

# **Atelier d'Héмато-Pathologie**

*Société Tunisienne de Pathologie*

*Société Tunisienne d'Hémato-Cancérologie*

*Roche Tunisie*

Hammamet, 4-5 Mars 2016

**N° 6**

**Lymphomes Médiastinaux**

**Antoine de Mascarel, Bordeaux**

« [antoine.demascarel@chu-bordeaux.fr](mailto:antoine.demascarel@chu-bordeaux.fr) »

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

**1980** : Primary mediastinal large B-cell lymphoma

**1986** : PMBCL with sclerosis

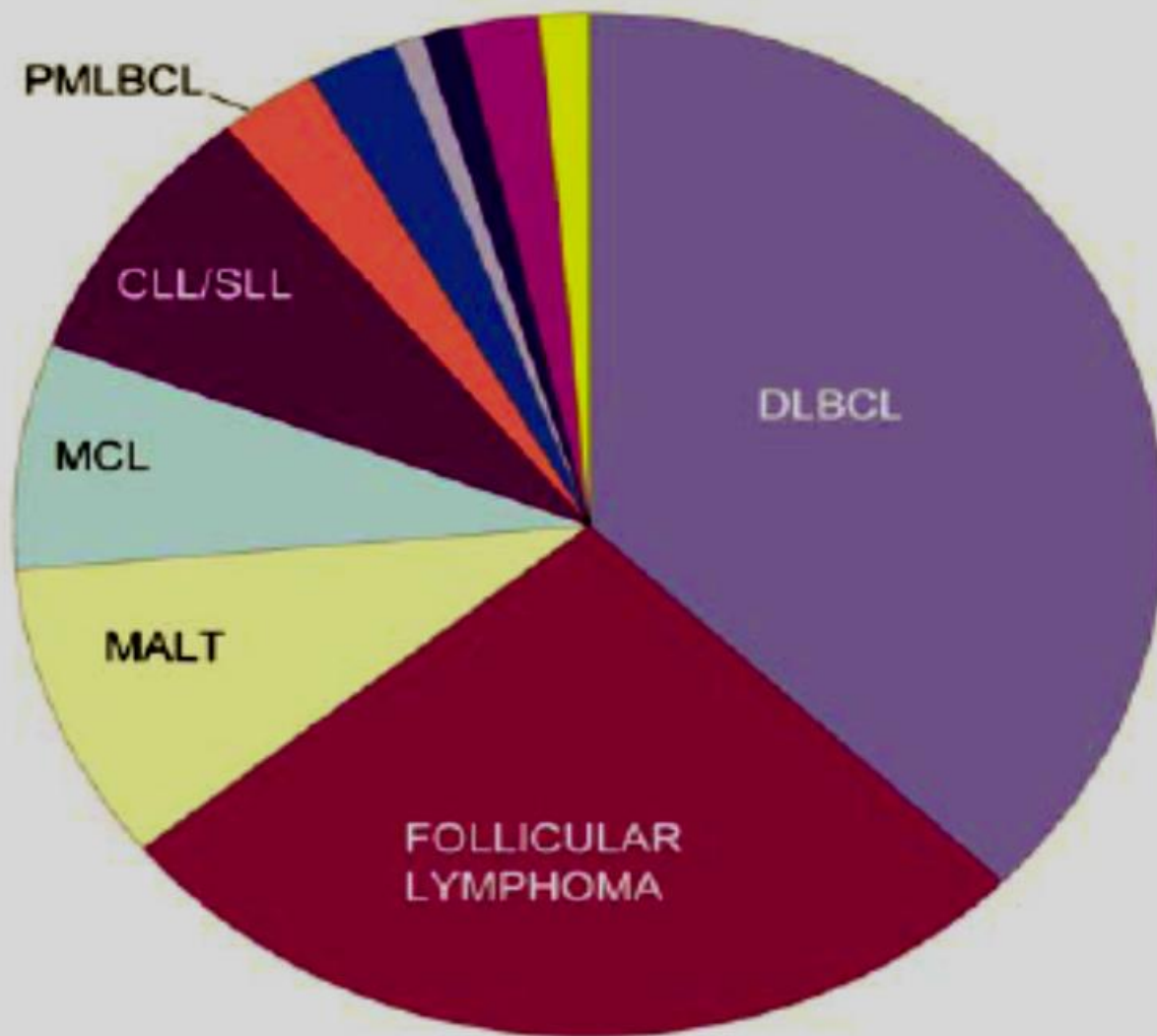
Mediastinal clear cell lymphoma

**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%**

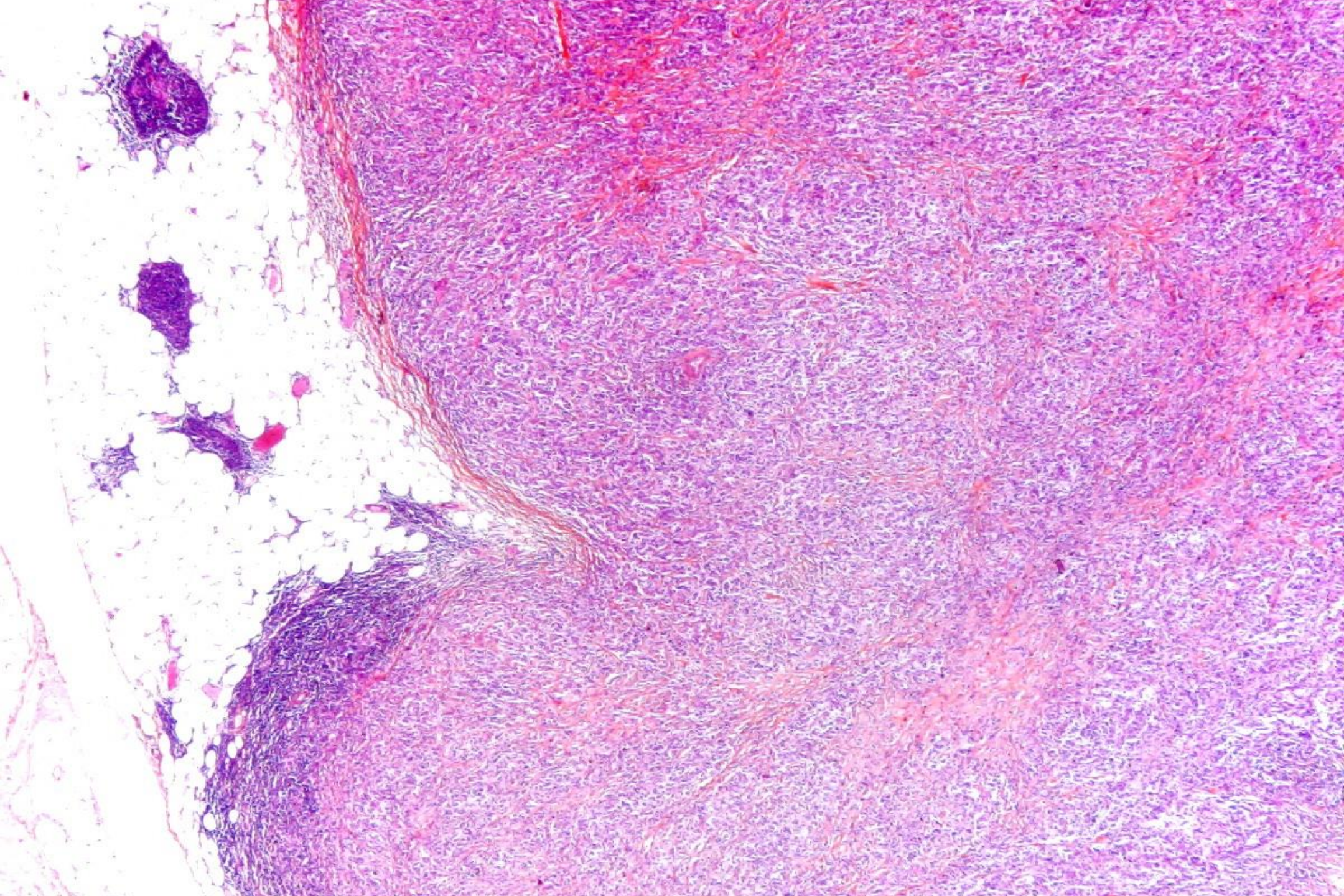
Splenic marginal zone 0.9%

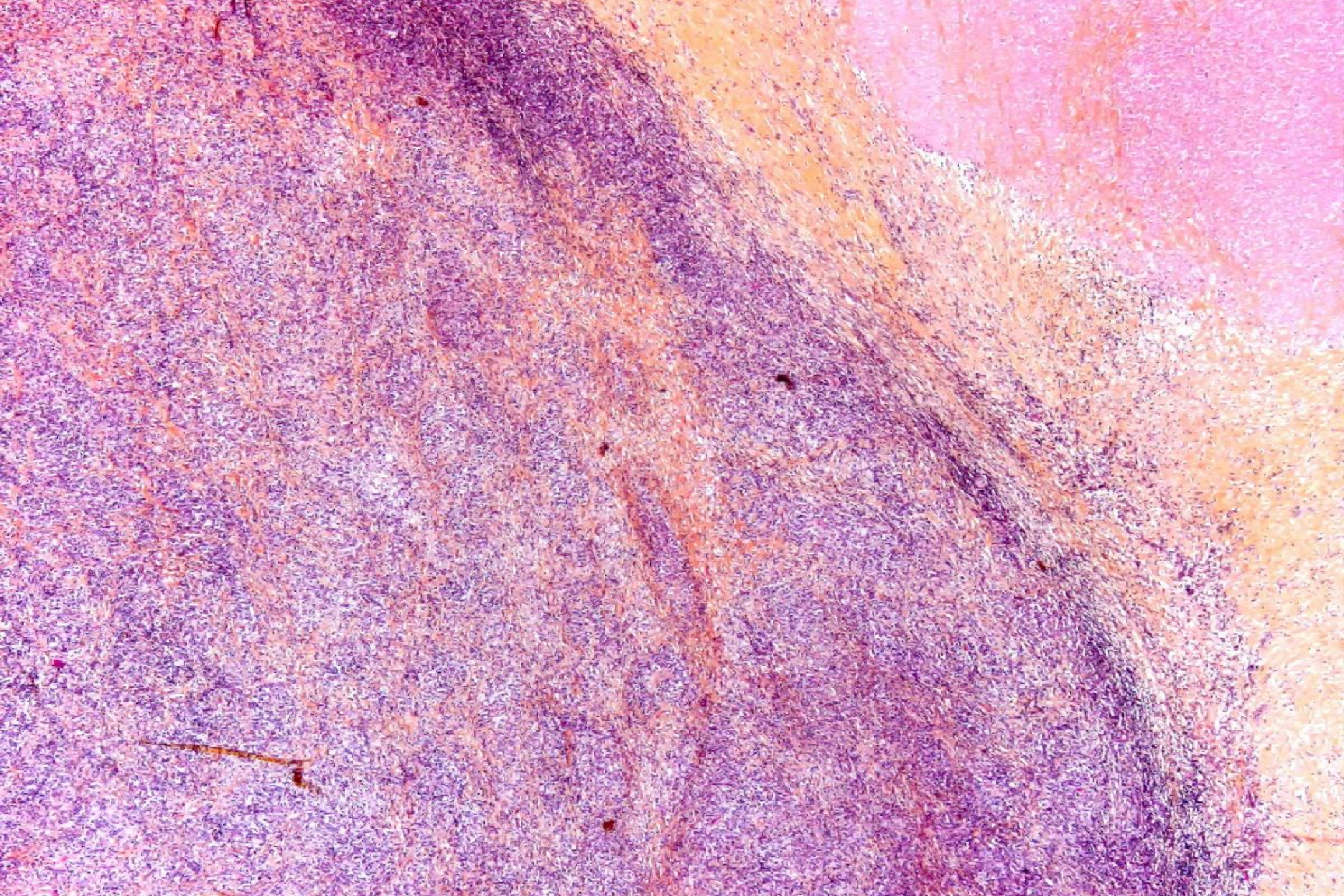
Nodal marginal zone 2%

Lymphoplasmacytic 1.4%

# PMLBCL

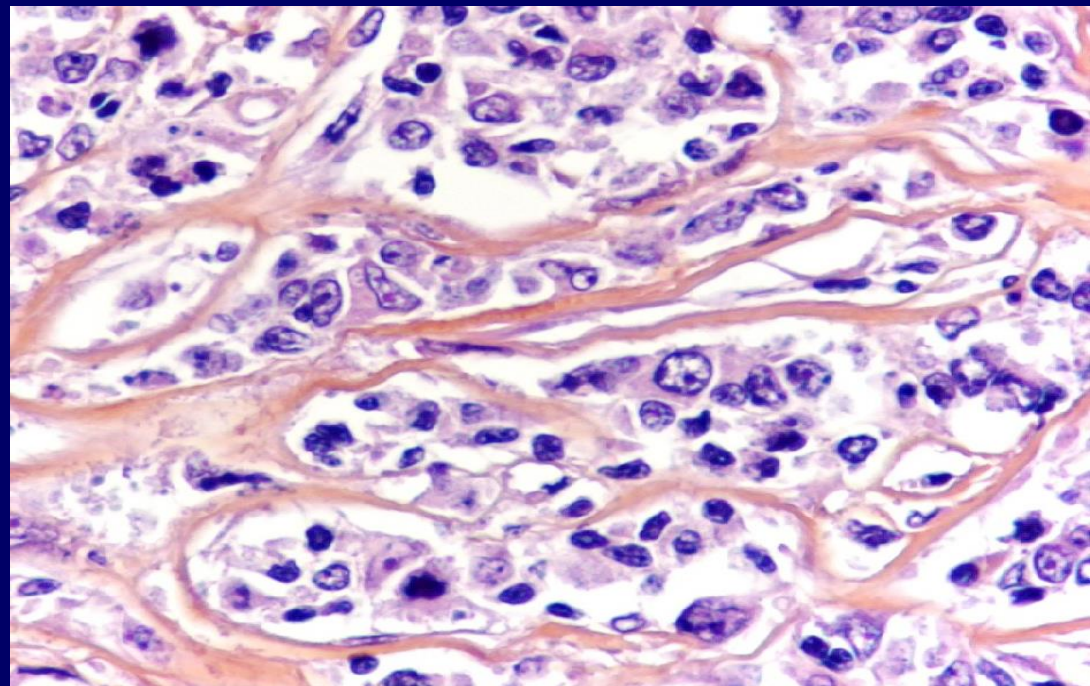
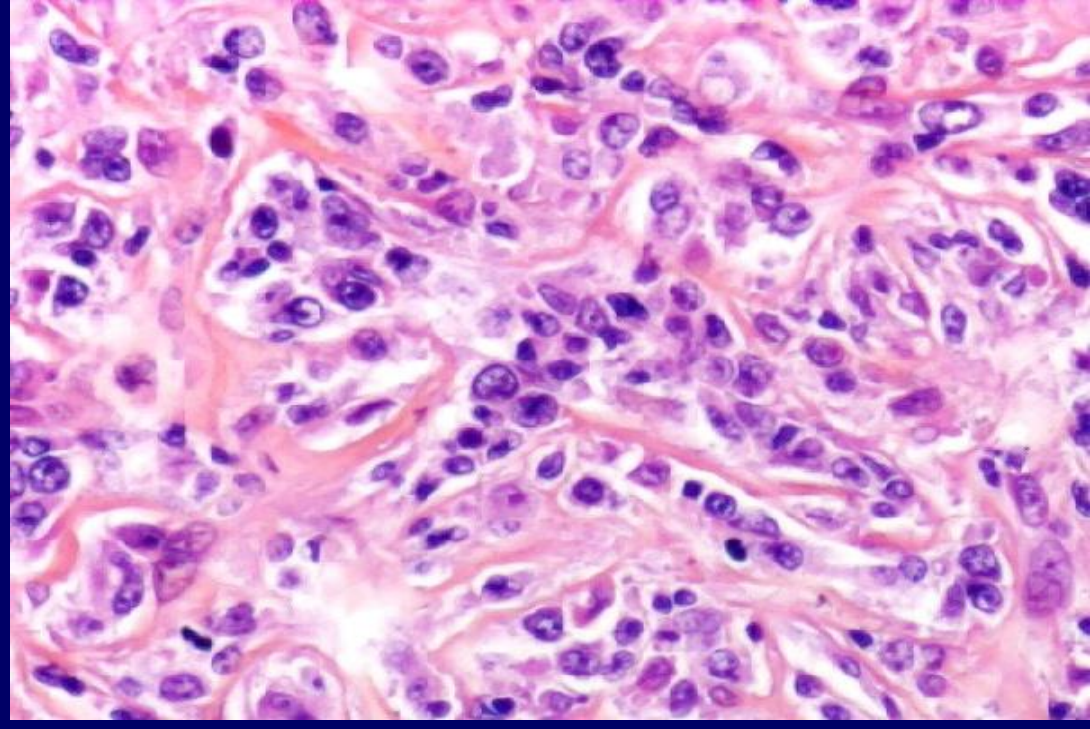
- 2,4 % des LNH
- âge moyen < 40 ans, 2F / 1M
- masse médiastinale (bulky: 50%:>10cm), stade I/II avec symptômes liés à compression/extension locale
- récurrence extra-ganglionnaire: ovaire, rein, surrénale, intestin, cerveau, peau
- BOM : + : 3%
- bonne réponse à la polychimiothérapie
- facteurs de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





