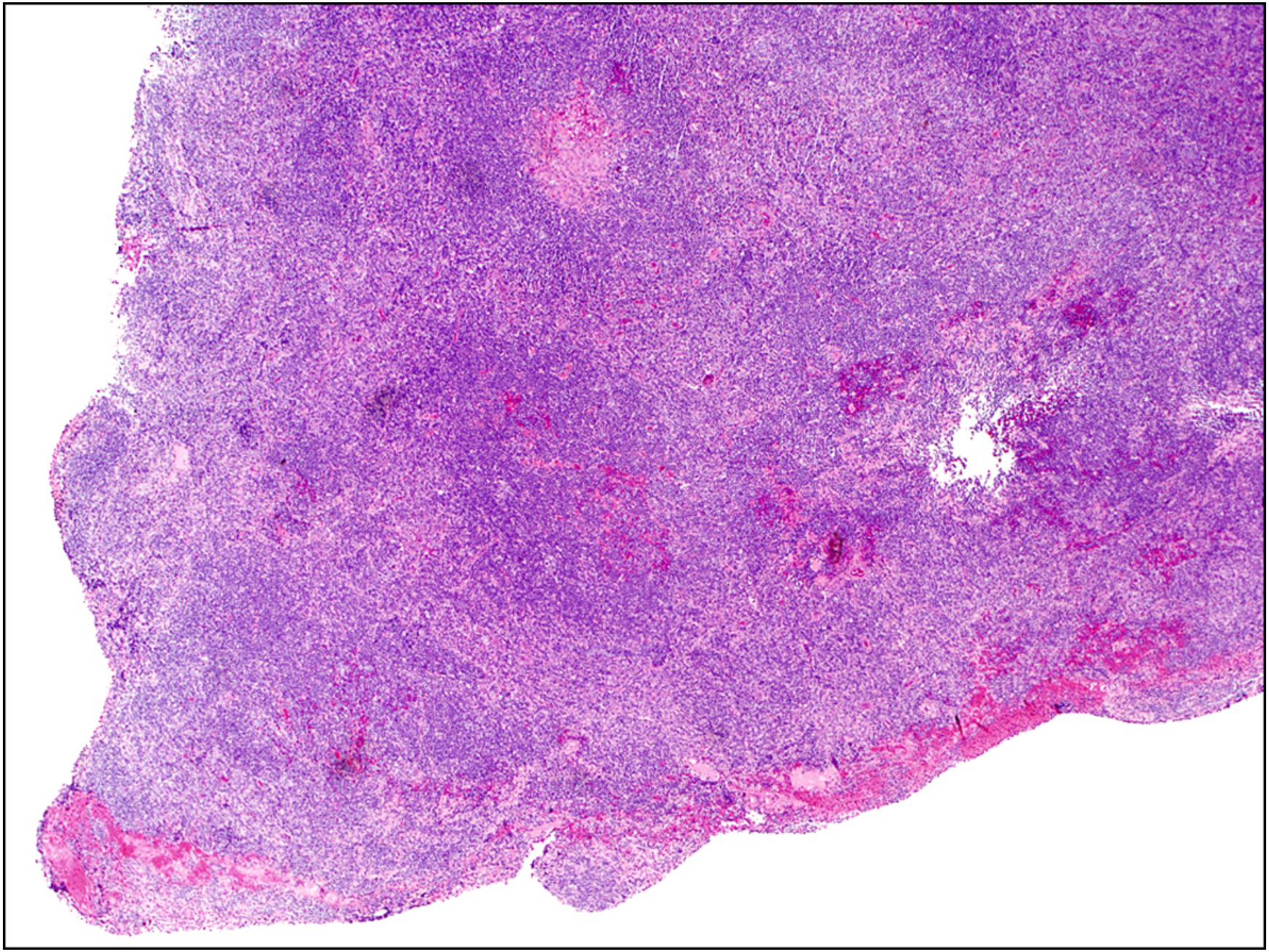
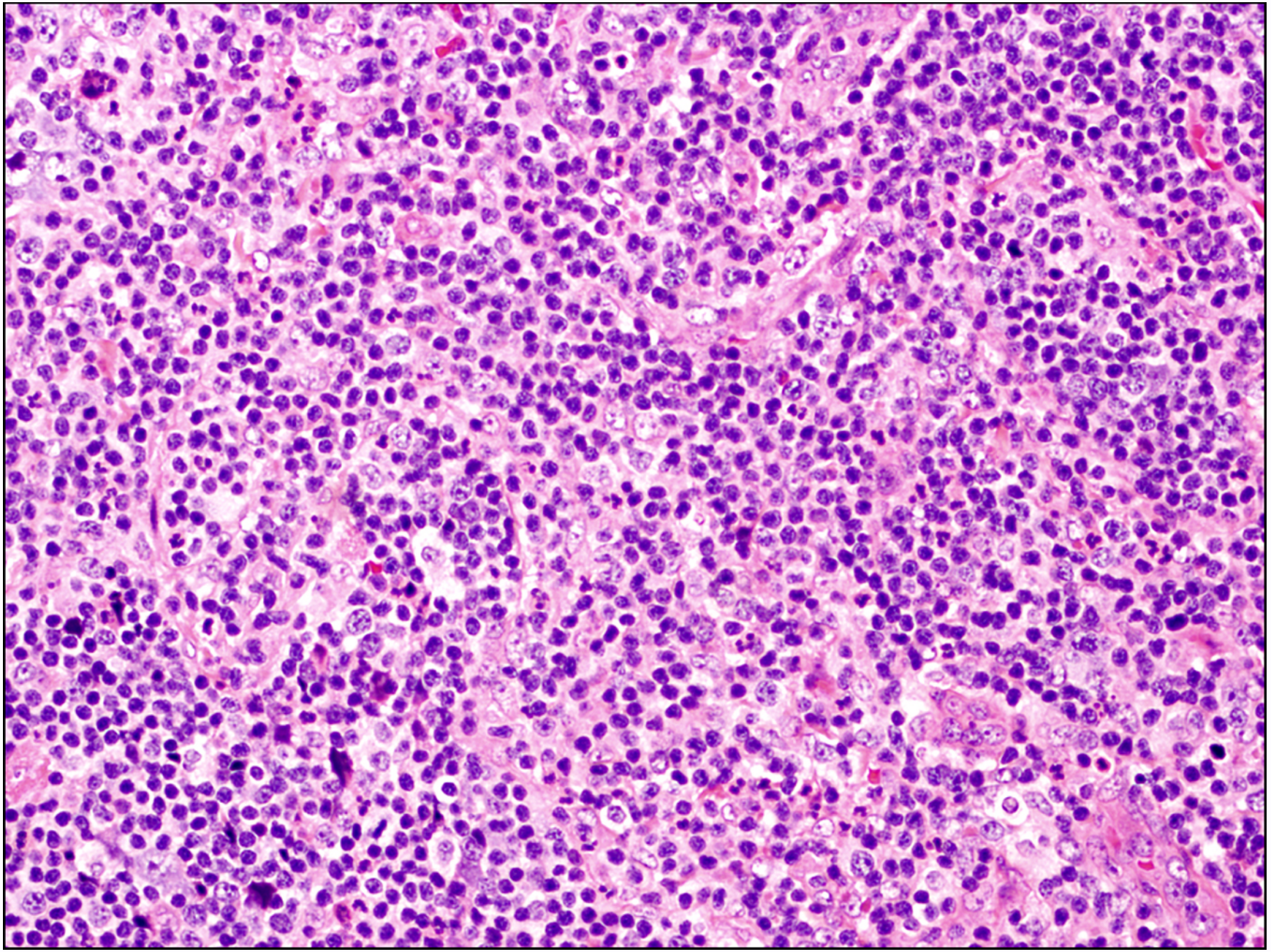
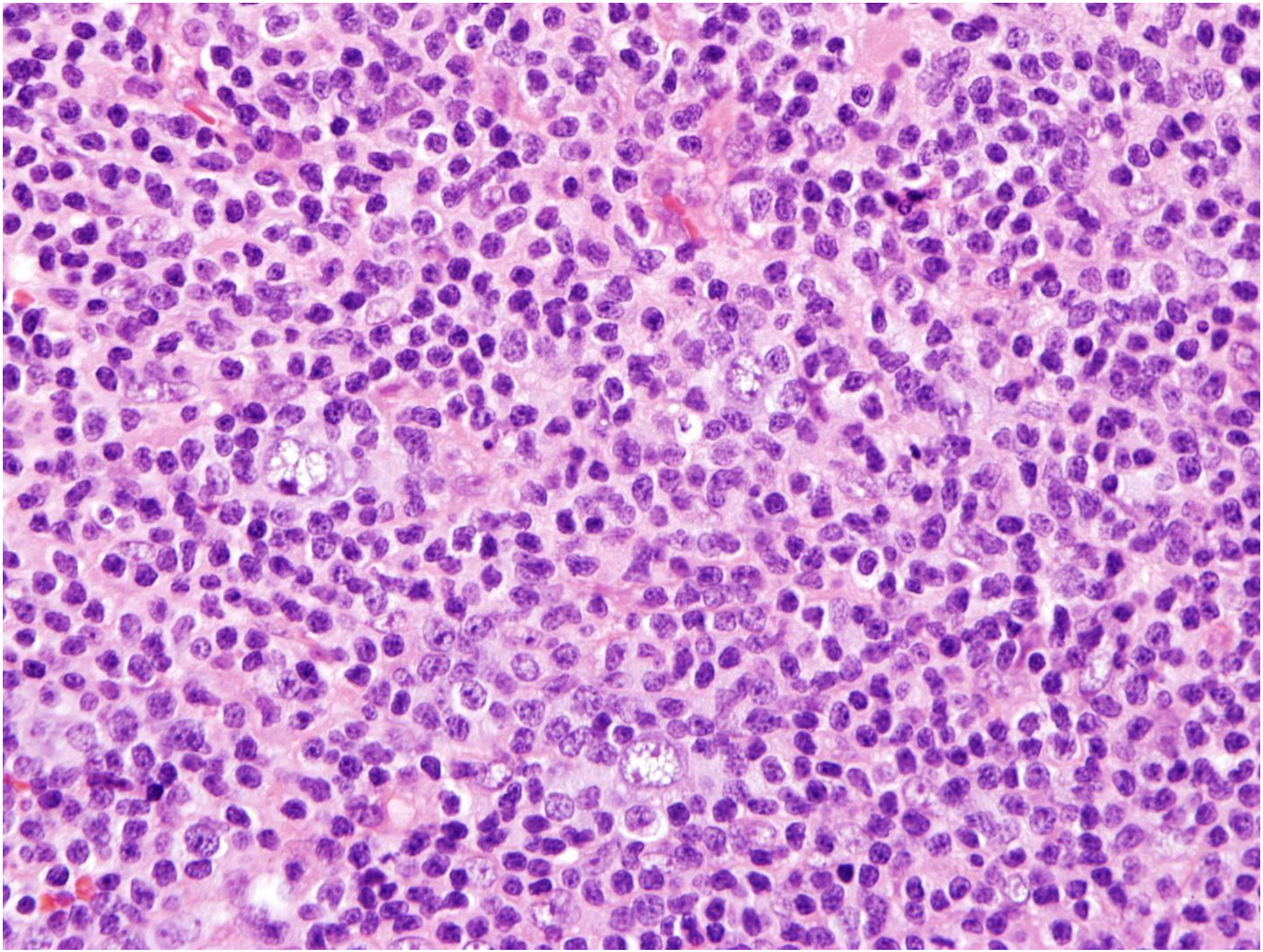
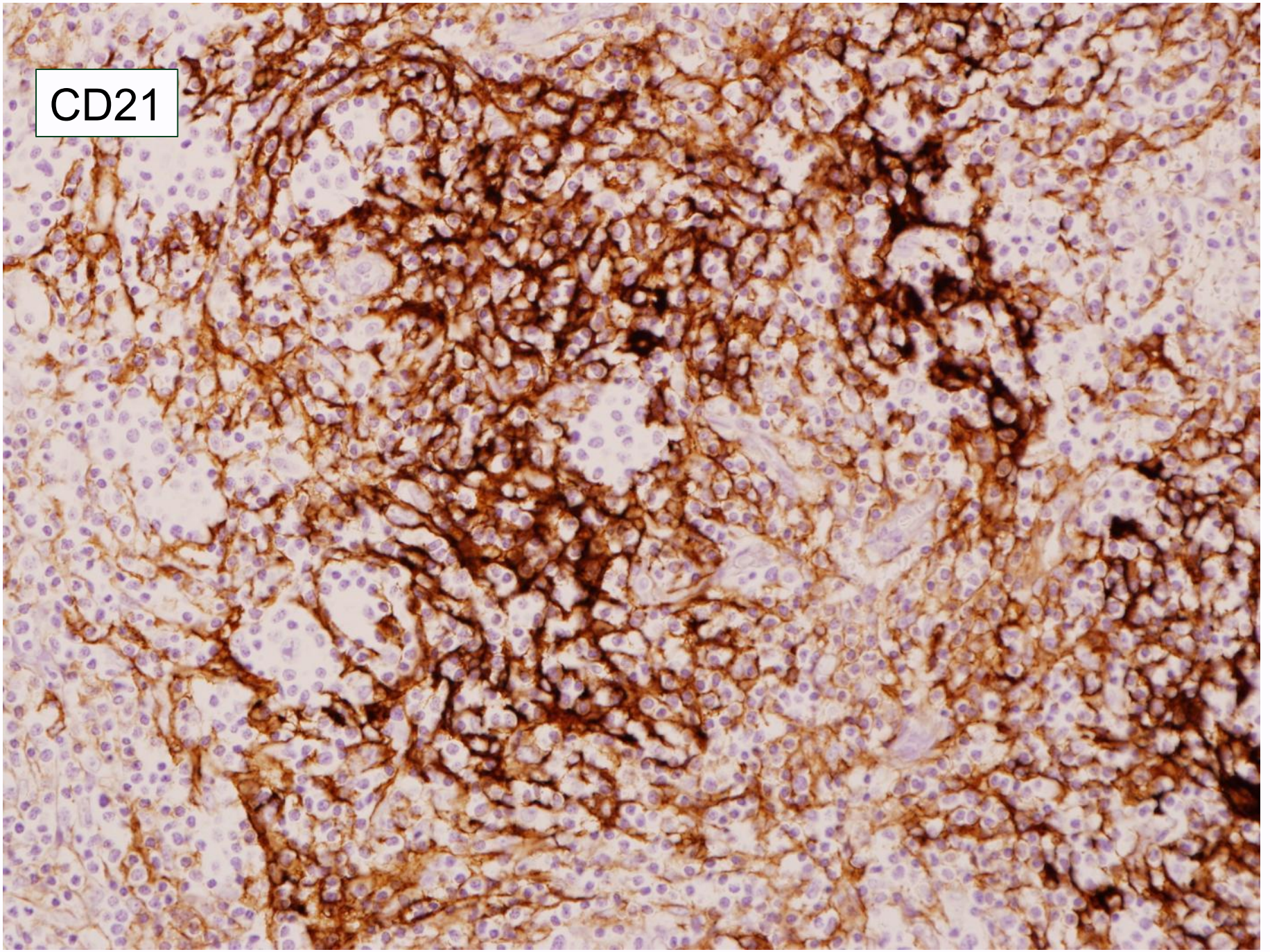
Case 14

