

Atelier d'Héмато-Pathologie

Société Tunisienne de Pathologie

Société Tunisienne d'Hémato-Cancérologie

Roche Tunisie

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N° 6

Lymphomes Médiastinaux

Antoine de Mascarel, Bordeaux

« antoine.demascarel@chu-bordeaux.fr »

Primary Mediastinal (Thymic) Large B-cell Lymphoma (PMLBCL)

1980 : Primary mediastinal large B-cell lymphoma

1986 : PMBCL with sclerosis

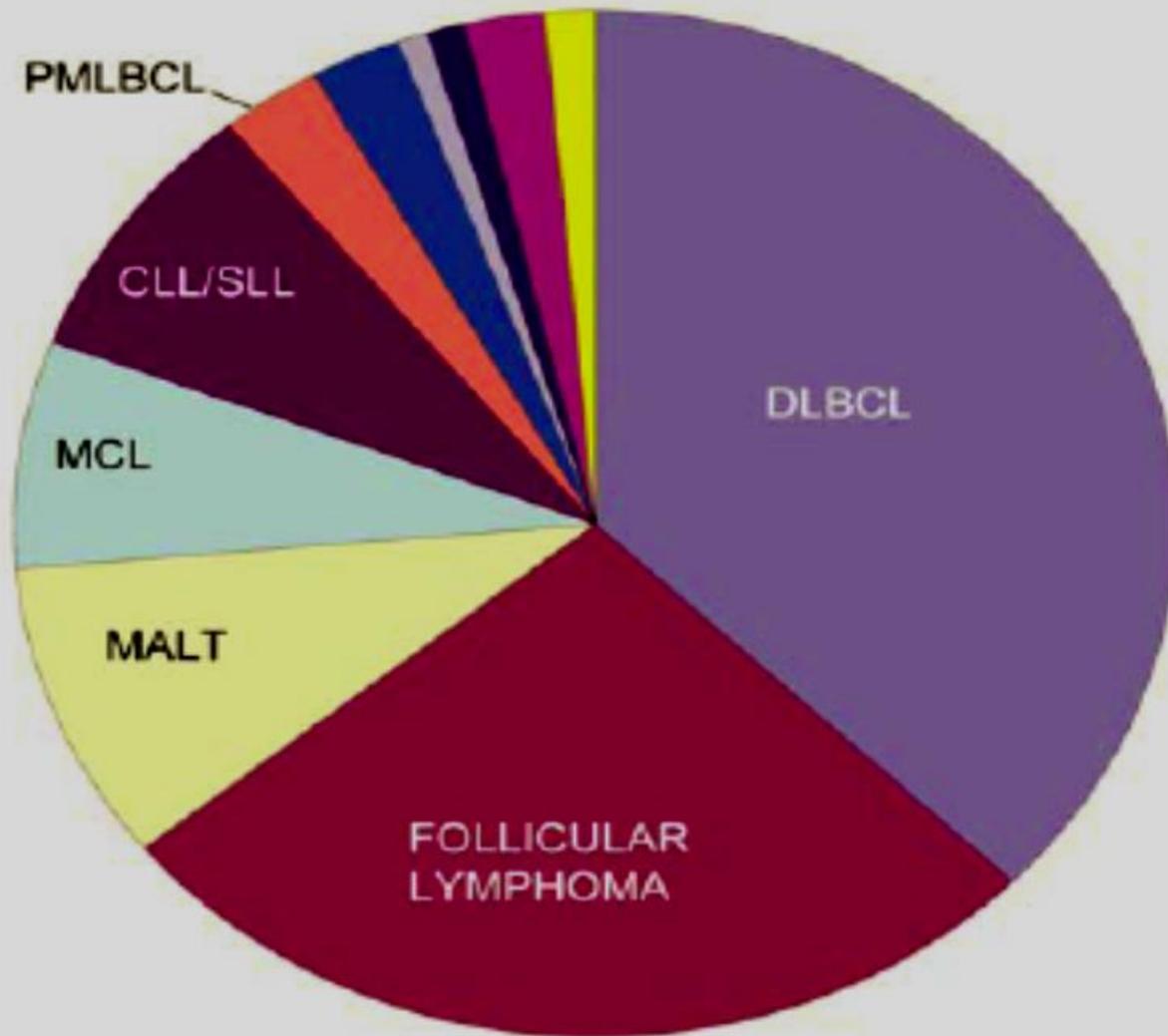
Mediastinal clear cell lymphoma

2001 : OMS : PMBCL

Definition WHO 2008

A diffuse large B-cell lymphoma arising in the mediastinum from putative thymic origin with distinctive clinical, immunophenotypic and genotypic features

Incidence of all aggressive B-cell L: 43%



Diffuse large B-cell 37%

Follicular 29%

MALT lymphoma 9%

Mantle cell lymphoma 7%

CLL/SLL 12%

Primary med large B-cell 3%

High Grade B, NOS 2.5%

Burkitt 0.8%

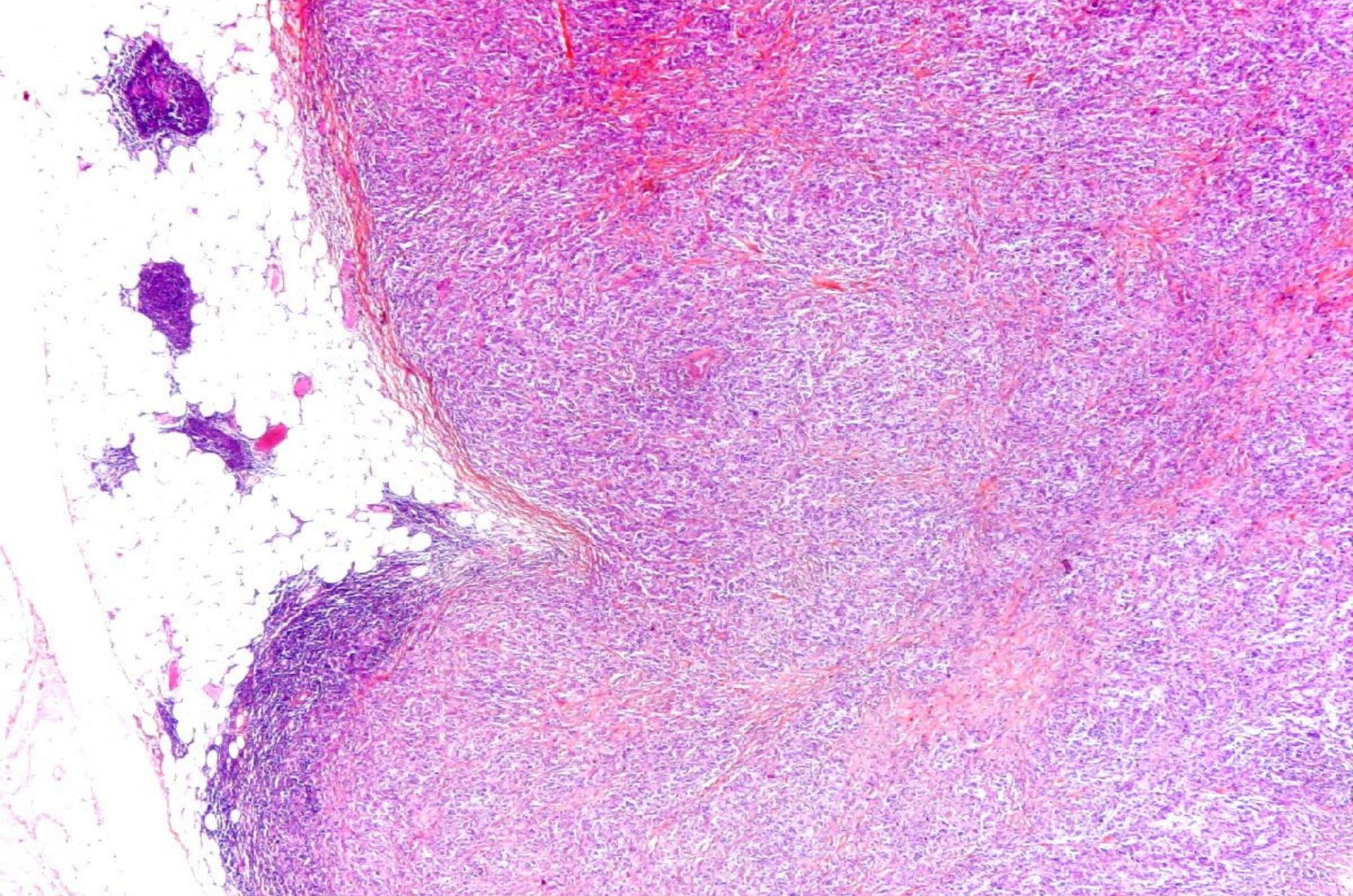
Splenic marginal zone 0.9%

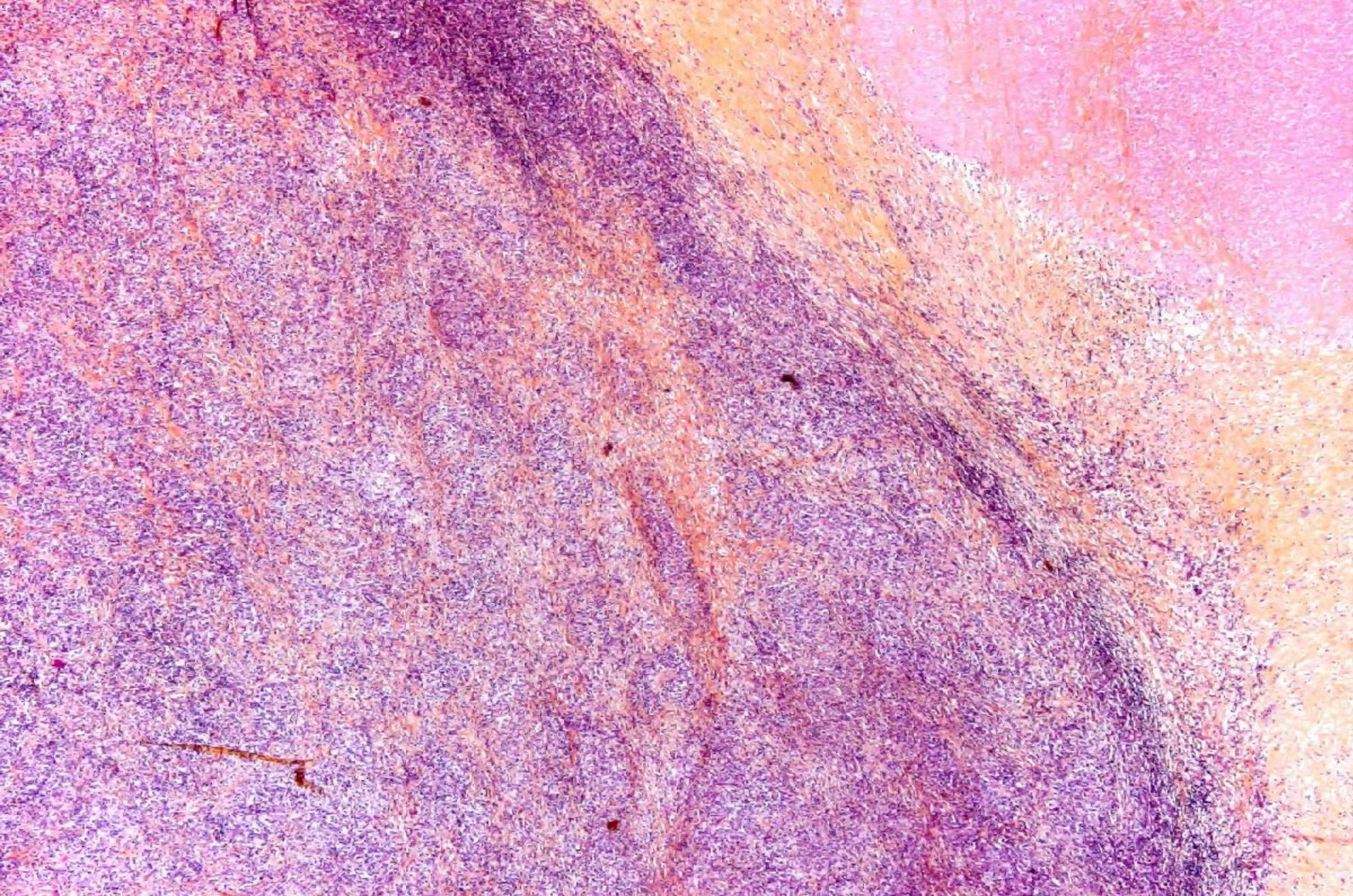
Nodal marginal zone 2%

Lymphoplasmacytic 1.4%

PMLBCL

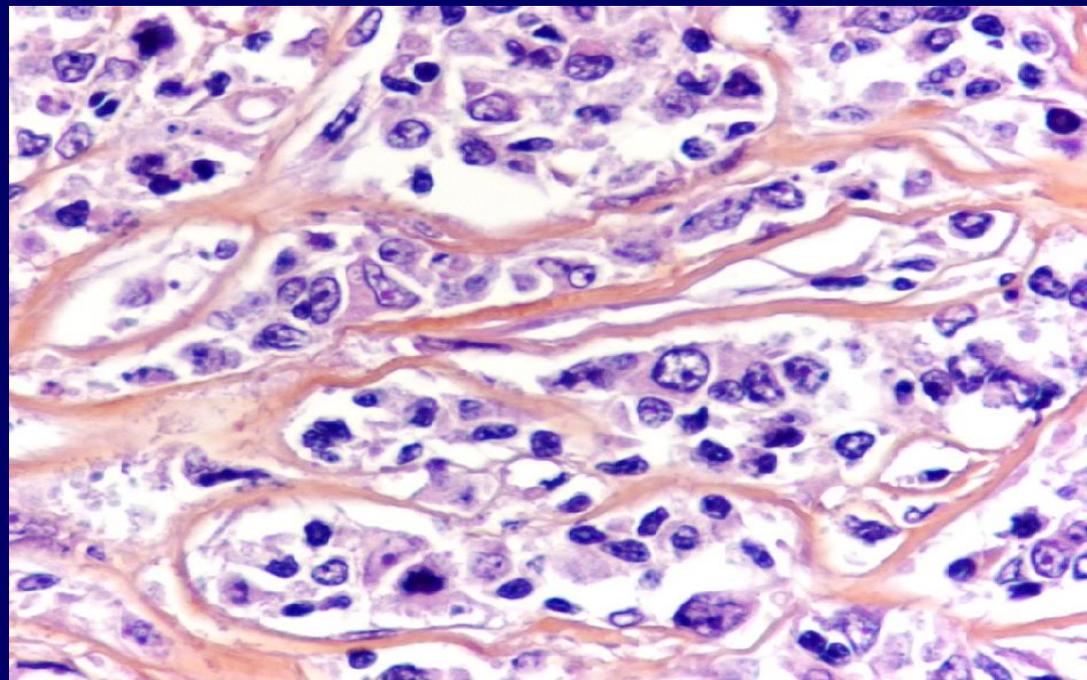
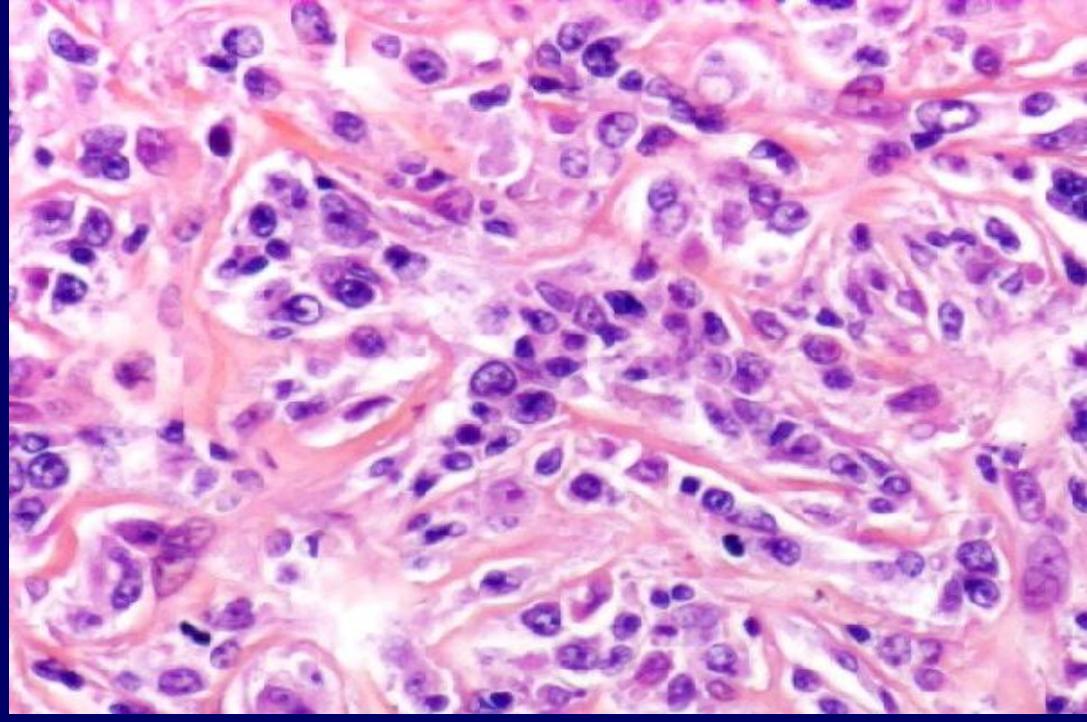
- 2,4 % des LNH
- âge moyen < 40 ans, 2F / 1M
- masse médiastinale (bulky: 50%:>10cm), stade I/II avec symptômes liés à compression/extension locale
- récurrence extra-ganglionnaire: ovaire, rein, surrénale, intestin, cerveau, peau
- BOM : + : 3%
- bonne réponse à la polychimiothérapie
- facteurs de mauvais pronostic: >10cm, pleurésie