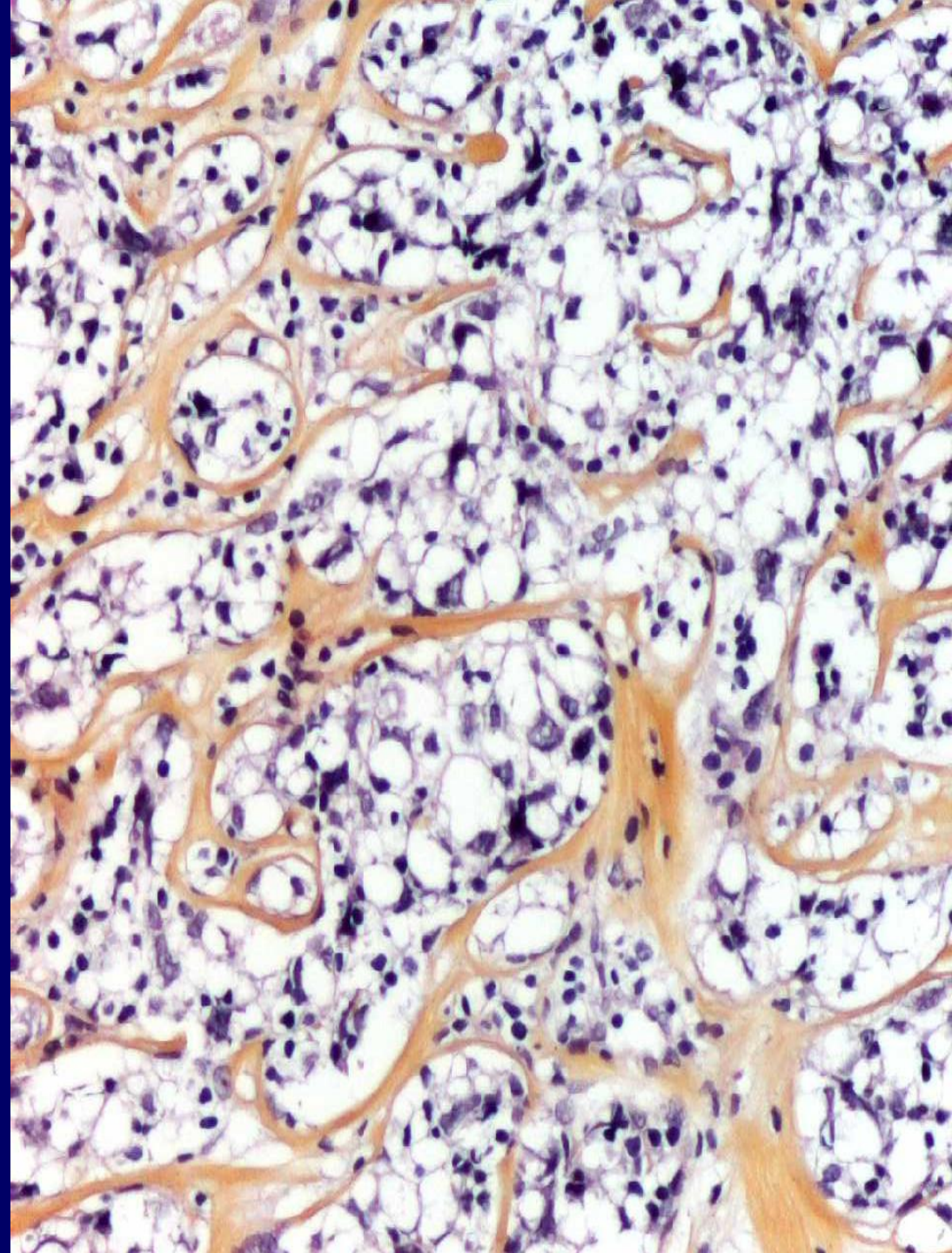
# PMLBCL

- 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%)



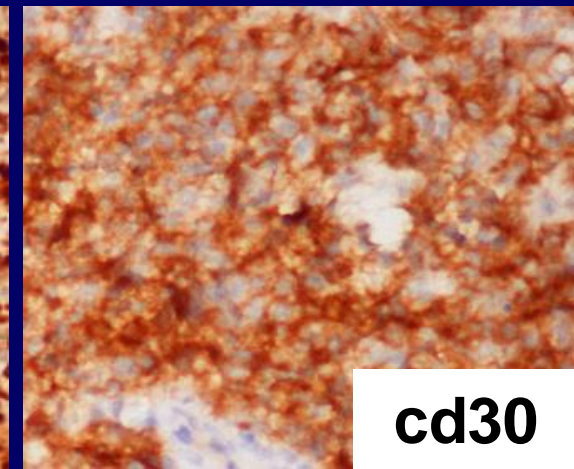
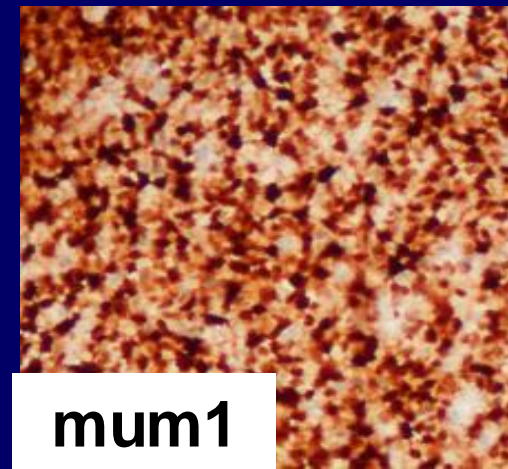
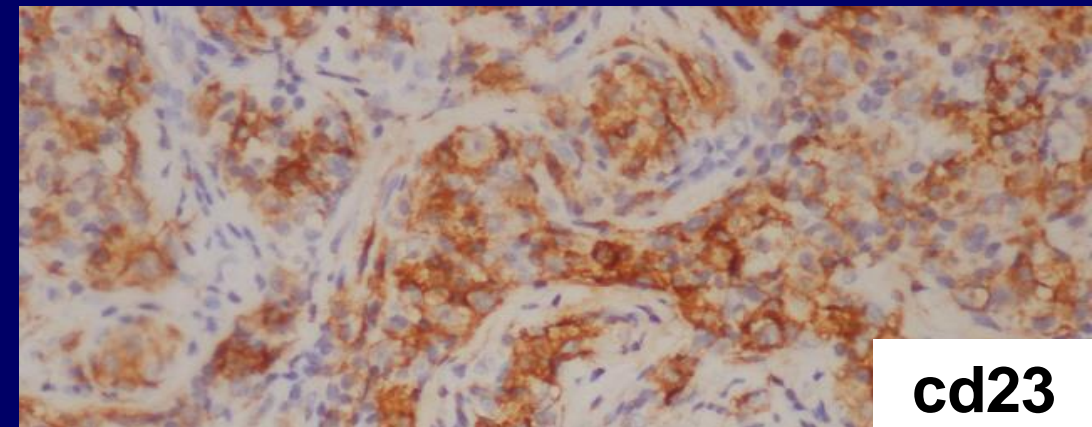
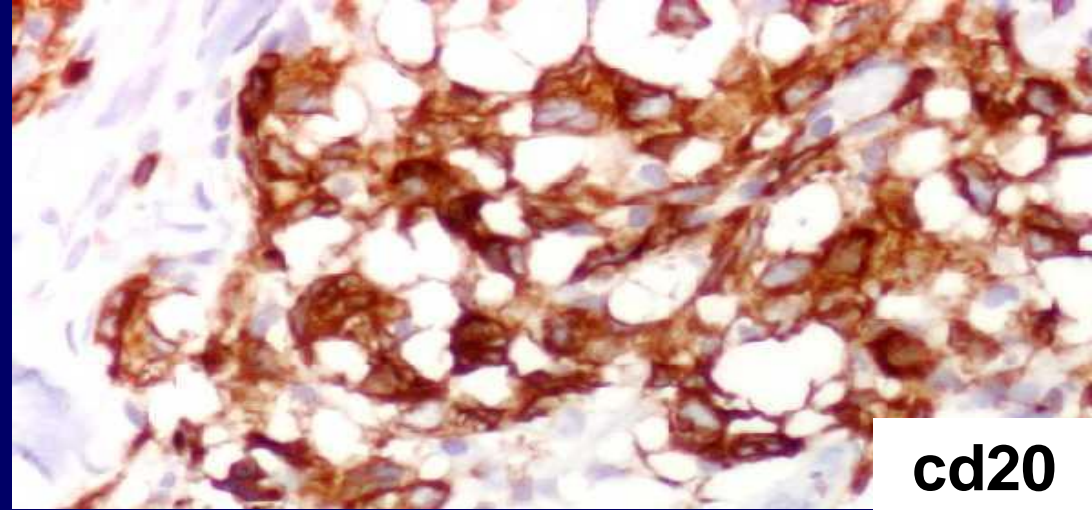


# PMBCL

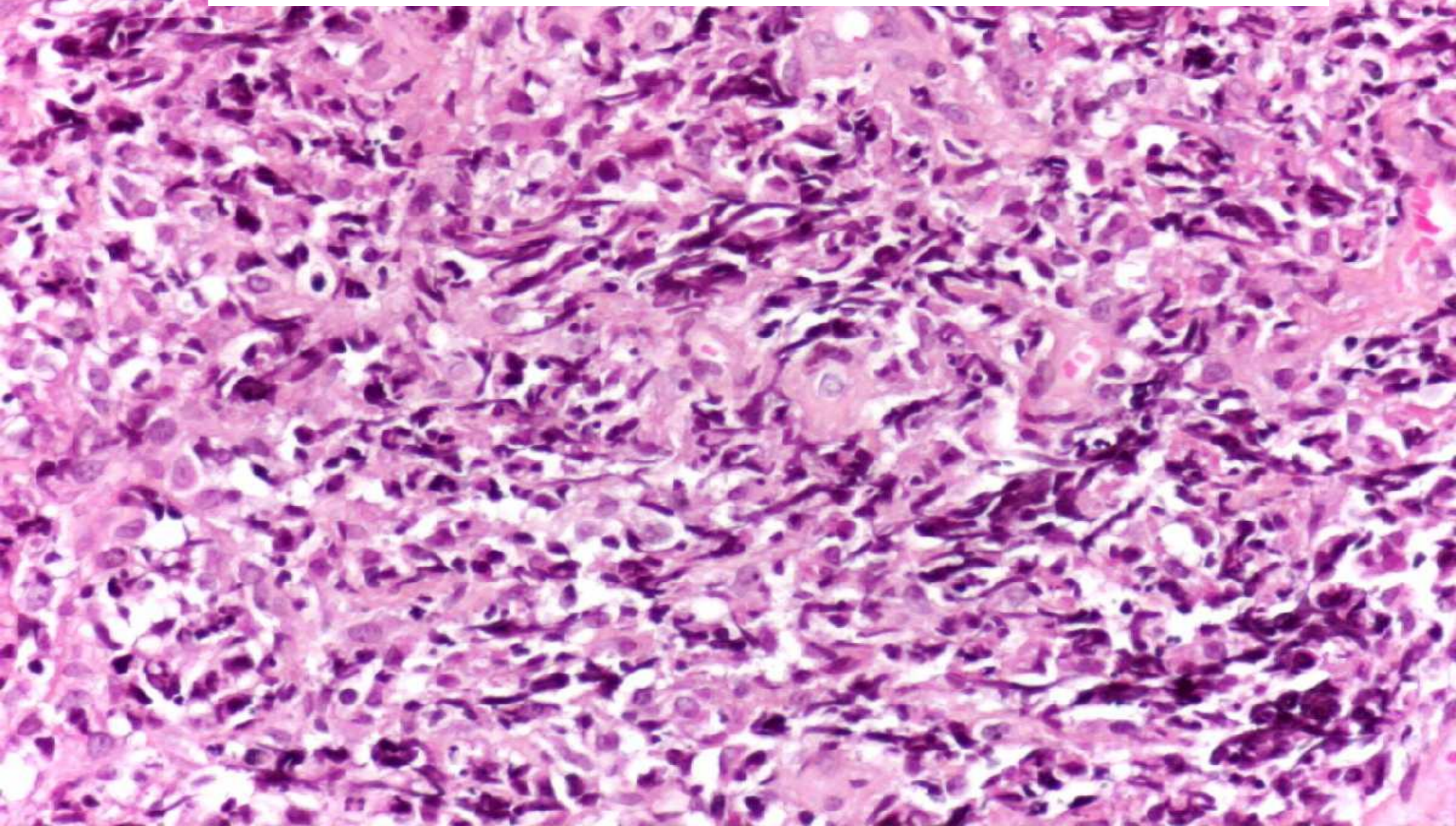
**+** : **CD20**, CD45,  
CD79a, PAX5, Oct2,  
Bob1, CD45 (60%),  
**CD30 (80%** variable  
intensity), **MAL**

