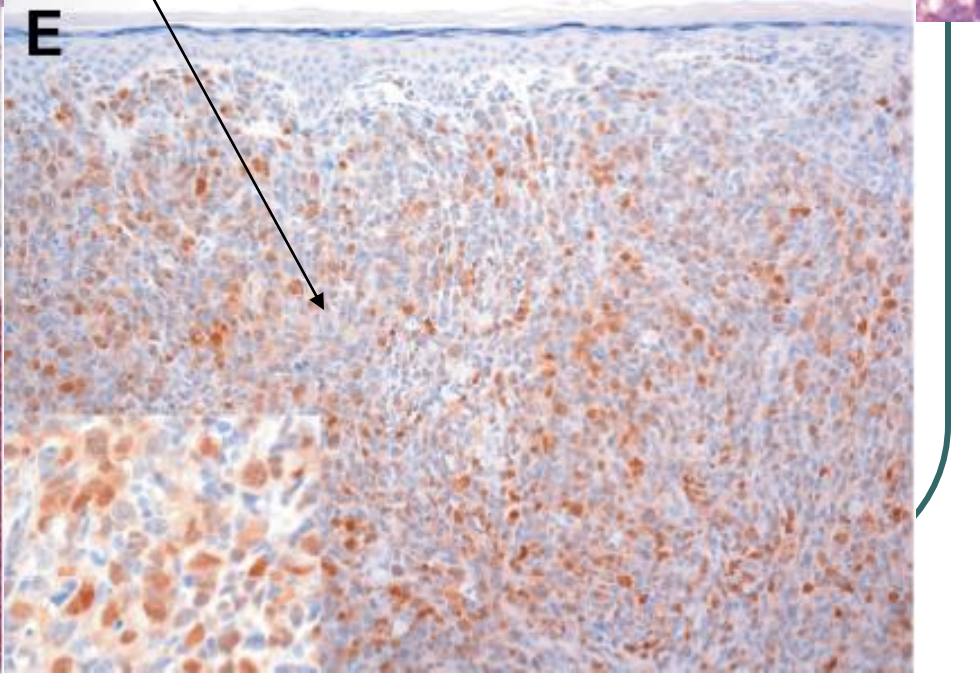
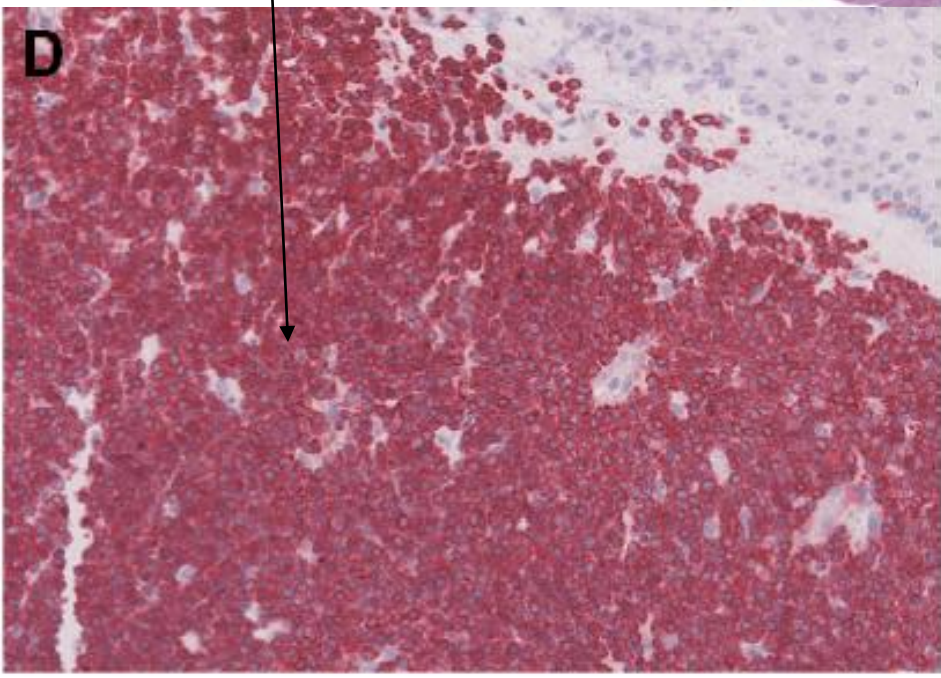
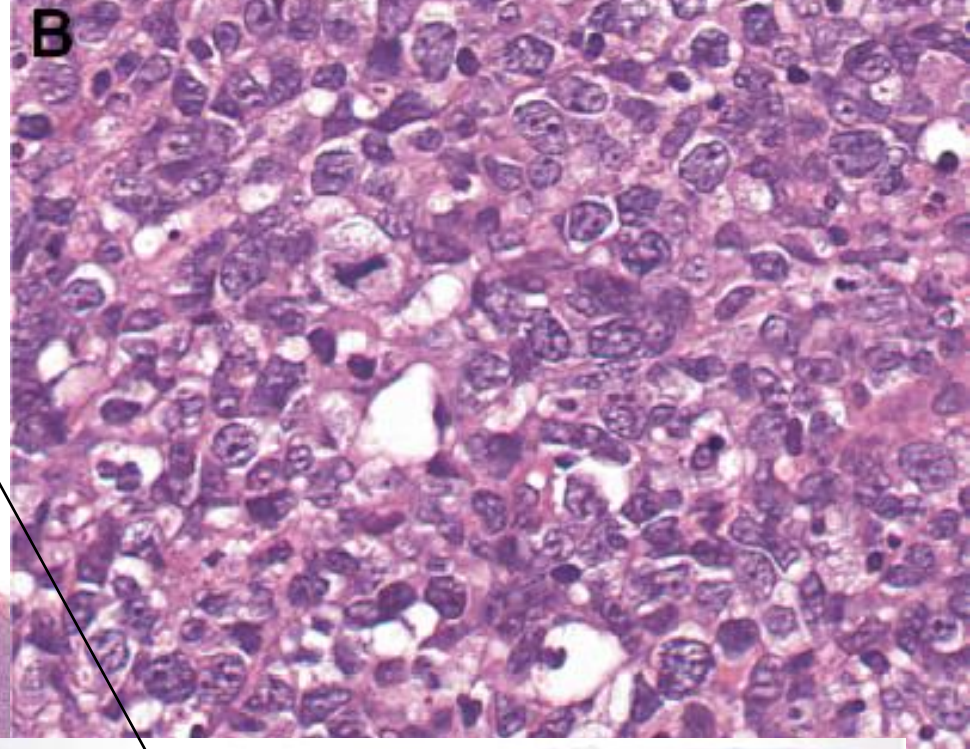
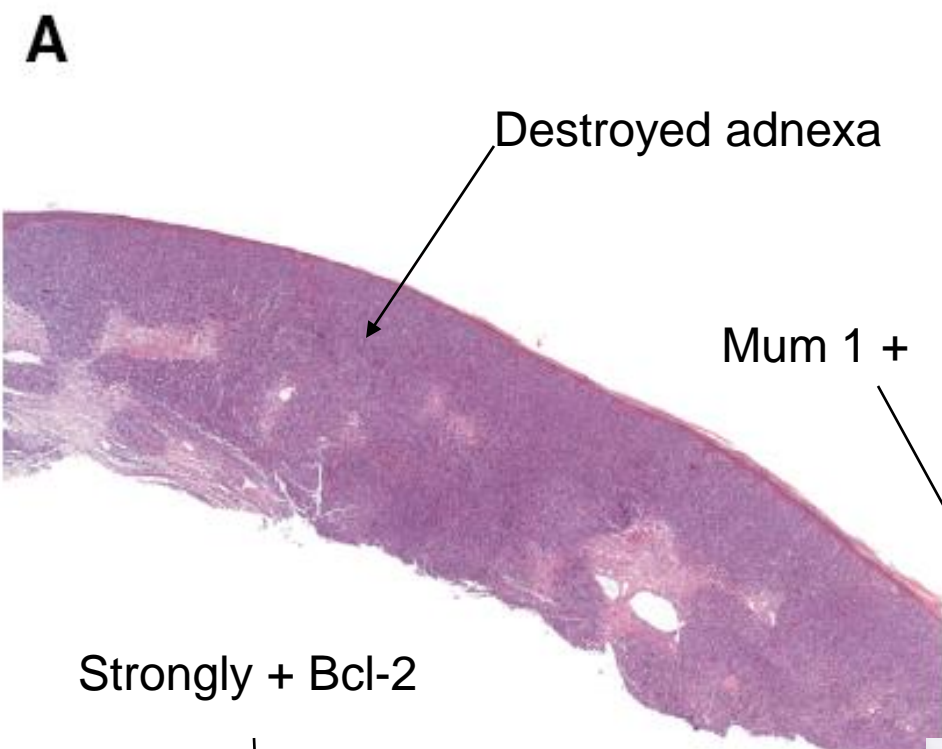
- Neoplasm of follicle center cells (centrocytes and centroblasts)
- Follicular, follicular and diffuse or a diffuse growth pattern
- Grading of primary cutaneous FCL as in its nodal counterpart based is not prognostically relevant.
- Few mitosis, and no tingible body macrophages seen (present in pseudolymphoma)
- CD19+, CD20+, CD22+, CD79a+, CD5-, CD43 -, bcl-2 -, bcl-6 +
- Bcl-2 gene rearrangement and t(14;18) chromosomal translocation are absent in most cases.
- 5-year survival of more than 95%
- Tx radiotherapy or surgical excision

