

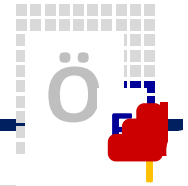
CUTAN LYMPHOMA

Csomor Judit

Patológus Kongresszus

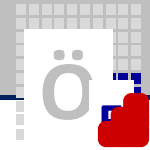
2013. Siófok







Primary cutaneous lymphoma --- klasszifikáció



- Mycosis fungoides **50 %**
- Sézary syndrome **3 %**
- CD30+ **20 %**
 - Anaplastic large cell
 - Lymphomatoid papulosis
- CD4+ small/medium pleomorphic
- T-cell lymphoma NOS **2 %**



- Follicle center lymphoma
- Diffuse large B-cell leg type
- Extranodal marginal zone



lymphoma of mucosa-associated lymphoid tissue

(MALT) **25%**

- CD8+ aggressive epidermotropic cytotoxic
- Subcutaneous panniculitis-like T-cell
- $\gamma\delta$ T-cell lymphoma

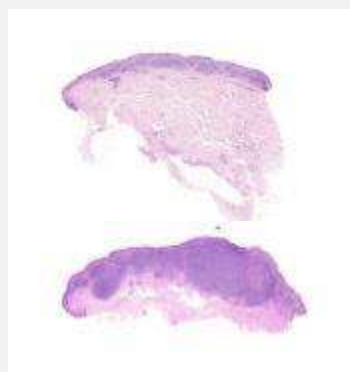
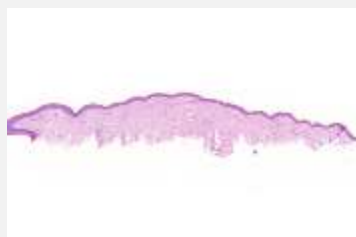
<1%

- Intravascular large B-cell
- Blastic plasmacytoid dendritic
- Extranodal NK/T-cell lymphoma nasal ty.
- Adult T-cell leukemia/lymphoma



MF

A - Ω , szekvenciális, perzisztáló, lassan progrediáló, 6/M/év,
összes lymphoma 2%



▼ MF

- MF - folt, vékony plakk = korai mycosis fungoides



?

diagnózis

- MF – vastag plakk, tumor



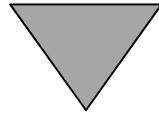
?

transzformáció

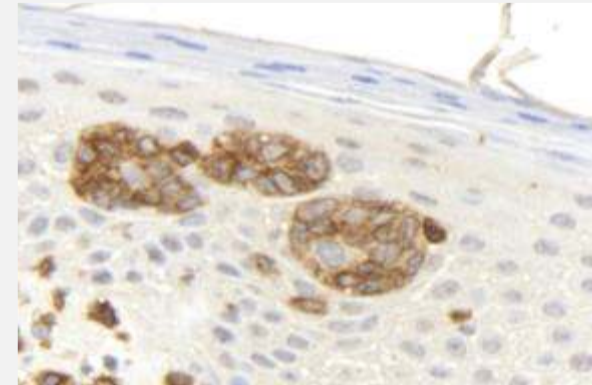
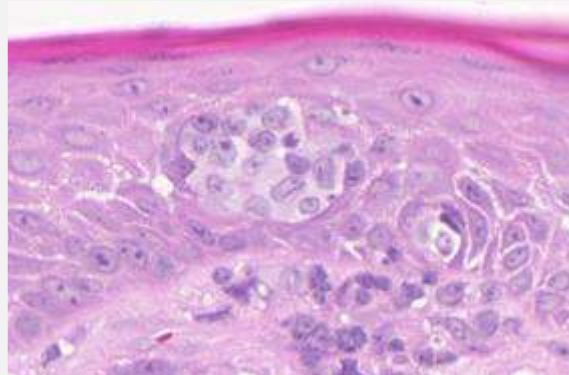
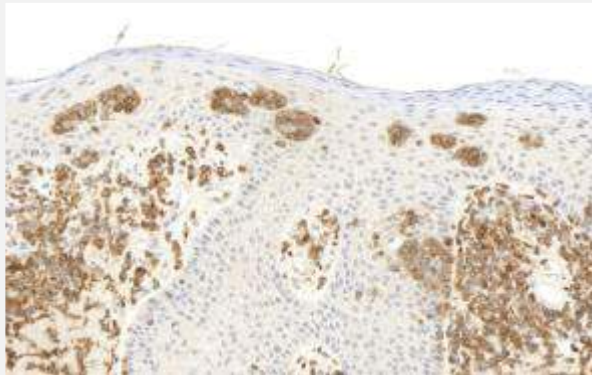


MF

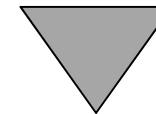
Pautrier abscessus



Majdnem specifikus



30-40 % fordul elő

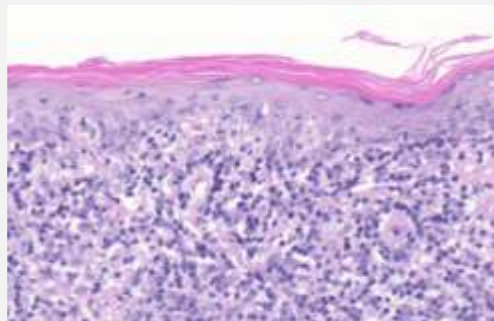
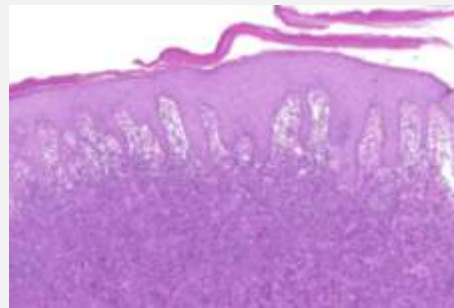
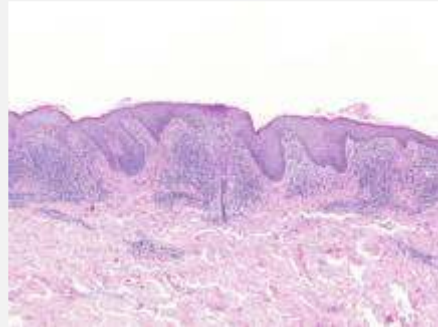
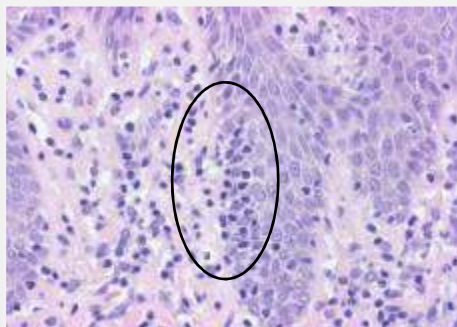
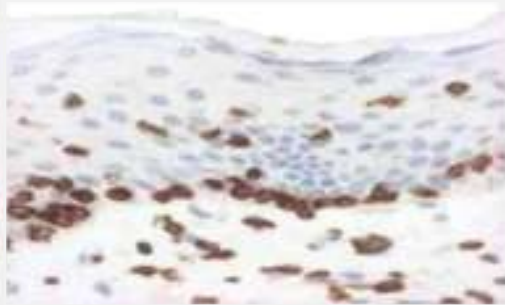
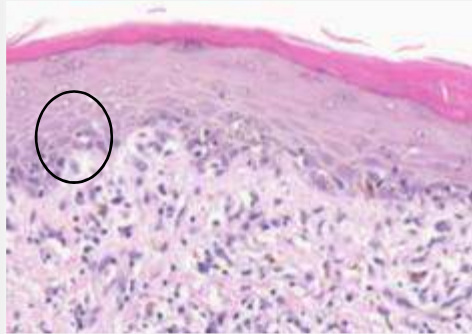
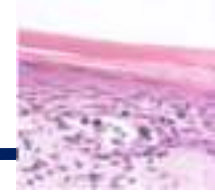