**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**

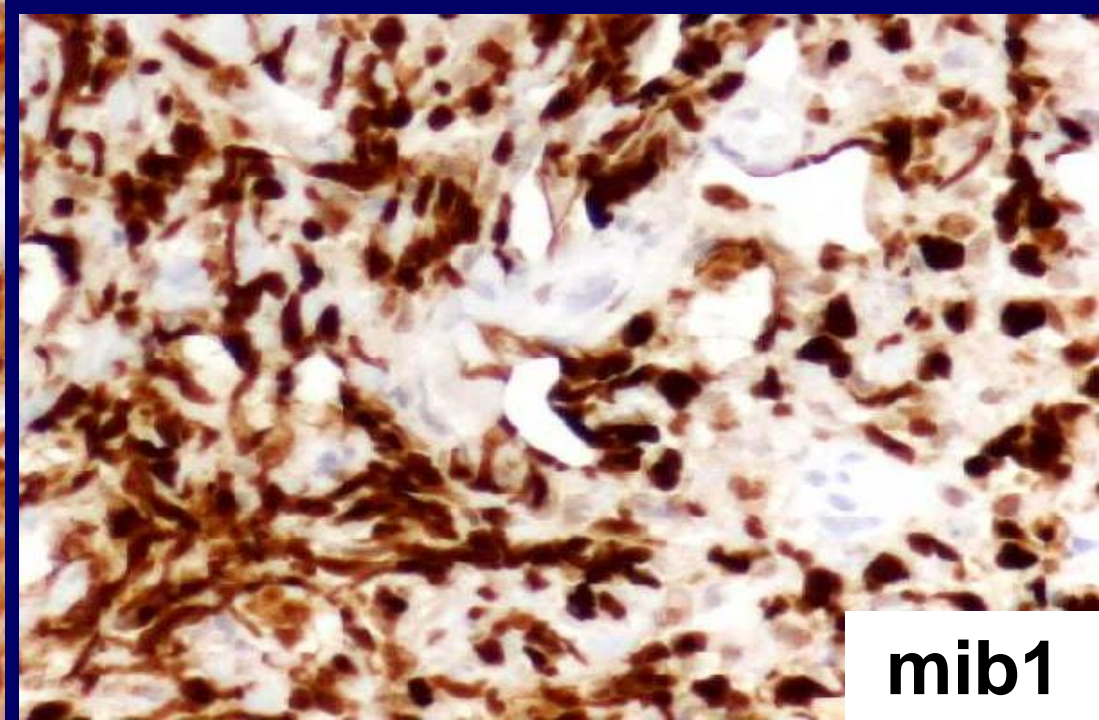
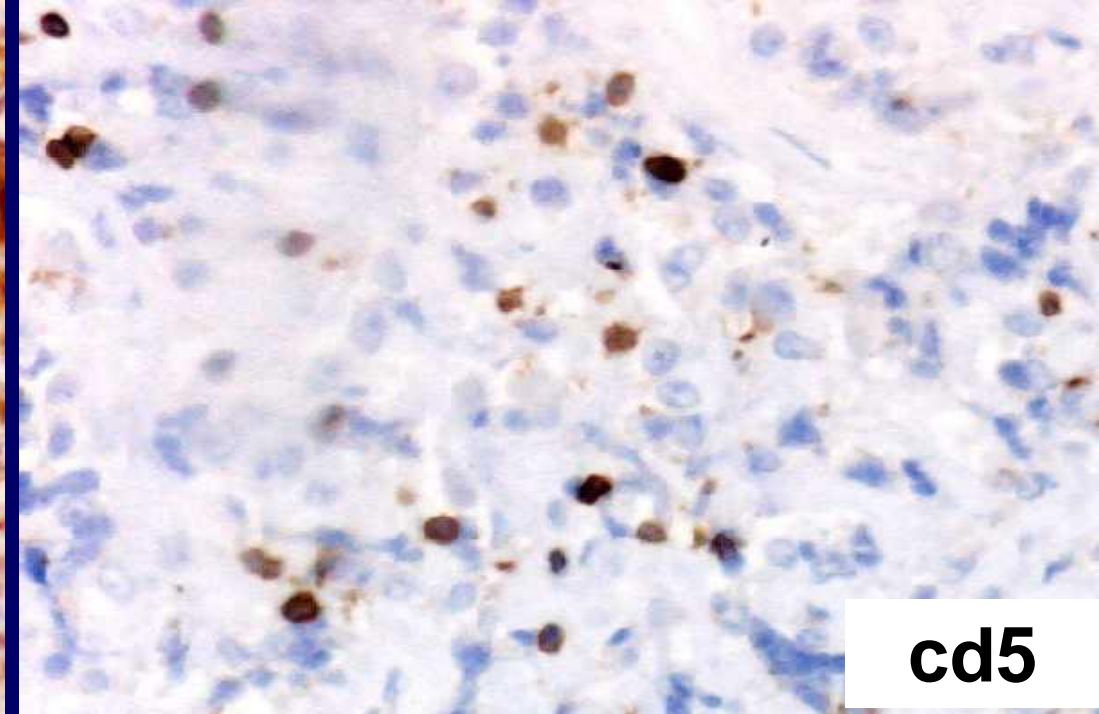
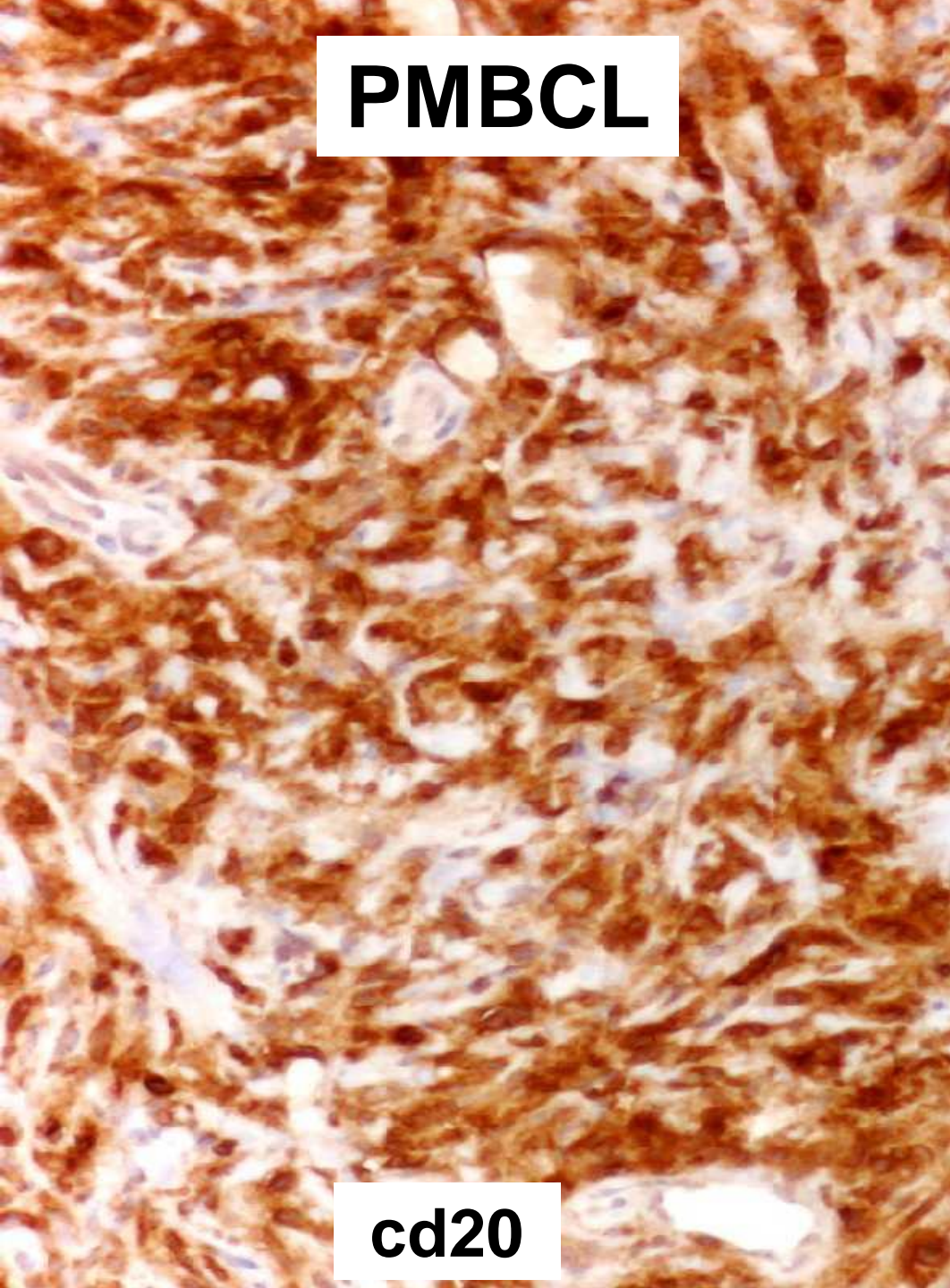


**PMBCL**

**cd20**

**cd5**

**mib1**

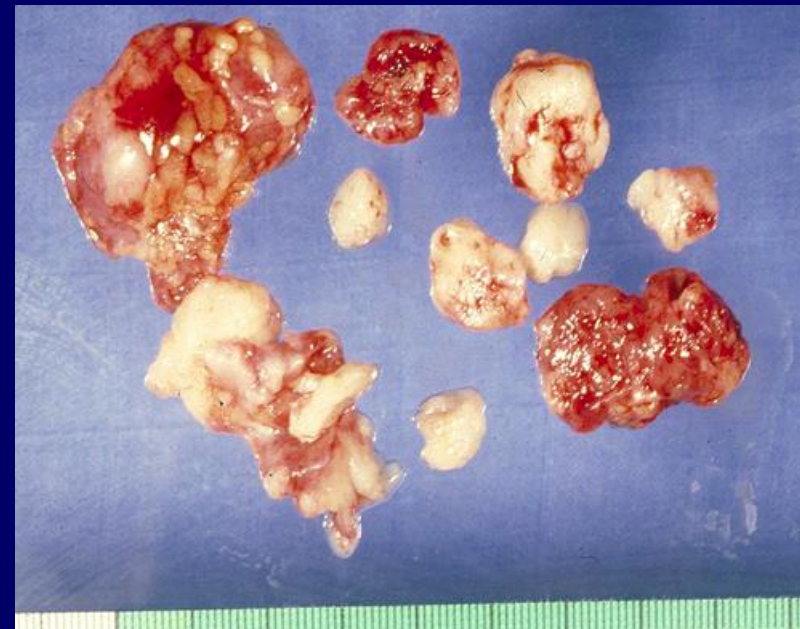


# **Lymphome à Grandes C/ B du Médiastin**

- 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  $\lambda$  T )

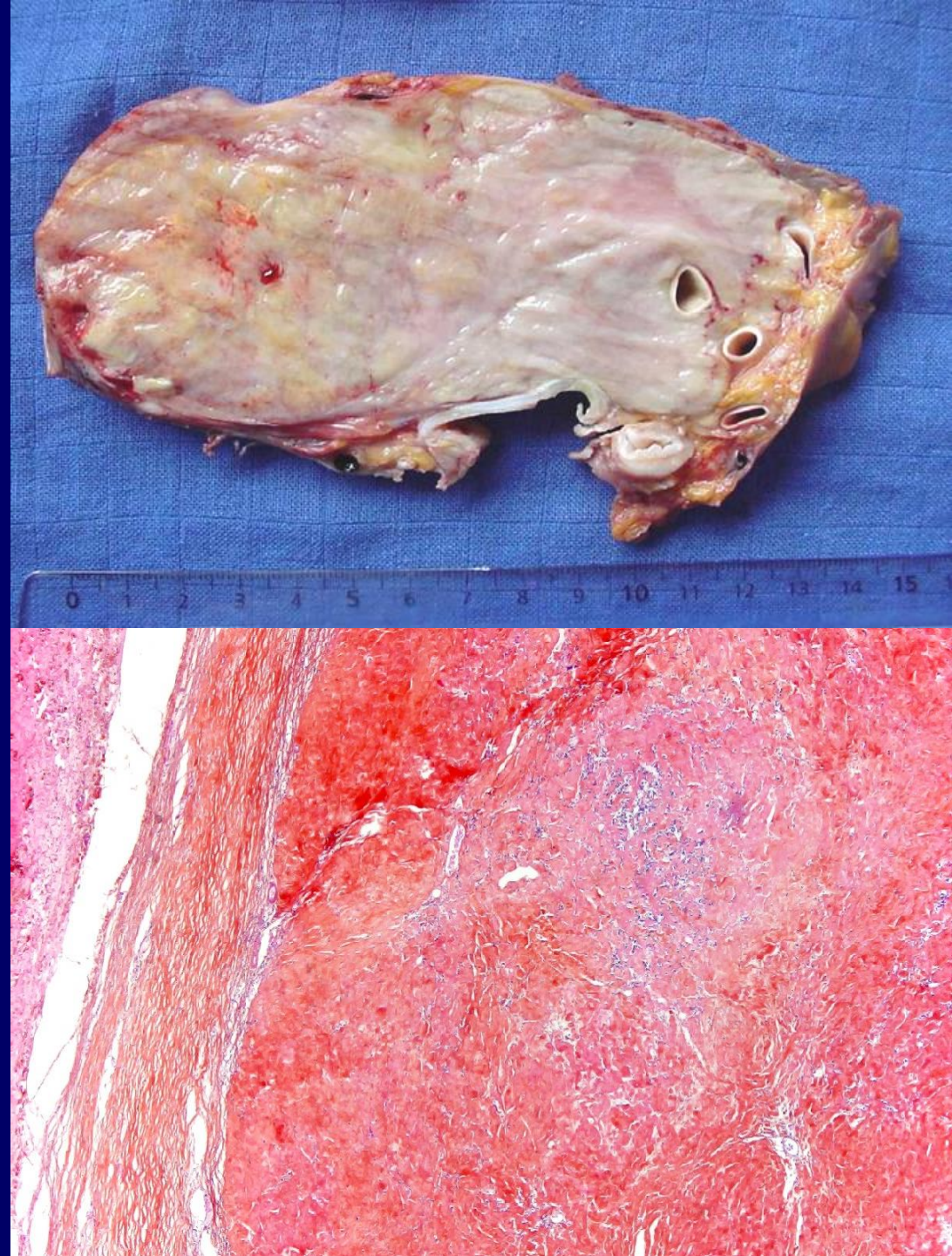
# Lésions Médiastinales Non Résécables avec Fibrose de l'Adulte