- A 71 year old female presented with B-symptoms and generalized lymphadenopathy. A lymph node biopsy was performed.





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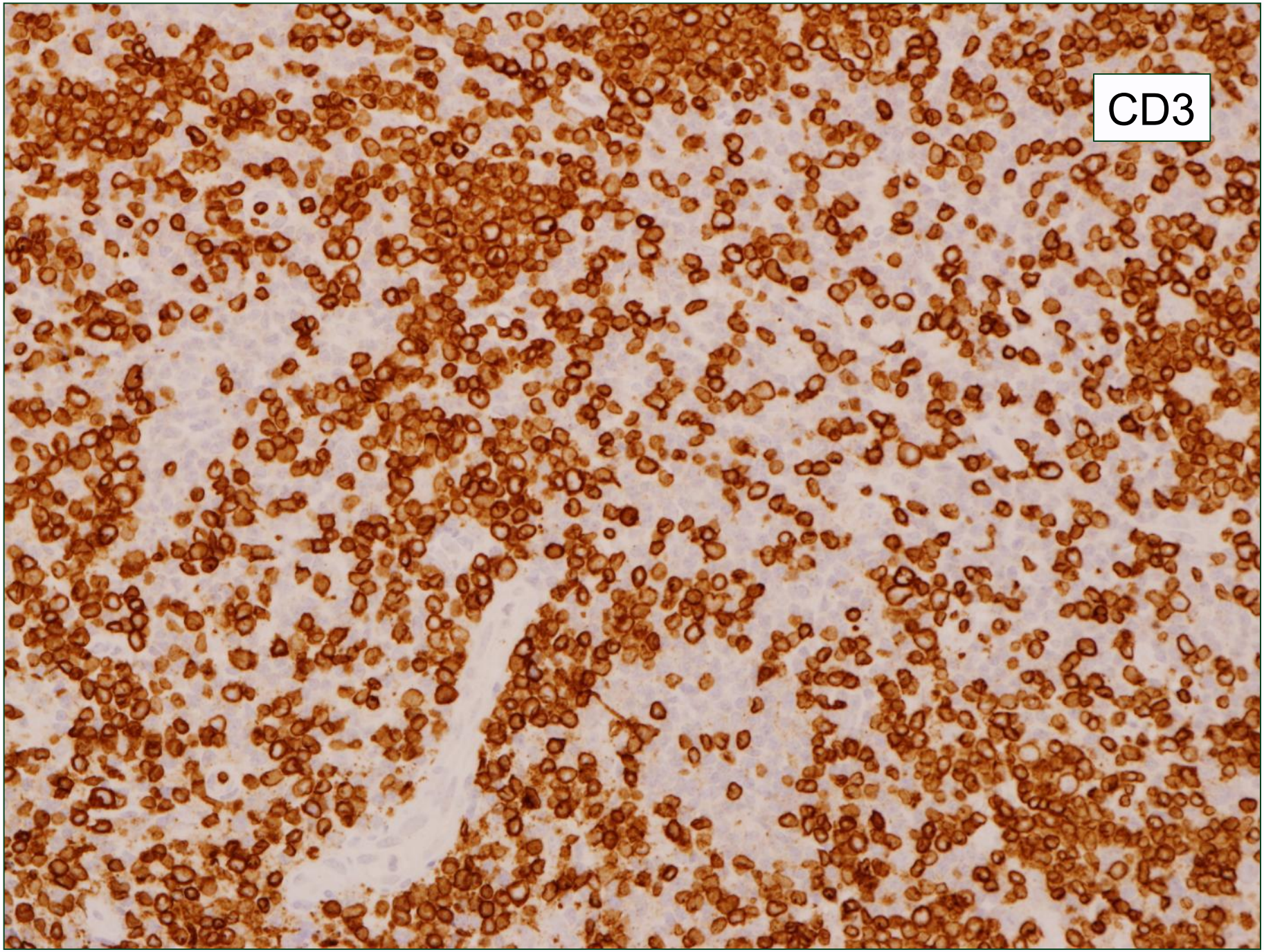
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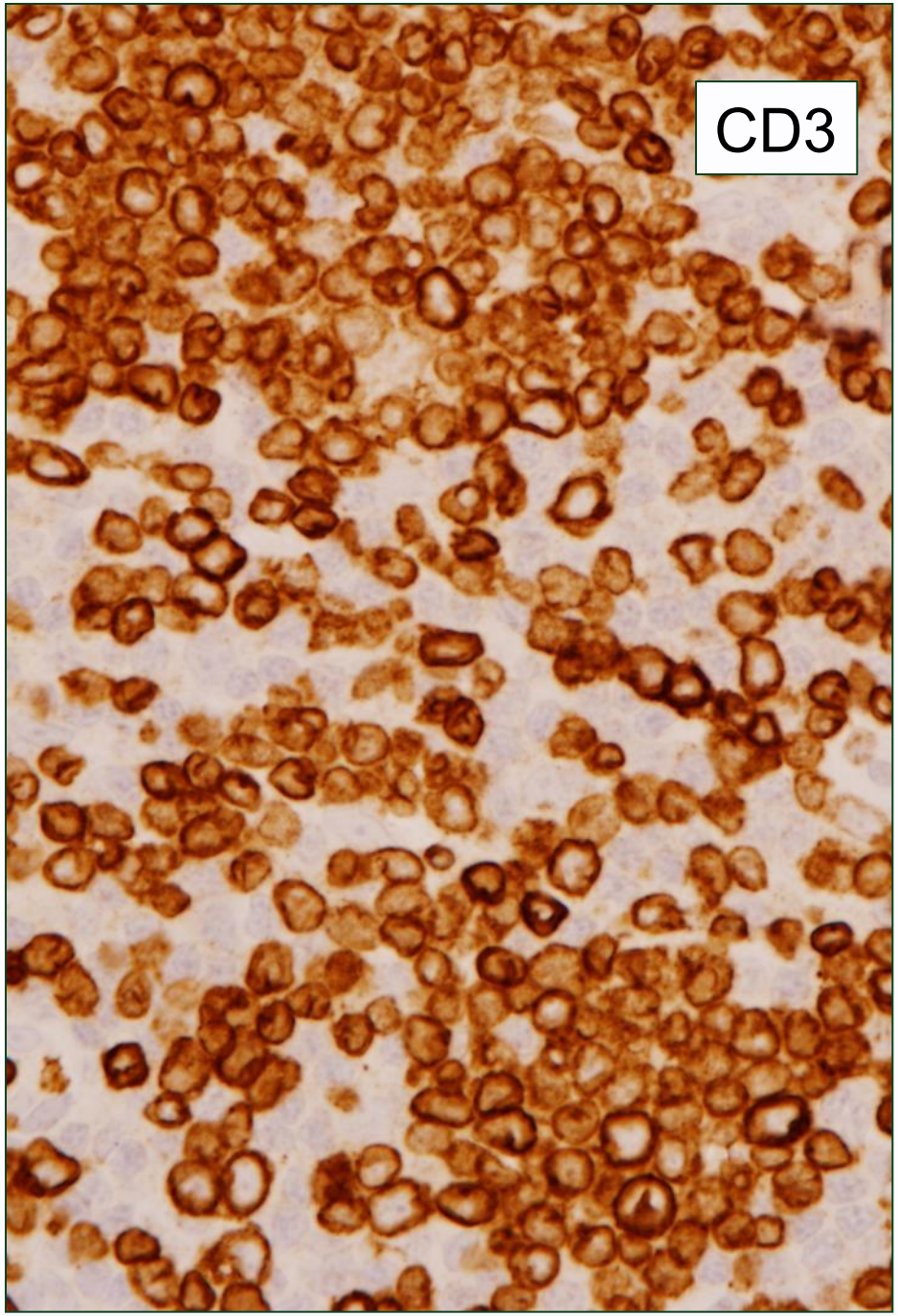