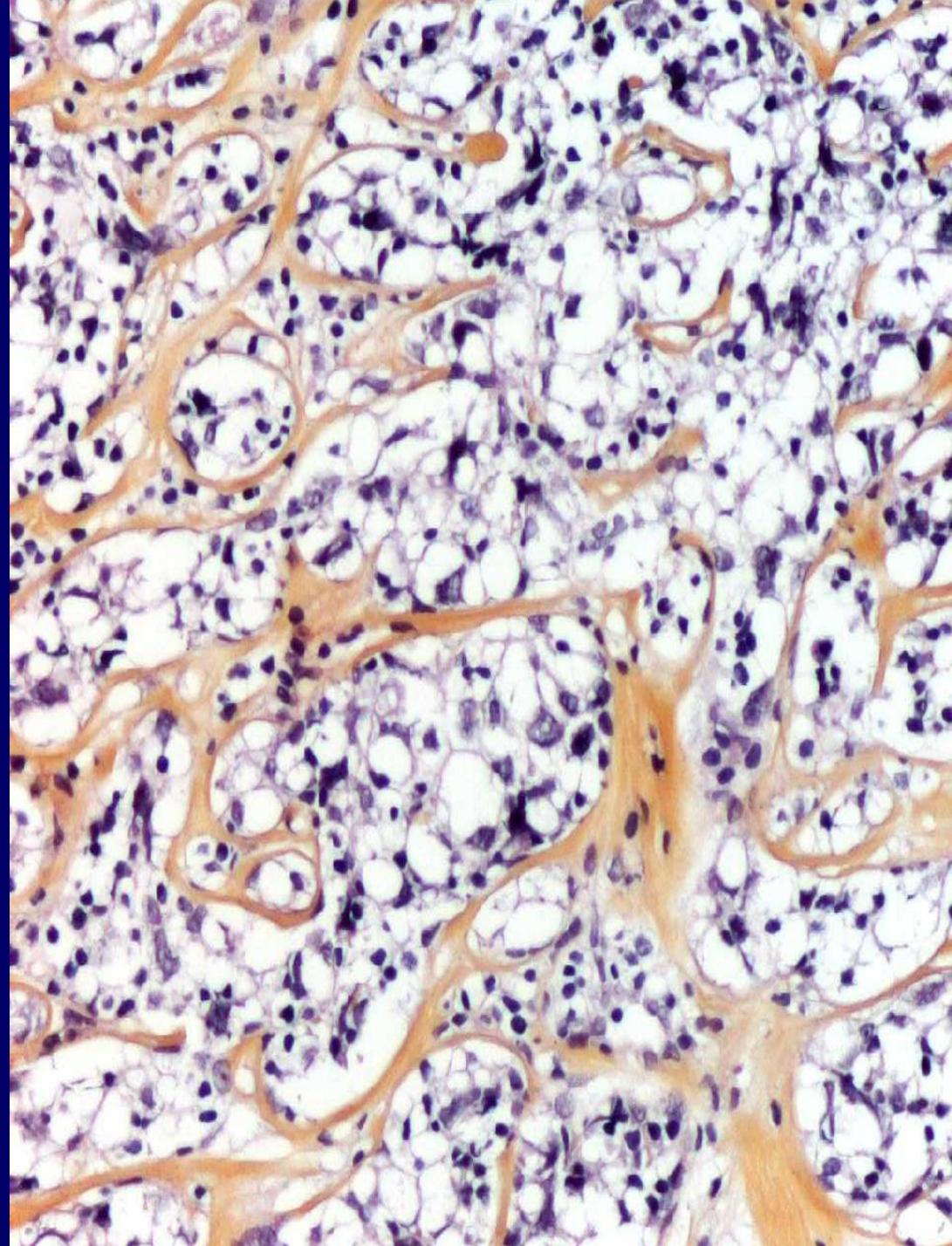
PMLBCL

- lymphome diffus avec **sclérose** “**compatimentante**”
- grandes cellules au noyau pléomorphe, lobulé et au cytoplasme abondant et clair (40%)
- parfois C/ RS-like
- nodularité focale (30%), aires de nécrose (25%)



PMLBCL

- lymphome diffus avec sclérose
“compartmentante”
- grandes cellules au noyau pléomorphe, lobulé et au **cytoplasme abondant et clair** (40%)
- parfois C/ RS-like
- nodularité focale (30%),
aires de nécrose (25%)

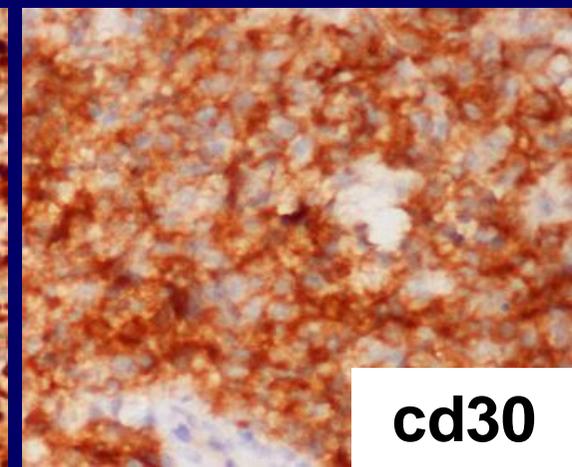
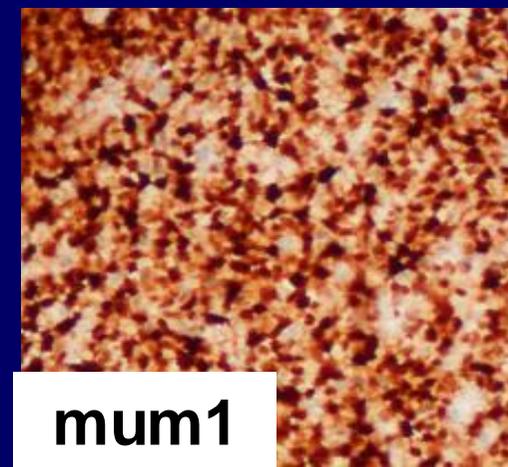
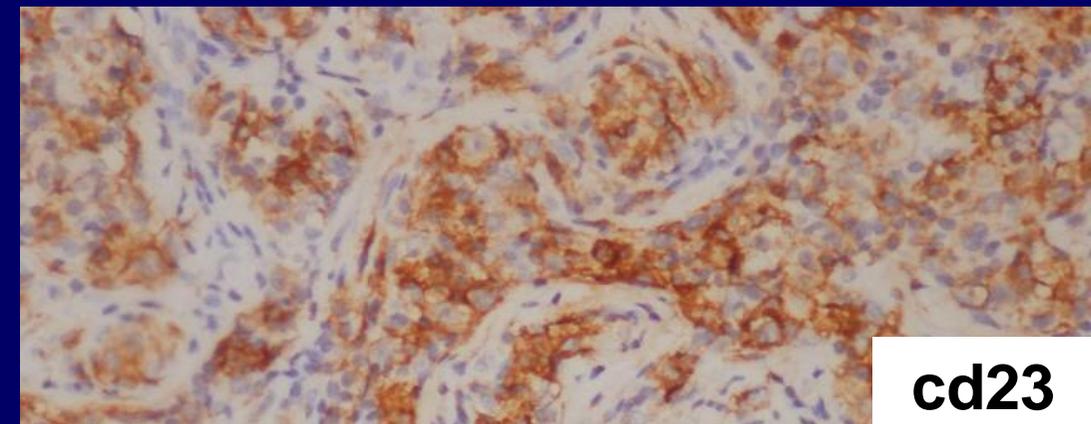
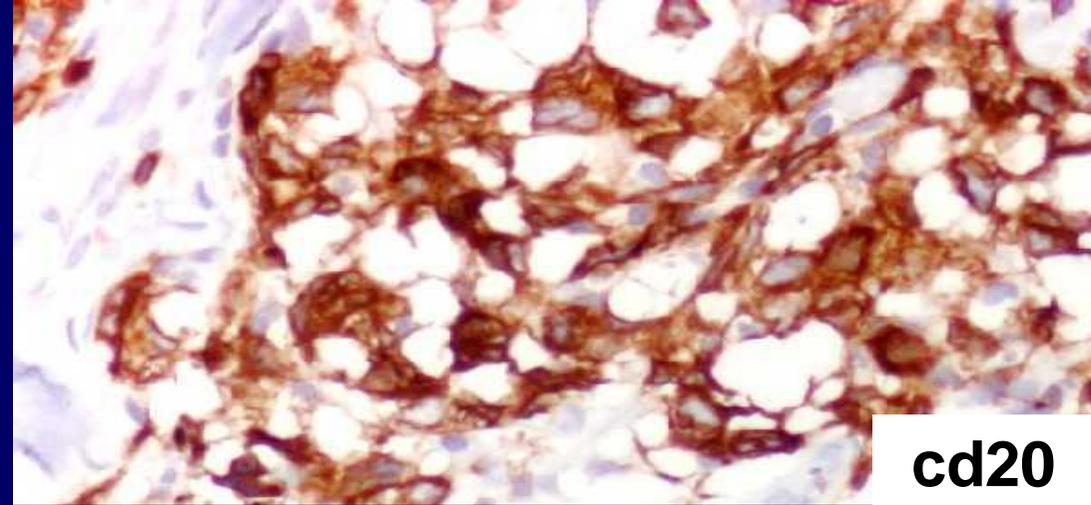


PMBCL

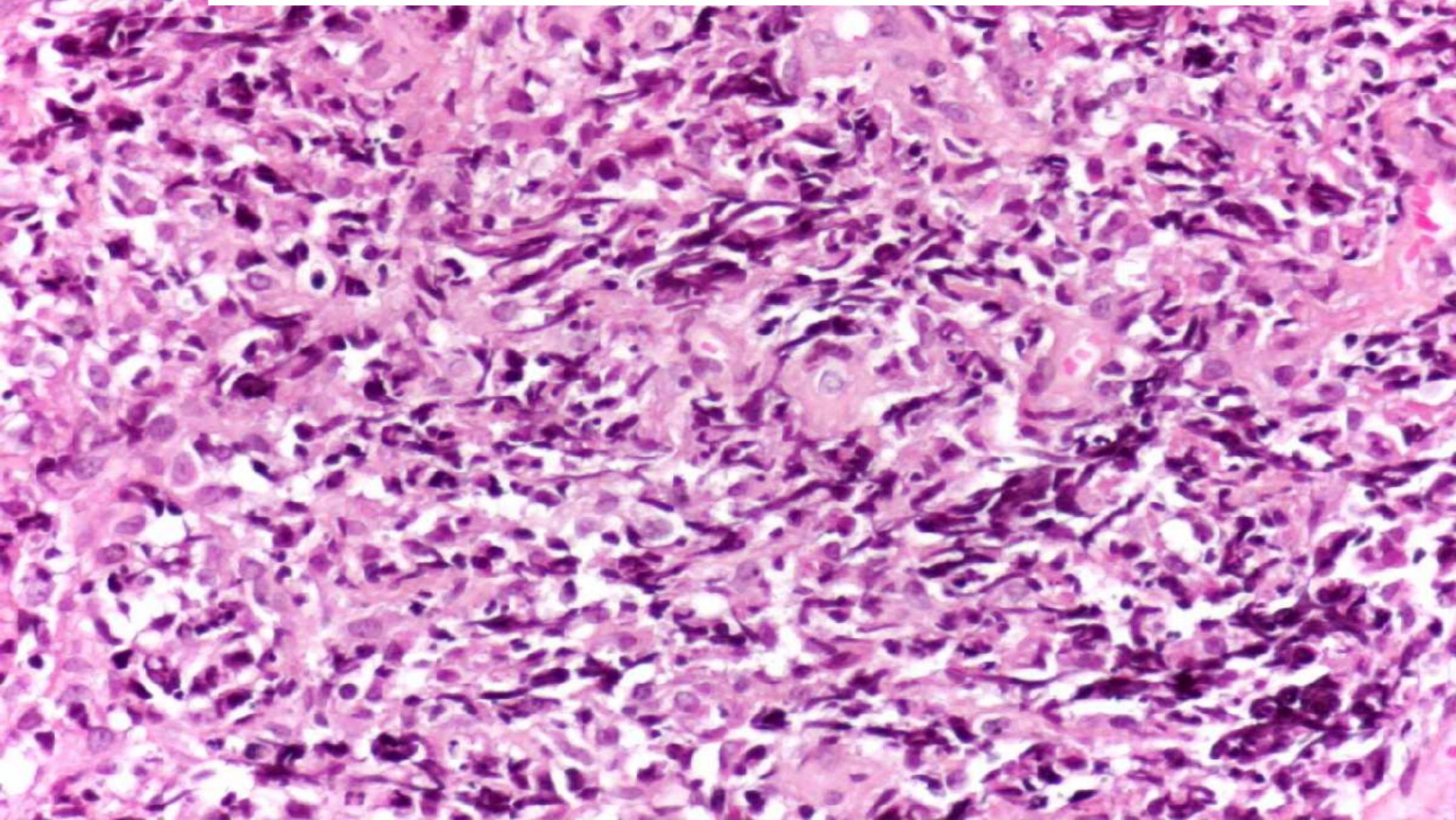
+ : **CD20**, CD45,
CD79a, PAX5, Oct2,
Bob1, CD45 (60%),
CD30 (80% variable
intensity), **MAL**