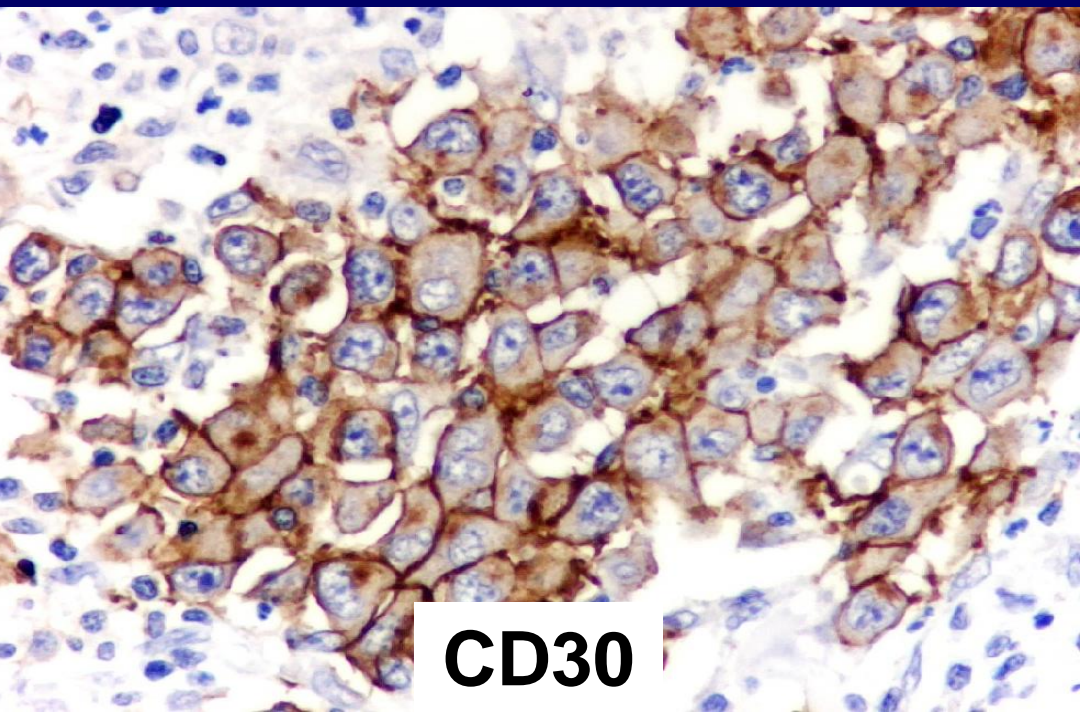
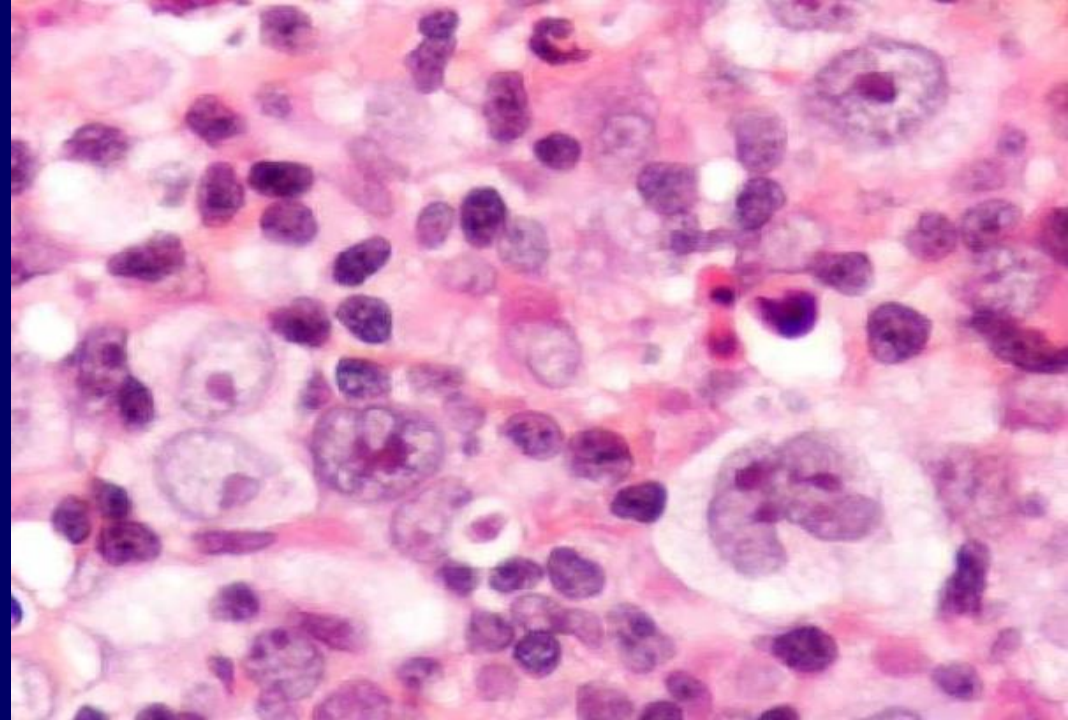
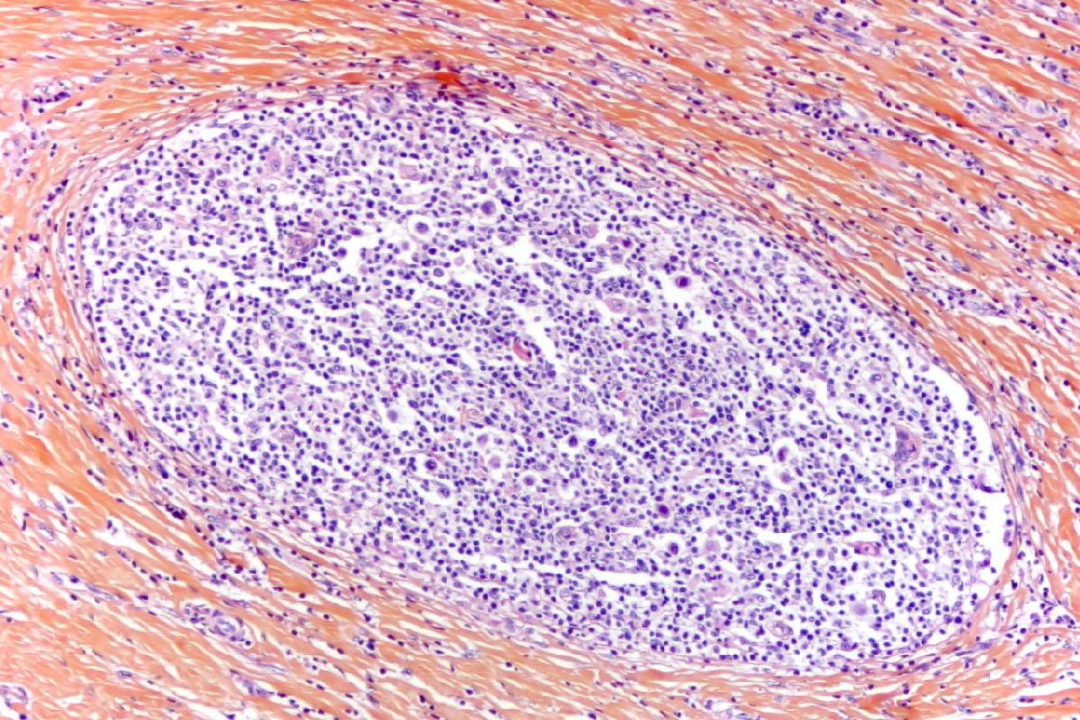
- adulte jeune
- volumineuse tumeur
- médiastinoscopie : biopsies difficiles



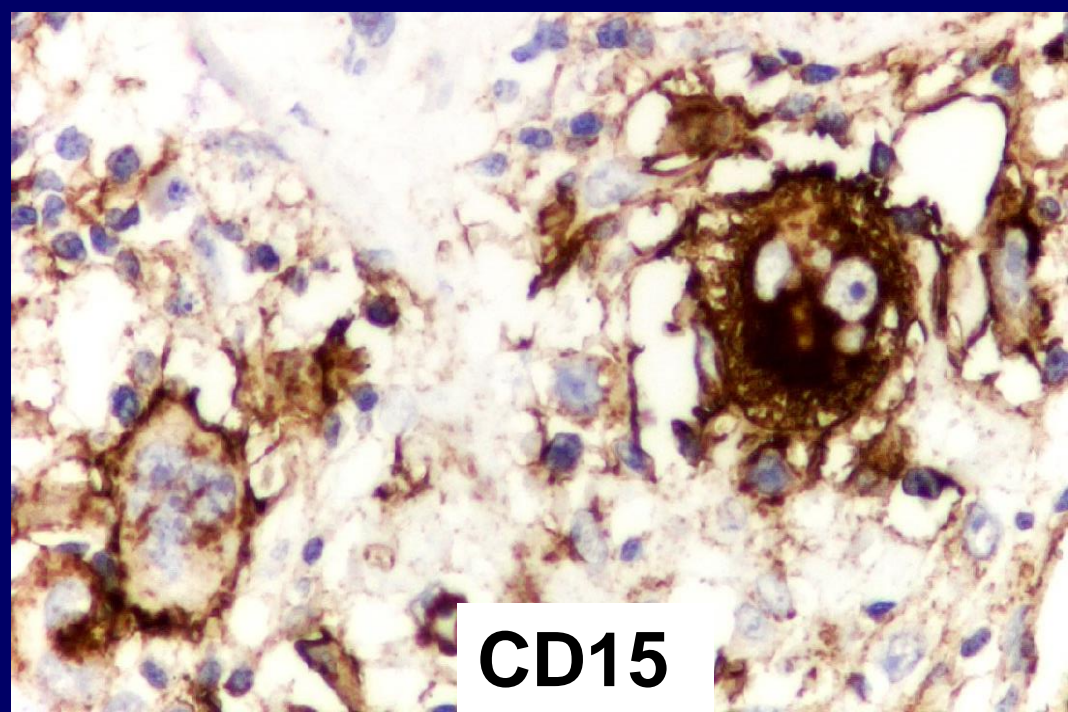
# Tumeurs du Médiastin avec sclérose

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





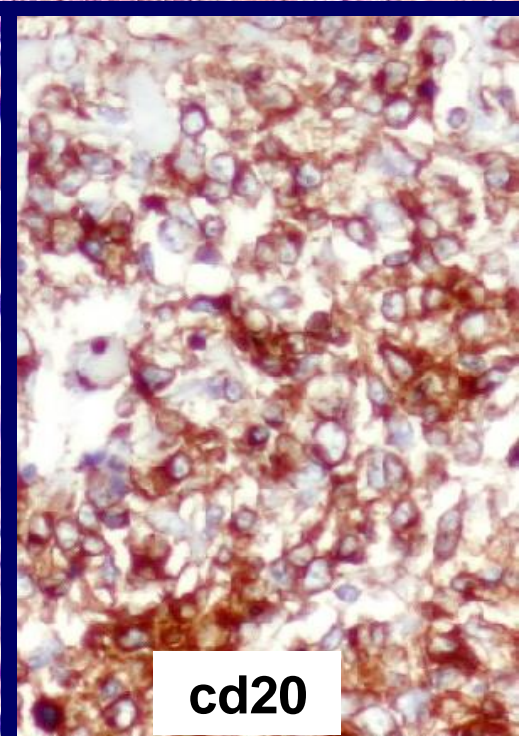
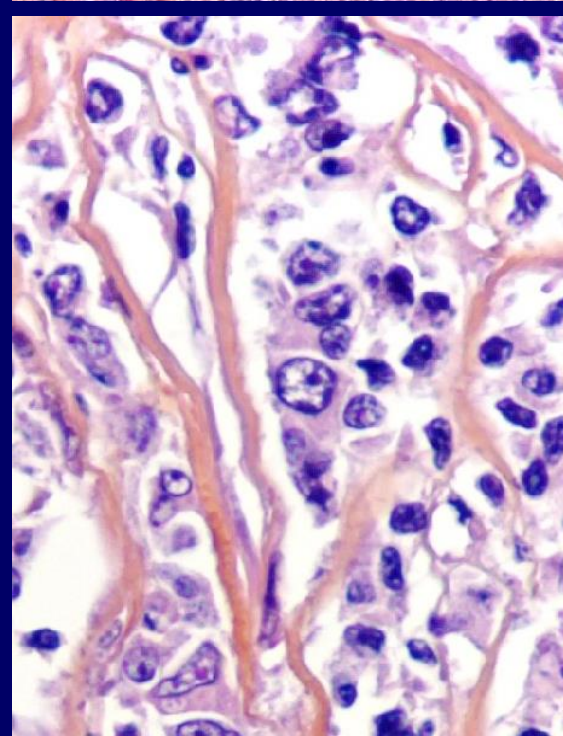
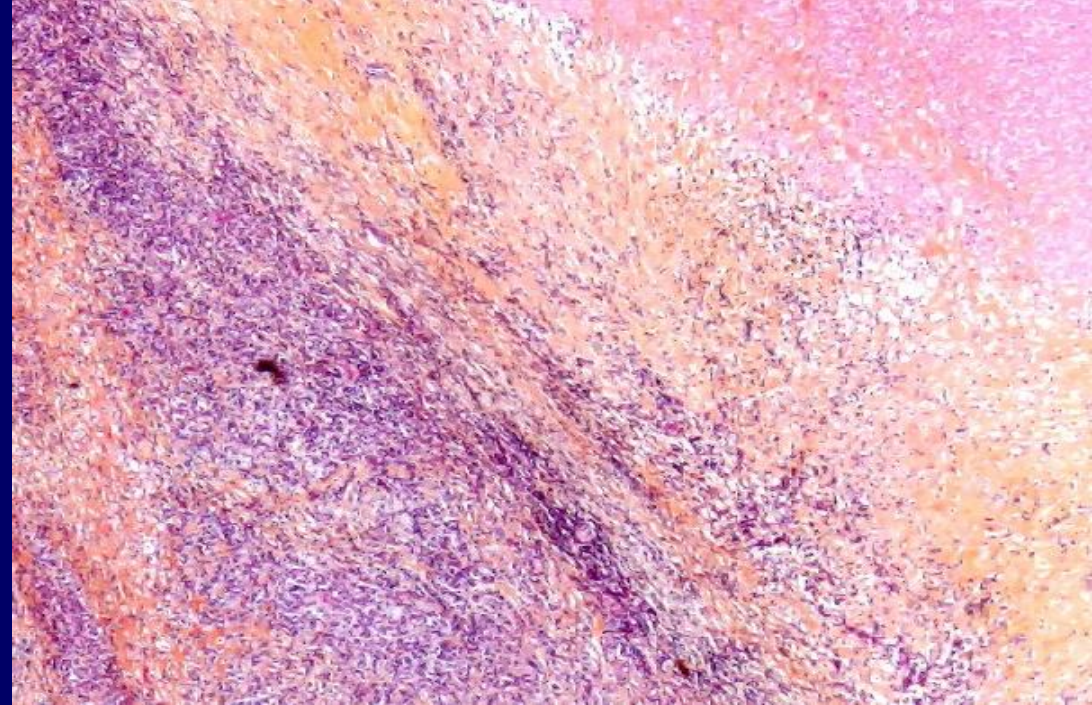
**CD30**