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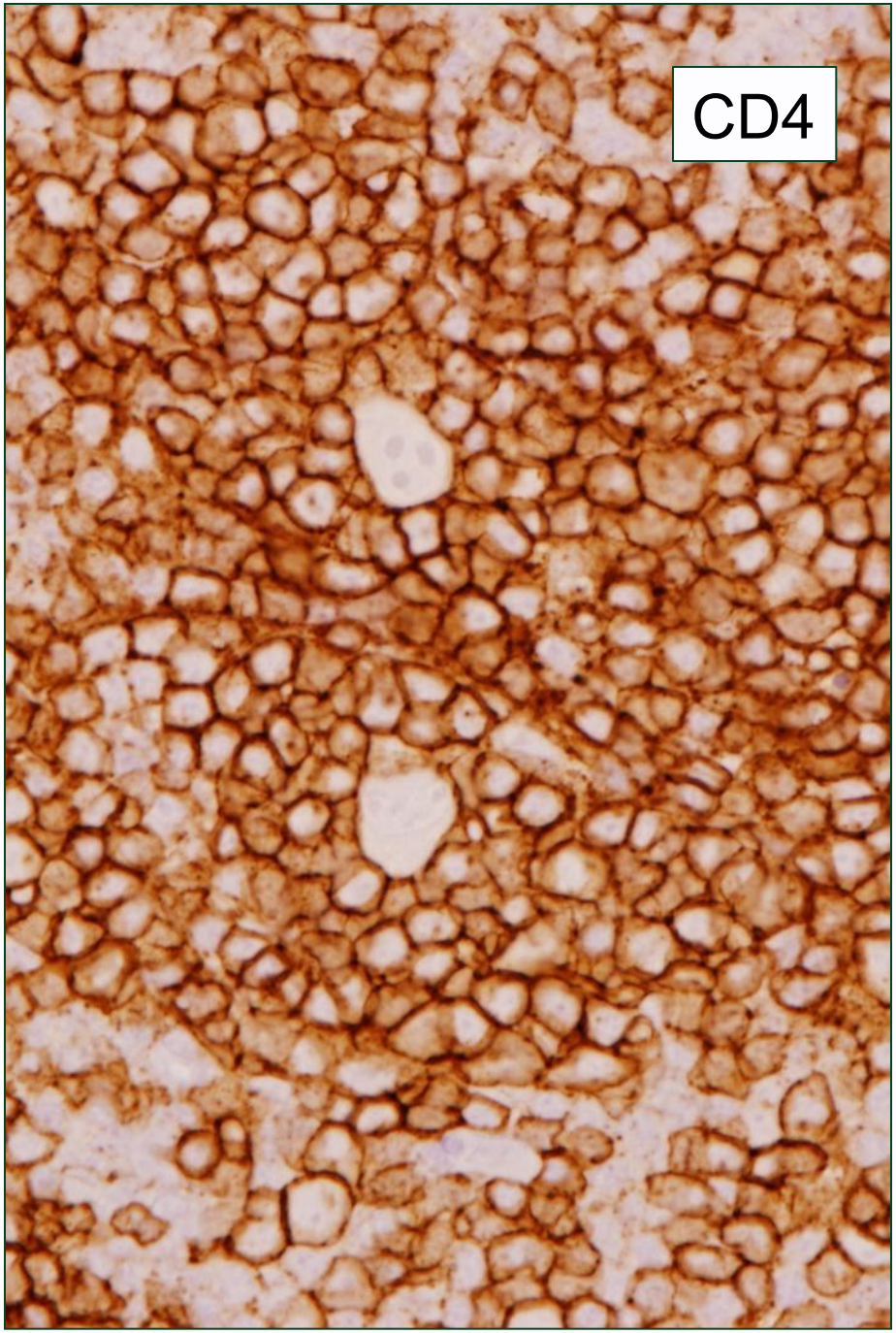
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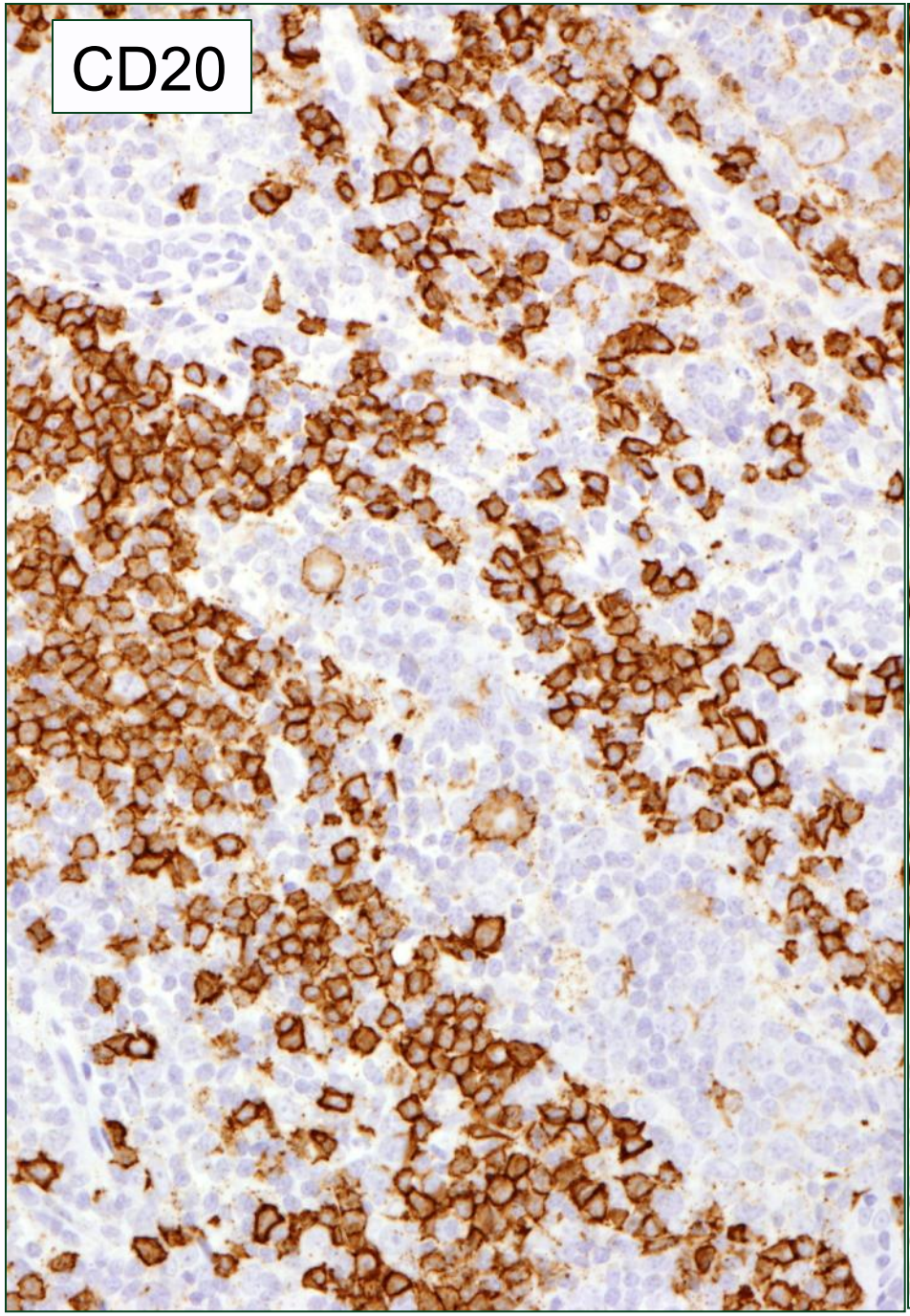
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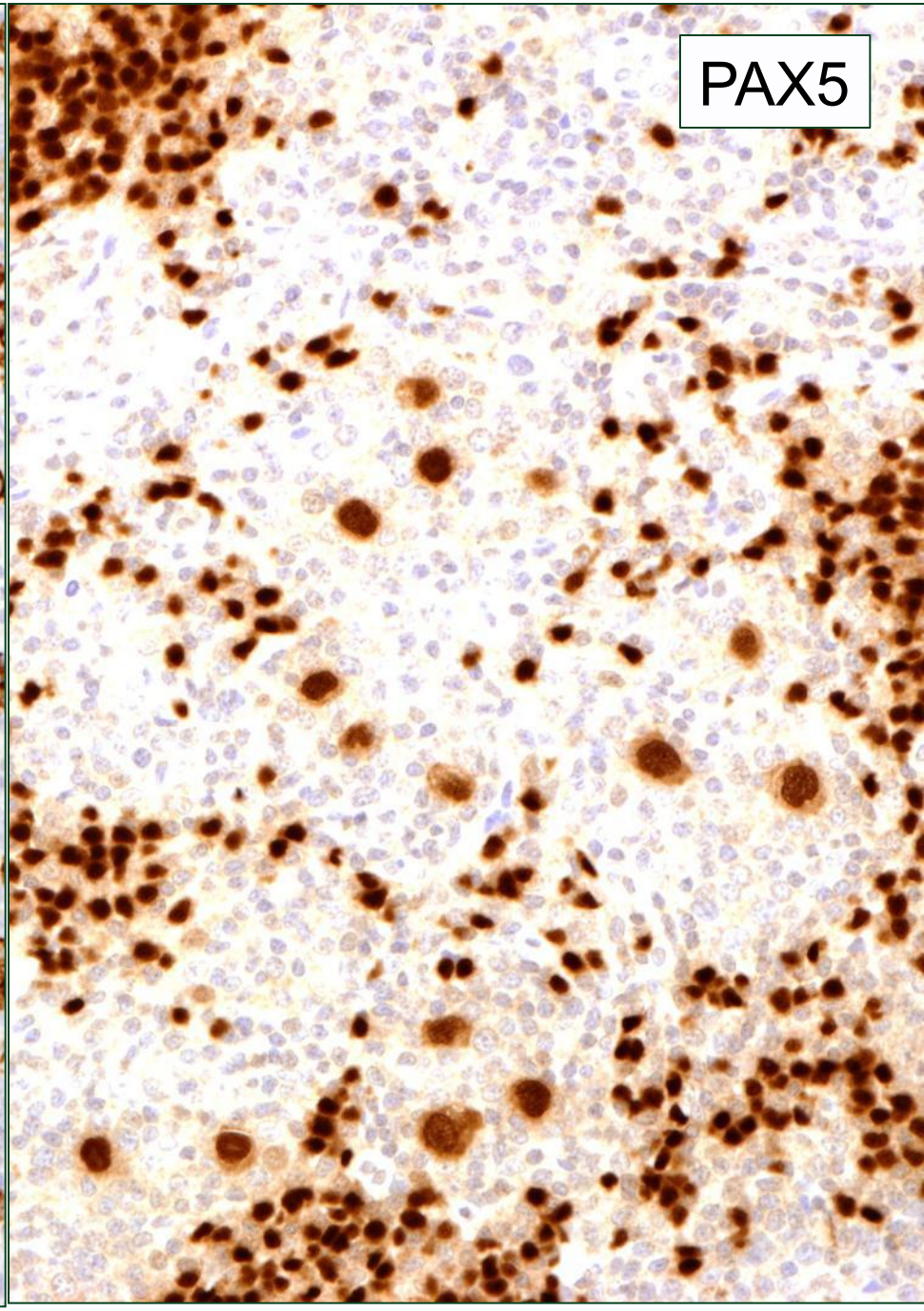
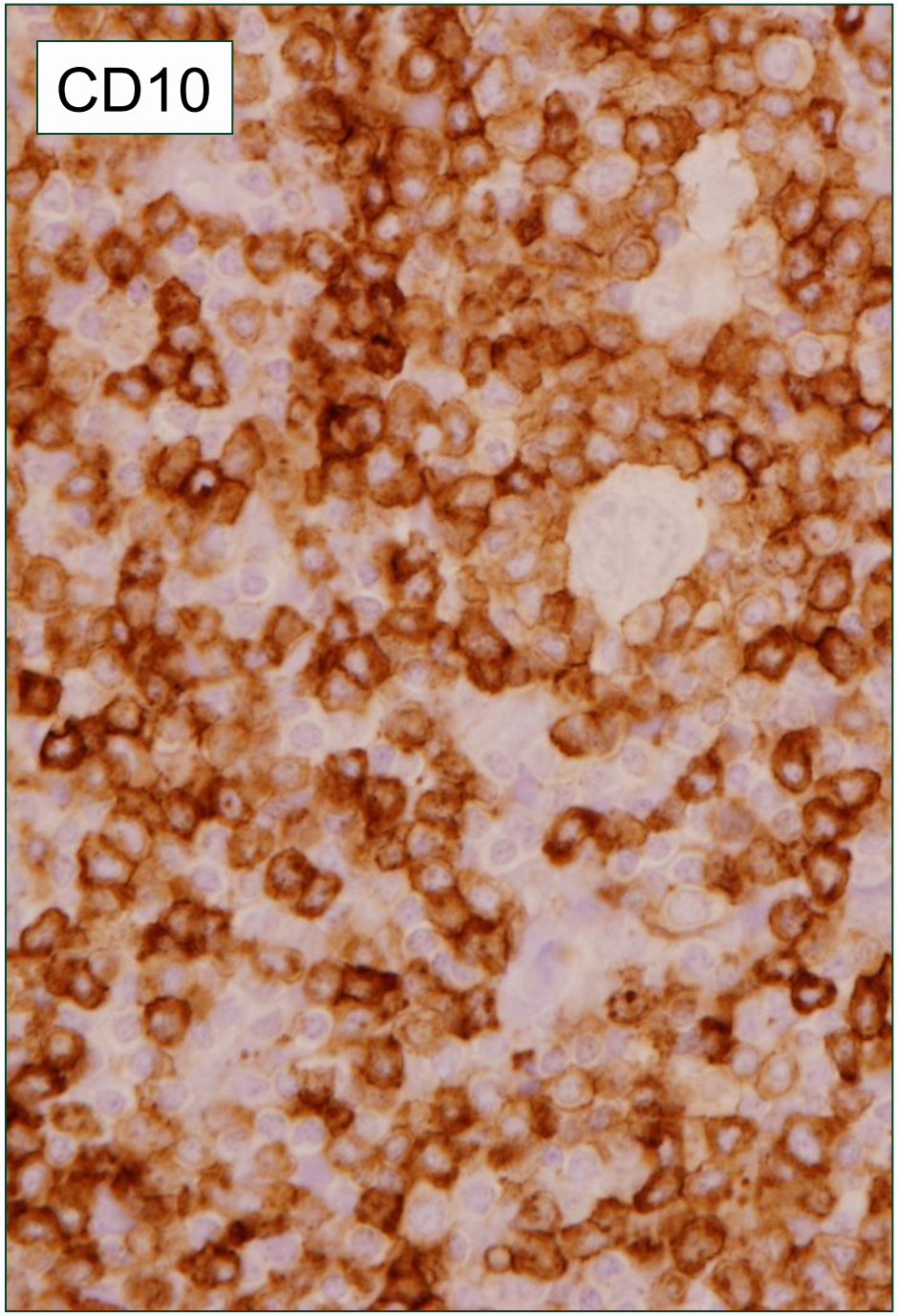