Variably + : CD10 (10-
30%), **CD23 (70%)**,
BCL6, BCL2 (60-
80%), MUM1 (75%)

- : **CD15**, CD21, CD68,
CD138, **EBV**



Masse médiastinale: « crash biopsy! »
IHC : cd20, Mib1, kératine, PCR B

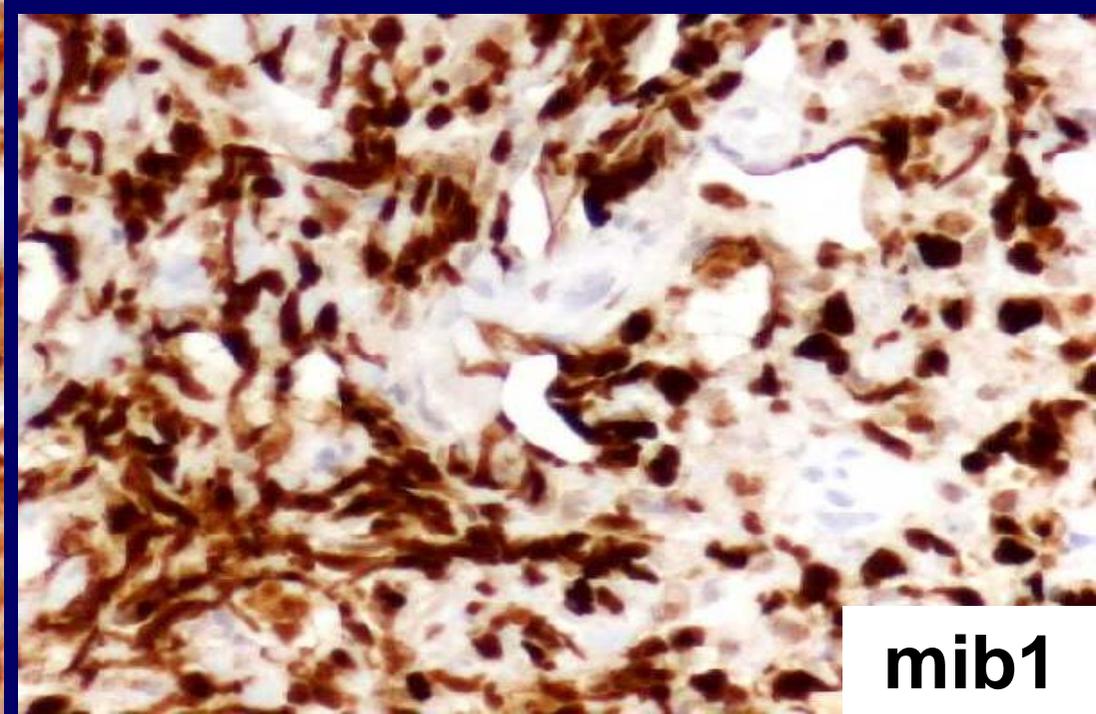
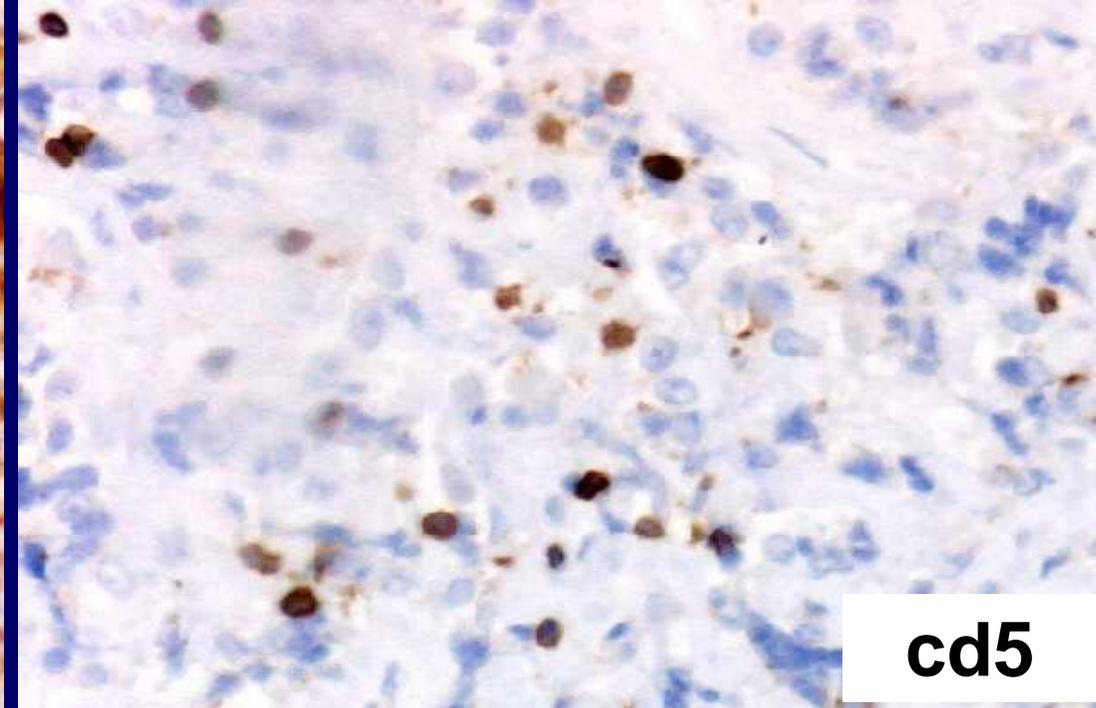
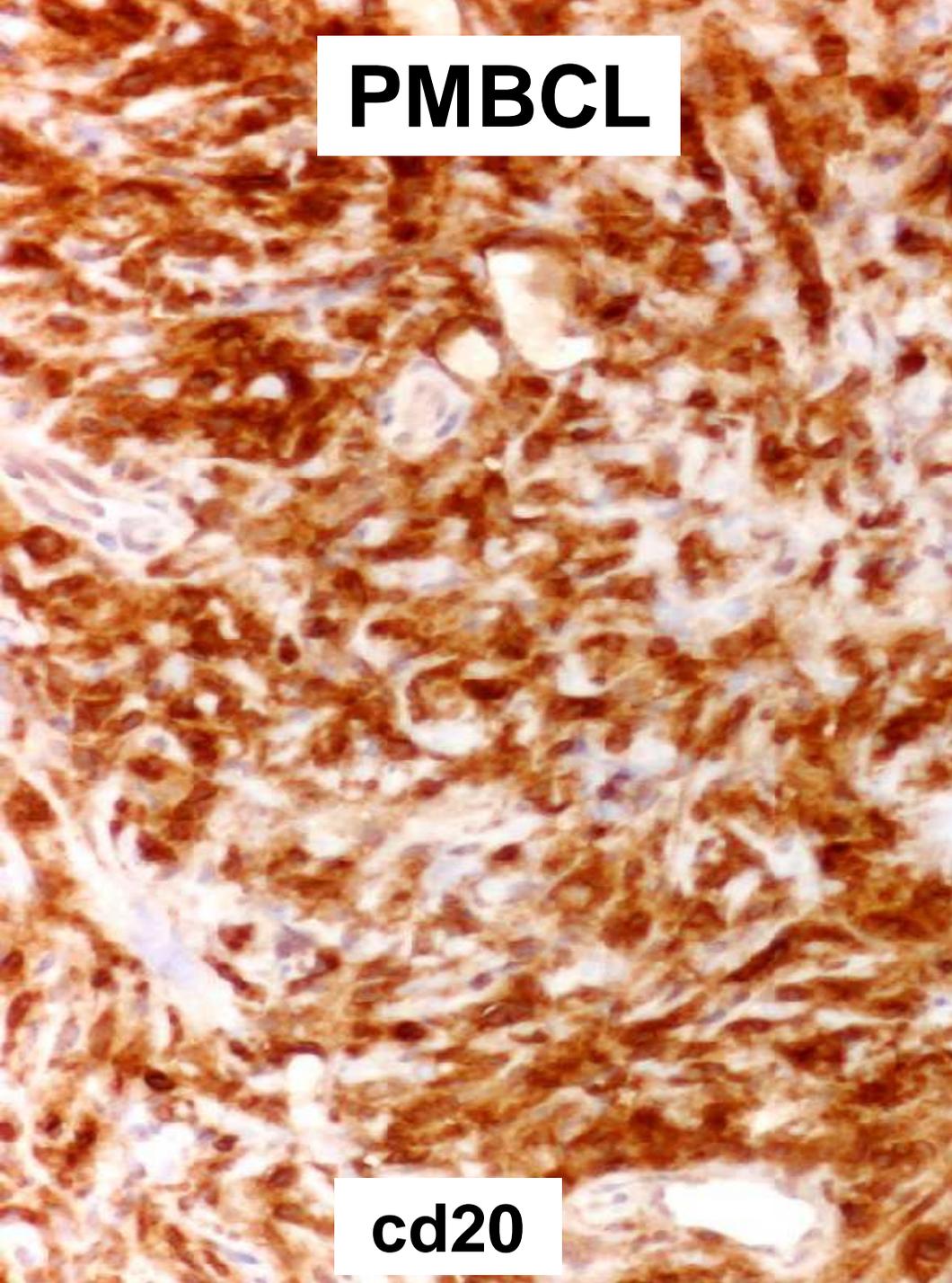


PMBCL

cd20

cd5

mib1



Lymphome à Grandes C/ B du Médiastin

- réarrangement des chaînes des Ig
- pas de translocation chromosomique spécifique
- réarrangement bcl 6 : 5%
- mutation bcl 6 : 10%
- bcl 2 non réarrangé
- gène MAL surexprimé : 7/9 / 0/8 DLBCL
(gène de différenciation des λ T)

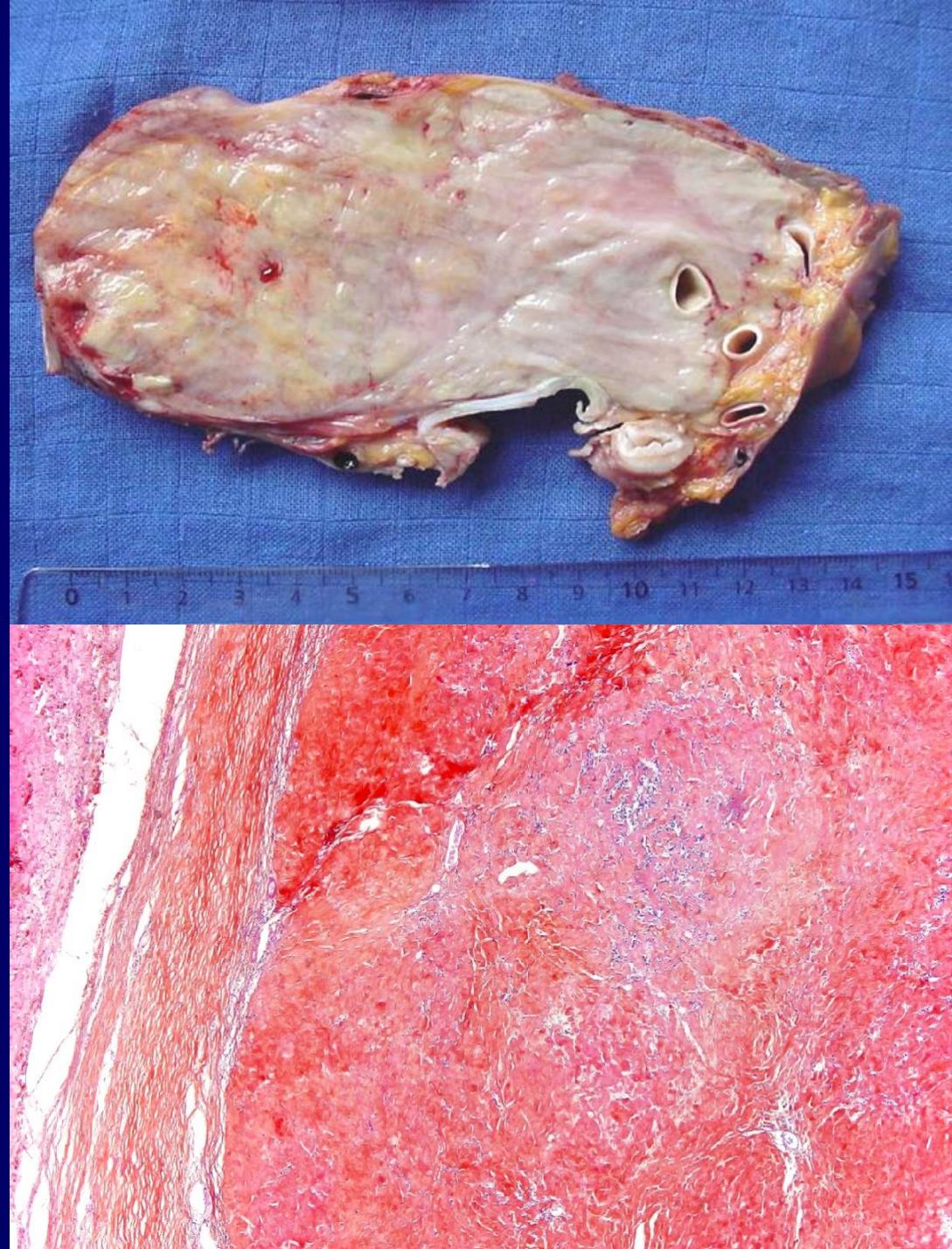
Lésions Médiastinales Non Résécables avec Fibrose de l'Adulte