Cutaneous diffuse large B-cell lymphoma (DLBCL)

- composed of large B cells (centroblasts and immunoblasts)
- DLBCL, leg-type and DLBCL, other
- DLBCL, leg type, is the most common variant, occurs on the leg and less frequently at other sites.
- DLCL other include; Tcell/histiocyte-rich DLBCL, plasmablastic lymphoma and others that do not fulfill the criteria for a DLBCL, leg-type

Diffuse large B-cell lymphoma (DLBCL), leg-type Histology

- Diffuse growth pattern, monomorphous infiltrate, entire dermis involvement, adnexal structures are usually destroyed
- The epidermis is often spared, Grenz zone
- Centrocytes are absent
- Mitotic figures can frequently be detected
- Nuclei are round with coarsely clumped chromatin.
- Minimal inflammatory component and little stromal reaction.

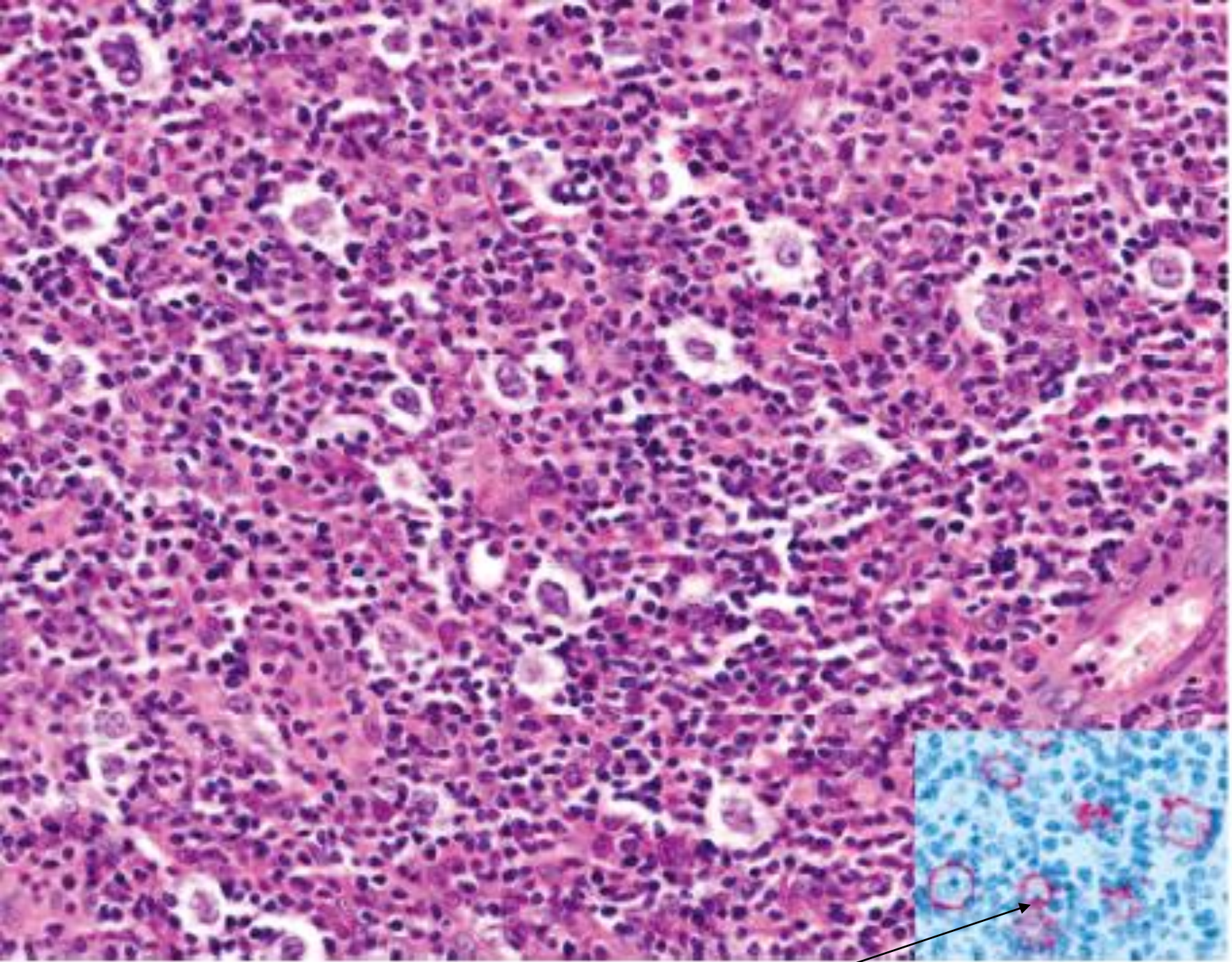


Diffuse large B-cell lymphoma (DLBCL), leg-type

- CD19+, CD20+, CD22+, and CD79a+ CD10-, Bcl-6 + in most cases.
- Combination + Bcl-2, and + MUM-1/IRF4 is characteristic regardless of site and distinguish from FCL diffuse type.
- The t(14;18) can be detected in **secondary cutaneous large B-cell lymphomas** but not in primary cutaneous diffuse large B-cell lymphomas.
- The 5-year survival 55%
- PCLBCLs on the leg have an inferior prognosis compared to PCLBCLs presenting at other
- Tx Radiotherapy and Rituximab

Diffuse large B-cell lymphoma, other

- Neoplastic large B-cells that lack the typical features of DLBCL, leg-type and do not conform to the definition of primary cutaneous FCL with diffuse growth pattern.
- T cell rich, histiocyte rich, plasmablastic (HIV) variants
- CD19+, CD22 +, and CD79a+, with light-chain restriction, negative for CD15 and CD30, which excludes Hodgkin lymphoma.
- 5 yr survival 65%
- Tx Radiotherapy and Rituximab



CD20 +



Intravascular large B-cell lymphoma (IVL)

- Rare highly malignant large-cell lymphoma with systemic spread

- Tumor cells in the lumina of small vessels, particularly capillaries and venules.

- **Skin and the nervous system are preferential sites of primary manifestation.**

- The tumor cells express B-cell markers in the vast majority of cases; rarely a T-cell phenotype is found

- **Patients with disease limited to the skin (cutaneous variant) have a significantly better outcome than the other patients with IVL. 3-year overall survival: 56% versus 22%**