Differenciál diagnózis - Spongiform abscessus (CD3 + CD4)
disproporcionált – epidermotrop lymphocytá atípusos,
nem kíséri spongiosis, T-sejt marker vesztés



MF Epidermotropismus (exocytosis)

+



tömeges
linearis
atípusos
halo

+

lichenoid
superficiális
infiltráció,
interface aktivitás
parakeratosis,
psoriasiform
hyperplasia,
superficiális
fibrosis,
atrófia



MF Korai MF - diagnózis, differenciál diagnosztika



Klinikai tünetek + histomorfológia + immunfenotípus + genotípus

2

2

1

1

Ekzema

Psoriasis

Parapsoriasis

Photodermatitis

Gyógyszerkiütés

CD3+,CD4+,
(CD3+, CD8+)

TCR
monoklonális

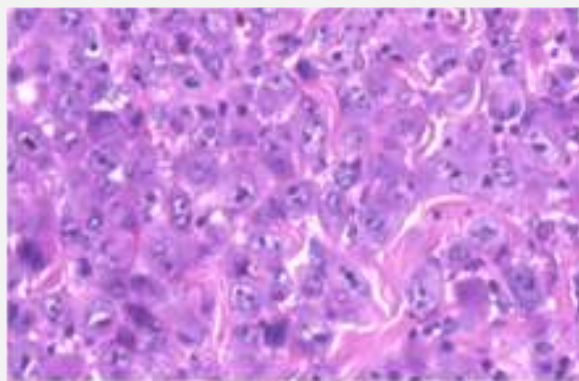
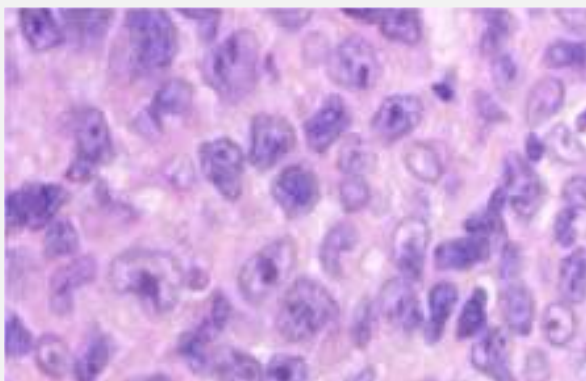
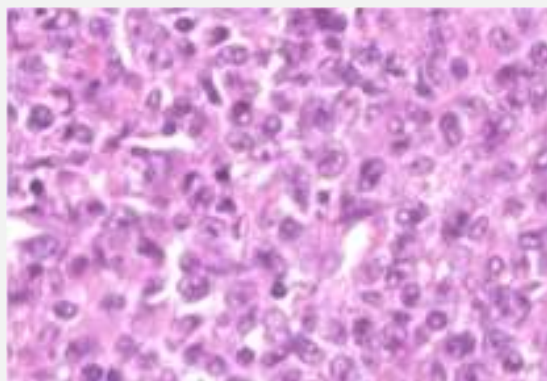
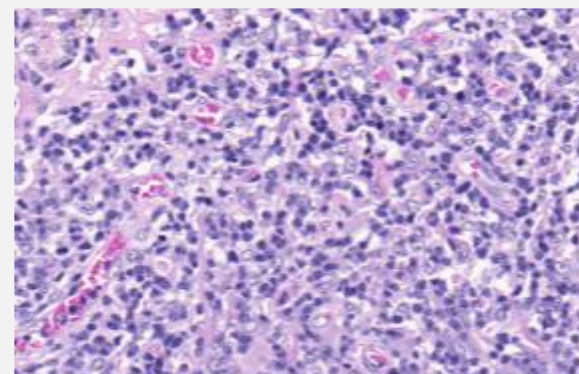
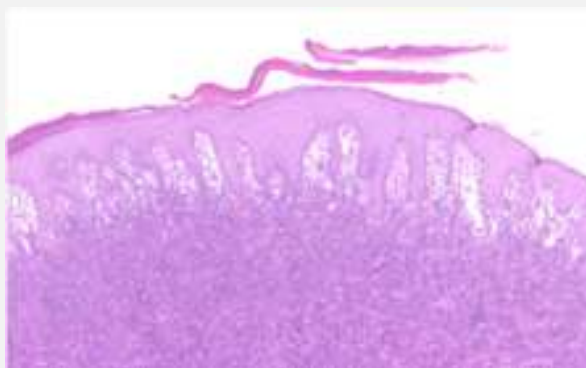
?

Tumorsejt : reaktív = 1 : 10



MF

Transzformáció > 30% nagysejt

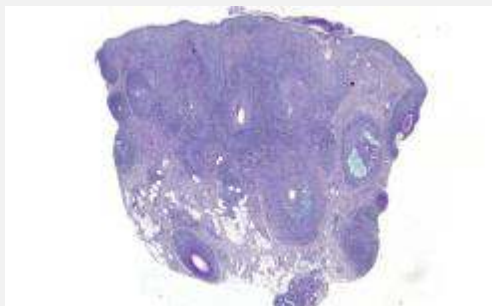


PTCL NOS / ALCL

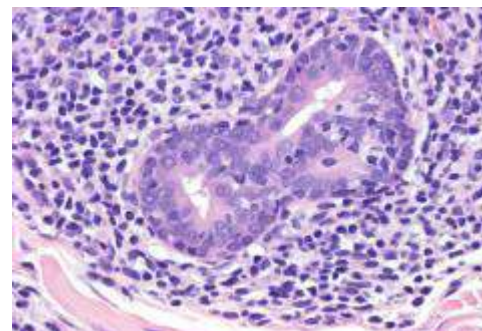
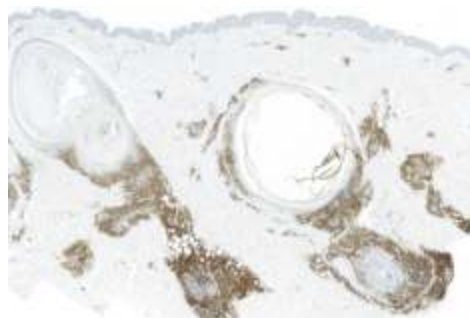