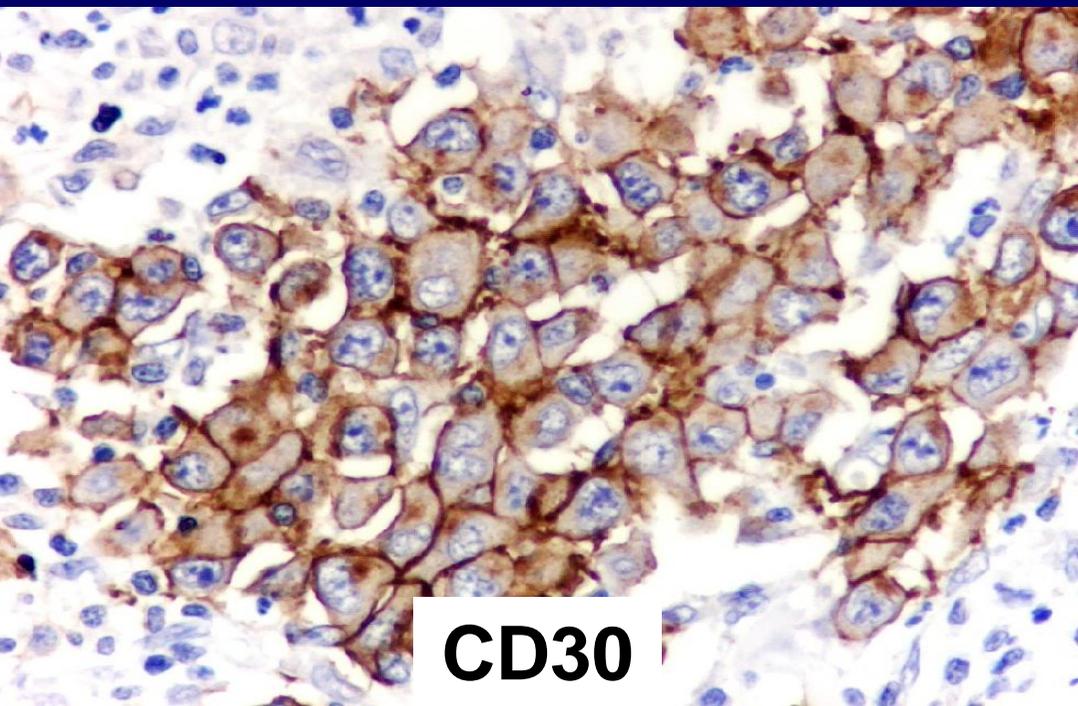
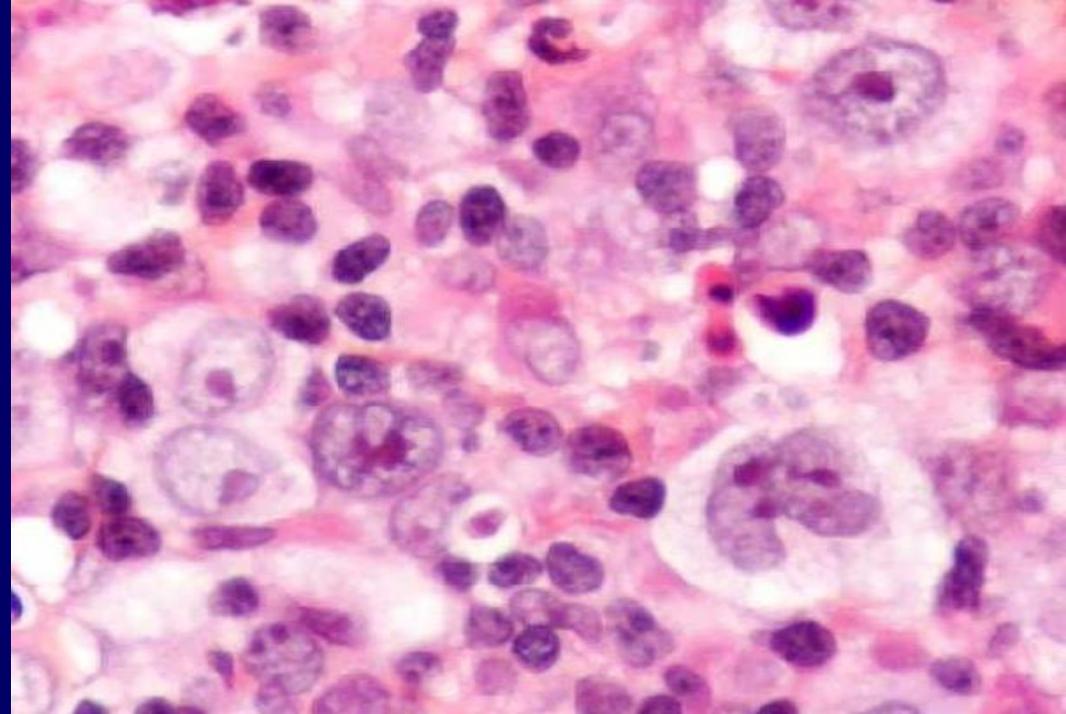
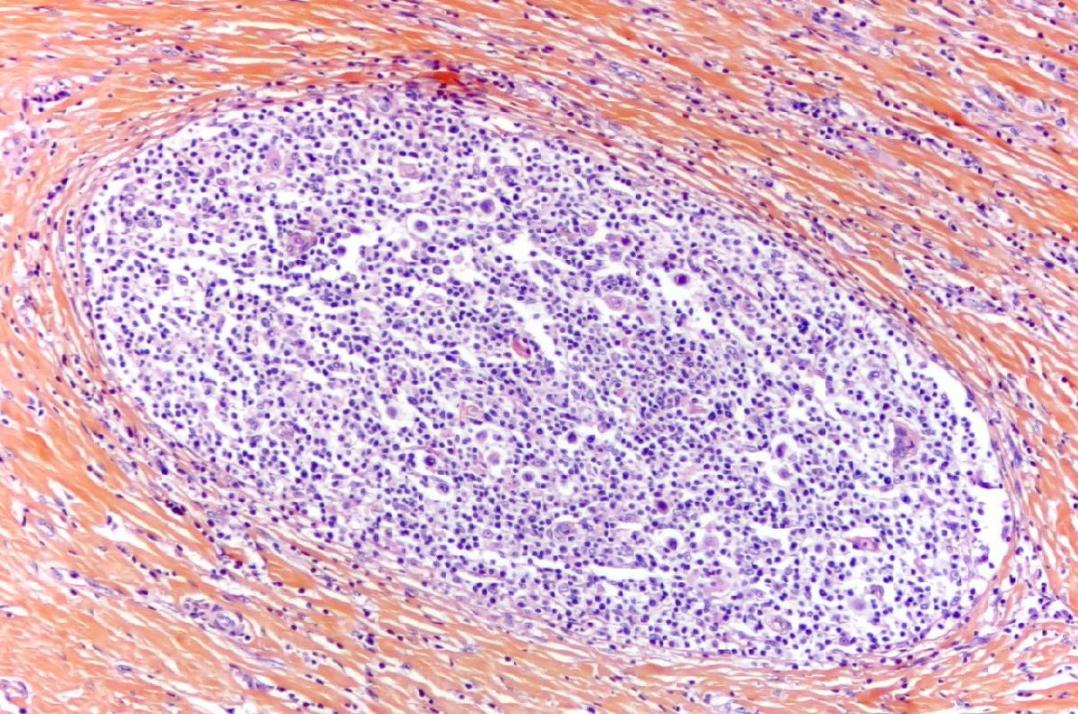
- adulte jeune
- volumineuse tumeur
- médiastinoscopie : biopsies difficiles



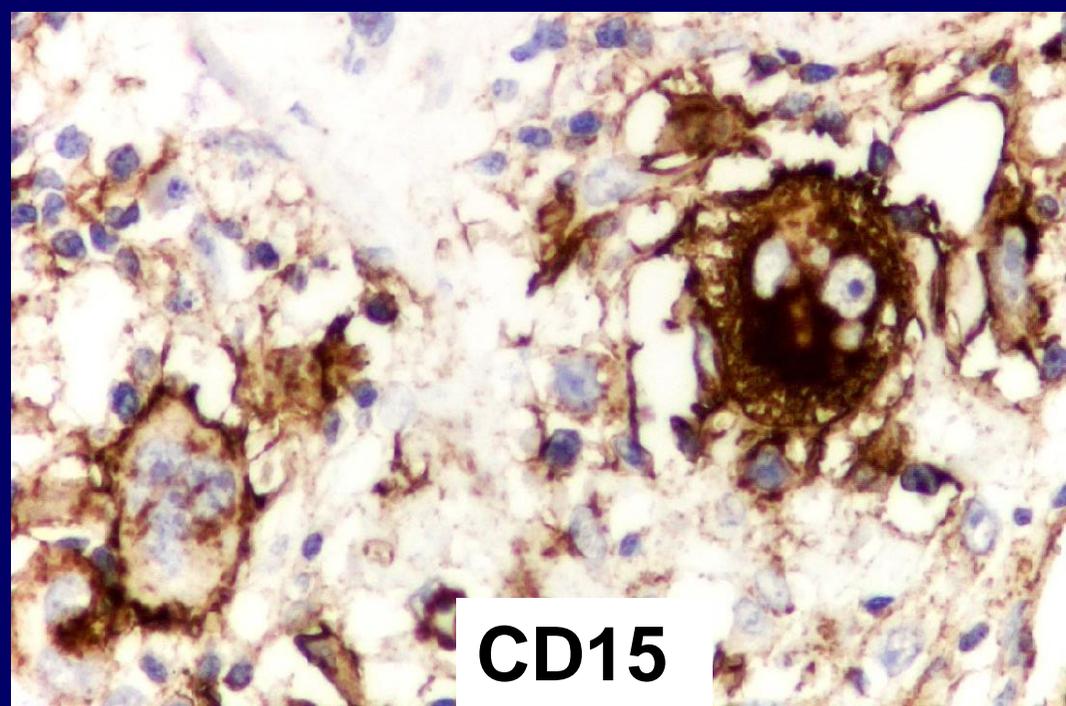
Tumeurs du Médiastin avec sclérose

- Lymphome de Hodgkin
- Lymphome à grandes cellules B du médiastin
- Grey zone H / DLBCL
- Séminome
- L lymphoblastique (jeune)
- Schwannome





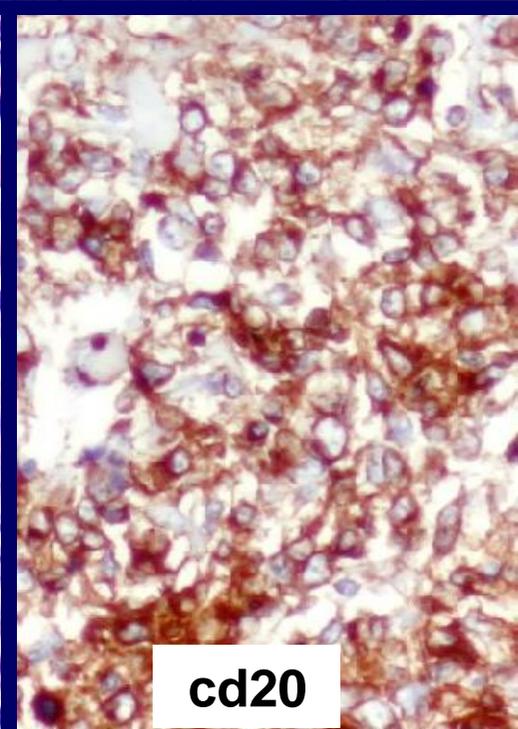
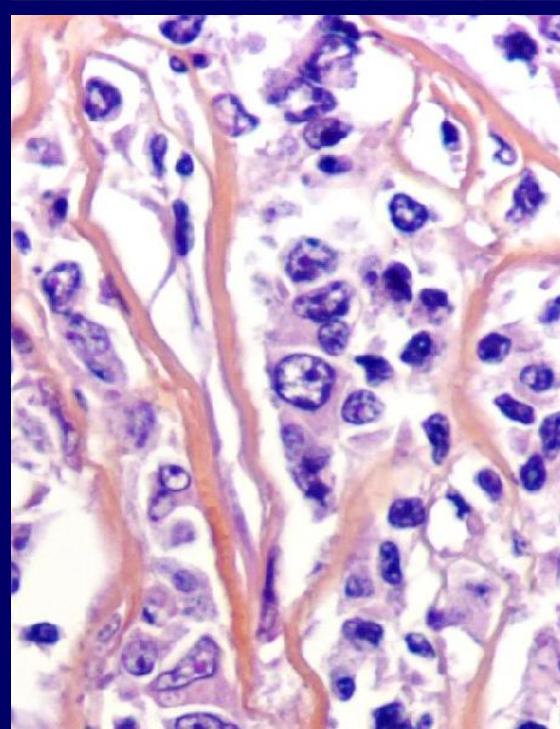
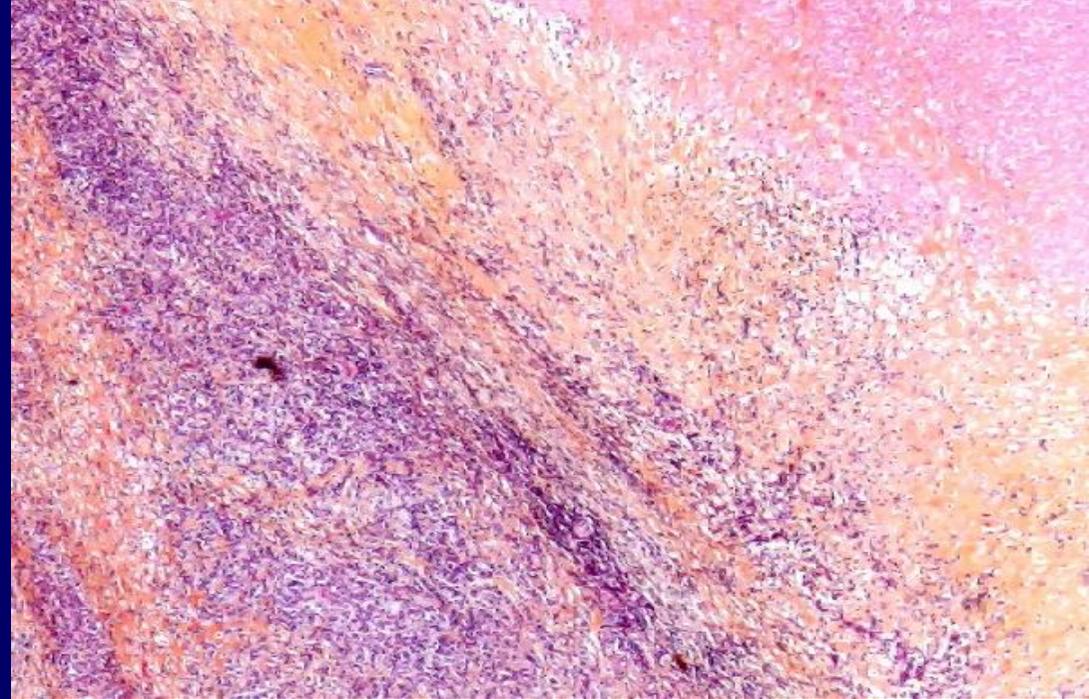
CD30



CD15

Tumeurs du Médiastin avec sclérose

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- **Lymphome à grandes cellules B du médiastin**
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- Schwannome



cd20

Tumeurs du Médiastin avec sclérose

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- **Grey zone H / DLBCL**
- Séminome
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- Schwannome