- Tx Multiagent chemotherapy

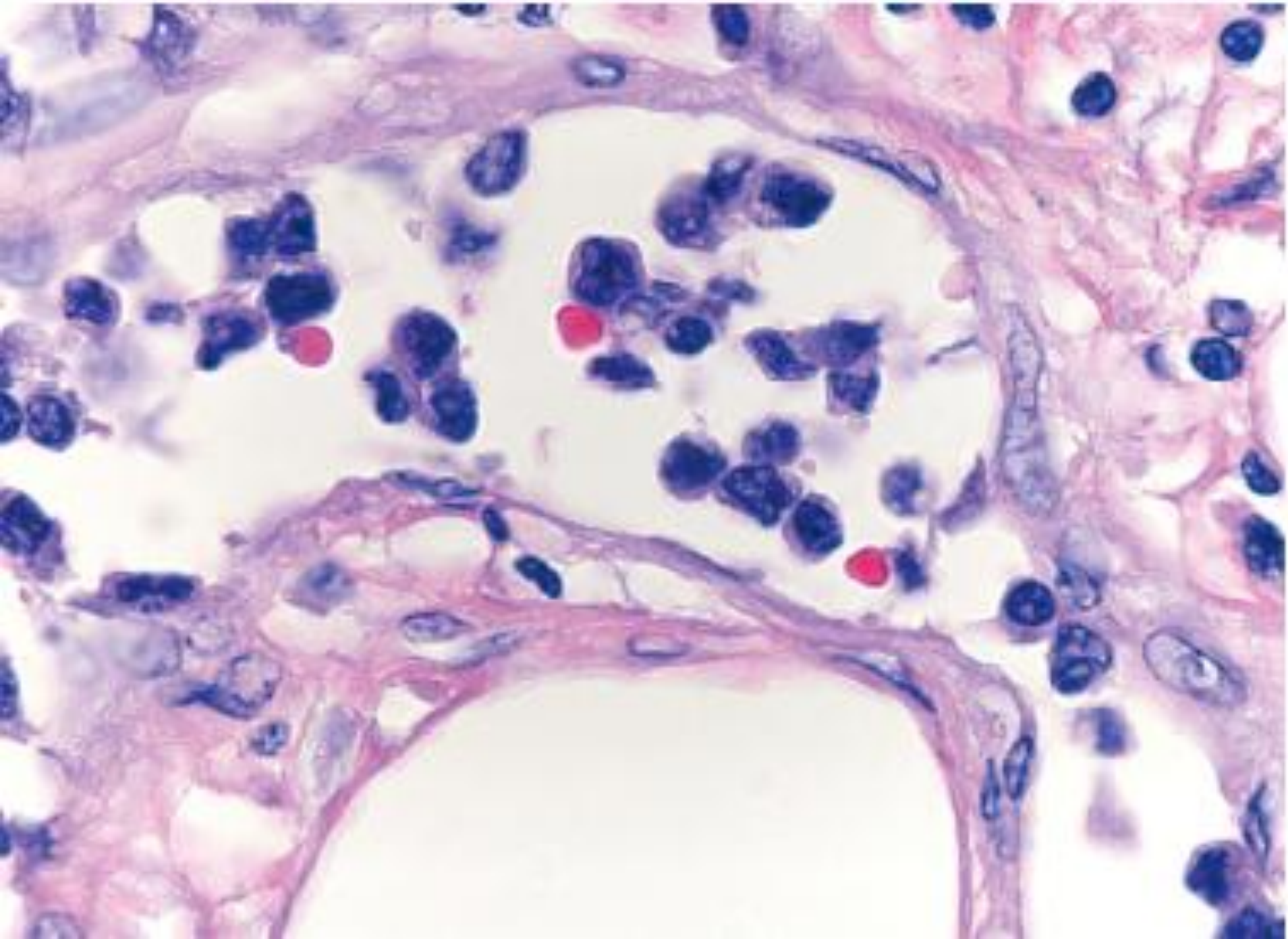


Table 3. Summary on phenotypical features of cutaneous B-cell lymphomas and pseudolymphomas

	Bcl-2	Bcl-6	CD10	(14; 18)	MUM1/IRF4
MZL/ICY	+	-	-	-	-
FCL	-	+	+/-	-	-
Secondary FCL	+	+	+	+	-
DLBCCL	+	+	-	-/+	+
PSL	-	+	+	-	-

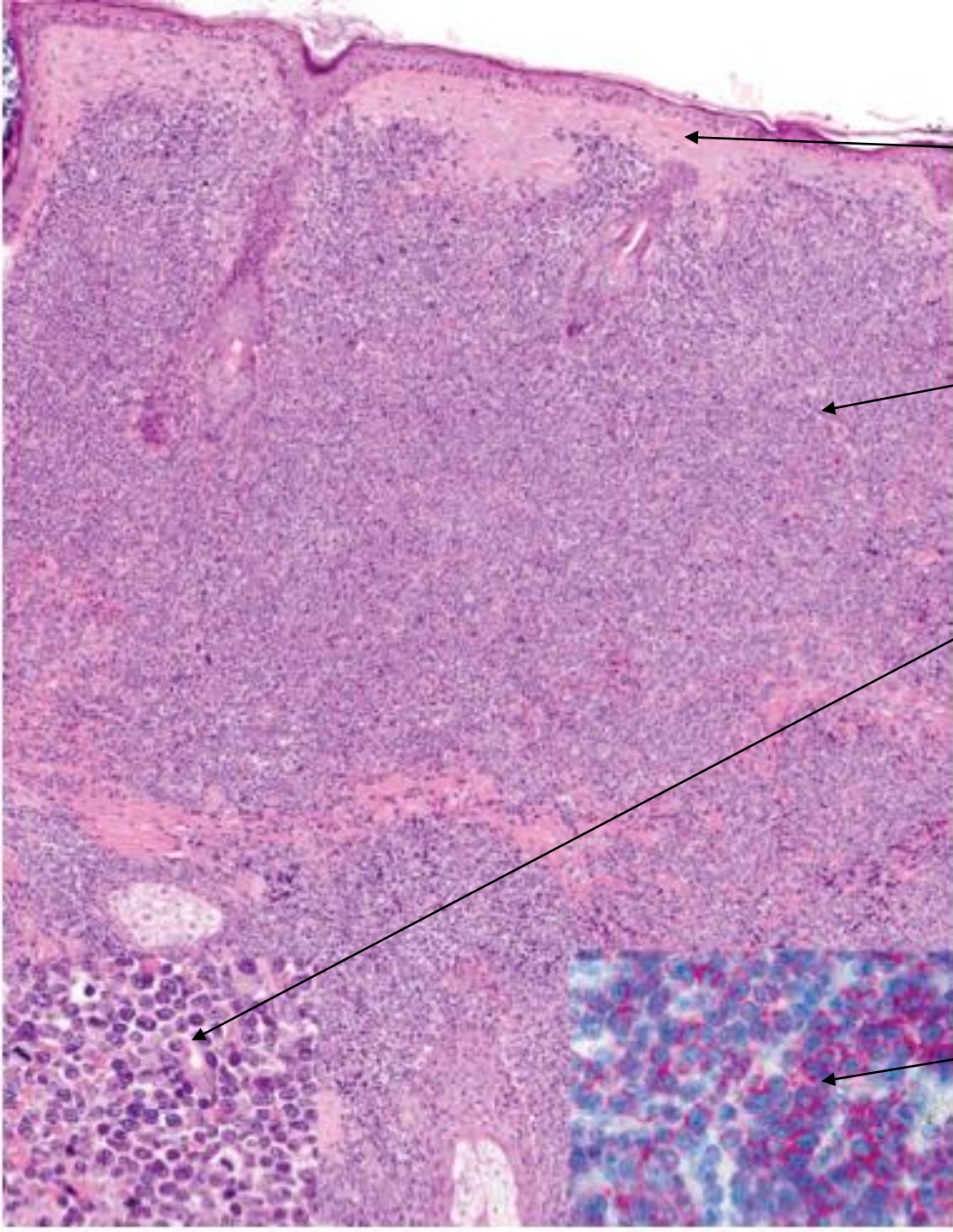
MZL/ICY, marginal zone lymphoma/immunocytoma; FCL, follicle center lymphoma; Secondary FCL secondary cutaneous FCL; DLBCCL, diffuse large B-cell lymphoma; PSL, pseudolymphoma or reactive lymphoid hyperplasia. The term pseudolymphoma is used by dermatologists as a synonym for any type of reactive infiltrate in the skin that is extensive enough to cause a tumor or nodule.