MF

Variánsok

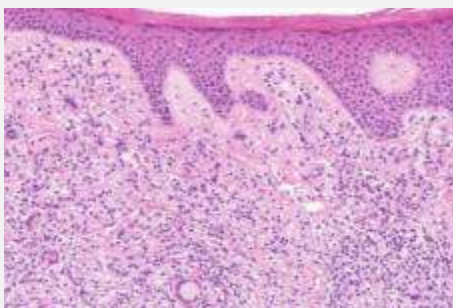


Unilézionális, folliculotrop, folliculáris mucinosissal

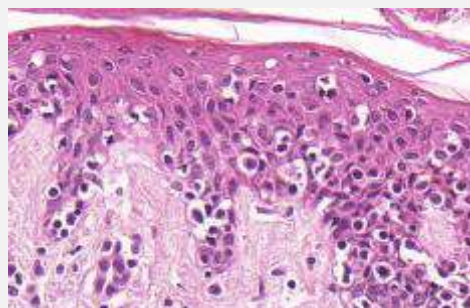


Folliculotrop, folliculáris plugginggal

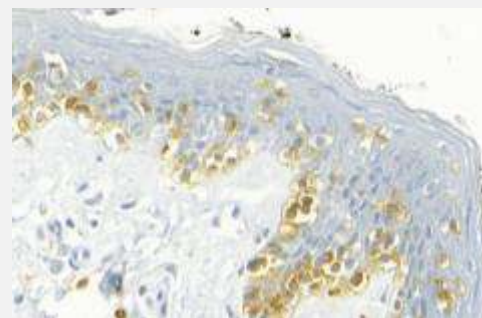
Syringotrop



Granulomatosus



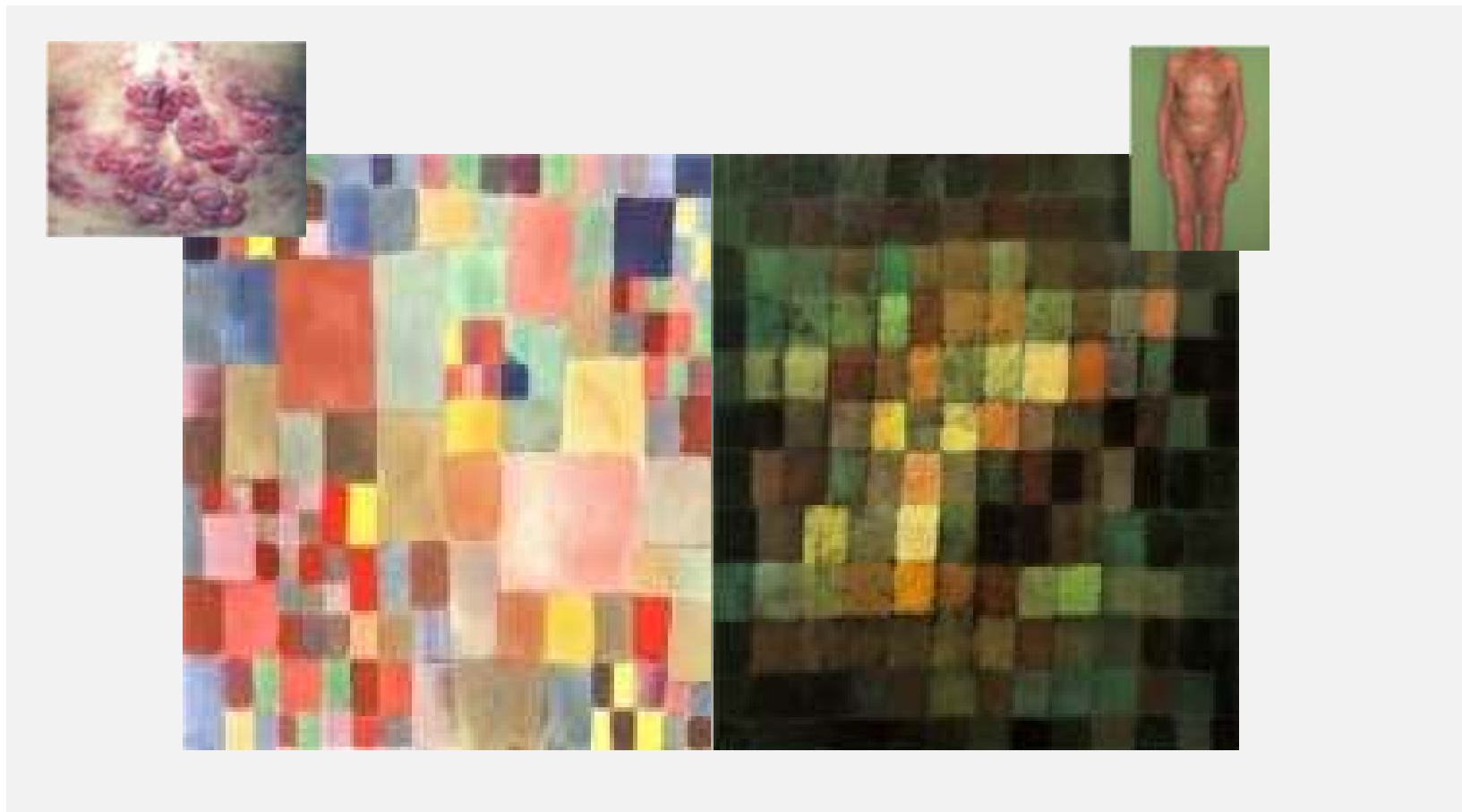
Pagetoid



▼ Sézary szindróma

Erythroderma + leukémiás vérkép + lymphadenomegalia

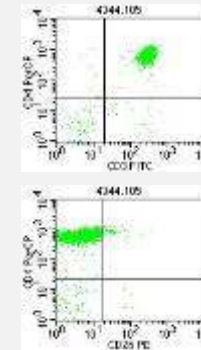
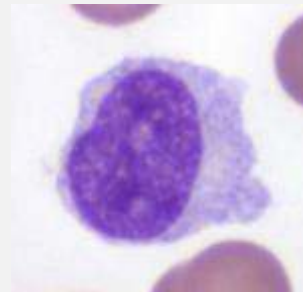
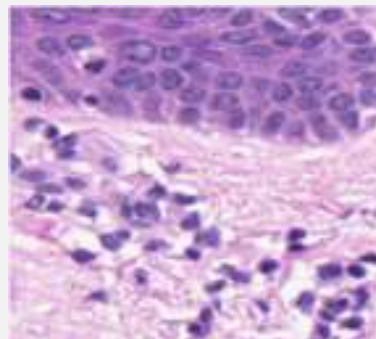
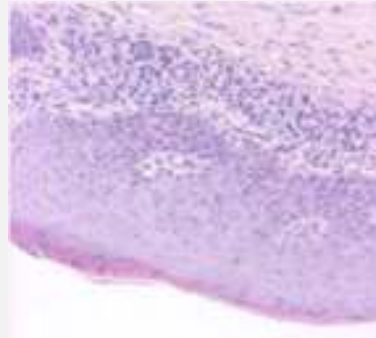
(MF / SS), MF és SS





SS

Erythroderma + lymphadenomegalia + keringő SS sejtek



**CD3+, CD4+, CD2+,
TCR $\alpha\beta$ +, CD5+, CD26-
CD4+, CD26- > 30%**



TCR monoklonális



> 1000/mm³

- dermatopathias
- lymphadenitis
- tumoros infiltráció

Lymphomatoid papulosis

CD30

- recidiváló, ulceronecroticus erupciók
- 5 hisztopatológiai altípus : A,B,C,D,E, 6p23.5
- Az esetek 10-20% ALCL vagy MF alakul ki
- CD30+, Tia+, CD4+, (LyP-D, E CD8+)