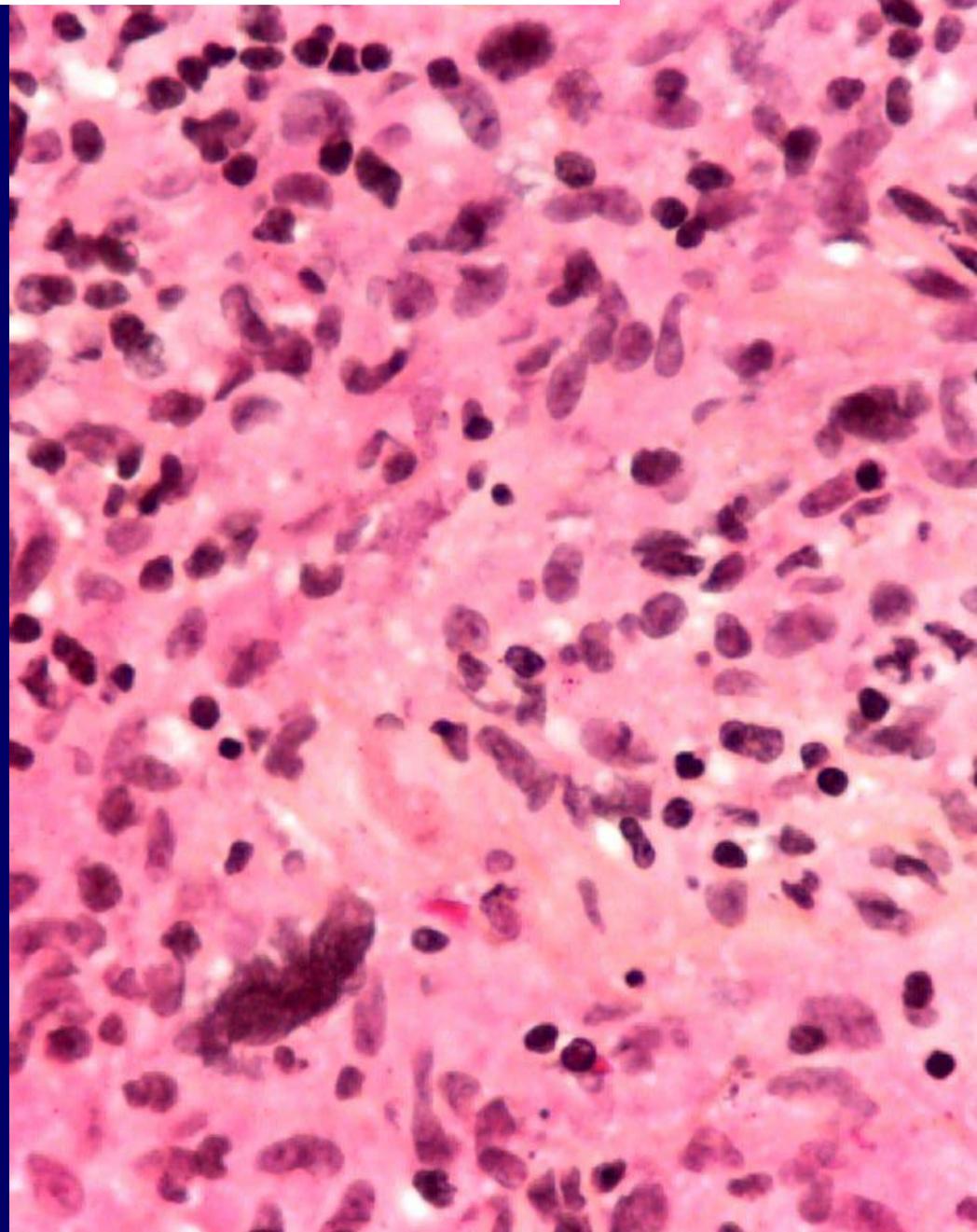
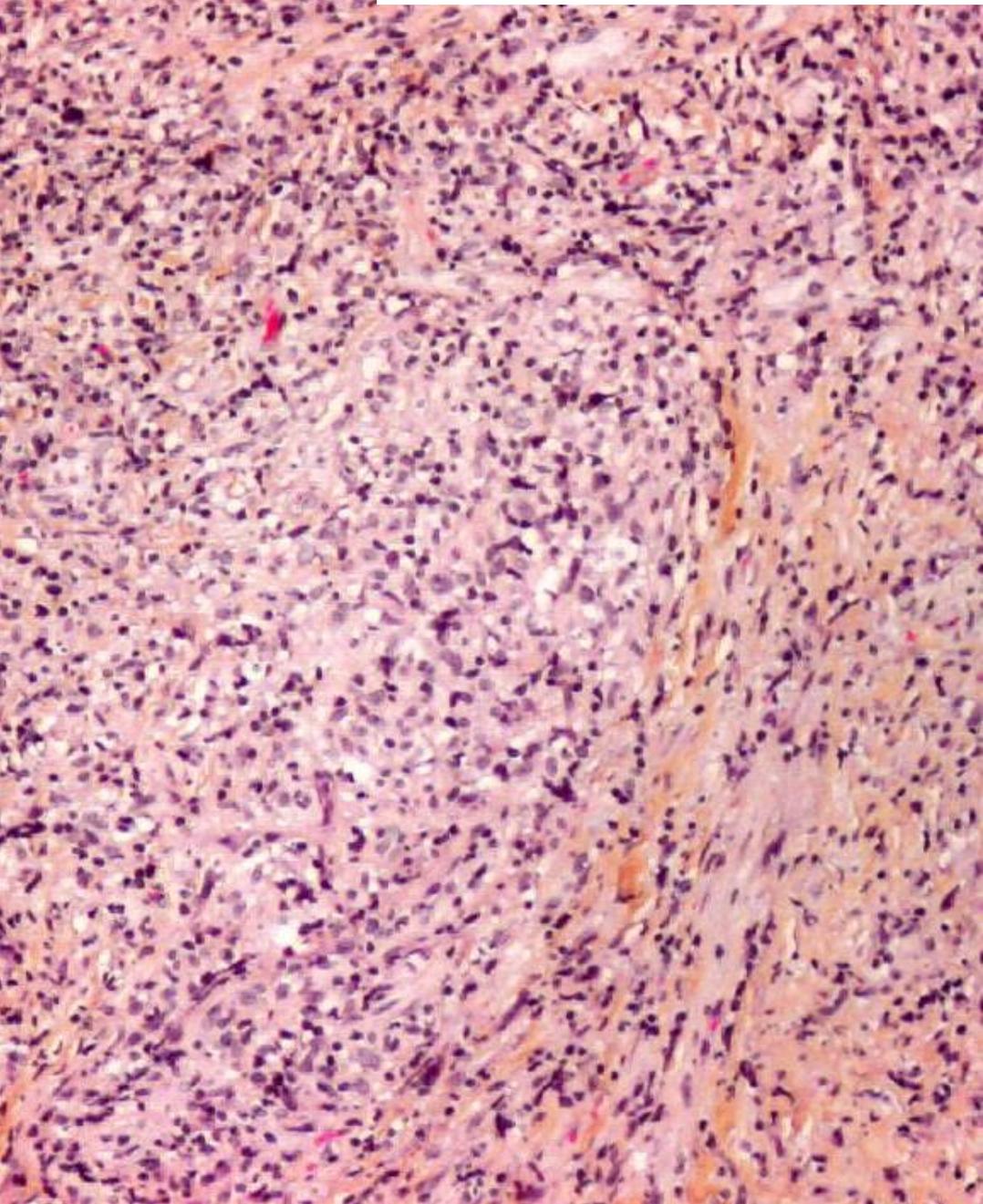
Definition WHO 2008

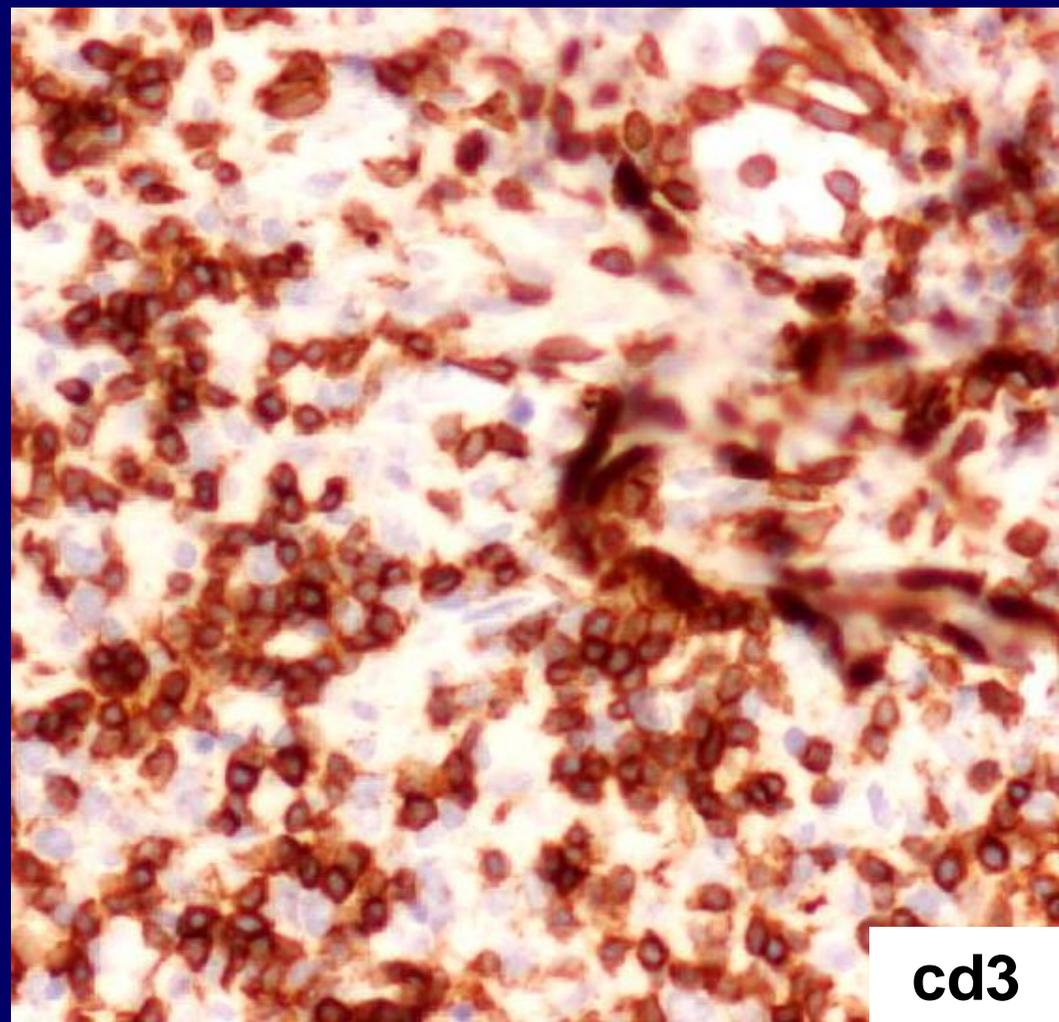
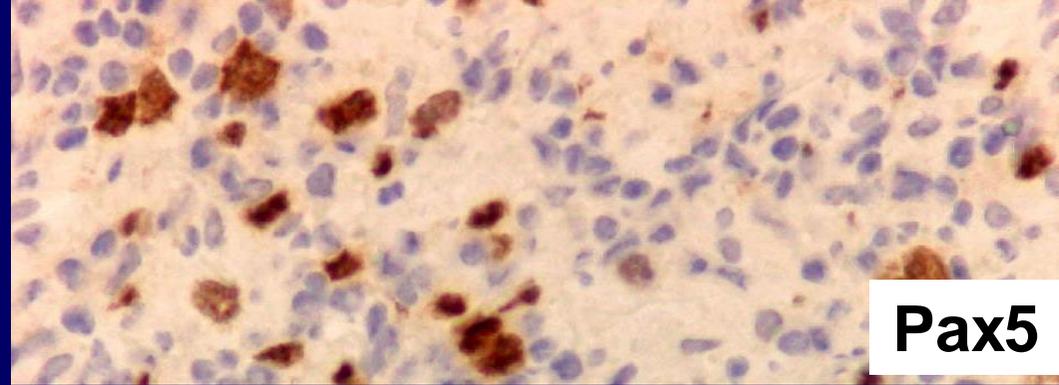
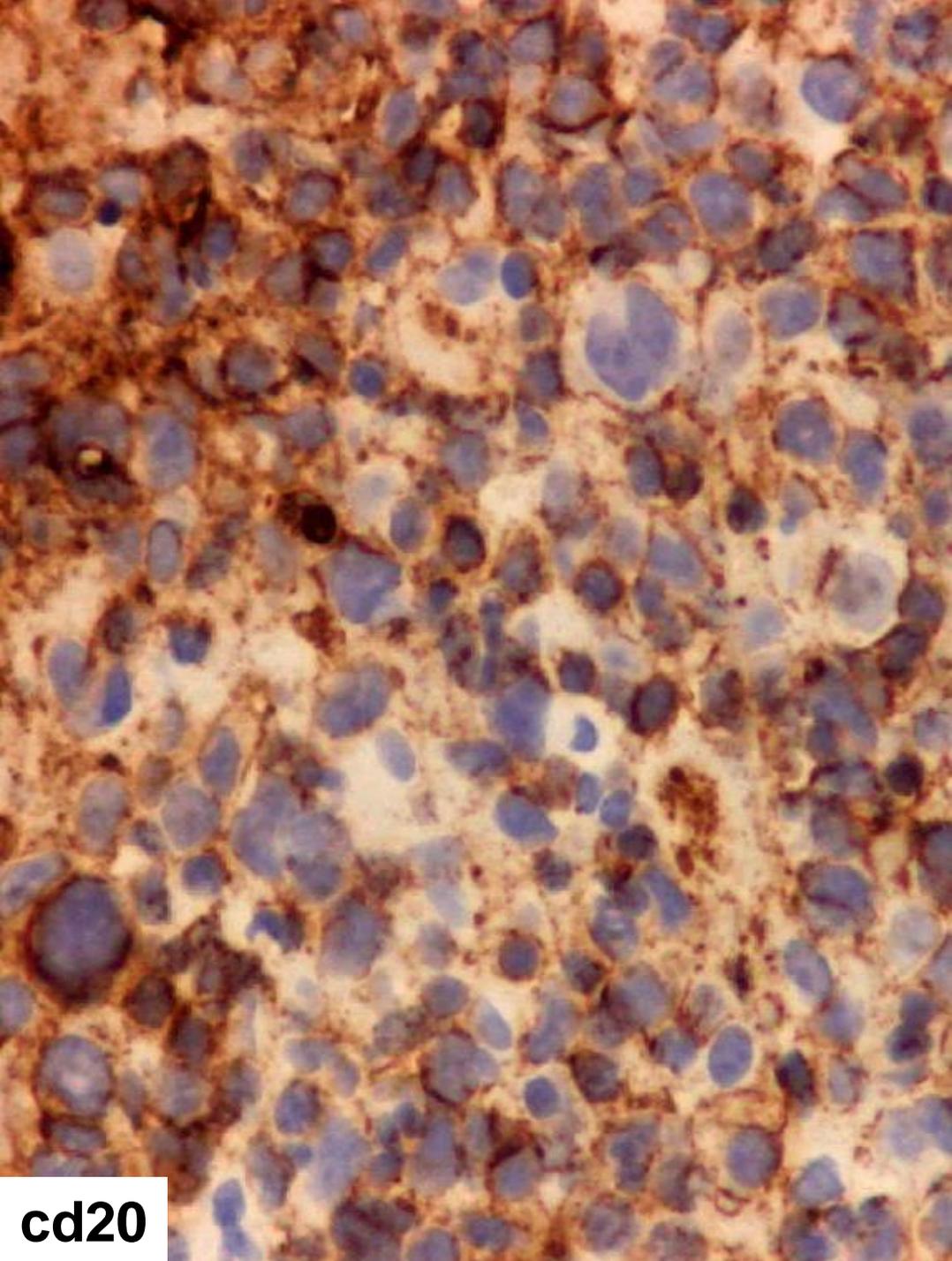
- Lymphome B qui montre un chevauchement clinique, morphologique et/ou immunophénotypique entre Hodgkin classique et DLBCL, en particulier le lymphome primitif médiastinal à grandes cellules B
- plus agressif que Hodgkin ou DLBCL. Pas de consensus thérapeutique