Blastic NK-cell lymphoma or CD4+/CD56+ hematodermic neoplasm

- Cell of origin is not yet completely elucidated
- Cytogenetically, they are related to dendritic cells.
- Skin involvement occurs in 87% of the patients and manifests with contusiform, brownish infiltrated plaques or nodules.
- The oral mucosa is commonly involved.
- The cells express CD4, CD56, CD123 and TCL-1 but are negative for other T-, B-, NK-cell, or myeloid markers.
- median survival, 14 months)
- Systemic chemotherapy



Bruiselike skin lesion

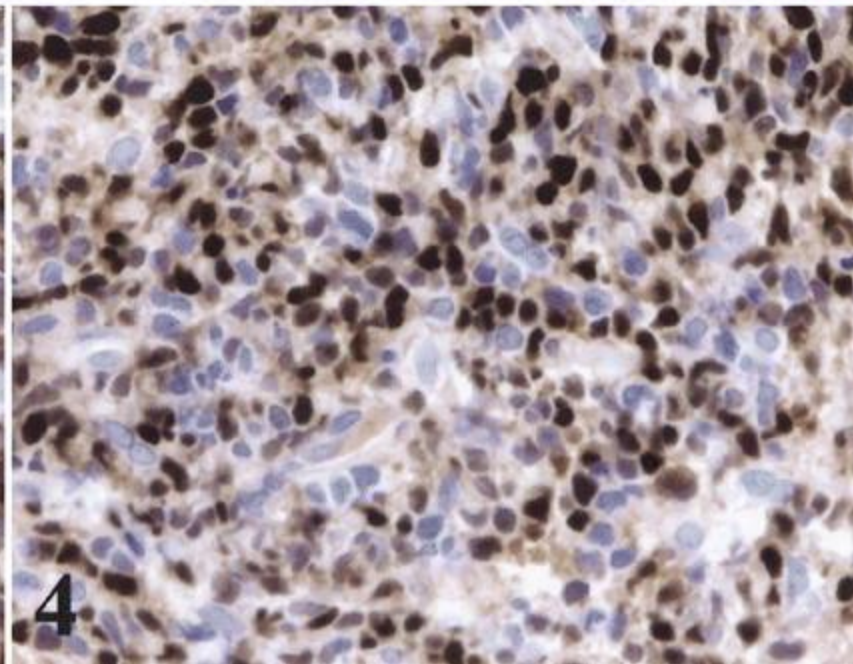
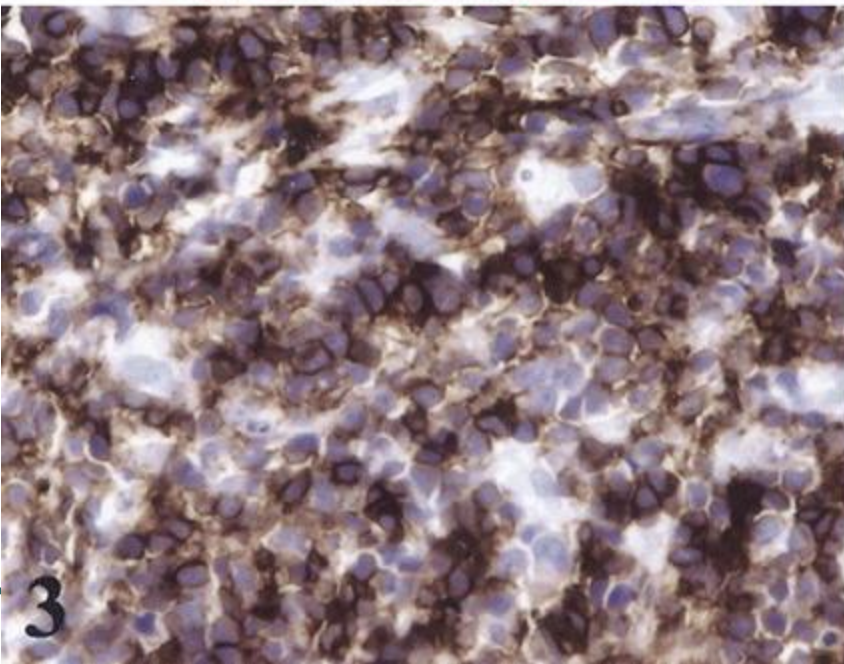
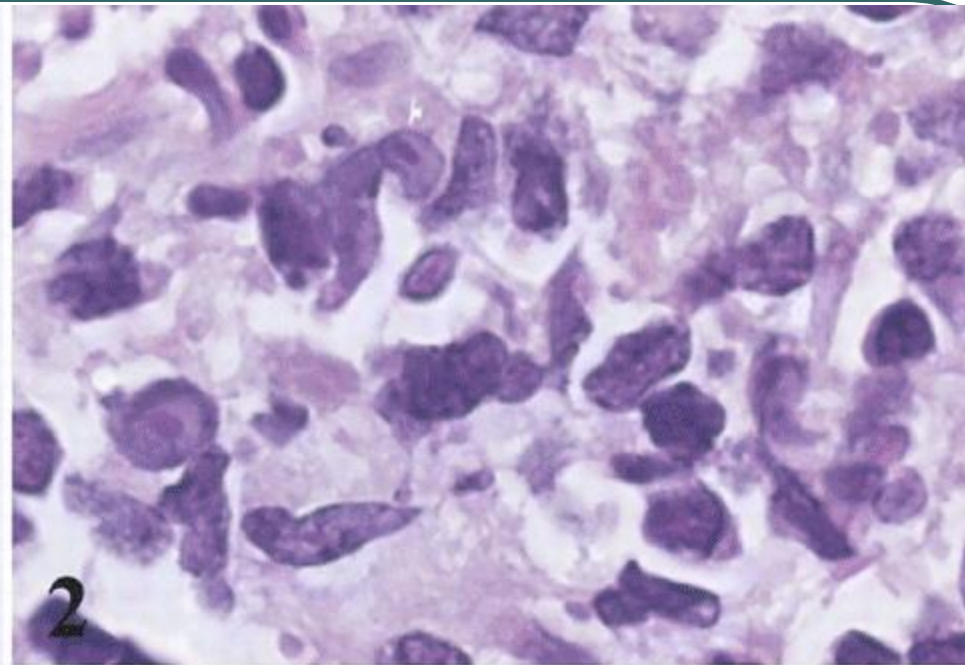
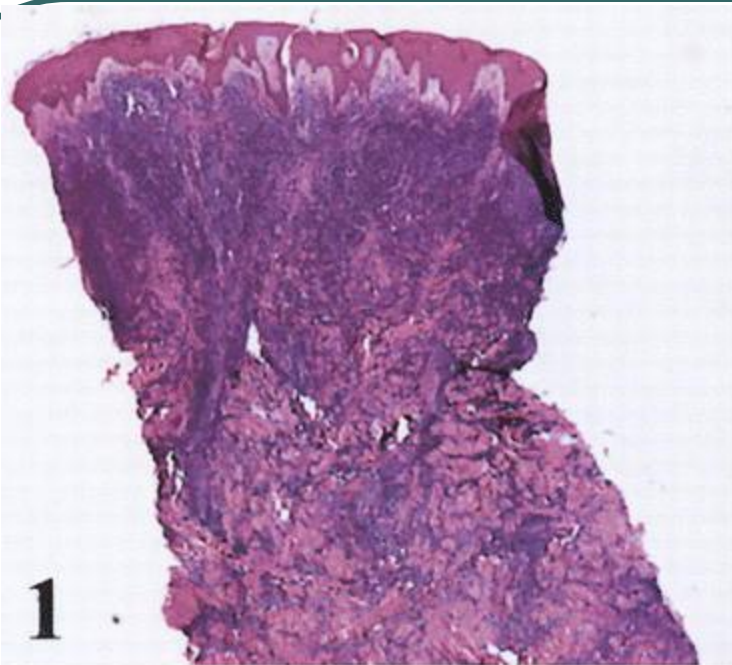