**CD15**

# Tumeurs du Médiastin avec sclérose

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



cd20



# Tumeurs du Médiastin avec sclérose

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

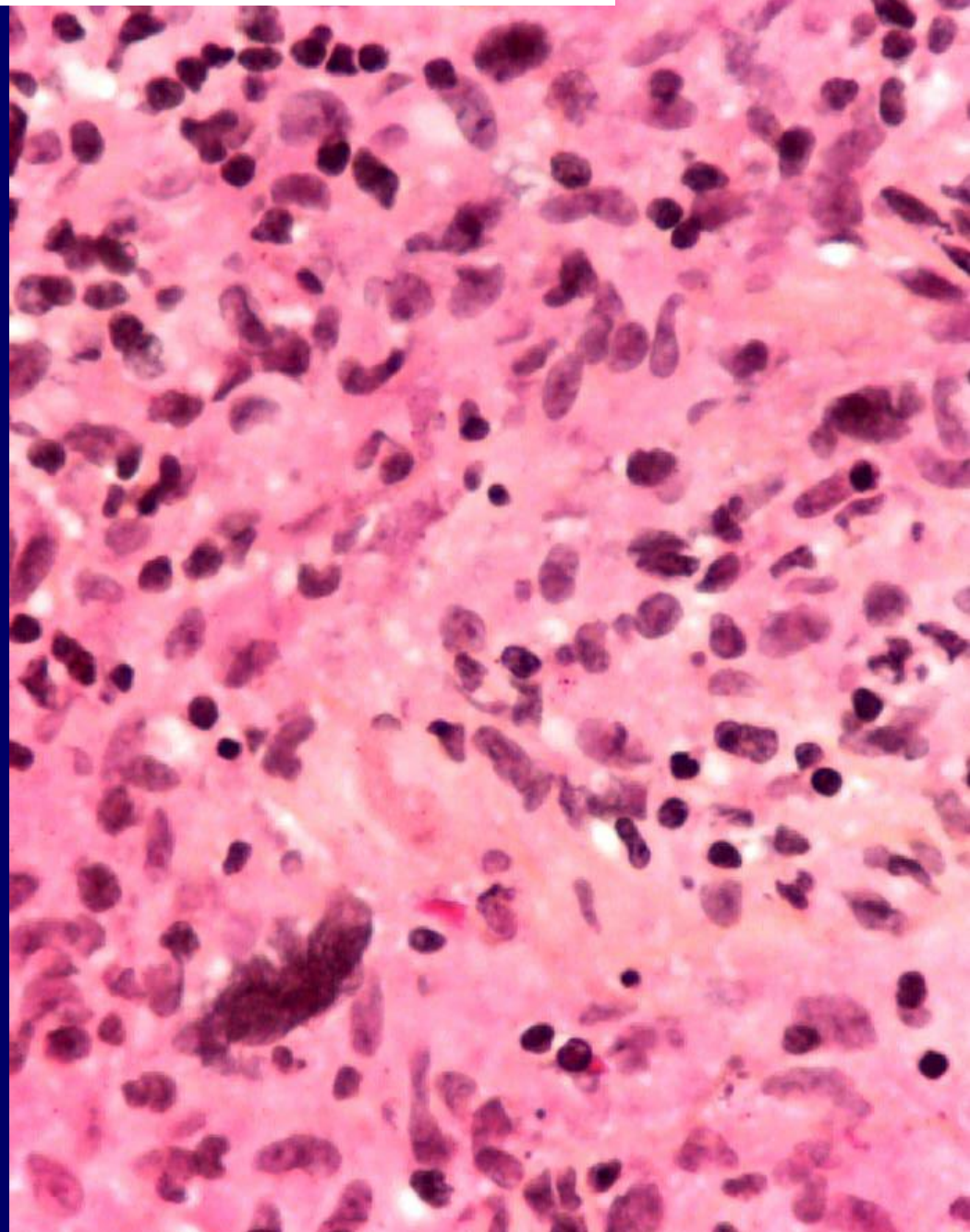
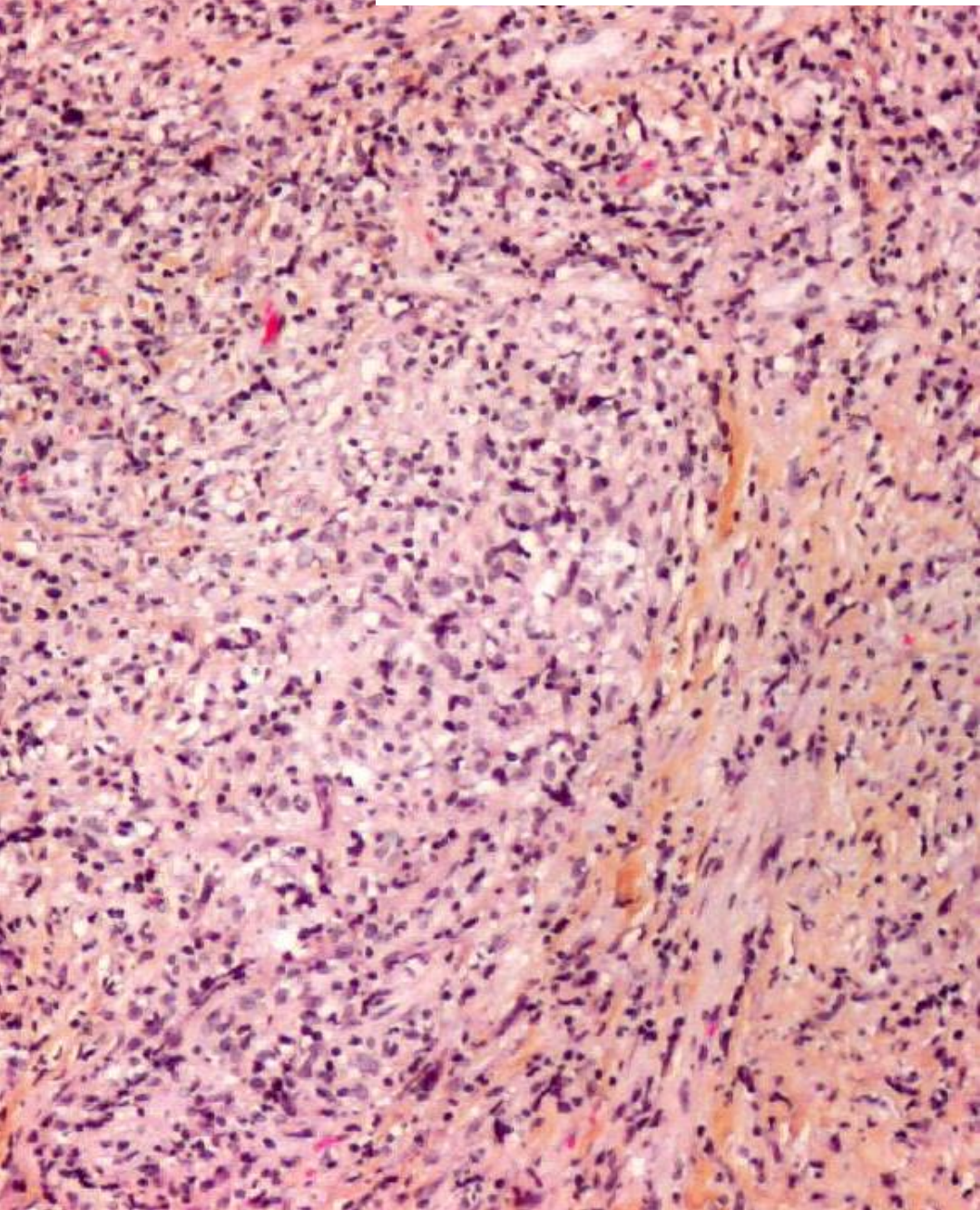
## Definition WHO 2008

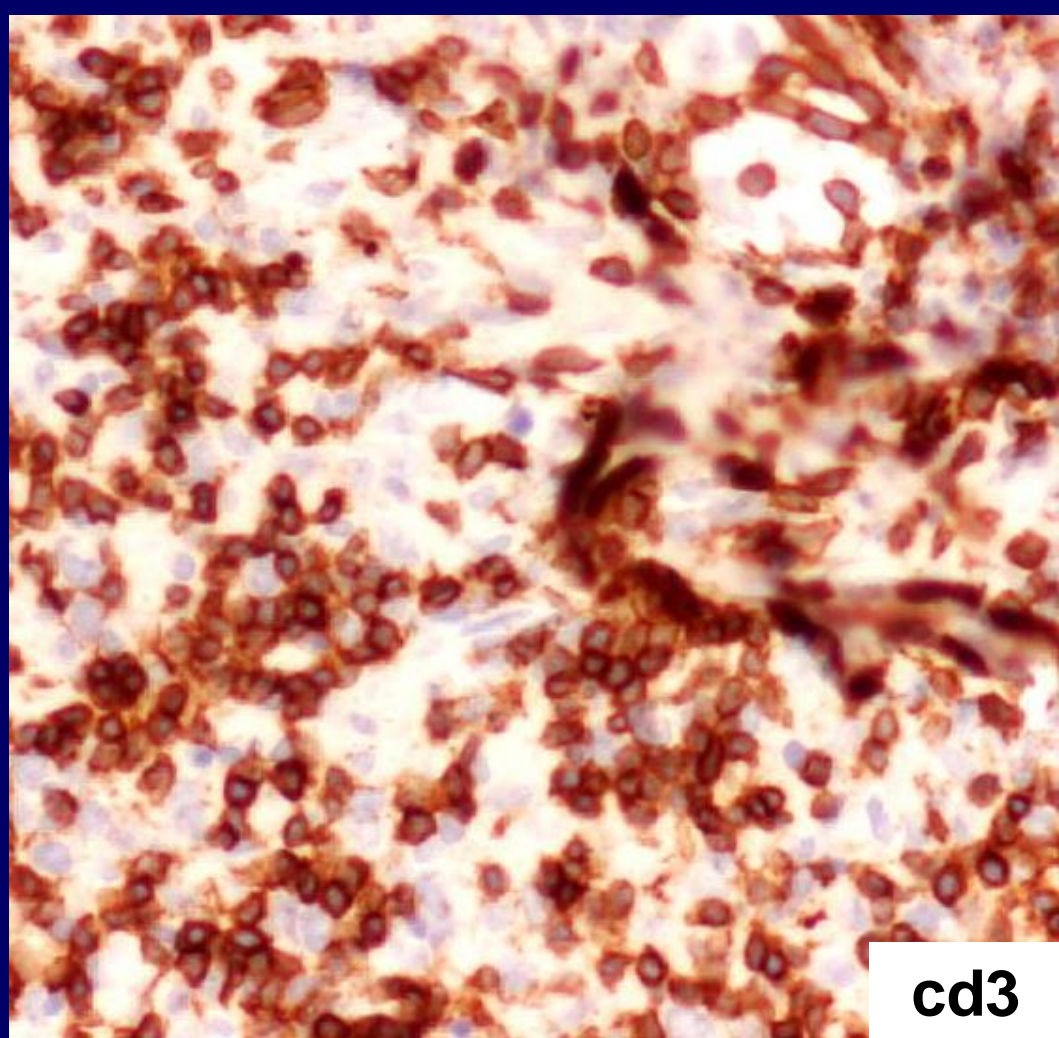
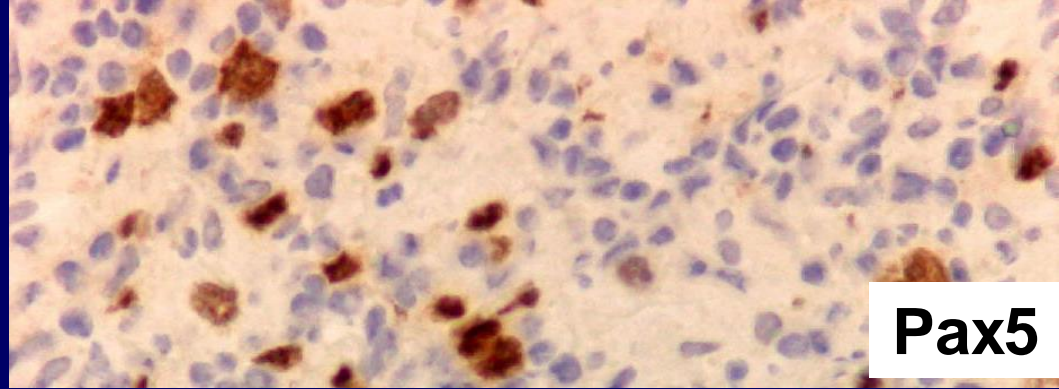
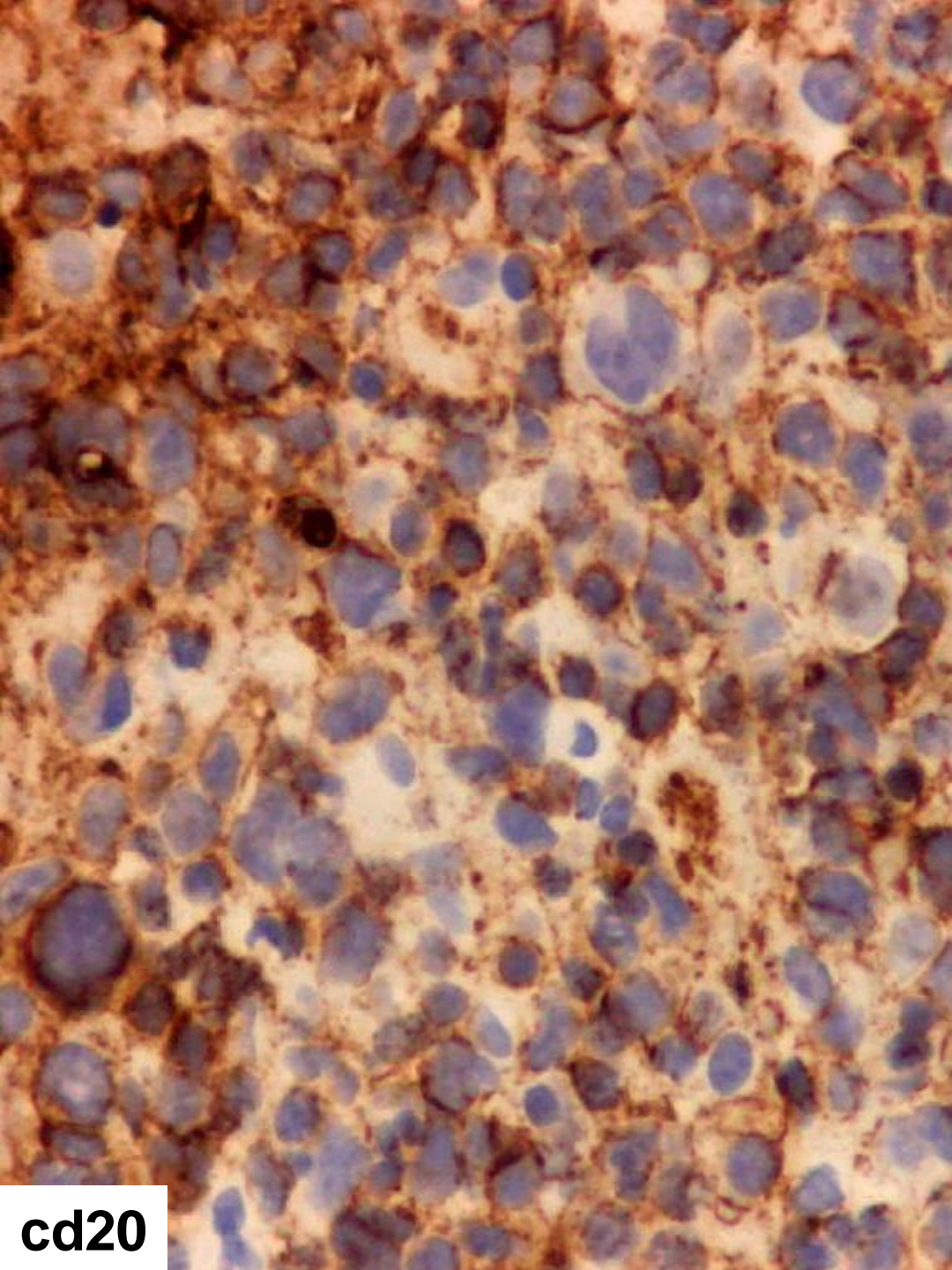
- Lymphome B qui montre un chevauchement clinique, morphologique et/ou immunophénotypique entre Hodgkin classique et DLBCL, en particulier le lymphome primitif médiastinal à grandes cellules B
- plus agressif que Hodgkin ou DLBCL. Pas de consensus thérapeutique