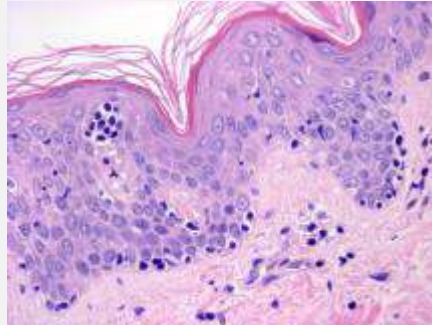
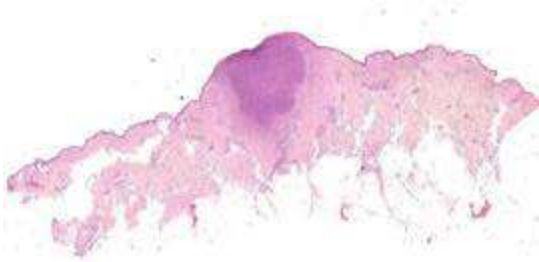


▼ LyP

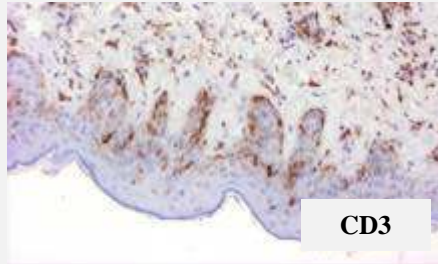
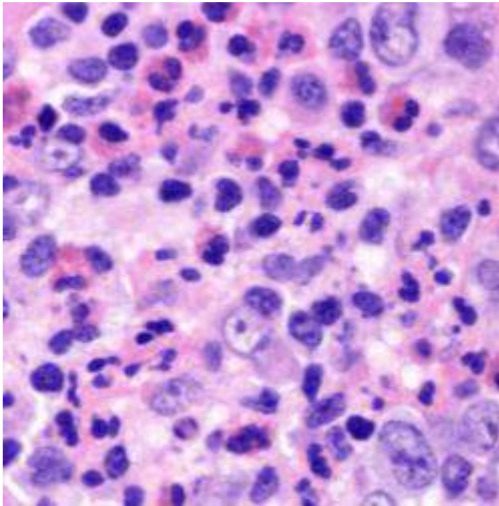
CD30

B

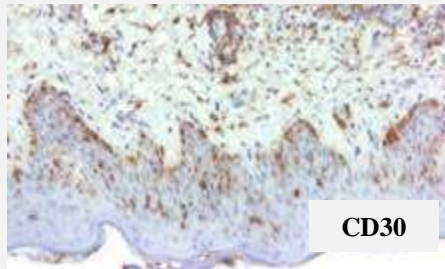
A



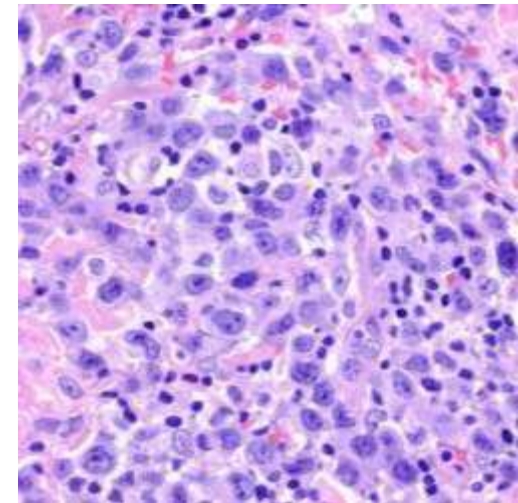
C



CD3



CD30



Hodgkin-szerű

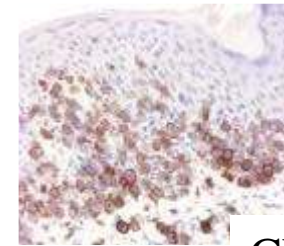
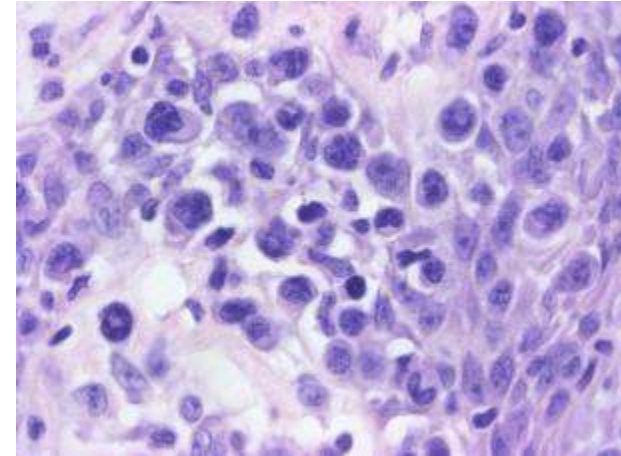
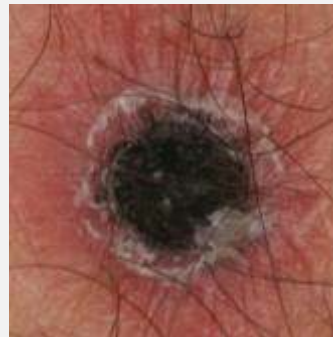
MF-szerű
CD30-/CD30+

ALCL-szerű

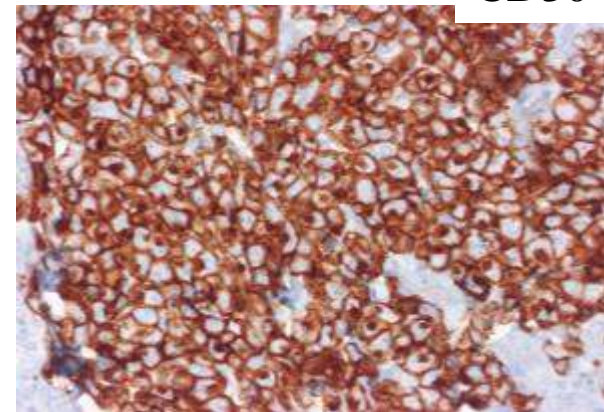
Primer cutan anaplasziás nagysejtes lymphoma

CD30

- soliter / multiplex papula, nodulus, tumor, ulceráció
- spontán regresszió előfordulhat
- Atípusos nagysejtek
- > 70% CD30+ expresszió
- CD3+/-, CD4+, CD30+, TIA+, ALK-
- 10 éves túlélés > 90%



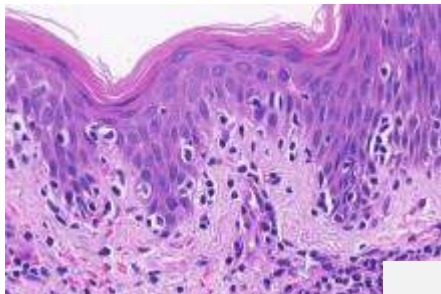
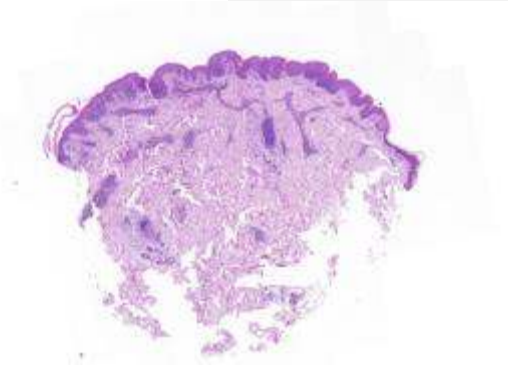
CD30



▼ LyP

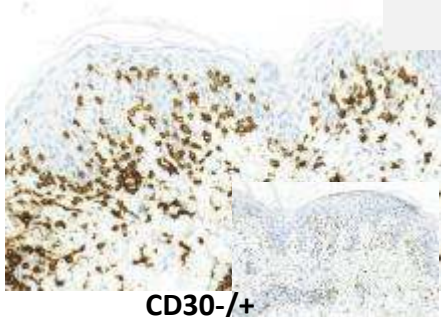
E

D



Vasculitis -
szerű

CD8+



CD30-/+

6p23.5

bifázisos

Differenciál
diagnózis

ALCL

LY

PLEVA

drug erupció

csípés

vírus inf.