PAX5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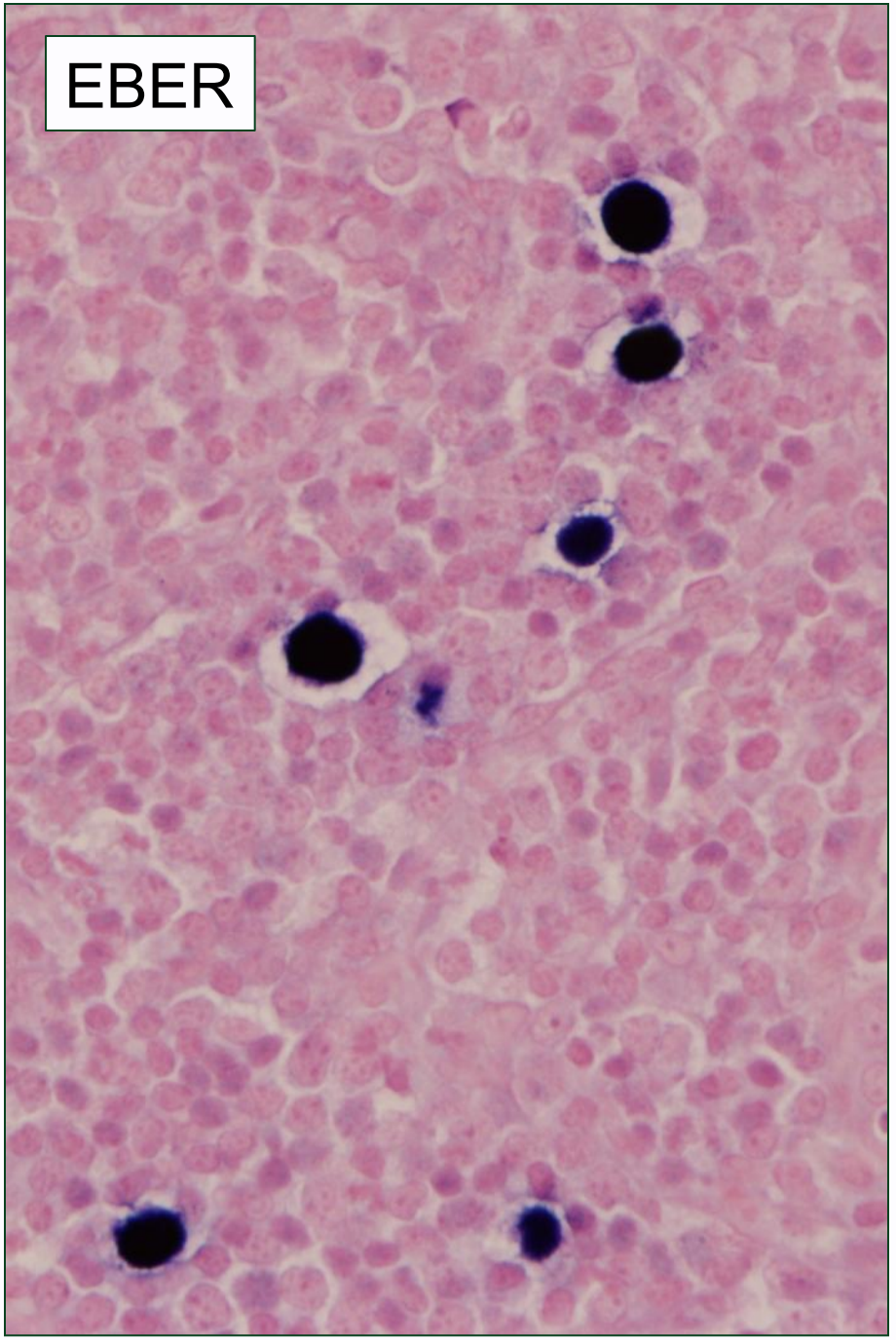
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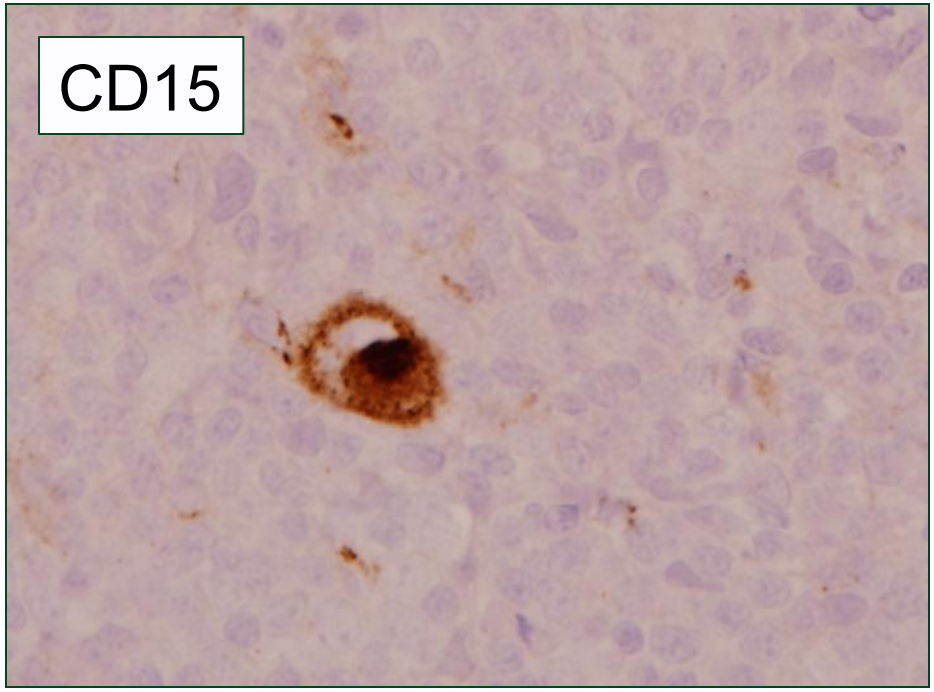
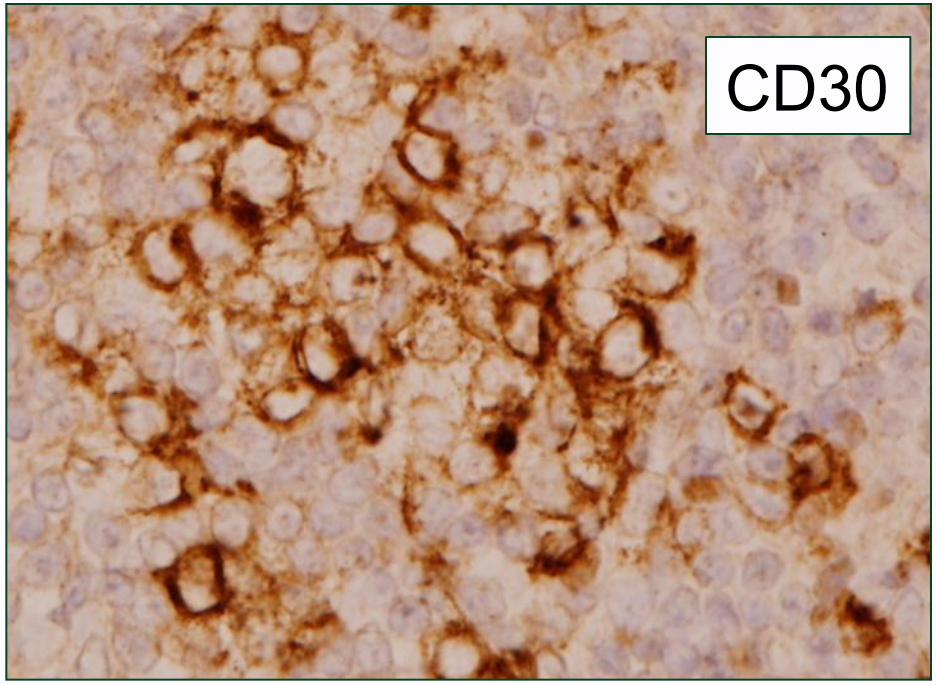