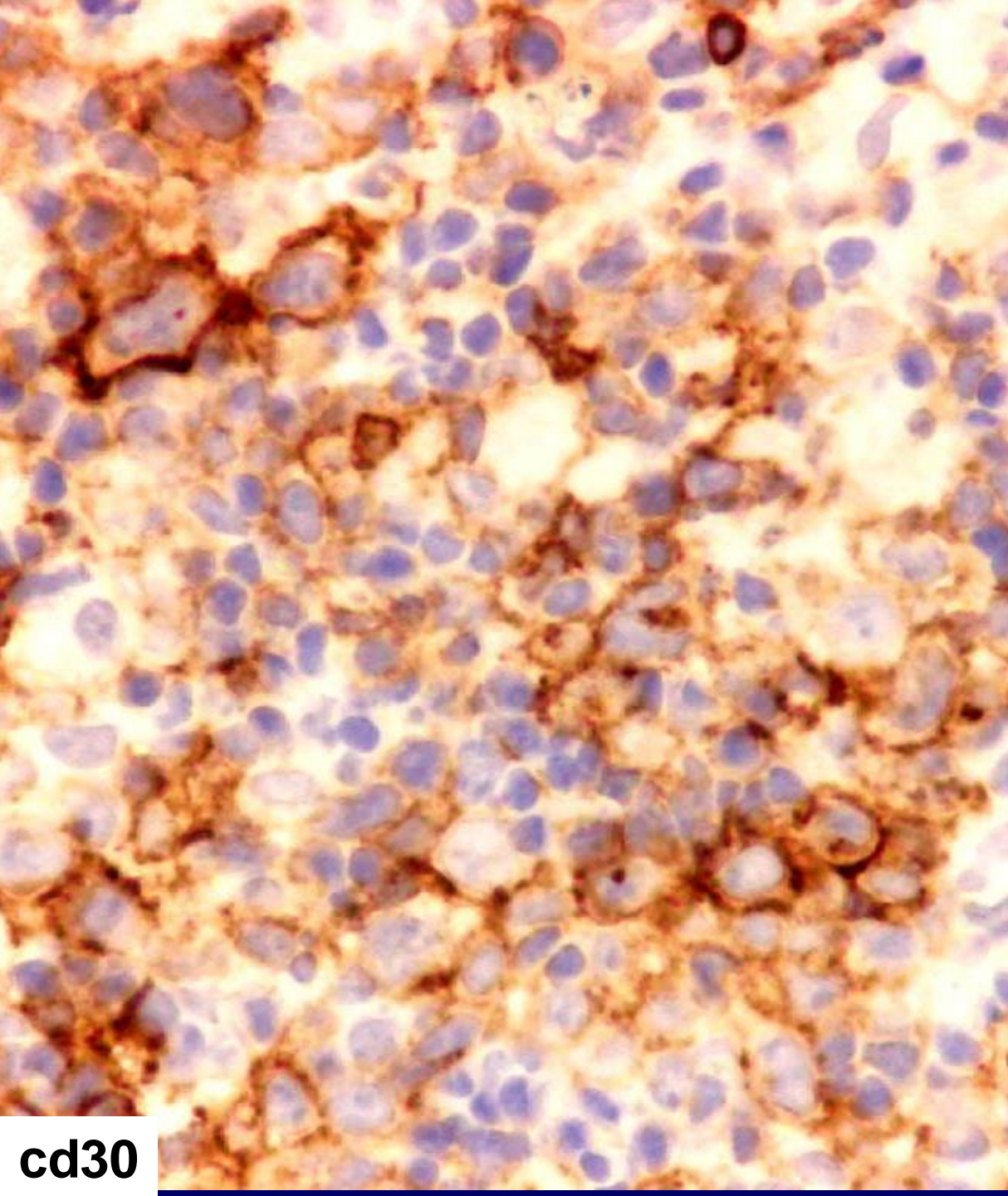
# Mediastinal Grey Zone Lymphoma

- **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

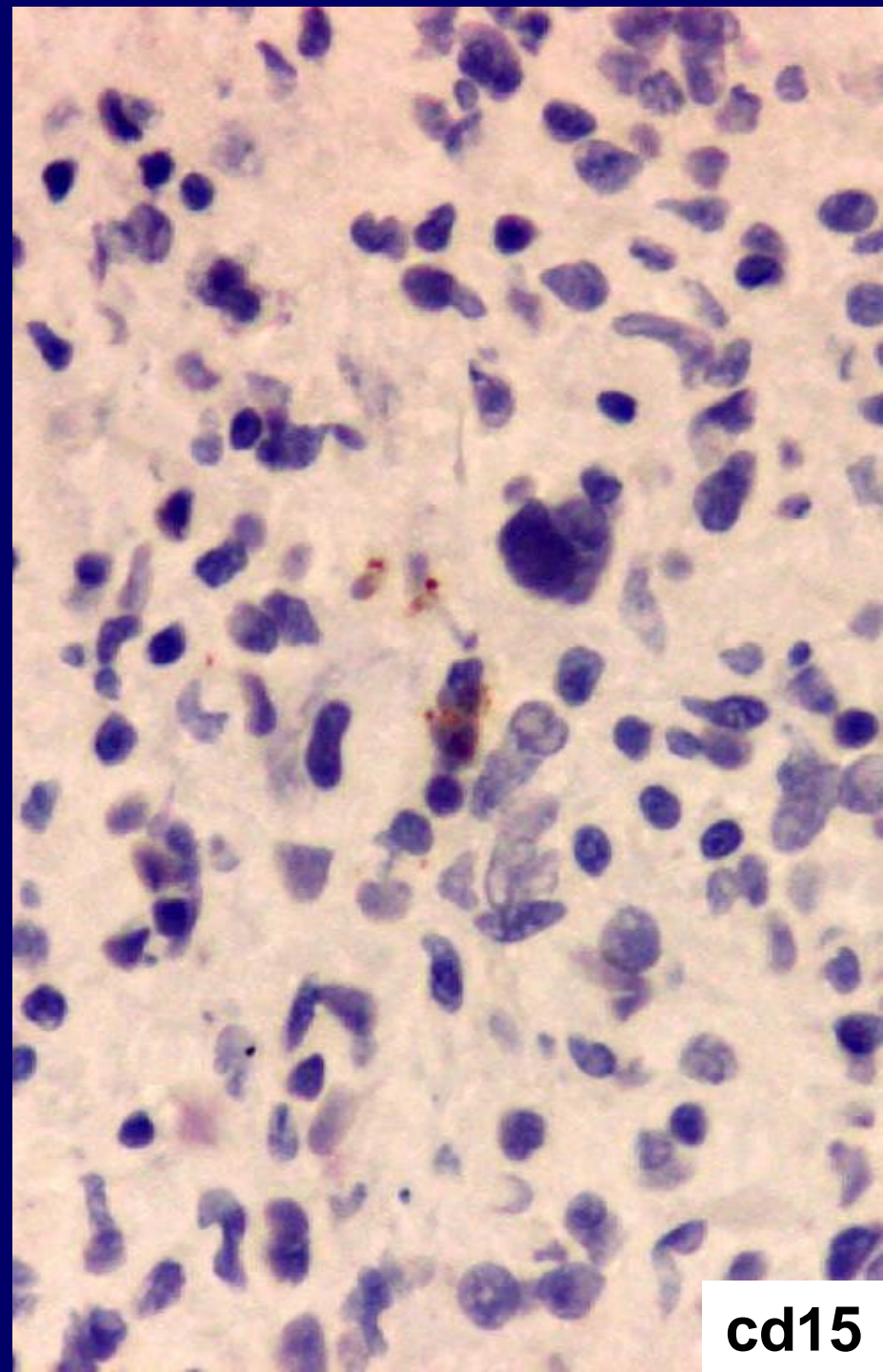
**F, 29 ans, tumeur médiastinale**







**cd30**

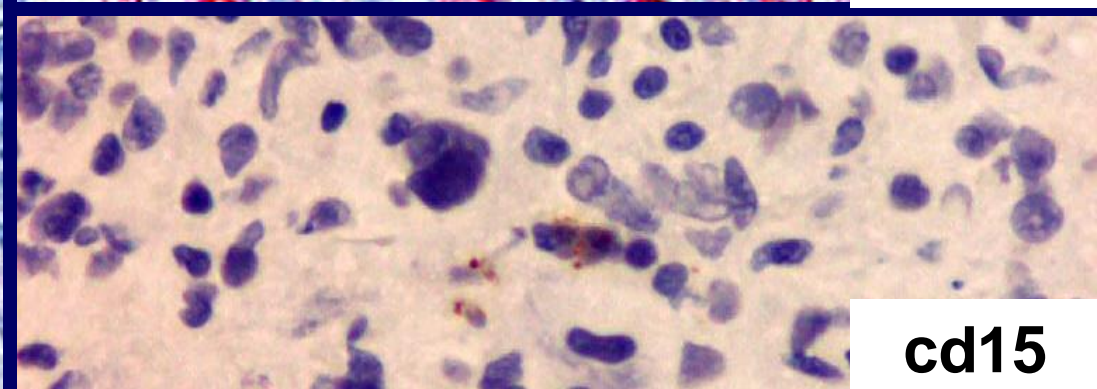
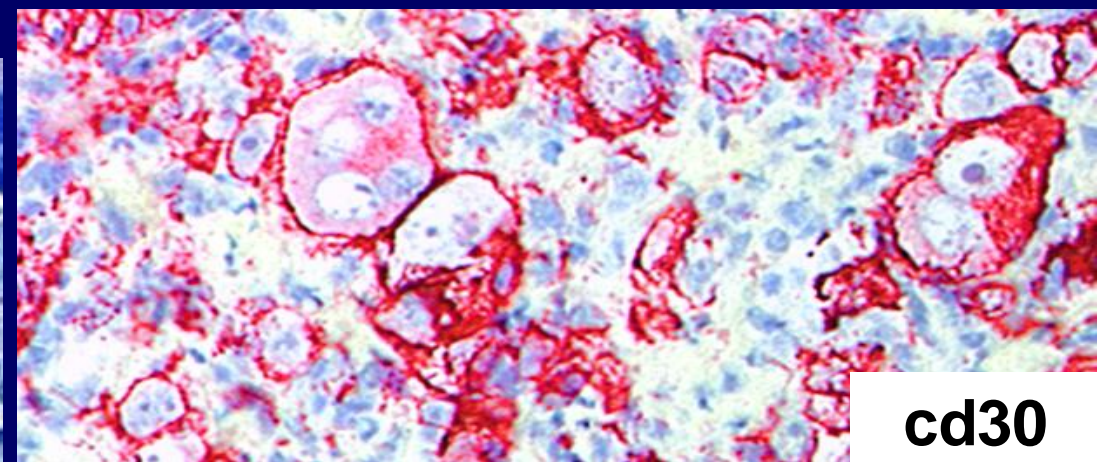
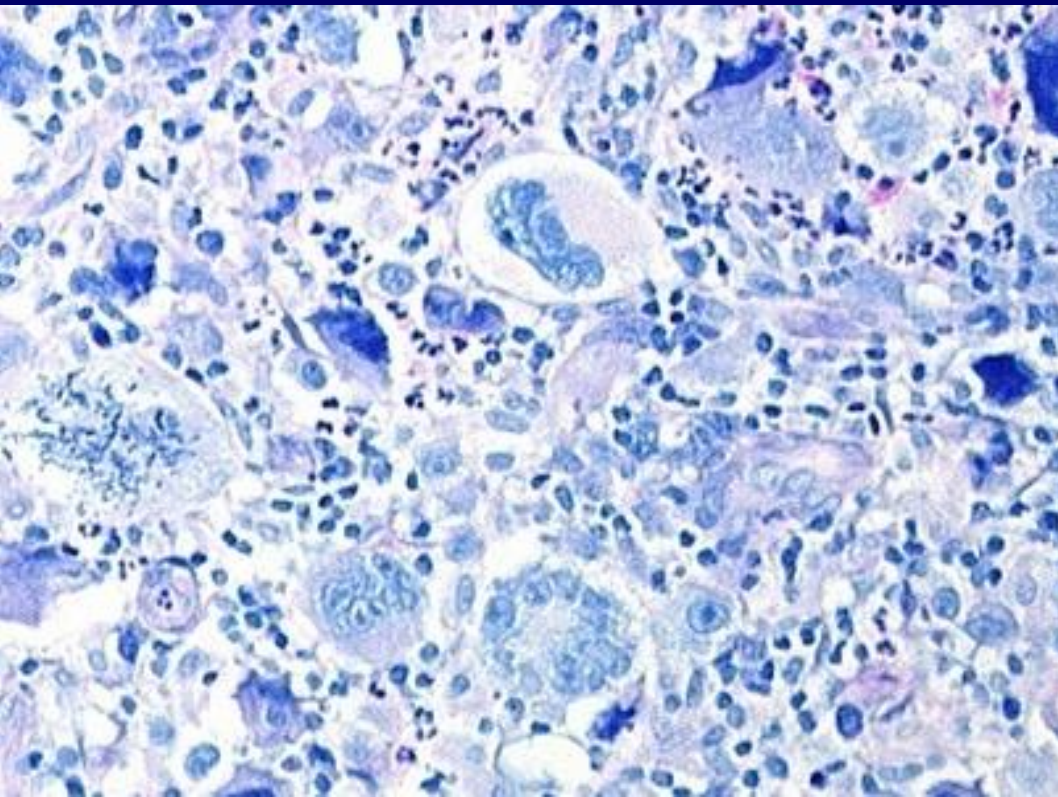
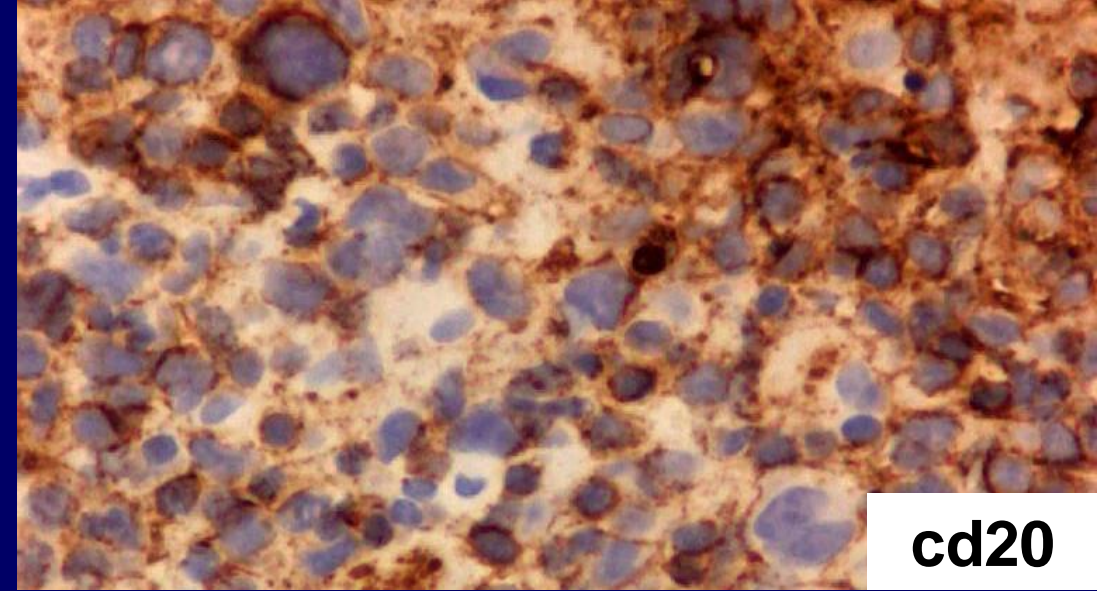