Grenz zone

Diffuse dermal infiltrate

Erythrocyte extravasation is a characteristic feature, explaining the bruise-like appearance.

CD 56 + tumor cells

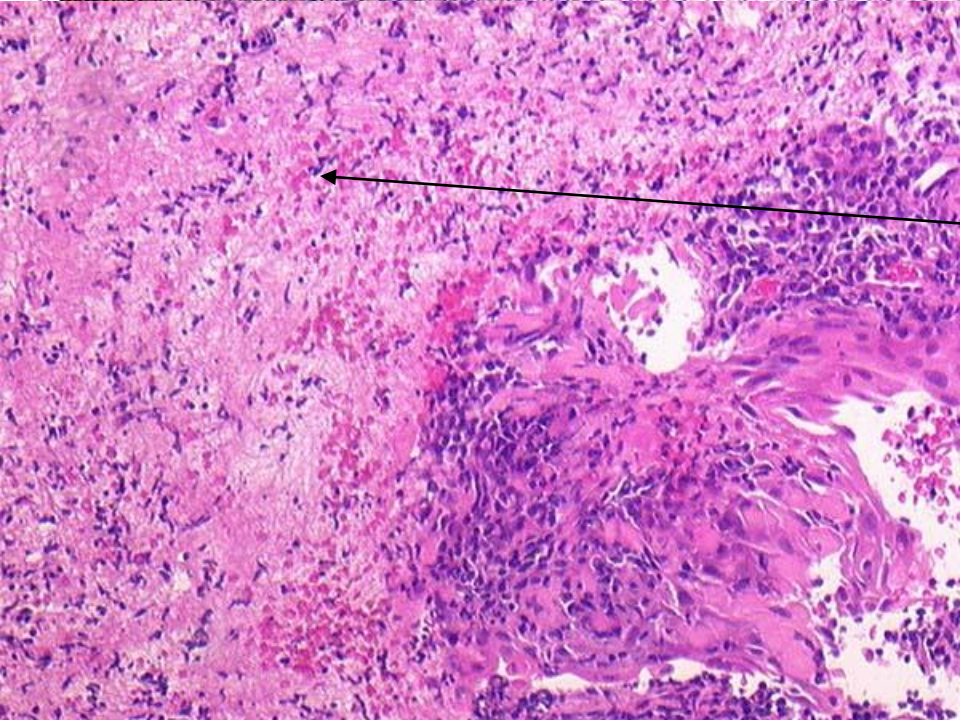
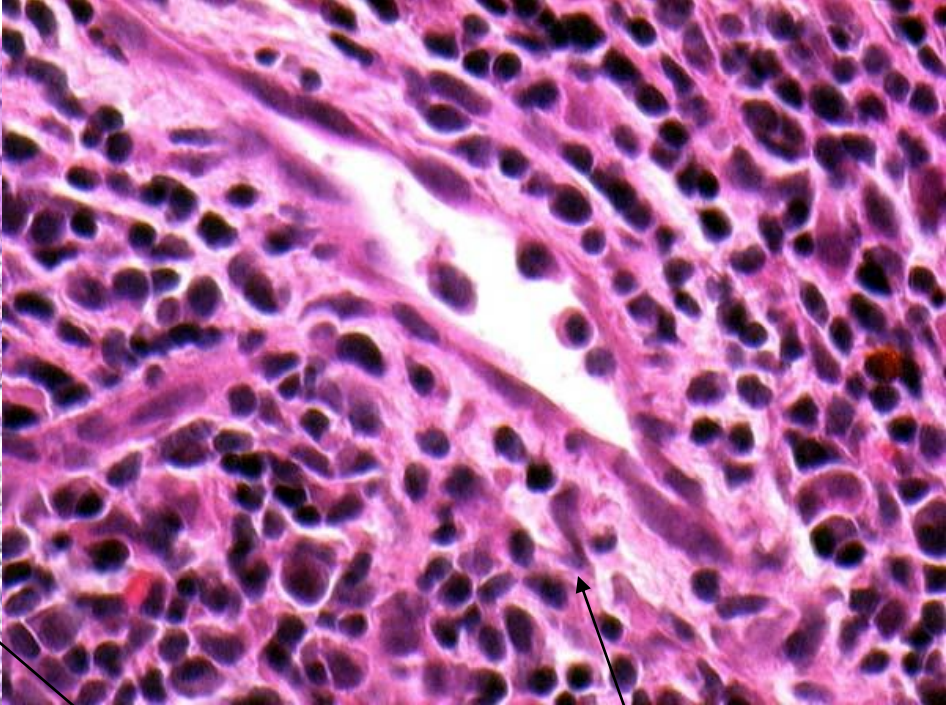
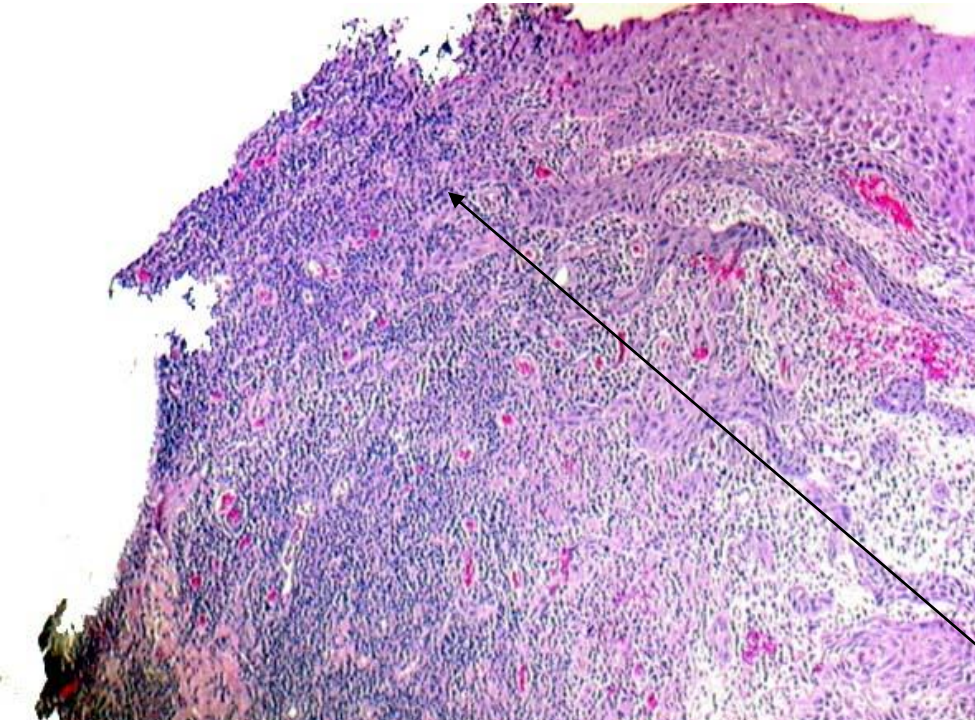