CD15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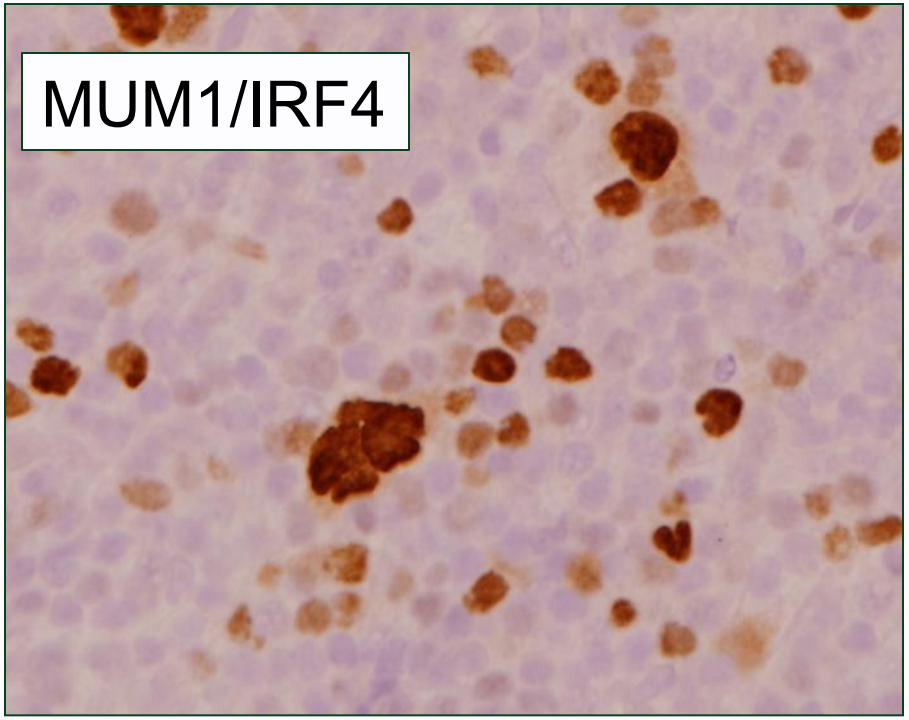
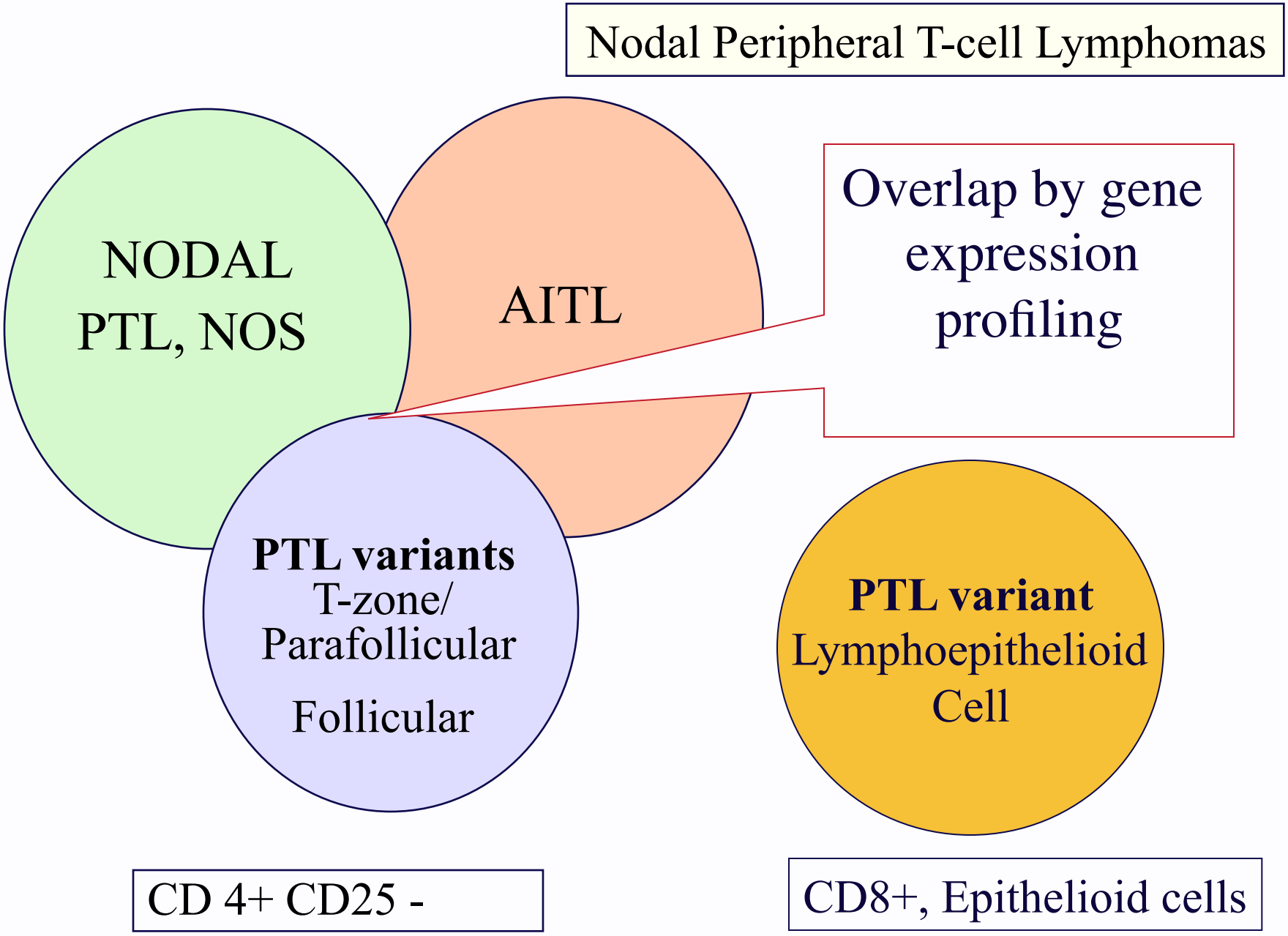