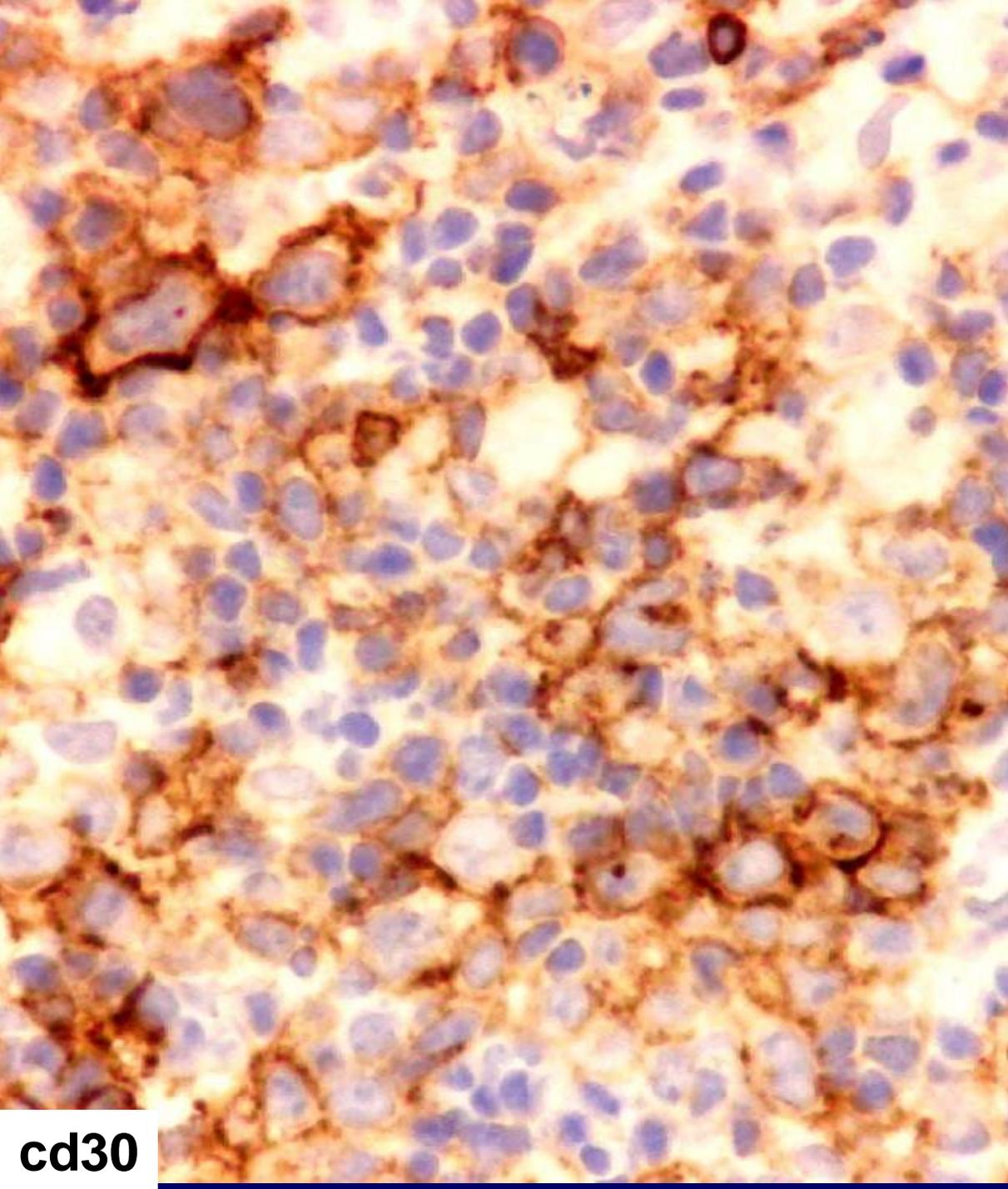
Mediastinal Grey Zone Lymphoma

- **CHL histology with**
 - Uniform strong expression of CD20
 - Intact B-cell program (OCT2, BOB1, PU1, PAX5)
 - Absence of CD15
- **PMBL histology with**
 - Absence of CD20 and other B-cell factors
 - Expression of CD30 & CD15
 - Presence of EBV

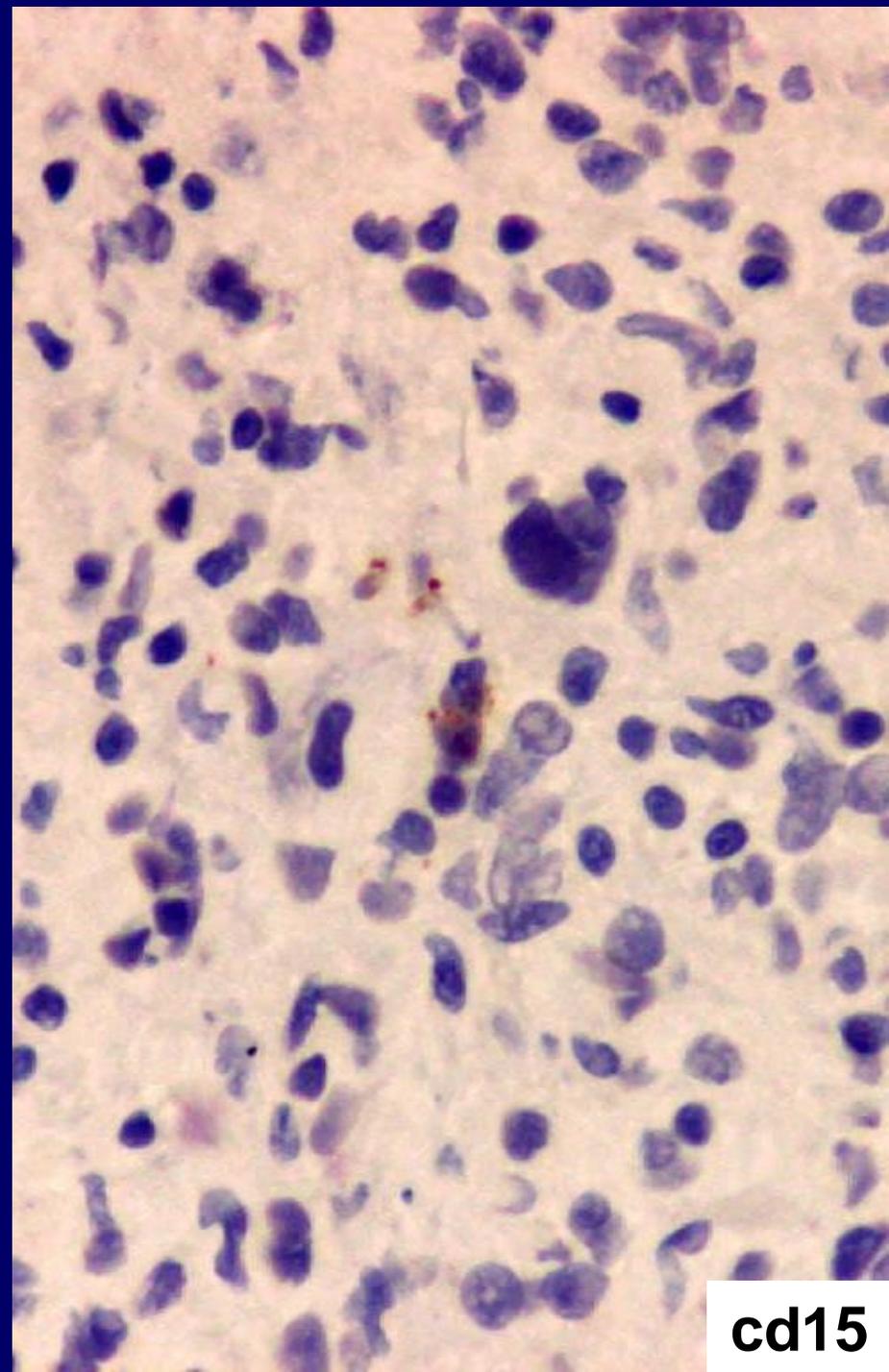
F, 29 ans, tumeur médiastinale







cd30

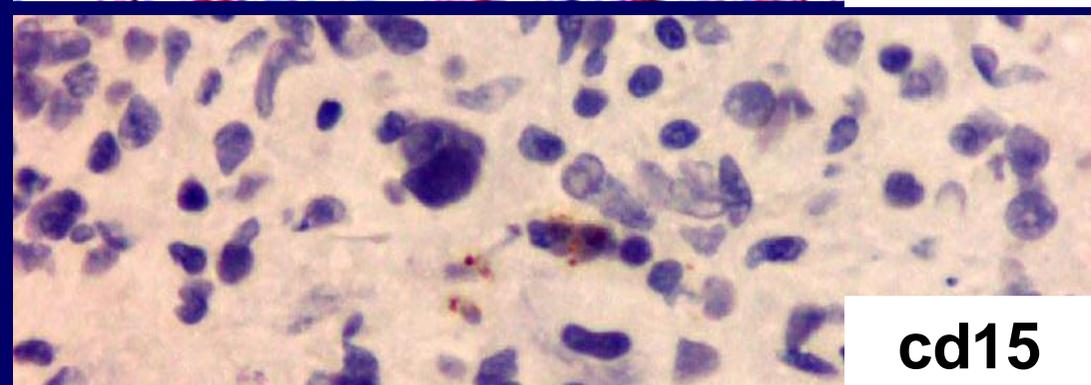
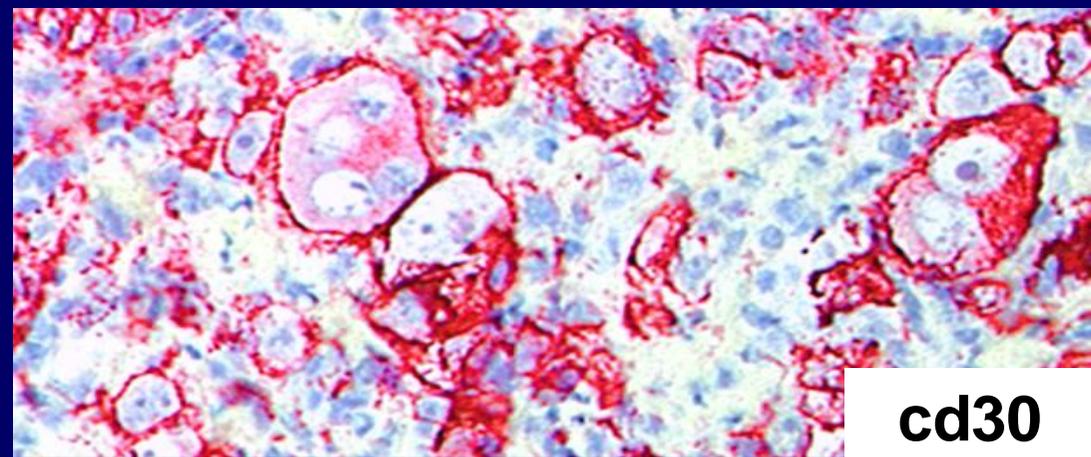
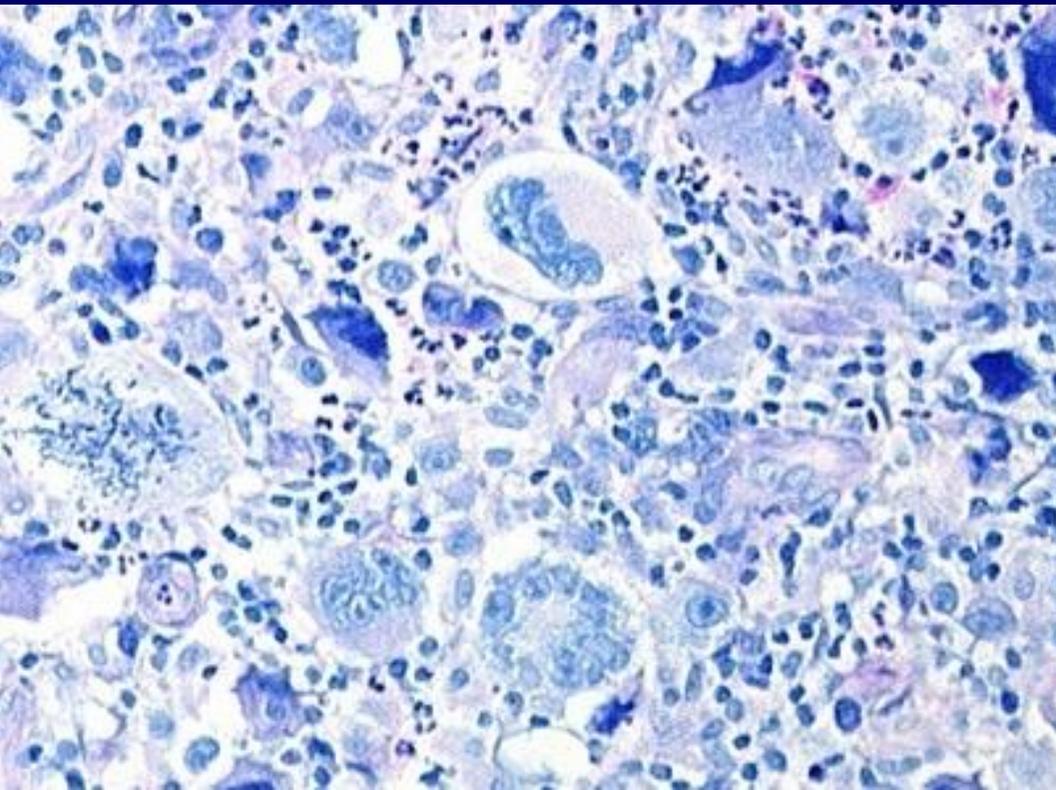
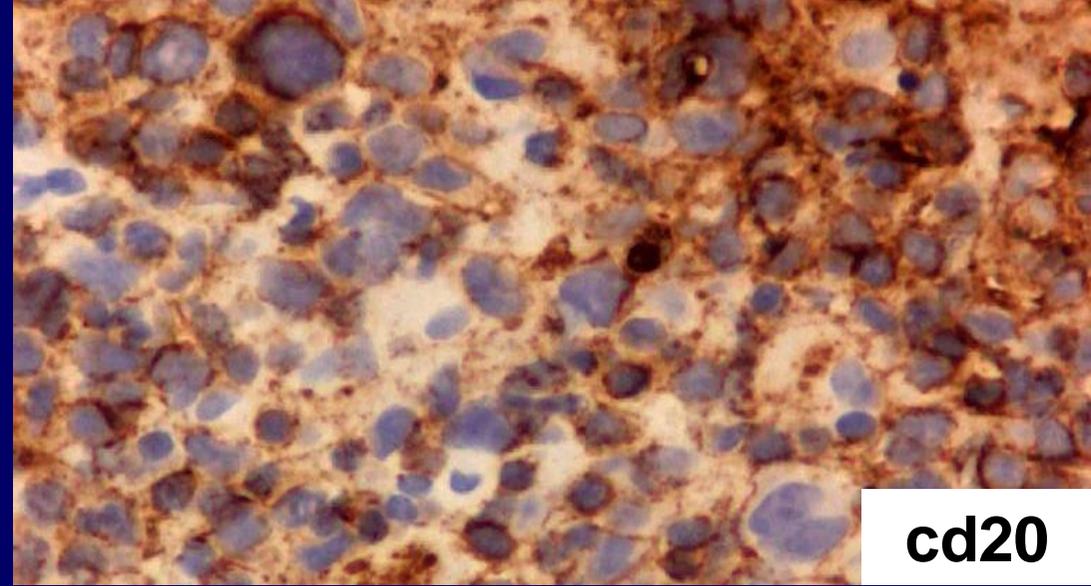