Primer cutan epidermotrop CD8 + perifériás T-sejtes lymphoma

Nagyon ritka

Kifekélyesedő plakk, tumor, generalizált bőrtünetek

Korai szisztémás (CNS, csontvelő) érintettség

Markáns epidermotropizmus, változatos sejtkép, érinvázió

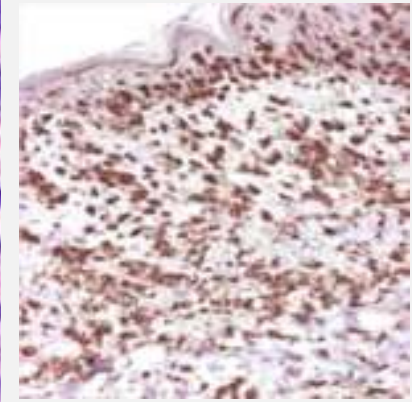
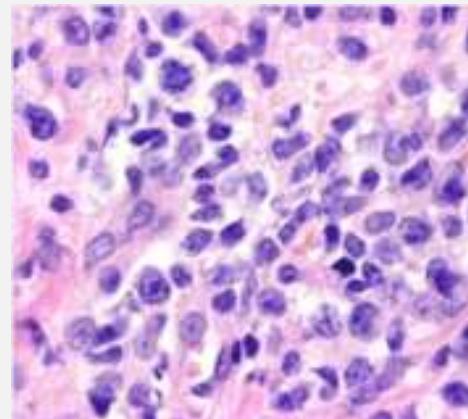
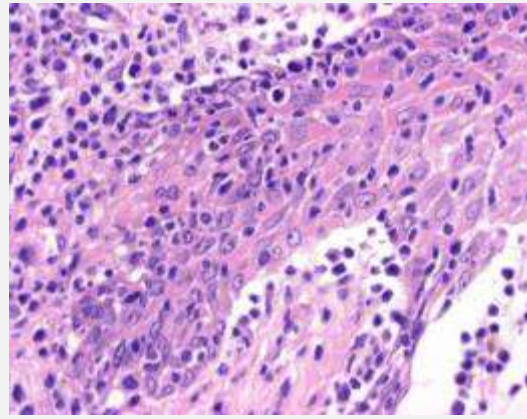
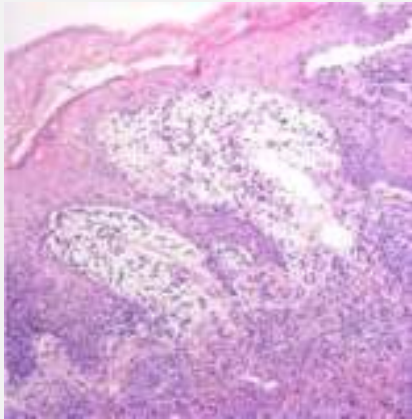
Rossz prognózisú, agresszív betegség, túlélés <5 év



Primer cutan epidermotrop CD8 + perifériás T-sejtes lymphoma



BF1+ ($\alpha\beta$), CD3+, CD8+, CD30-, CD56-/+, TIA+,
CD45RA+, CD45RO- fenotípus



Berti E, Santucci M, Marzano AV, Nofal A, Diwan H.

Agresszív MF, MF tumeur d'emblee, Pagetoid reticulosis Ketrón-Goodman type

[J Clin Pathol.](#) 2008 Jun;61(6):770-2. doi: 10.1136/jcp.2007.054585.

Rare provisional entity: primary cutaneous aggressive epidermotropic CD8+ cytotoxic T-cell lymphoma in a young woman.

[Csomor J](#), [Bognár A](#), [Benedek S](#), [Sinkó J](#), [Fekete S](#), [Krenács I](#), [Matolcsy A](#), [Reiniger L](#).

[Eur J Dermatol.](#) 2012 Sep-Oct;22(5):690-1. doi: 10.1684/ejd.2012.1823.

Primary cutaneous aggressive epidermotropic CD8+ T-cell lymphoma: report of two cases with no evidence of systemic disease.

[Szepesi Á](#), [Csomor J](#), [Rajnai H](#), [Erős N](#), [Wikonkál N](#), [Kárpáti S](#), [Matolcsy A](#), [Marschalkó M](#).

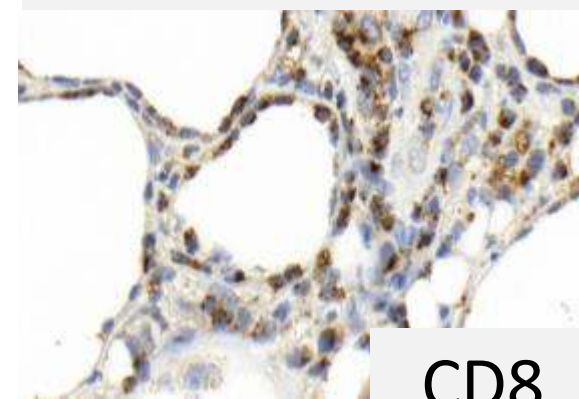
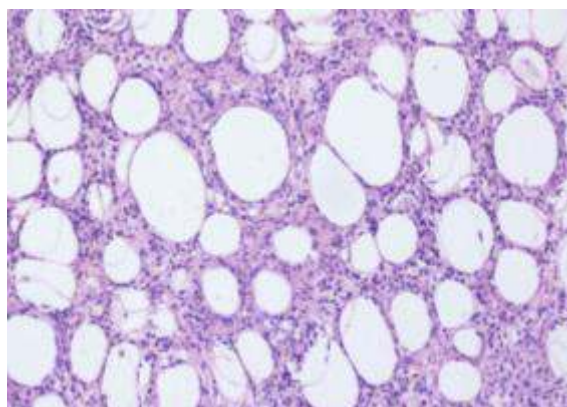
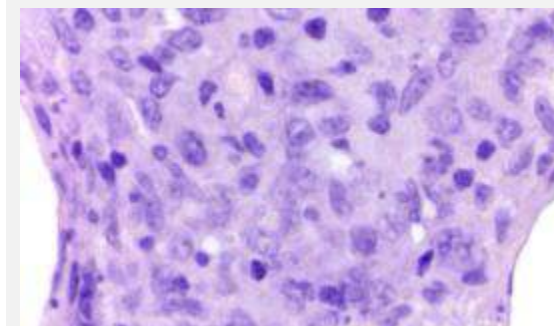
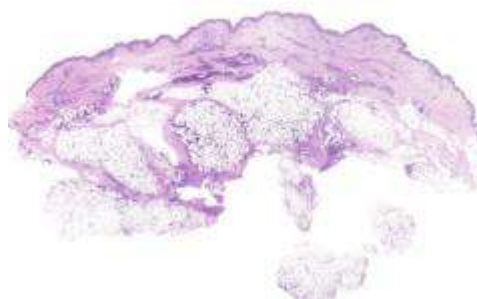