MUM1/IRF4

Case 14

- Diagnosis: Angioimmunoblastic T-cell lymphoma, with EBV-positive cells mimicking classical Hodgkin's lymphoma


Nodal Peripheral T-cell Lymphomas
NODAL
PTL, NOS
AITL
Overlap by gene expression profiling
PTL variants
T-zone/
Parafollicular
Follicular
PTL variant
Lymphoepithelioid
Cell
CD 4+ CD25 -
CD8+, Epithelioid cells

Angioimmunoblastic T-cell Lymphoma

- Initially thought to be an abnormal reactive process, *a disorder of immune regulation*
  - *Later defined as a form of peripheral T-cell lymphoma*
- The clinical syndrome is a nearly universal part of the disease definition
- One would be hesitant to make the diagnosis in the absence of the characteristic clinical picture

Angioimmunoblastic T-Cell Lymphoma

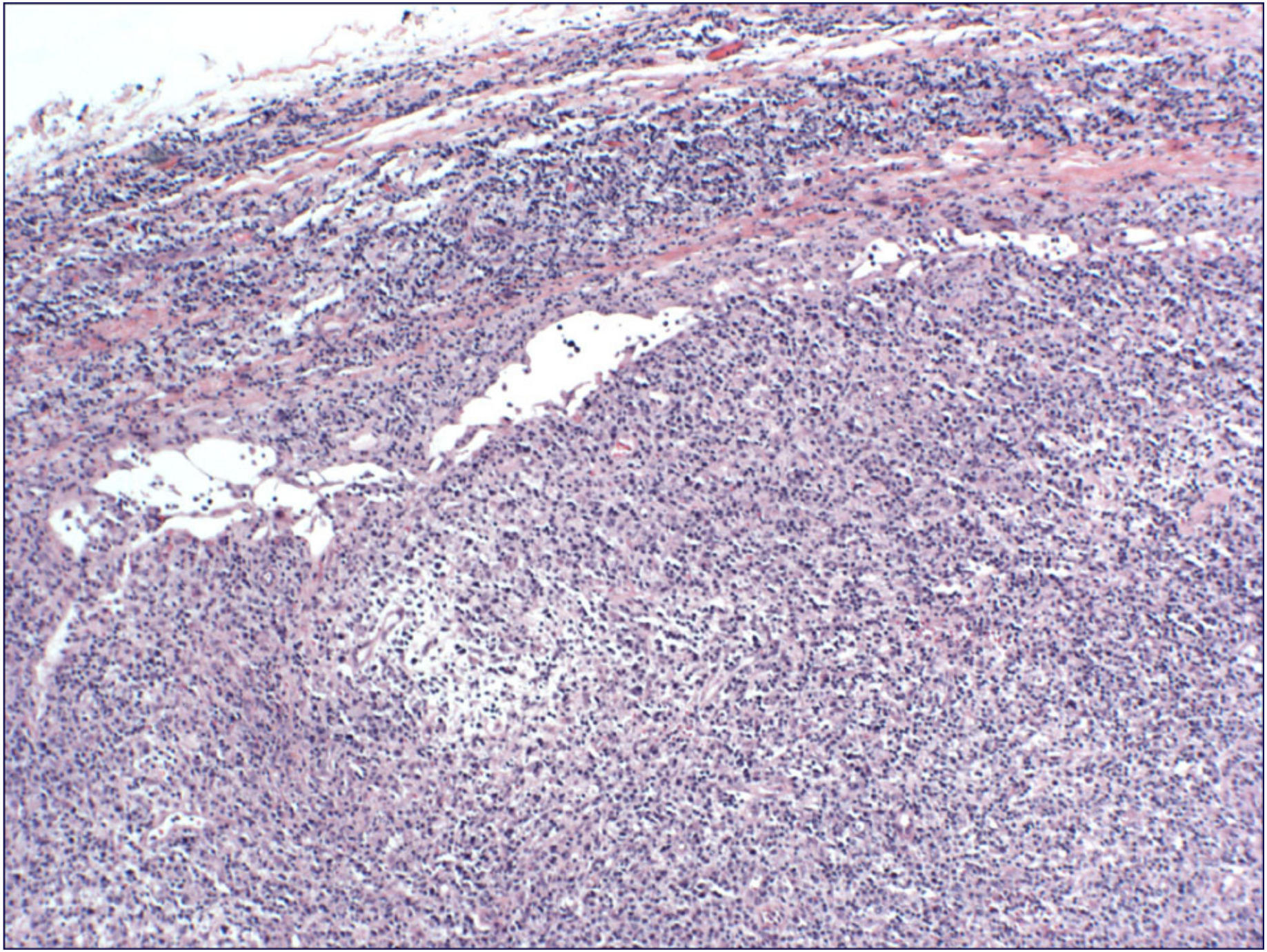
Clinical Features:

- Older adults, generalized lymphadenopathy
- Hepatosplenomegaly
- Skin rash, effusions, fever,
- Polyclonal hypergammaglobulinemia, hemolytic anemia
- Diverse constitutional signs & symptoms
- Aggressive clinical course, high risk of infectious complications with treatment

Angioimmunoblastic T-Cell Lymphoma

Pathologic Features:

- Arborizing vasculature
- Atypical T-lymphocytes with clear cytoplasm
- Scattered B-immunoblasts
- Plasmacytosis, eosinophils
- Regressed or absent follicles in most cases
- Some cases have follicular hyperplasia



Angioimmunoblastic T-Cell Lymphoma

Immunophenotypic features

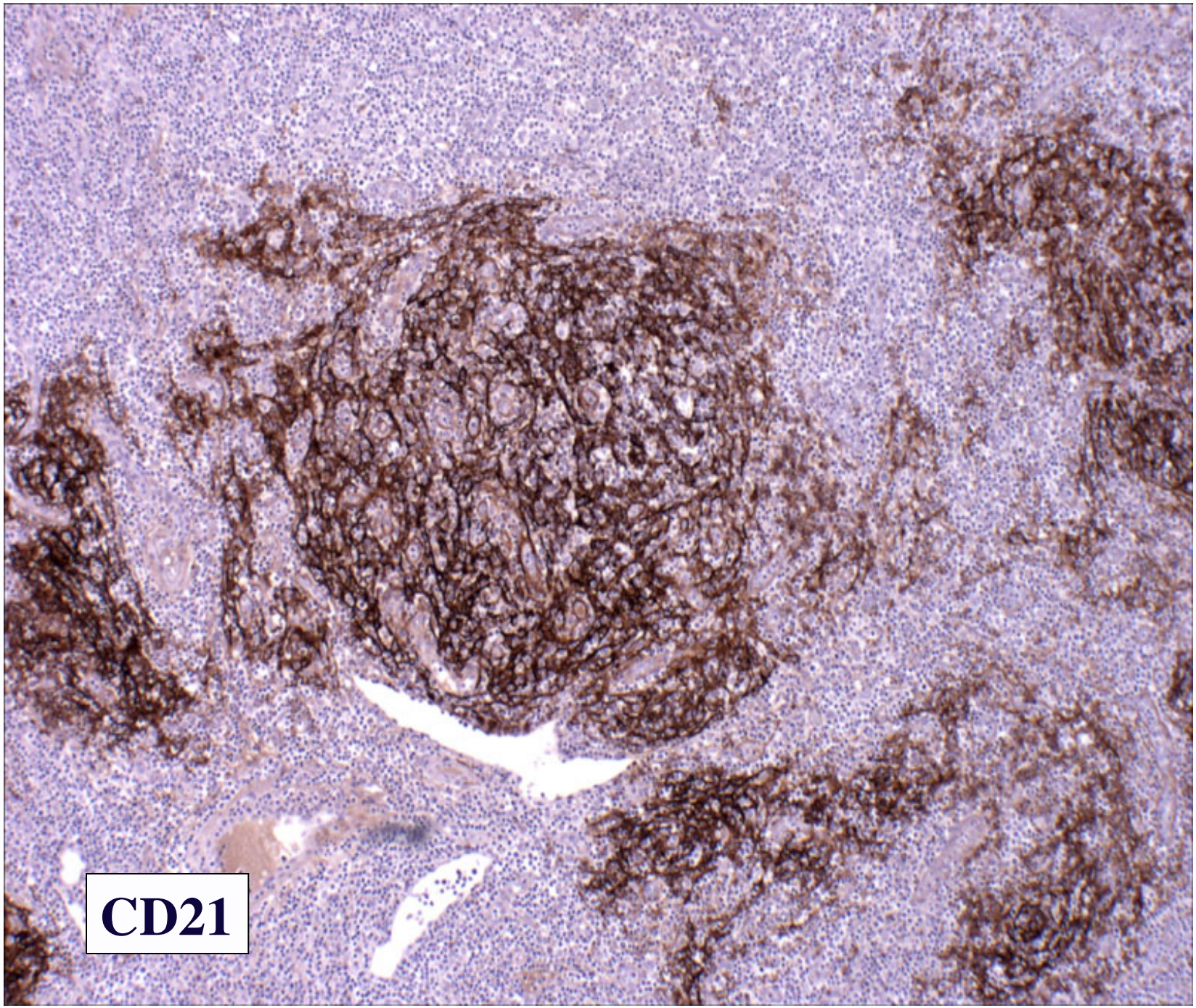
- CD4+, CD10+, PD-1+, BCL-6- T-cells (T_{FH})
- Expression of CXCL 13
- Extensive FDC CD21+ meshwork surrounding high endothelial venules (HEV)
- Scattered large B-cell blasts, usually EBV+
- Polyclonal/ rarely monoclonal plasma cells