**cd15**

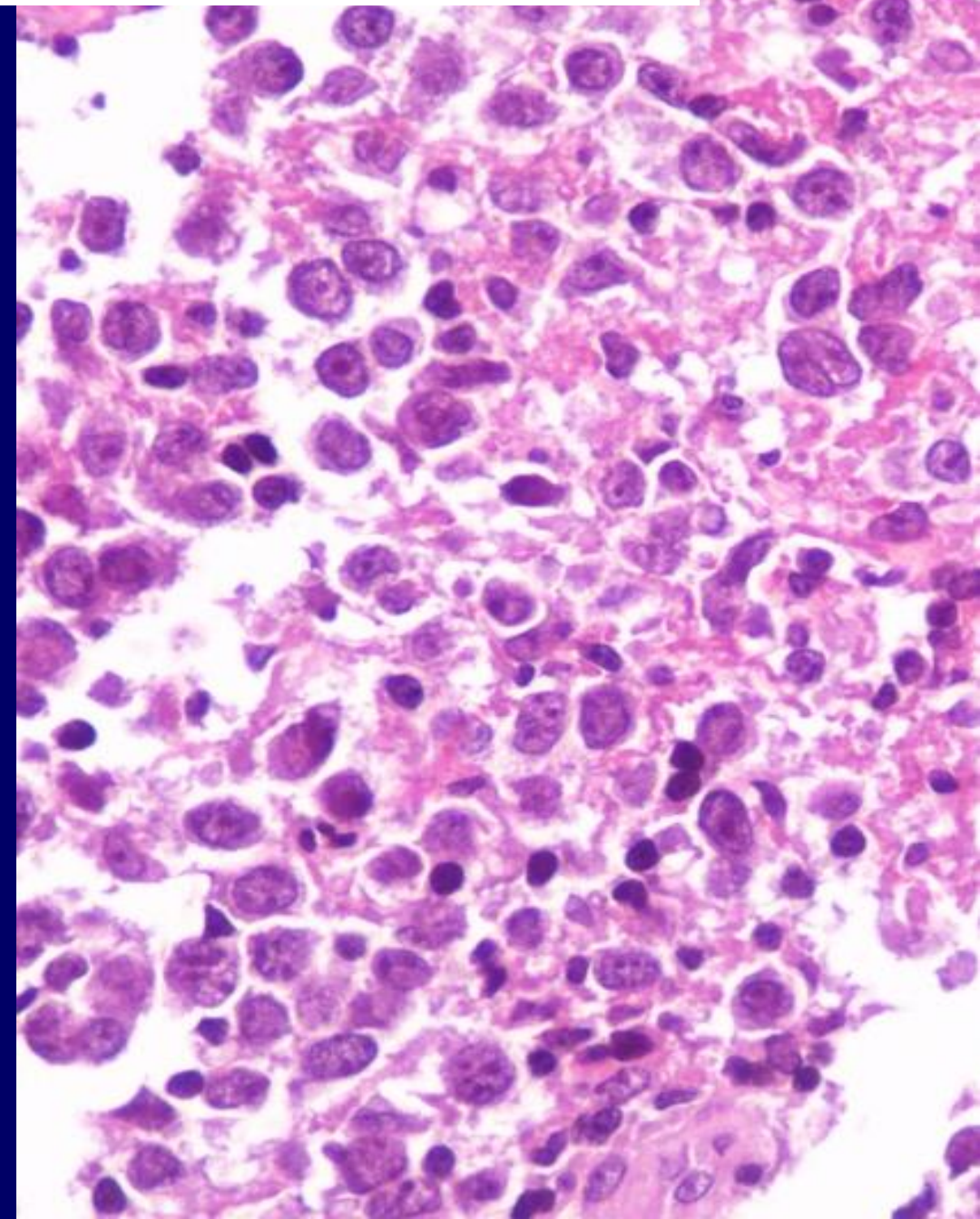
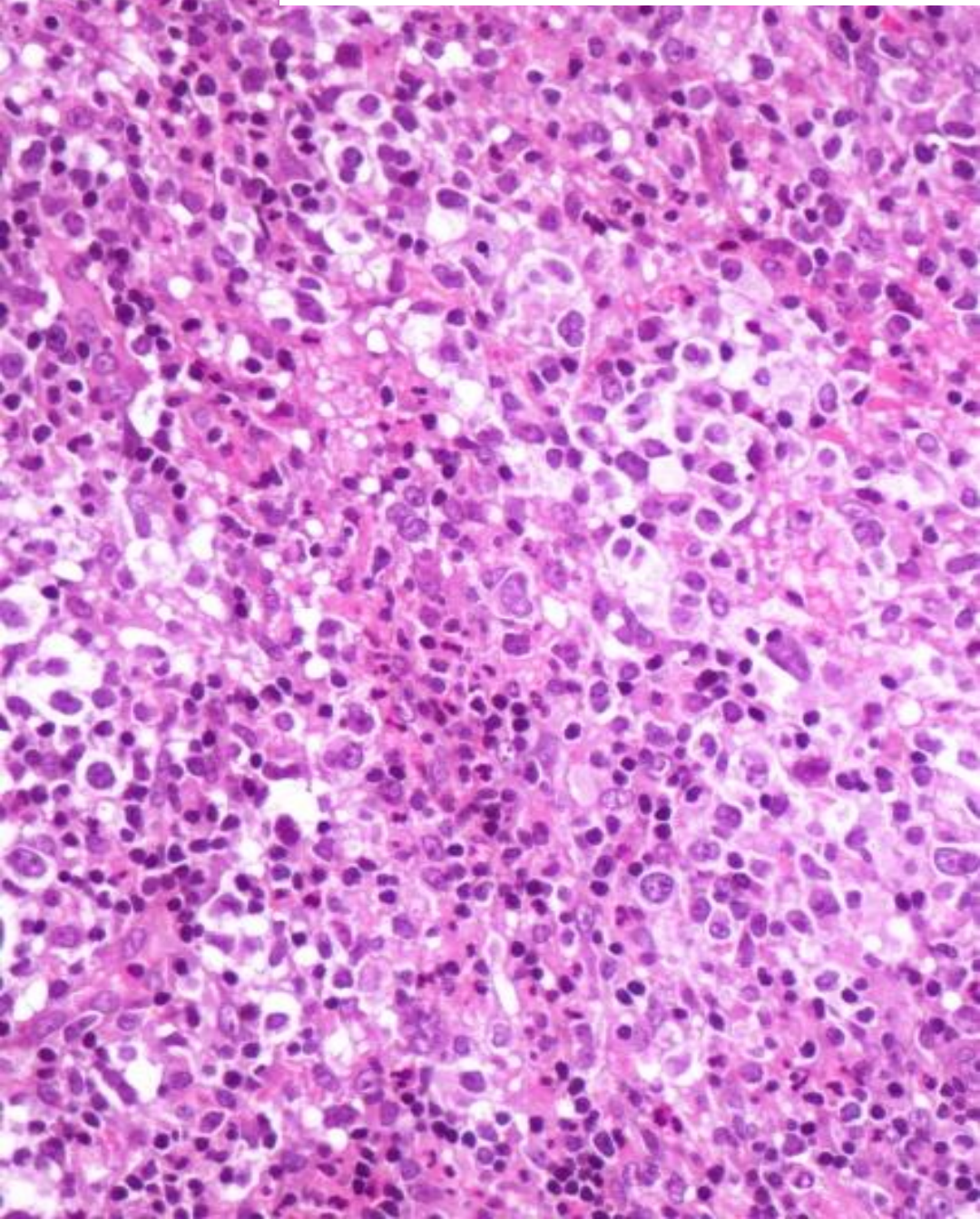
# Mediastinal Grey Zone Lymphoma