Subcutan panniculitis-like T-sejtes lymphoma



- szoliter/ multiplex plakk, subcutan csomó
- alsó végtag
- $\alpha\beta$, CD3+, CD8+, TIA1+, kis/ középnagy pleiomorph sejtek
- + macrophagok,
- karyorrhexis,
- cytophagocytosis
- necrosis
- nincs granulocytá
- jó prognózis

DD lupus panniculitis
 $\gamma\delta$ p. c. T-sejtes ly



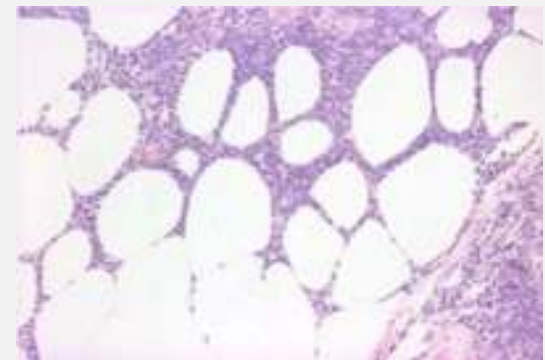
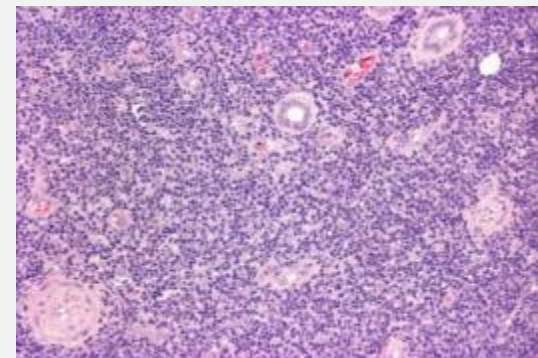
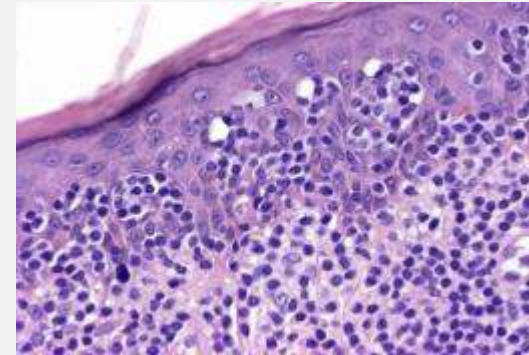
CD8



Primer cutan γ/δ T-sejtes lymphoma



- Disszeminált plakk, nodulus, gyorsan kifeléyesedő tumor
- Haemophagocytosis szindróma
- Necrosis
- Angioinvázió
- Középnagy hyperchrom CD3+, $\gamma\delta$, 4-,5-,8-/+, TdT-
- Rossz prognózis





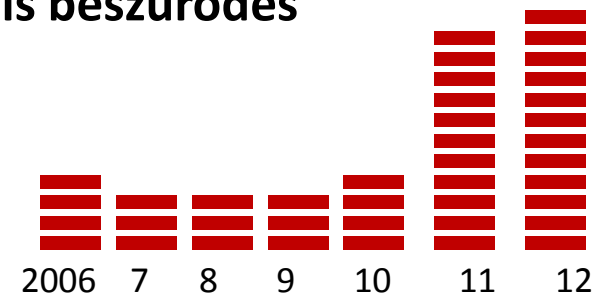
Primer cutan CD4+ kis/középnagy T-sejtes lymphoma



- Ritka (?)
- A tumort nem előzi meg folt, plakk stádium
- Általában a felső testfél szoliter tumora
- Kevert sejtösszetételű, sok reaktív B-sejtet tartalmazó, kis-középnagy

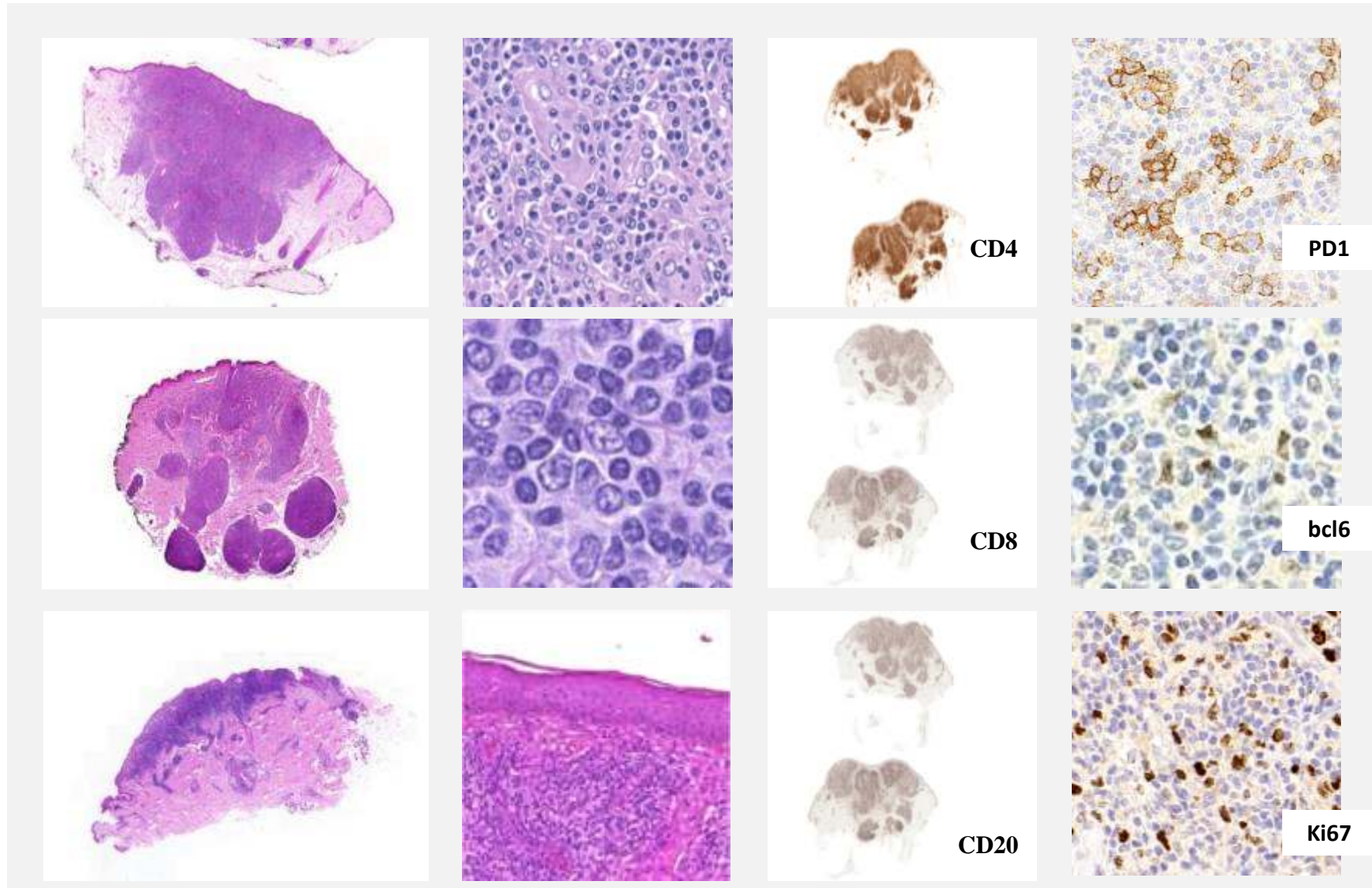
T-sejt dominanciájú sejtekből álló dermális beszűrődés