CD 56 +

TdT +

Extranodal NK/T-cell lymphoma, nasal type

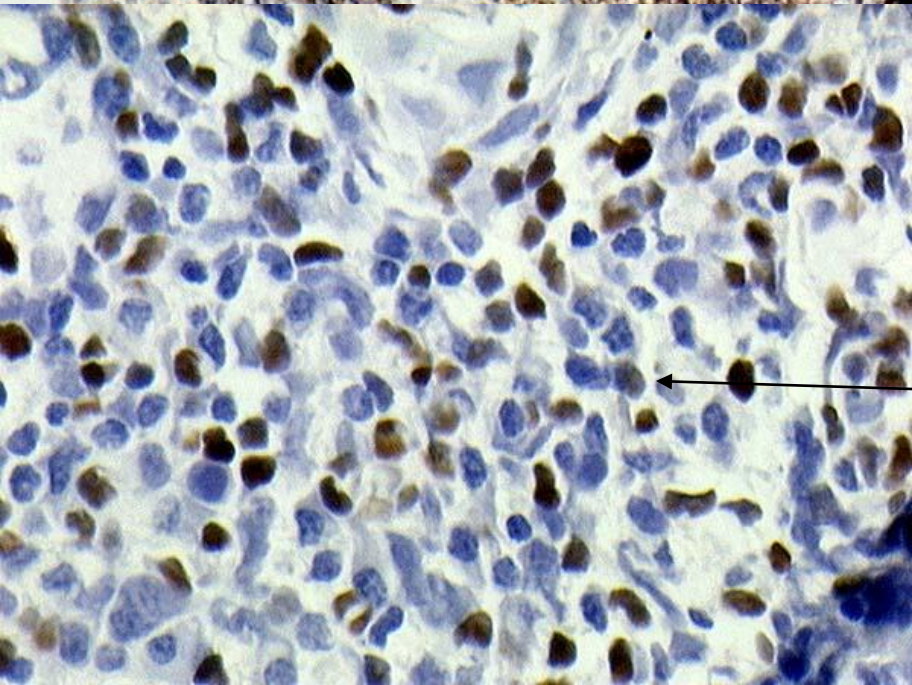
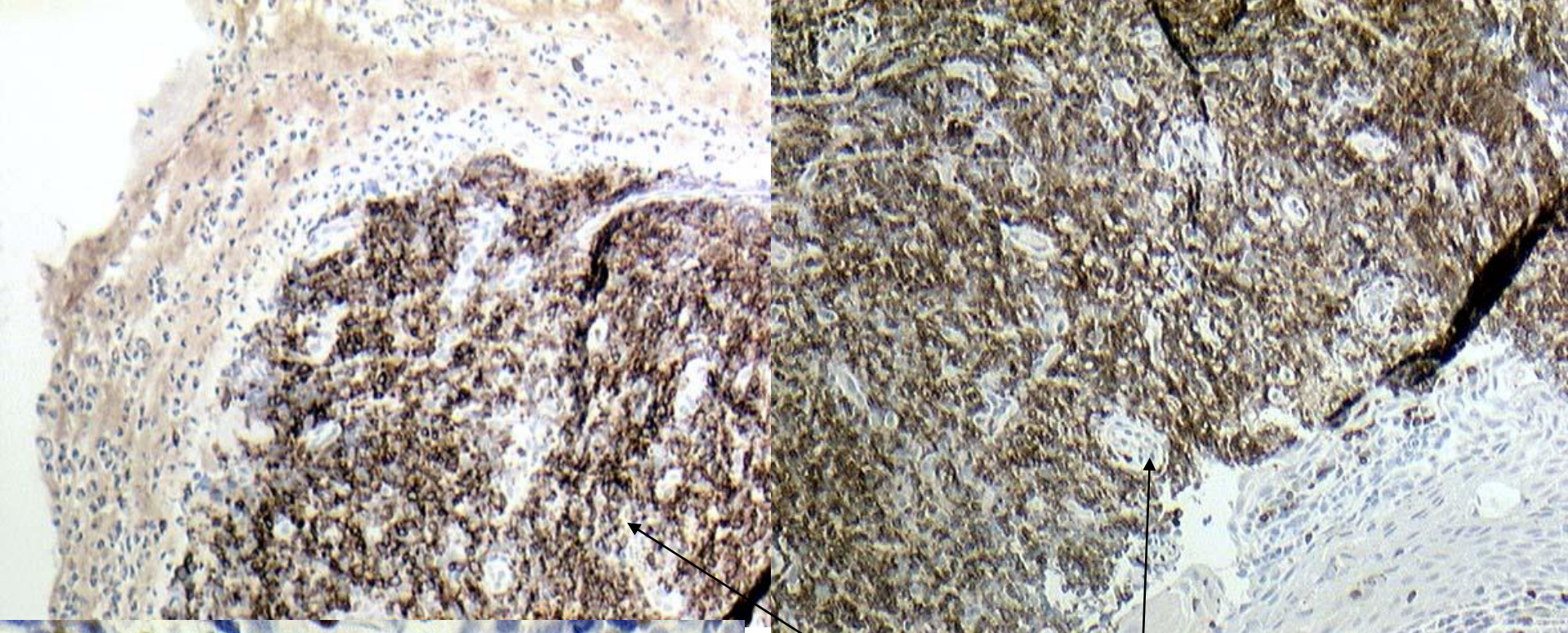
- Adults, males, Asia, Central America, and South America.
- Multiple plaques or tumors preferentially on the trunk and extremities, ulceration is common.
- Dense infiltrates in dermis and often the subcutis.
- Epidermotropism, prominent angiocentricity and angiodestruction and extensive necrosis.
- Small to large cells with irregular or oval nuclei, moderately dense chromatin, and pale cytoplasm, histiocytes, plasma cells, and eosinophils can be seen.
- Express CD2, CD56, cytoplasmic CD3, and cytotoxic proteins (TIA-1, granzyme B, perforin), but lack surface CD3.
- In rare CD56- cases detection of EBV by in situ hybridization and expression of cytotoxic proteins are required for diagnosis.
- EBV is expressed almost in all cases, suggesting a pathogenetic role of this virus



Ulceration

Necrosis

Angiocentric infiltration



CD56 +

CD3+ (cytoplasmic)

EBV (EBER/ISH) +

Extranodal NK/T-cell lymphoma, nasal type

- Nearly always EBV+ lymphoma of small, medium, or large cells usually with an NK-cell, or more rarely a cytotoxic T-cell, phenotype.
- The skin is the second most common site of involvement after the nasal cavity/nasopharynx
- Skin involvement may be a primary or secondary manifestation
- Since both groups show an aggressive clinical behavior and require the same type of treatment, distinction is not useful.
- median survival of 27 months was reported, compared with 5 months for patients presenting with cutaneous and extracutaneous disease.
- Tx systemic chemotherapy

Conclusion

- The new WHO/EORTC classification of cutaneous lymphomas, employs a terminology compatible with systemic lymphomas but also reflects the organ-specific peculiarities of cutaneous lymphomas.

References

- **1)WHO-EORTC classification for cutaneous lymphomas** Blood, 15 May 2005, Vol. 105, No. 10, pp. 3768-3785.
- **2)WHO/EORTC classification of cutaneous lymphomas 2005: histological and molecular classification** Journal of Cutaneous Pathology
Volume 32 Page 647 - November 2005