histologie de CHL avec

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



**H, 35 ans, tumeur médiastinale antérieure**

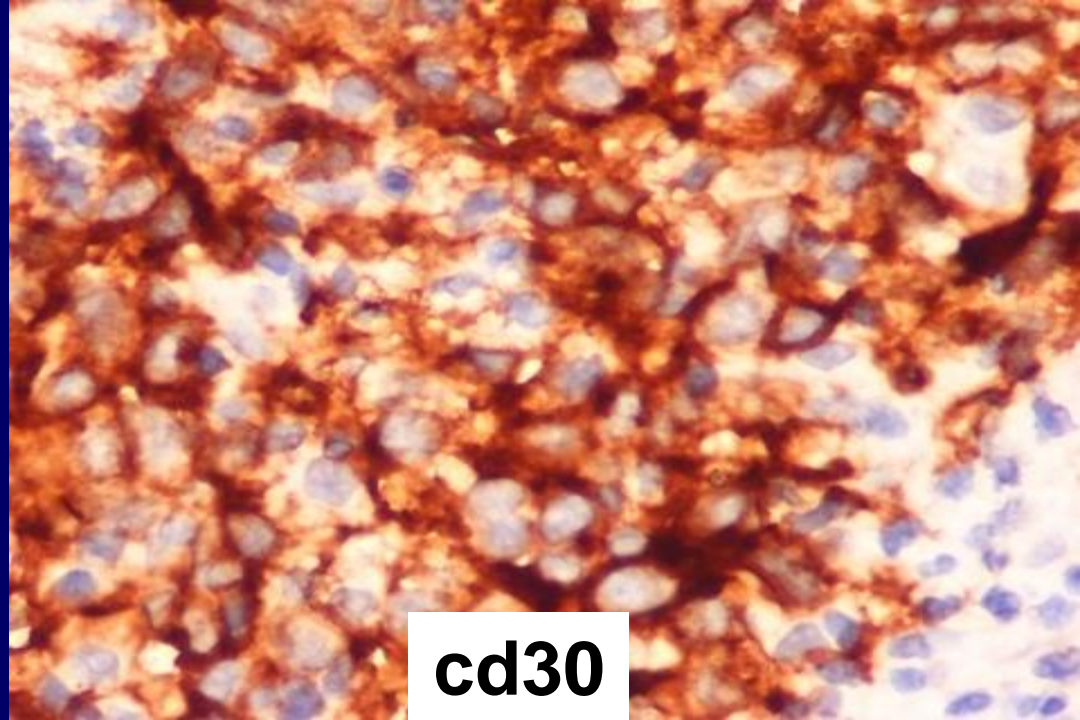


# Mediastinal Grey Zone Lymphoma

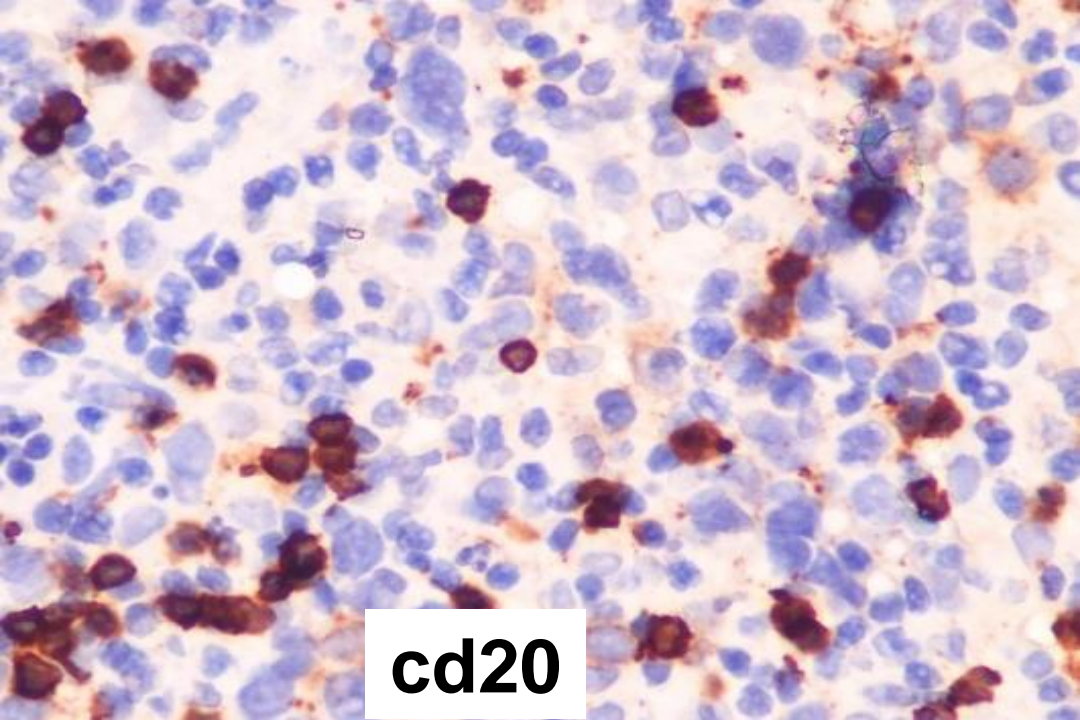
histologie de PMBL avec

- : cd20 , cd79a, OCT2,  
BOB1, PAX5

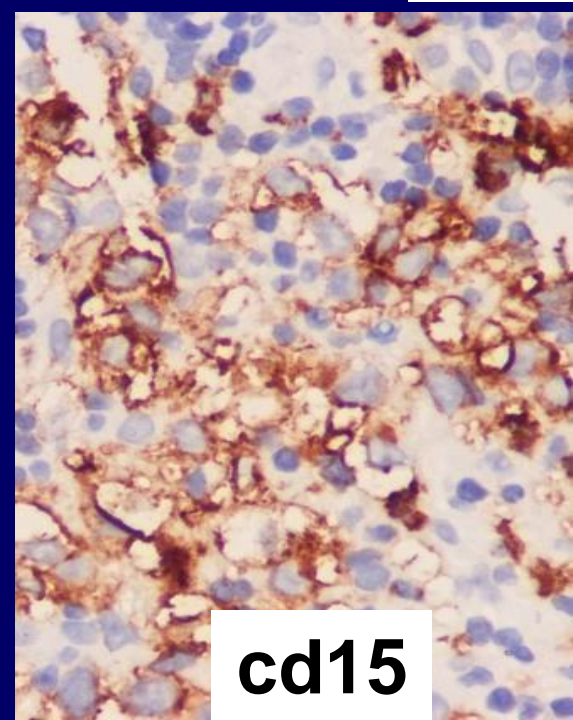
+ : cd30 , cd15 , EBV



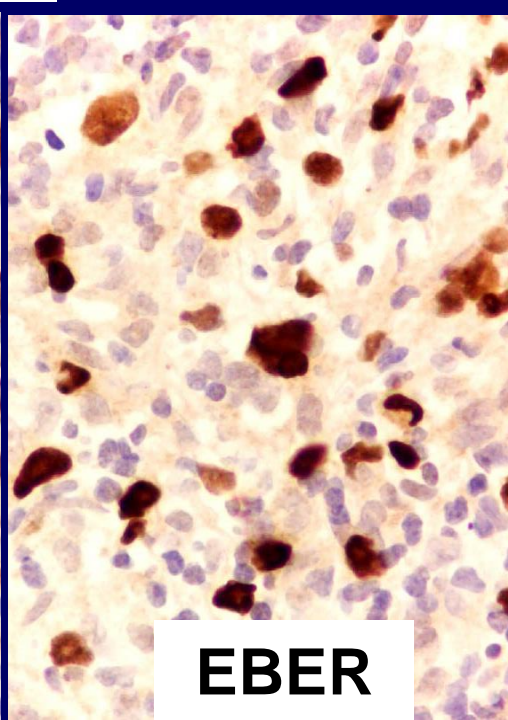
cd30



cd20



cd15

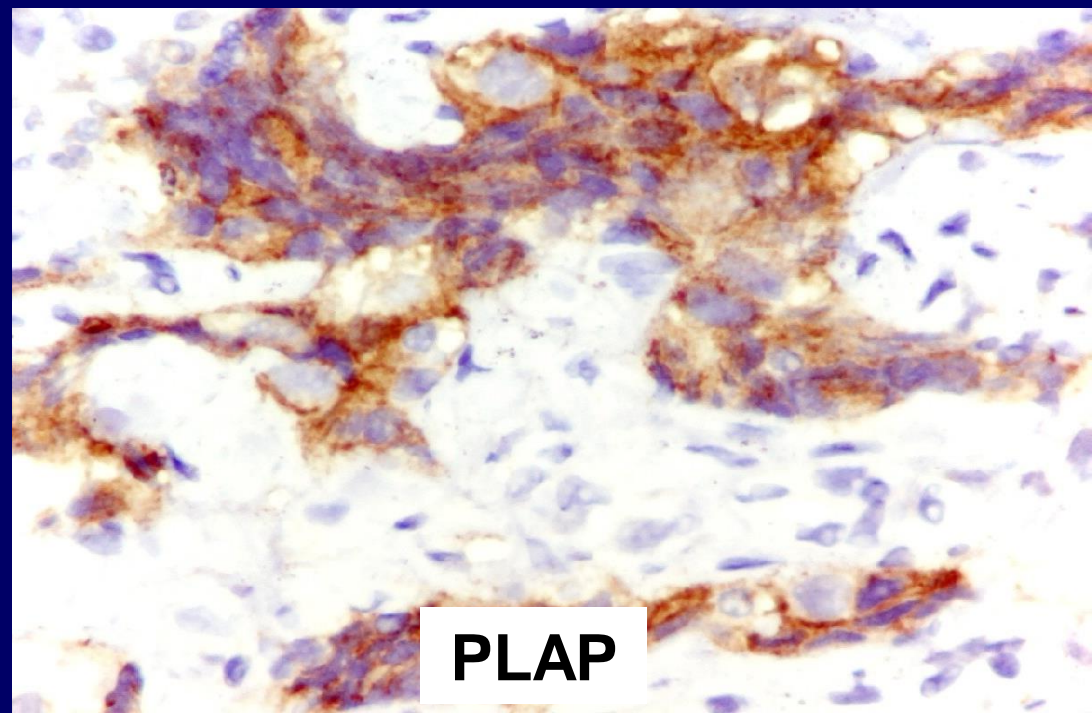
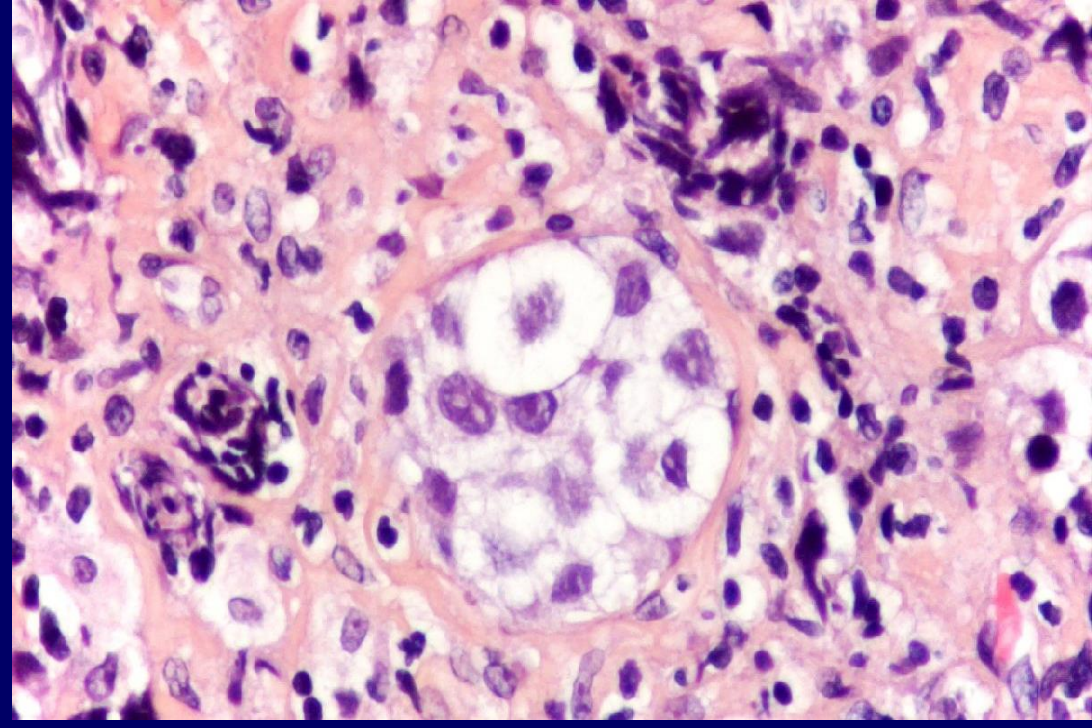


EBER



# Tumeurs du Médiastin avec sclérose

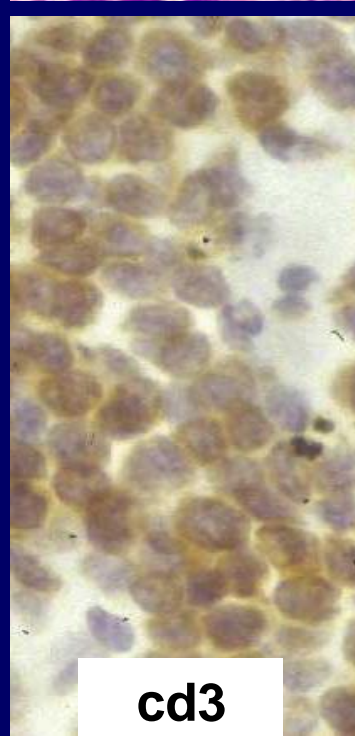
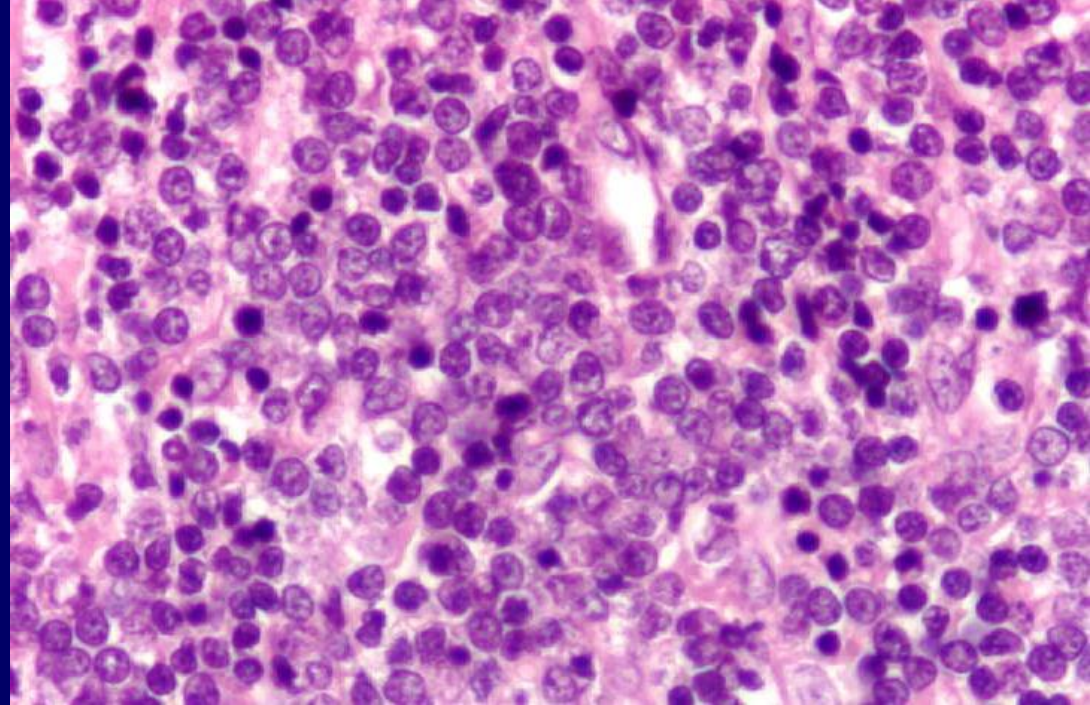
- Lymphome de Hodgkin
- Lymphome à grandes cellules B du médiastin
- **Séminome**
- Lymphome lymphoblastique (jeune)
- Schwannome



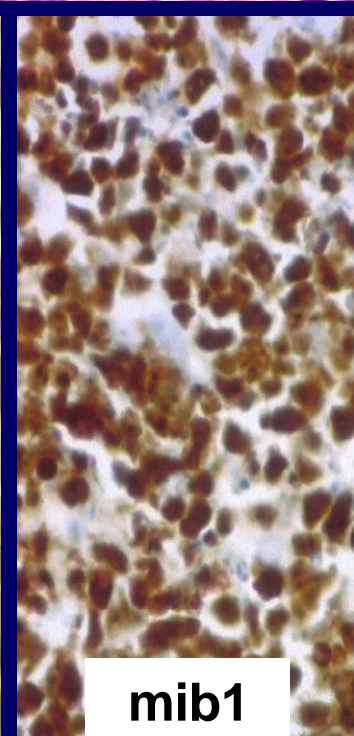
PLAP

# Tumeurs du Médiastin avec sclérose

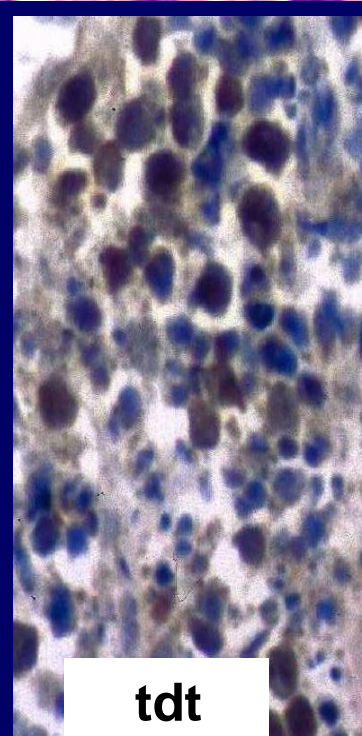
- Lymphome de Hodgkin
- Lymphome à grandes cellules B du médiastin
- Séminome
- **Lymphome lymphoblastique T (jeune)**
- Schwannome



cd3



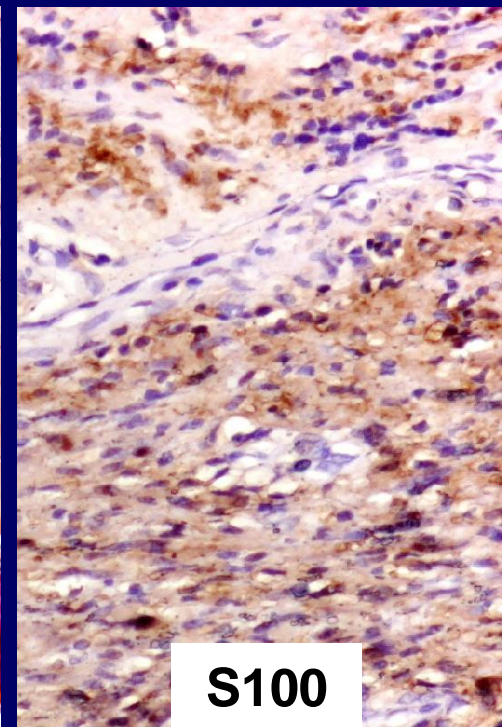
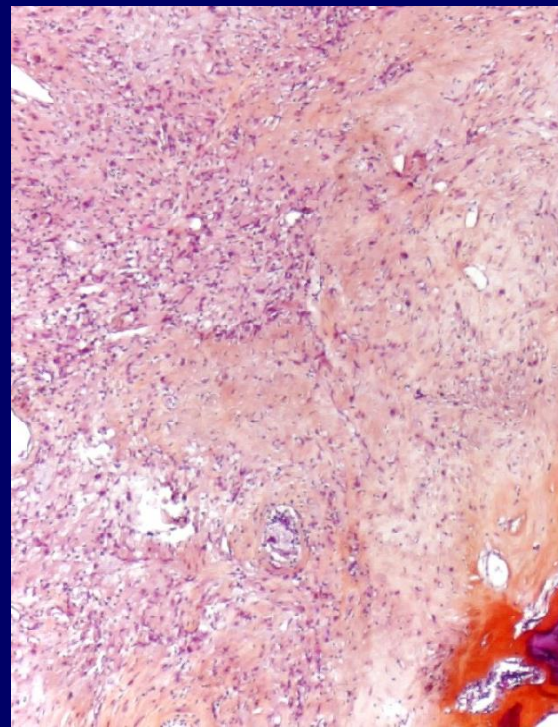
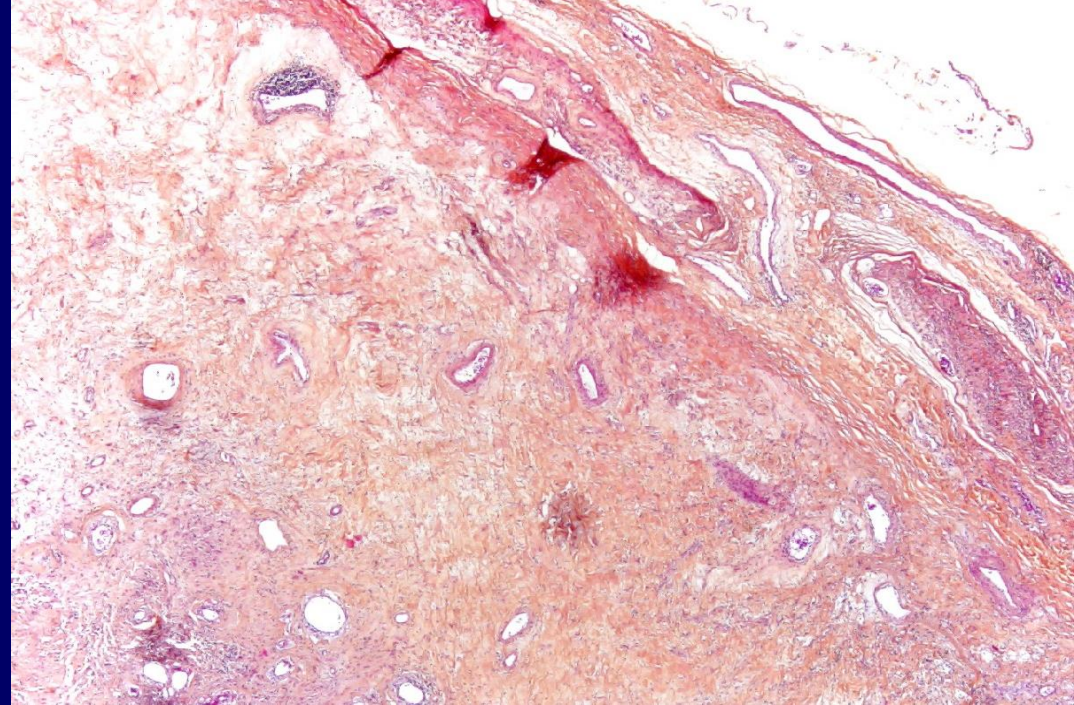
mib1



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