- Folliculáris helper T-sejt eredetű
- Kíváló prognózis



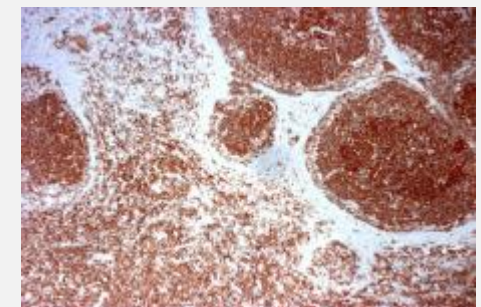
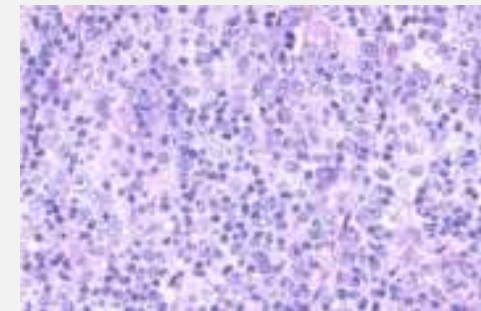
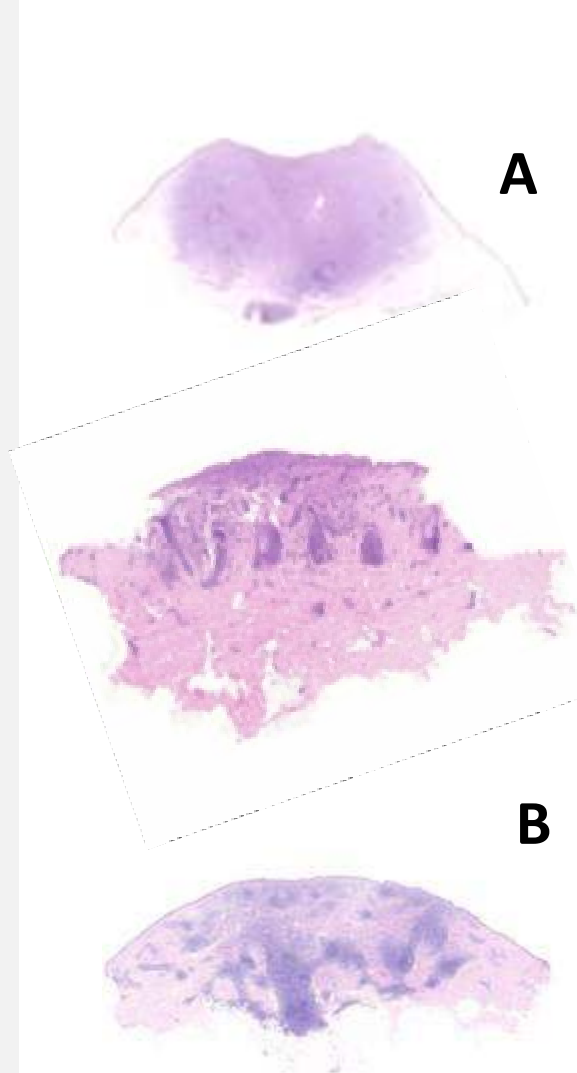


Primer cutan CD4+ kis/középnagy T-sejtes lymphoma



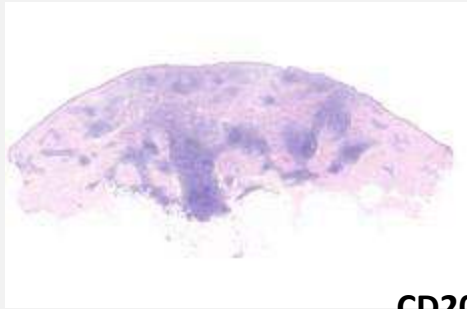
Primer cutan marginális zóna lymphoma

- Szoliter papula, plakk, nodulus, tumor – törzs, végtagok
- Post centrum germinativum memória sejtek: CD10 -, bcl-6-, bcl-2+, MUM1-
+
reaktív folliculusok, plasmasejtek, immunoblastok, histiocyter sejtek
- Borrelia infekció?
- Spontán regresszió
- 5-éves túlélés: 100 %