Genotypic features

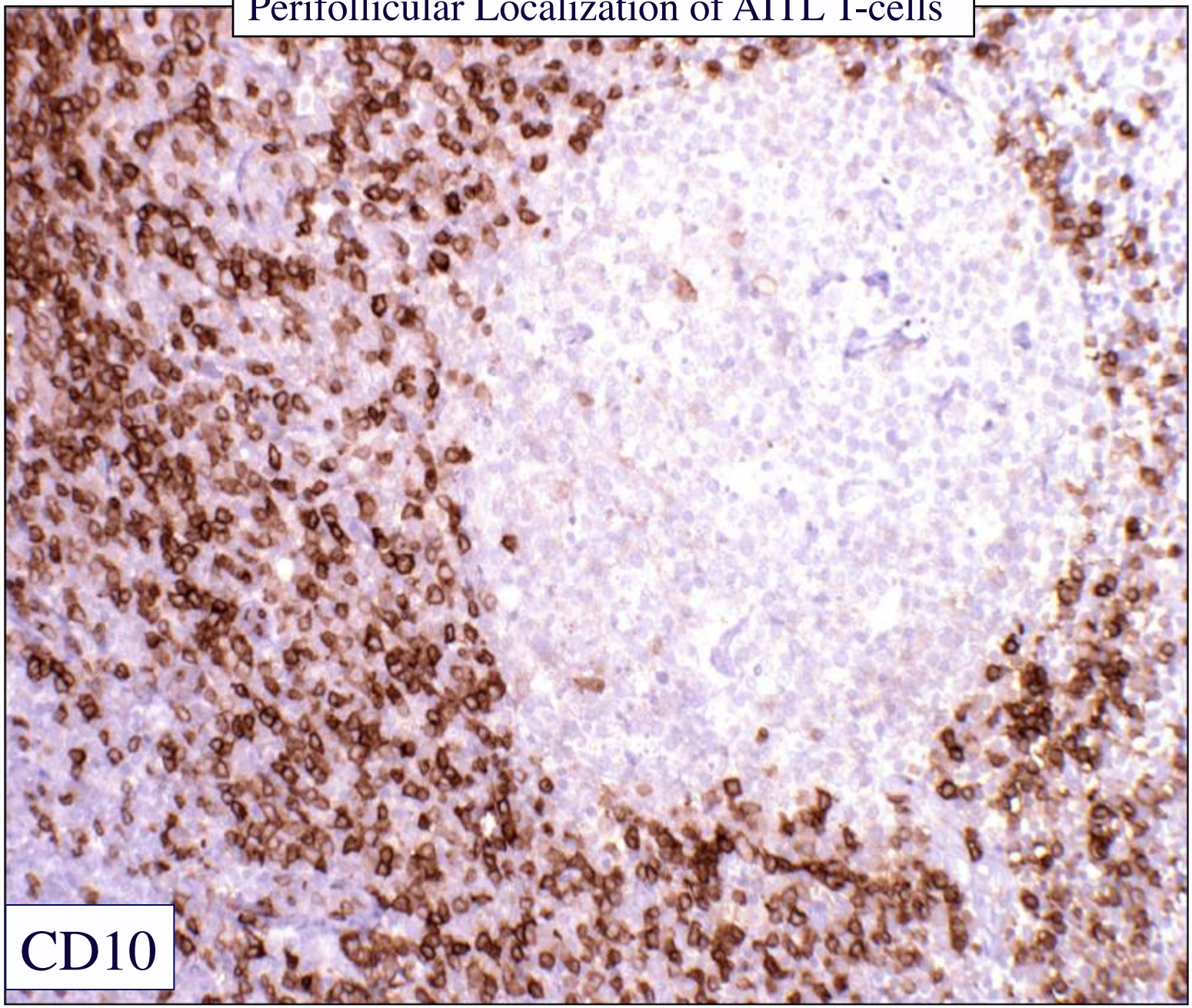
- > 90% TCR rearranged; 10-40% IG rearranged

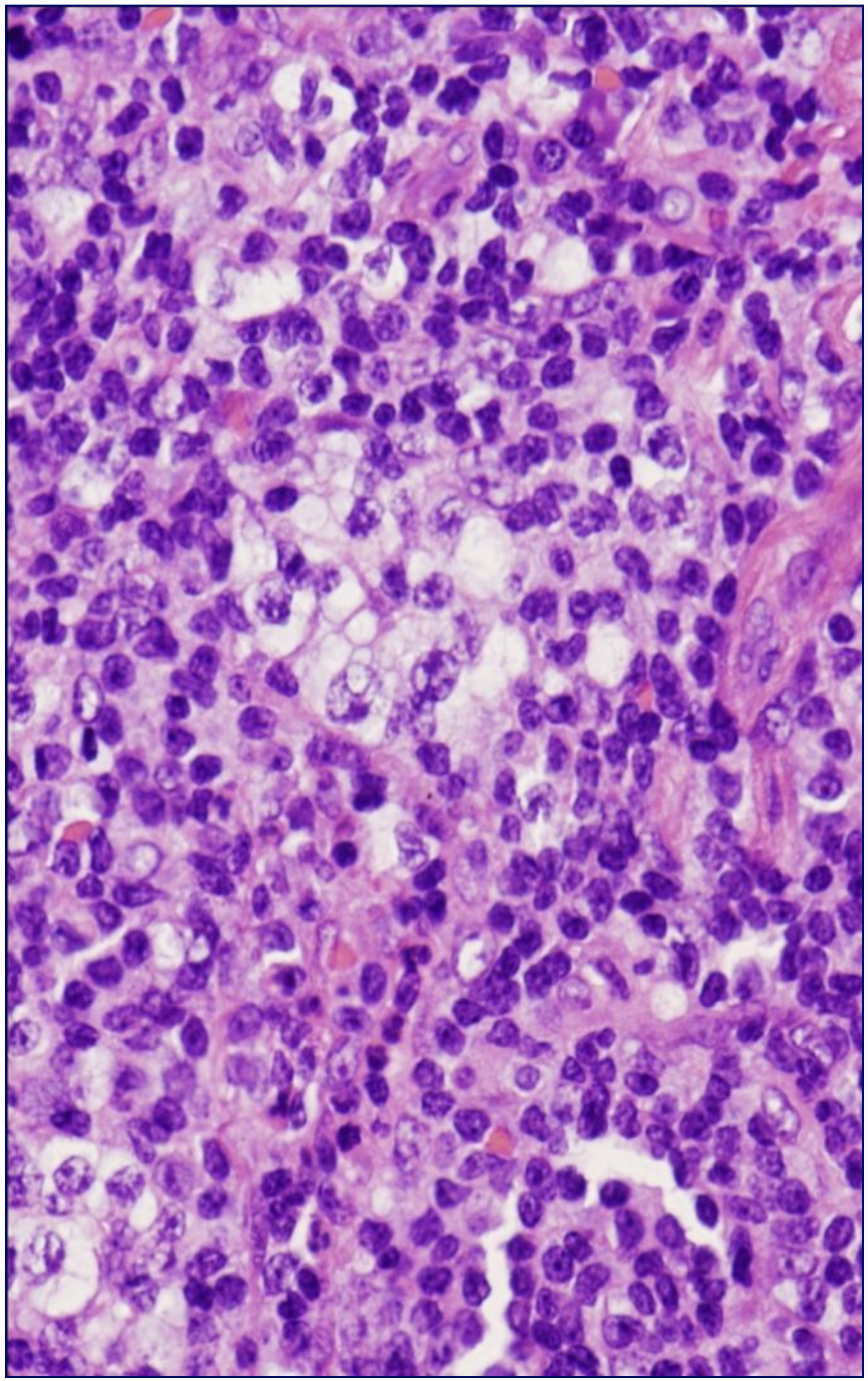
CXCL13 expression explains many aspects of AITL pathology