cd15

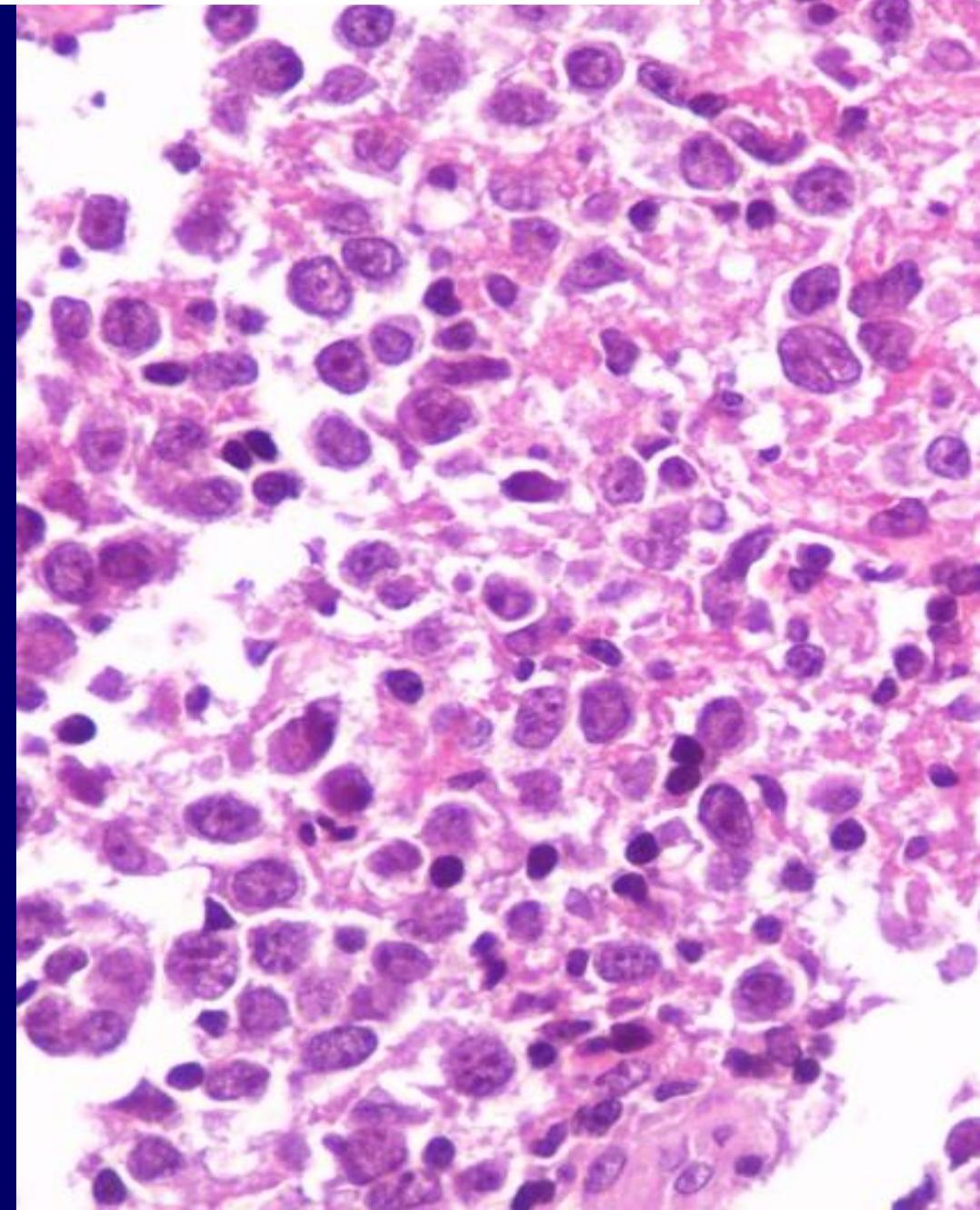
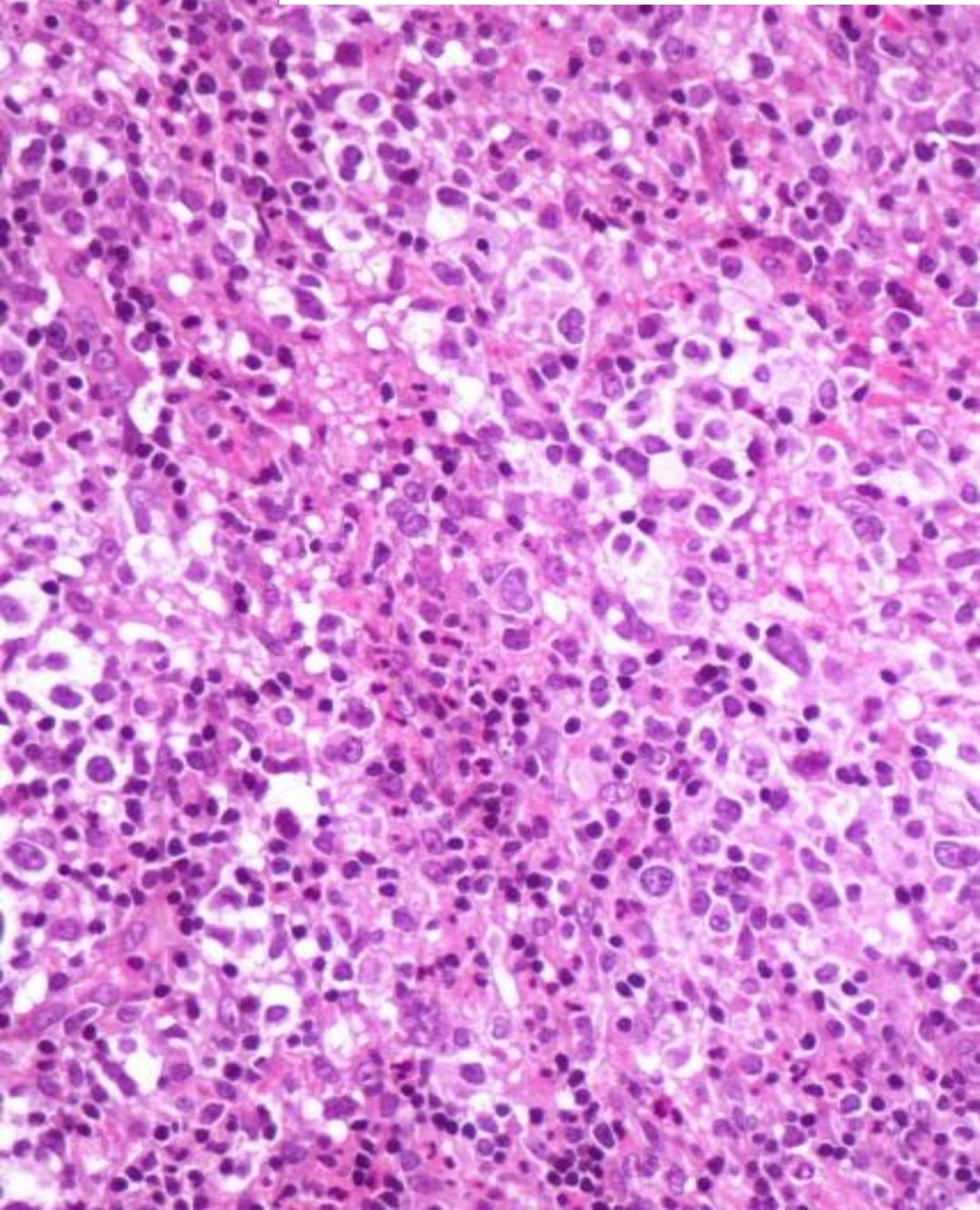
Mediastinal Grey Zone Lymphoma