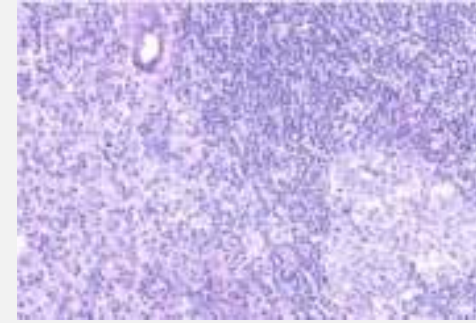
Primer cutan marginális zóna lymphoma



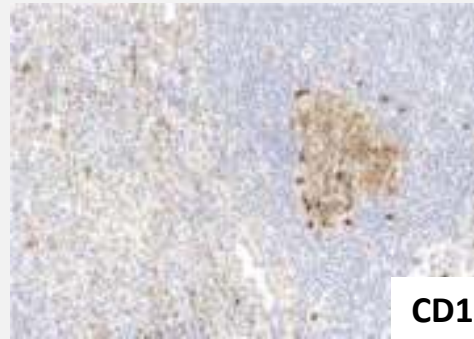
CD20



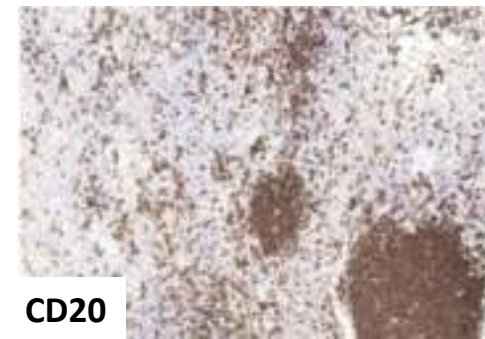
CD10



CD20



CD10



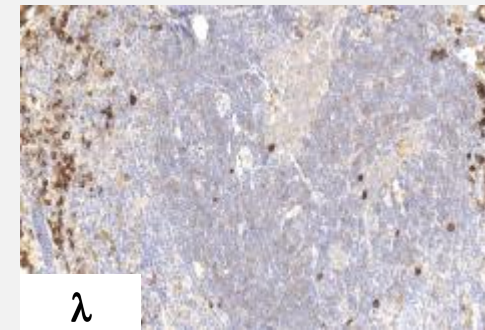
CD20



CD3



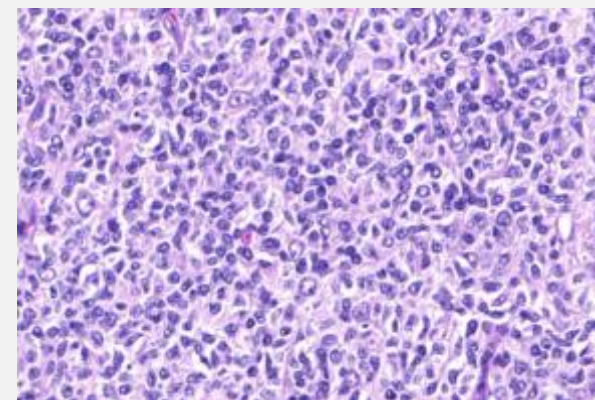
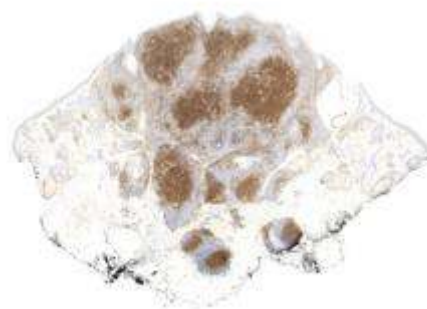
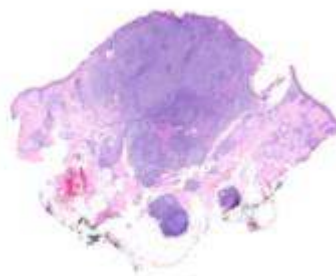
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Primer cutan folliculus center lymphoma

- Plakk, tumor
- Nyak, törzs
- folliculáris, folliculáris + diffúz / diffúz mintázat
- Centrocyták /centroblastok
- reaktív sejtek
- CD10+, bcl-2-, bcl-6+, MUM1 -
- 5-éves túlélés: > 95%,





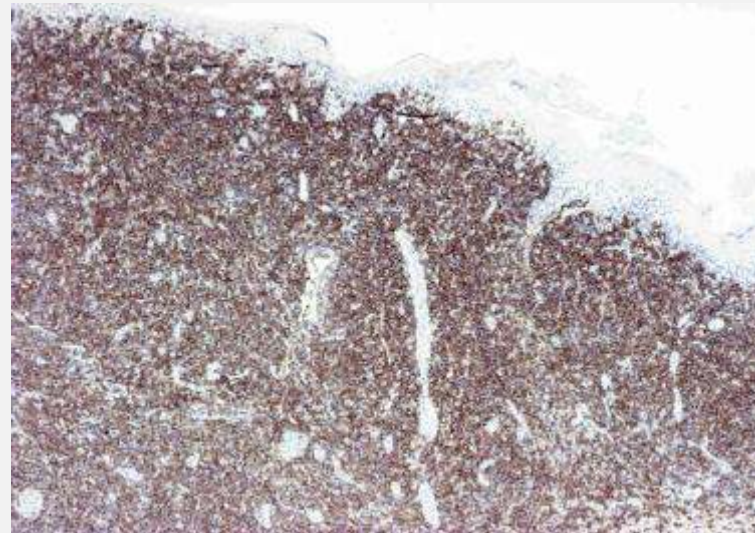
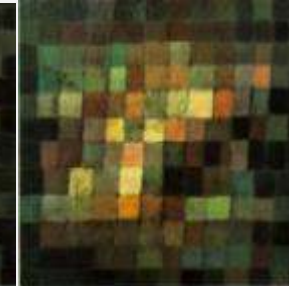
Primer cutan diffúz nagy B-sejtes lymphoma, leg type



DLBCL
nonGC



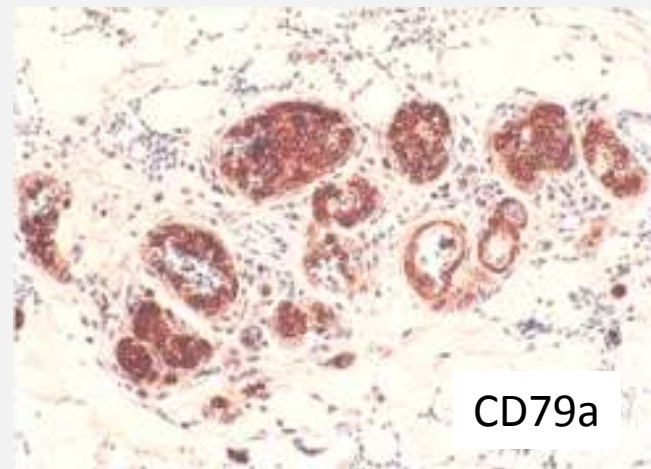
DLBCL
LEG



CD20+, CD79+, CD10-, MUM1+, bcl6+/-

Intravasculáris nagy B-sejtes lymphoma

- macula, plakk, panniculitis- vagy erythema nodosum-szerű nodulusok, teleangiectasia, alsó végtag, törzs
- 2 fő forma:
 - 1./ klasszikus európai: bőr, CNS, szolid szervek
 - 2./ ázsiai variáns: malignus histiocytosis-szerű forma : hepatosplenomegalia, hemophagocytosis szindróma
- CD20+, CD79a+
- 5-éves túlélés: 22-65%

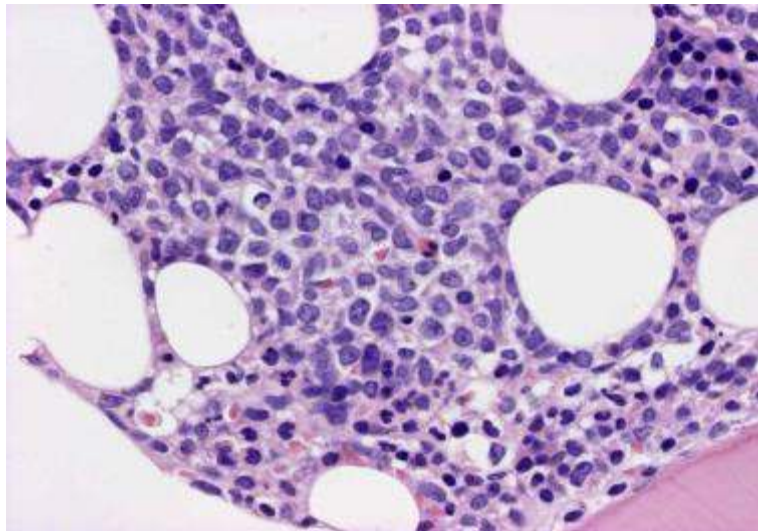


[Pathol Oncol Res.](#) 2008 Sep;14(3):281-4. doi: 10.1007/s12253-008-9051-y. Epub 2008 May
Prolonged survival using anti-CD20 combined chemotherapy in primary prostatic intravascular large B-cell lymphoma.

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Blastos plasmocytoid dendriticus sejtes tumor

- Lilás-barna macula, papula, nodulus, tumor, leukemia, nyirokcsomó, csv infiltráció
- dense dermalis és subcutan infiltrátum
- CD4+, CD43+, CD56+, CD45+, TdT-, CD68+/-
- median túlélés: 14 hónap



[J Neurooncol.](#) 2010 Apr;97(2):301-4. doi: 10.1007/s11060-009-0026-y. Epub 2009 Oct 2.

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KÖSZÖNET



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Weiss Éva, Kiss Renáta, Terdik Ilona**