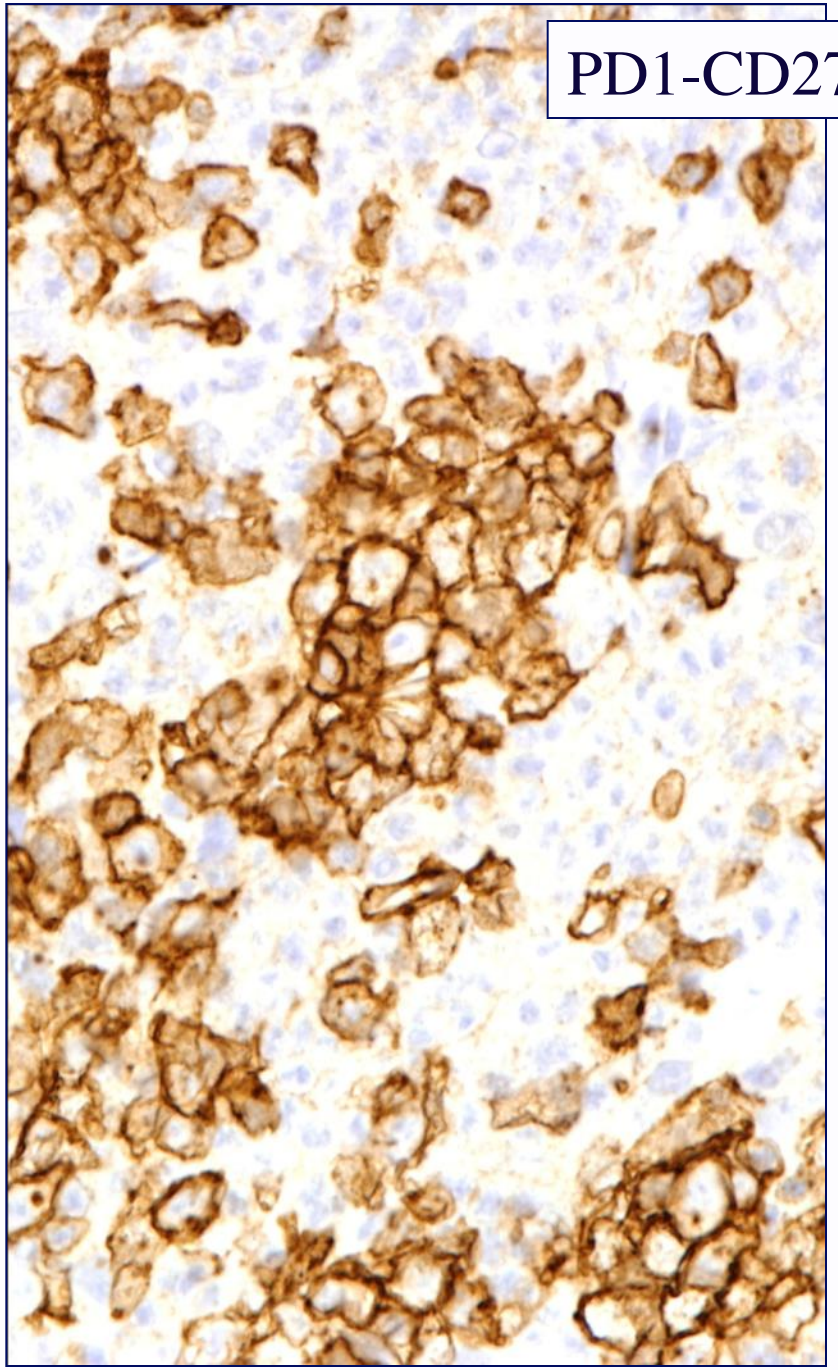
- CXCL 13 causes induction and proliferation of follicular dendritic cells
- CXCL 13 is involved in B-cell recruitment to LN's and activation of B-cells
  - CXCL13 is required for the adhesion and arrest of B-cells on HEV's
- Explains expansion of B-cells in a T-cell lymphoma
  - B-cells are both EBV+ and EBV-

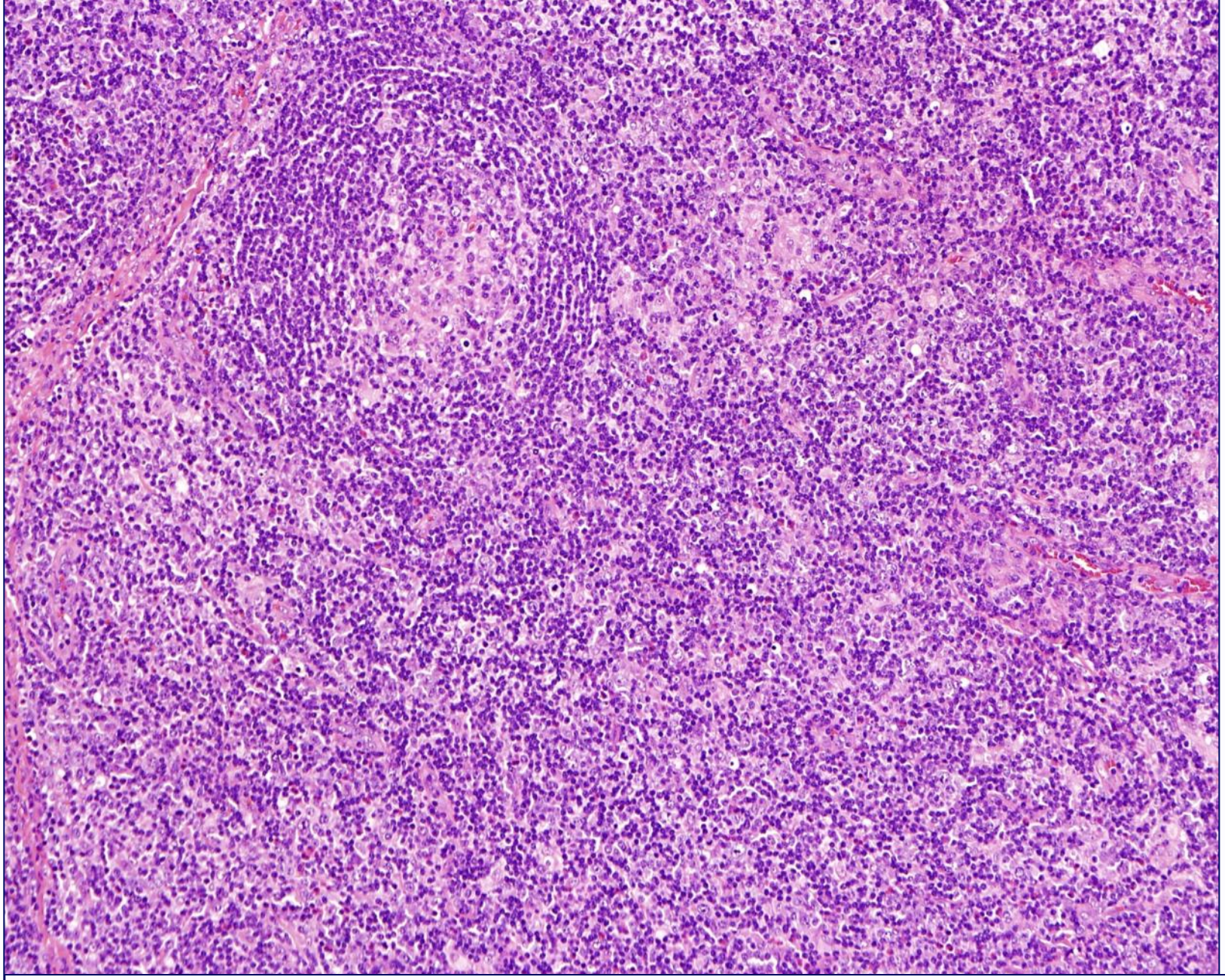

CD21

Perifollicular Localization of AITL T-cells

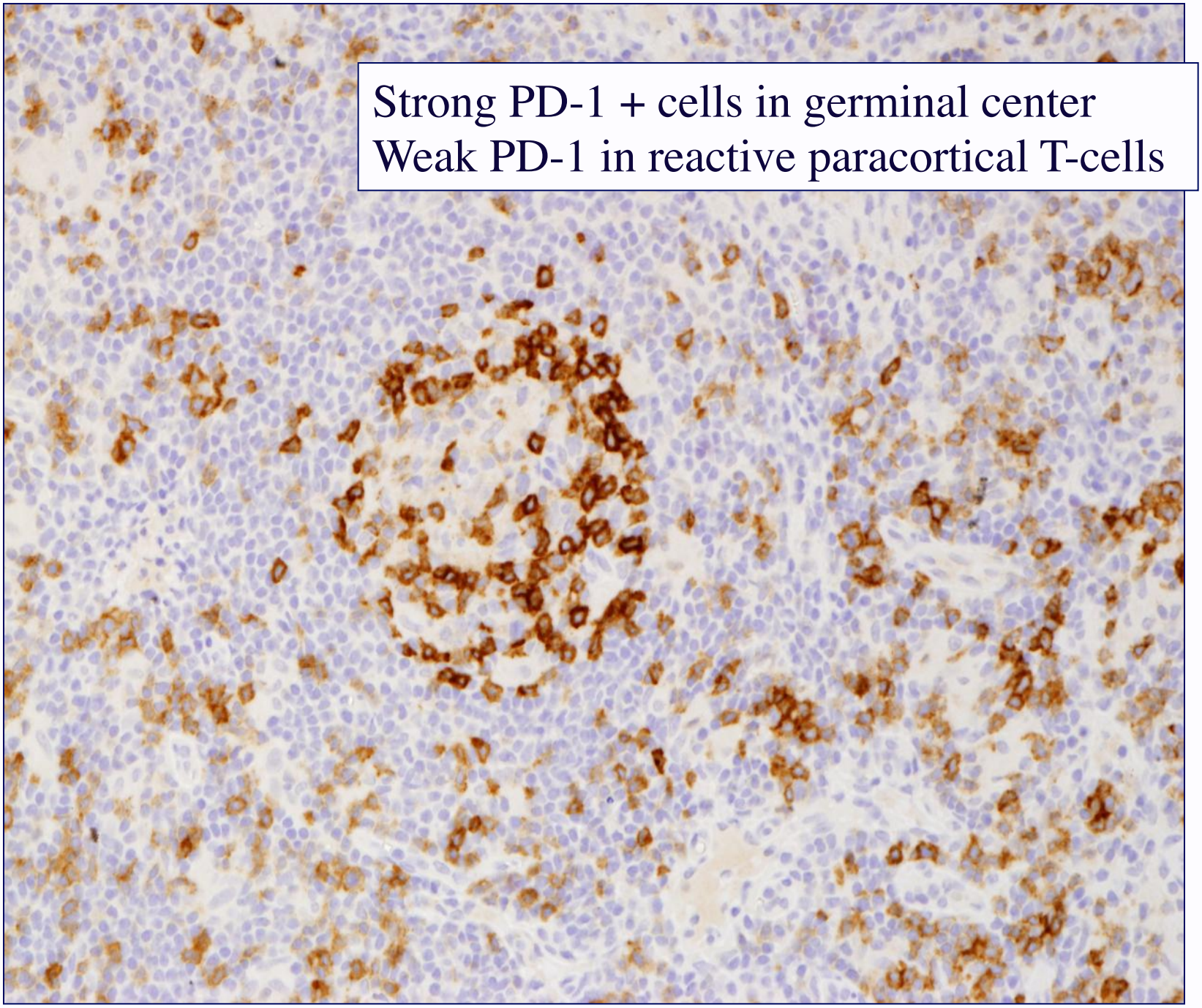





PD1-CD279



Reactive paracortical hyperplasia – 18 yo drug hypersensitivity


Strong PD-1 + cells in germinal center
Weak PD-1 in reactive paracortical T-cells