histologie de CHL avec

- CD20 fort et uniforme
- programme B intact (OCT2, BOB1, PAX5)
- CD30 +, CD15 -



H, 35 ans, tumeur médiastinale antérieure

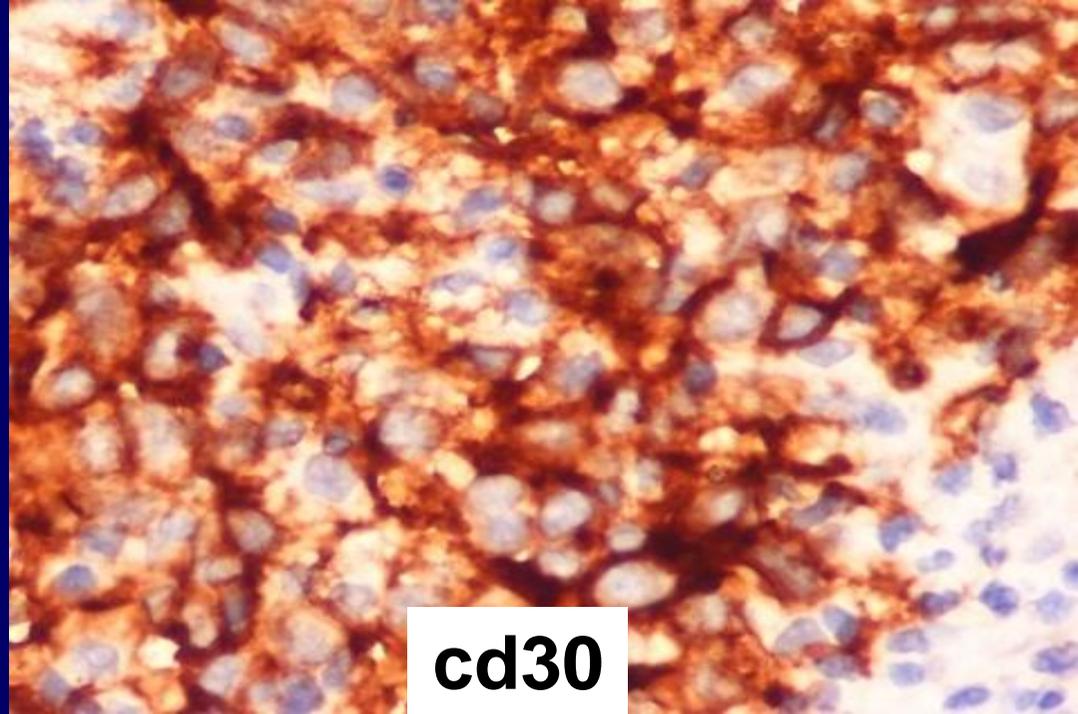


Mediastinal Grey Zone Lymphoma

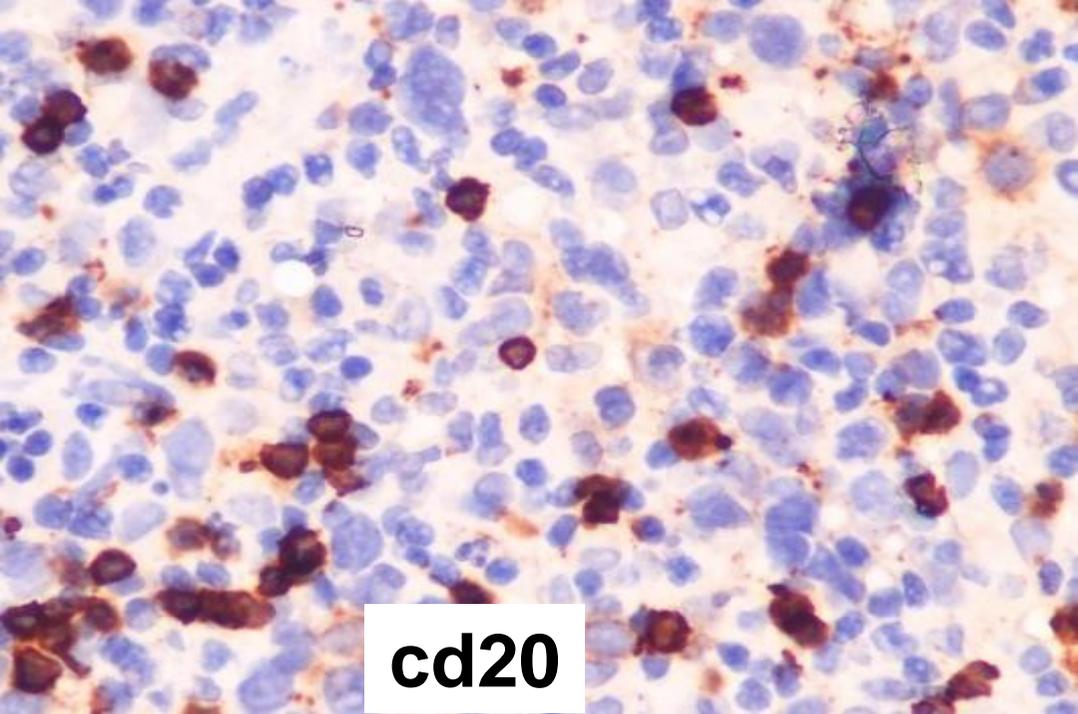
histologie de PMBL avec

- : cd20 , cd79a, OCT2,
BOB1, PAX5

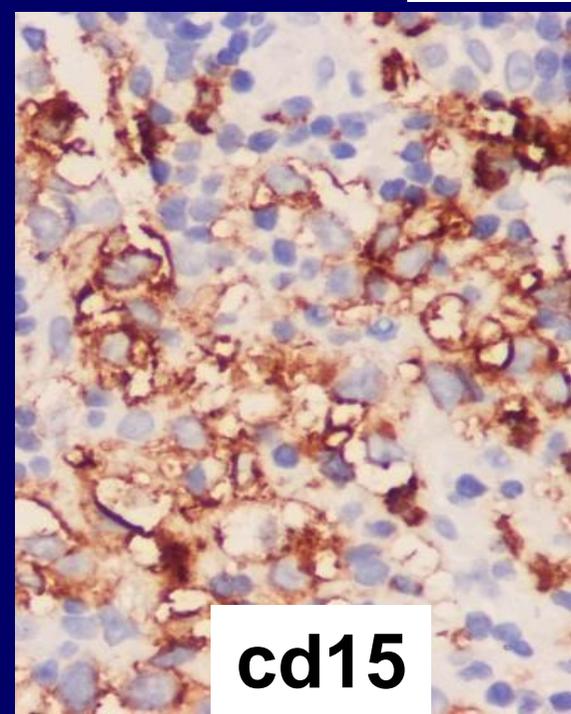
+ : cd30 , cd15 , EBV



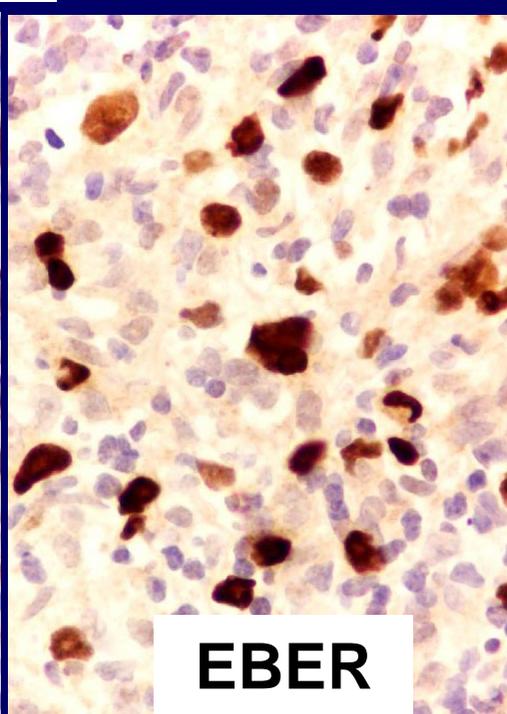
cd30



cd20



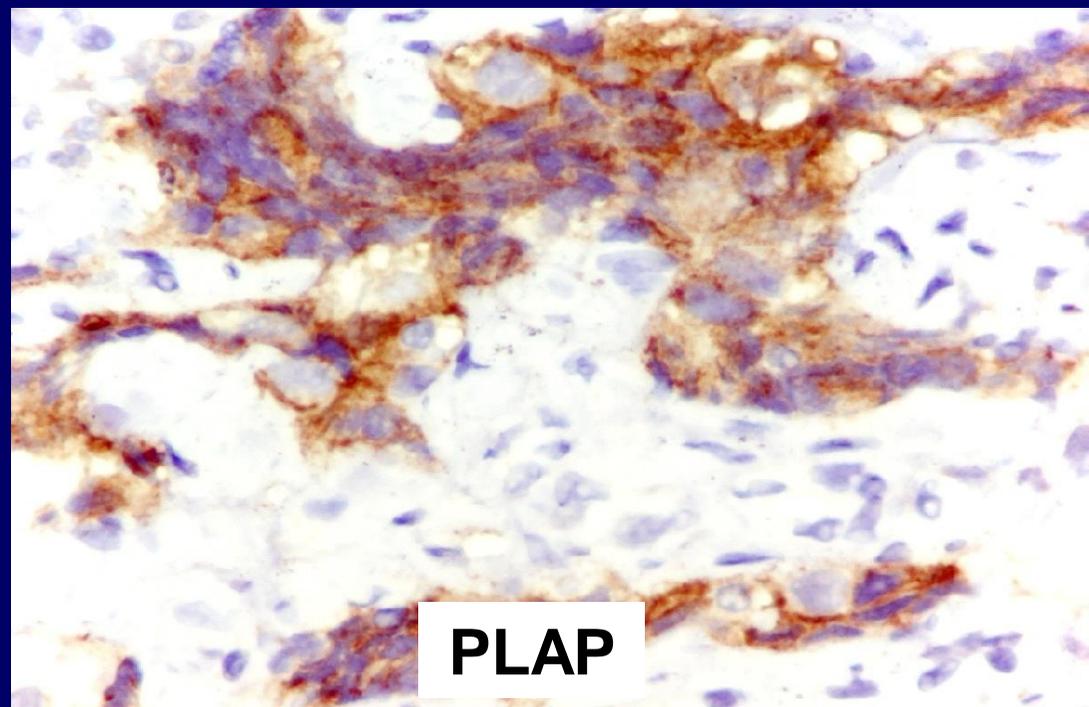
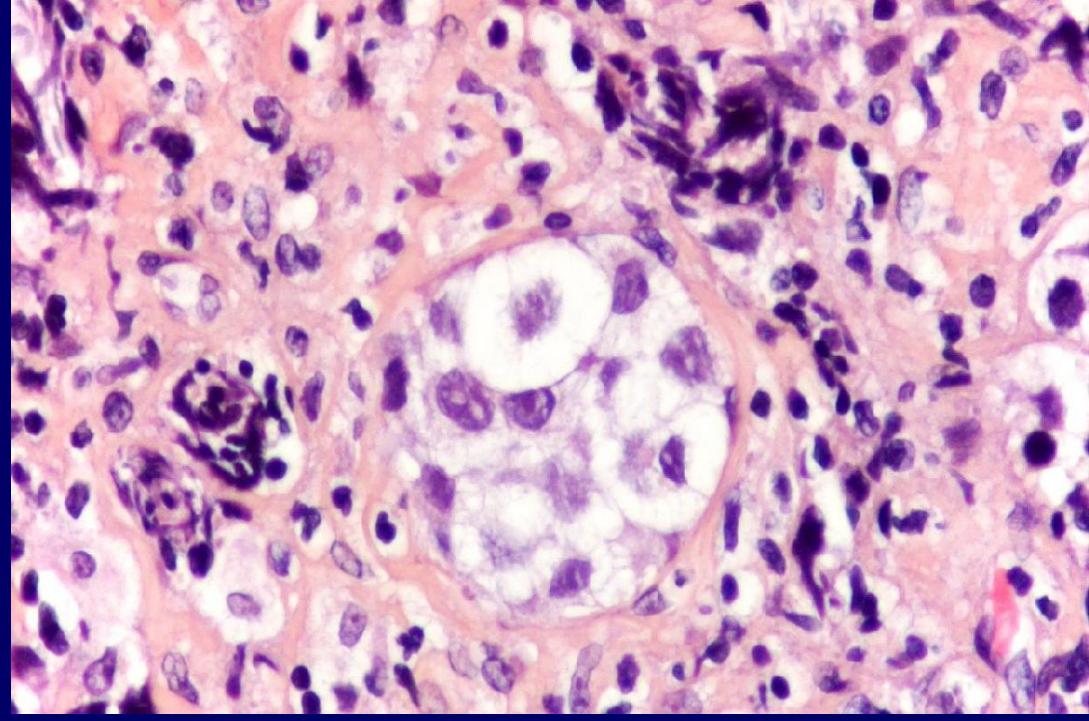
cd15



EBER

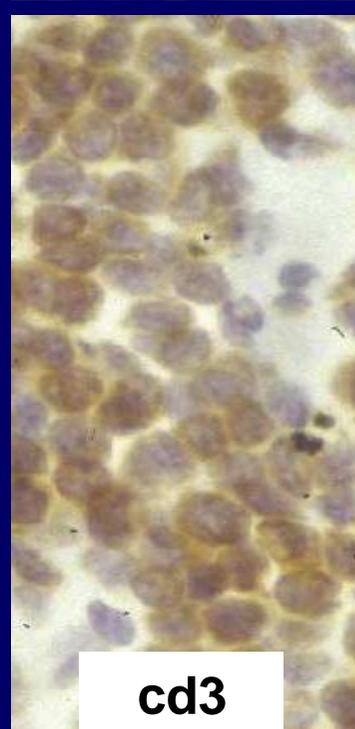
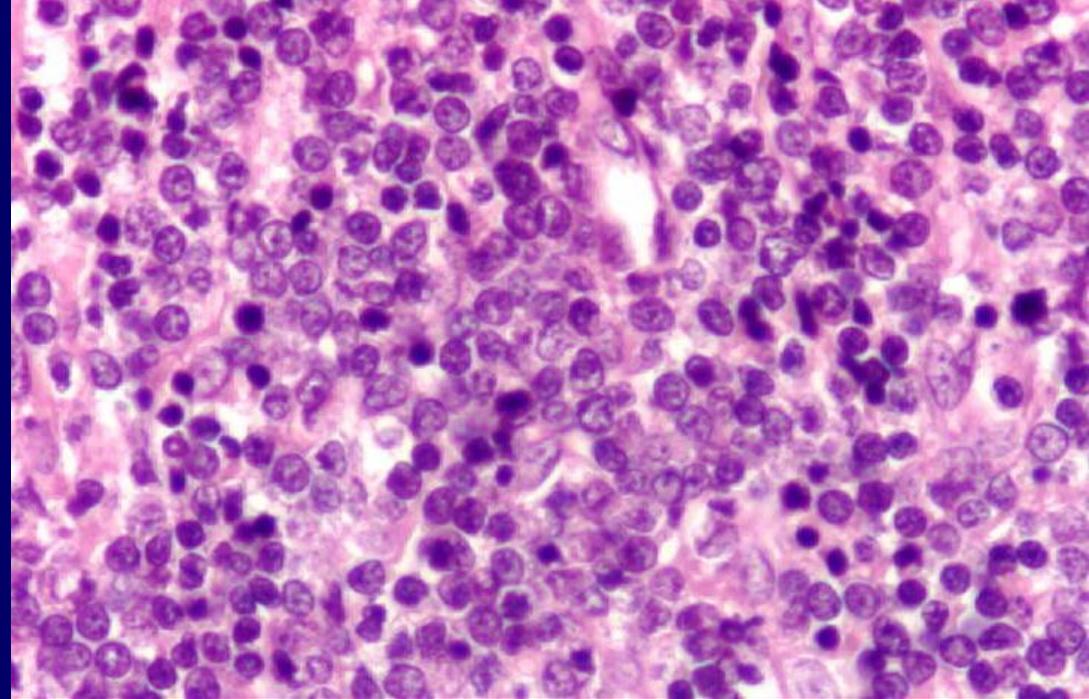
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- Lymphome à grandes cellules B du médiastin
- **Séminome**
- Lymphome lymphoblastique (jeune)
- Schwannome

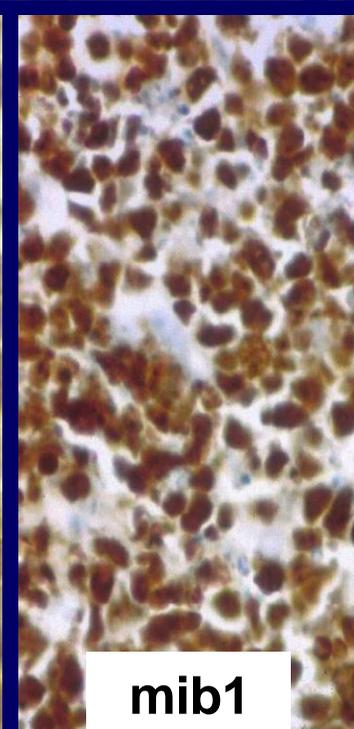


Tumeurs du Médiastin avec sclérose

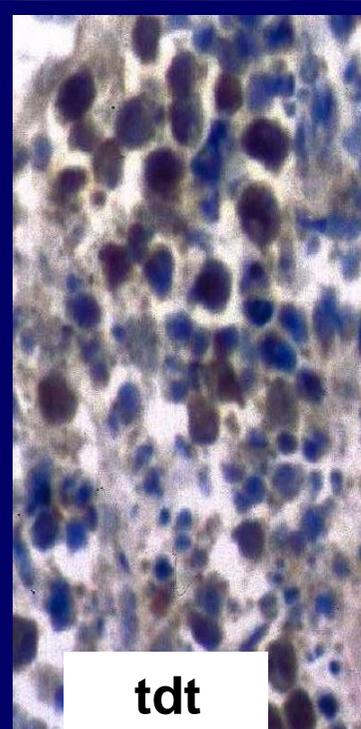
- Lymphome de Hodgkin
- Lymphome à grandes cellules B du médiastin
- Séminome
- **Lymphome lymphoblastique T (jeune)**
- Schwannome



cd3



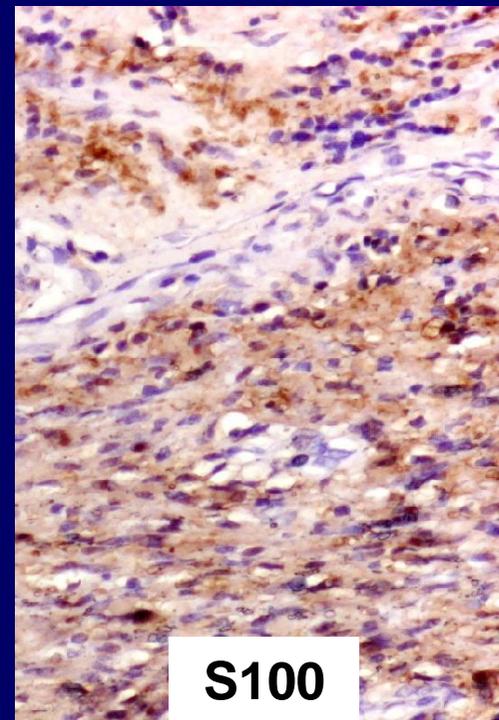
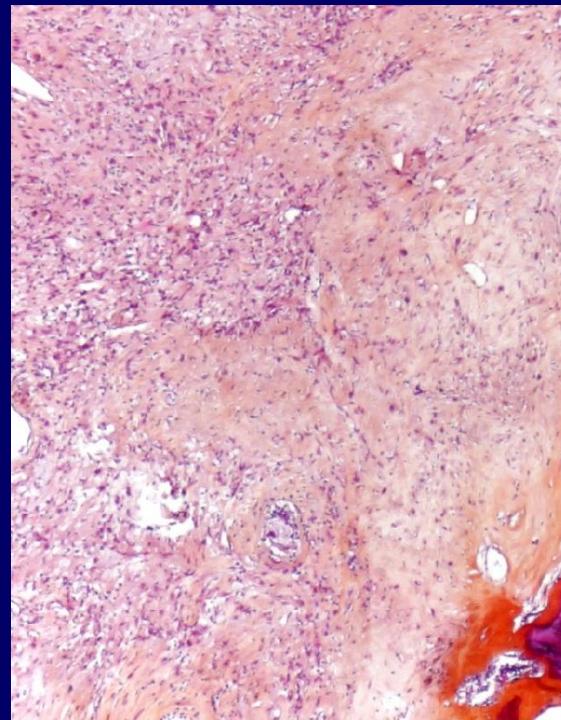
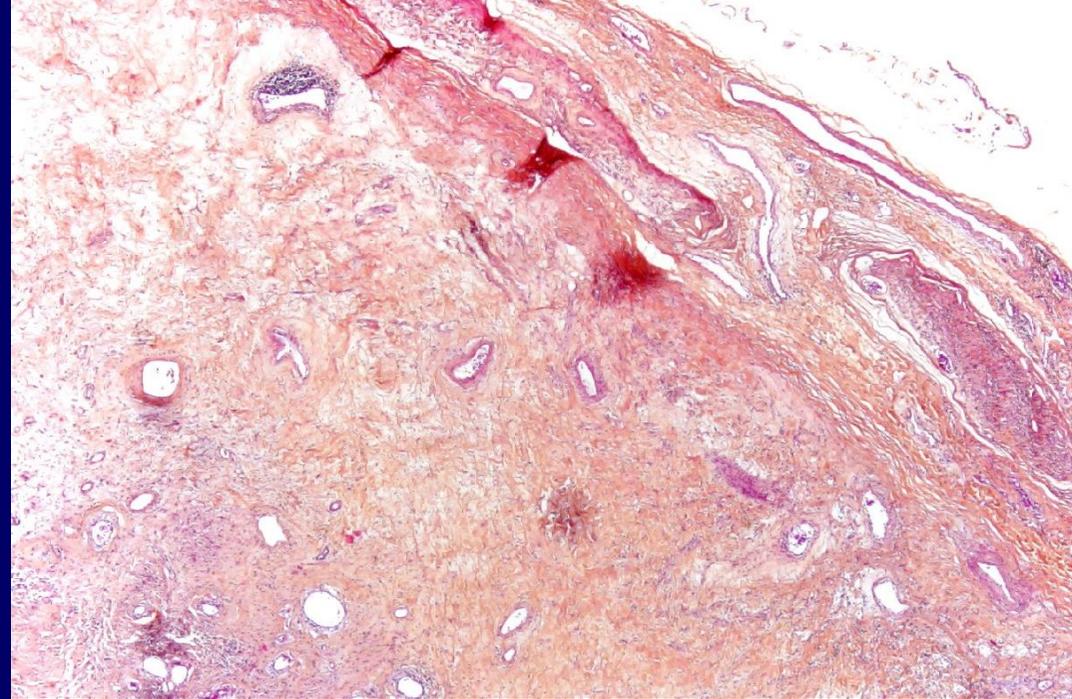
mib1



tdt

Tumeurs du Médiastin avec sclérose

- Lymphome de Hodgkin
- Lymphome à grandes cellules B du médiastin
- Séminome
- Lymphome lymphoblastique (jeune)
- **Schwannome**



S100

Rôle du Pathologiste dans la Prise en Charge des Lymphomes

- privilégier la morphologie qui dépend
 - prélèvement
 - fixation
- **corrélation clinique**
- IHC minimale
- PCR (B / T), HIS (EBER), FISH