

Spleen

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Spleen 50-250 gm (median 150 gm)

Functions:

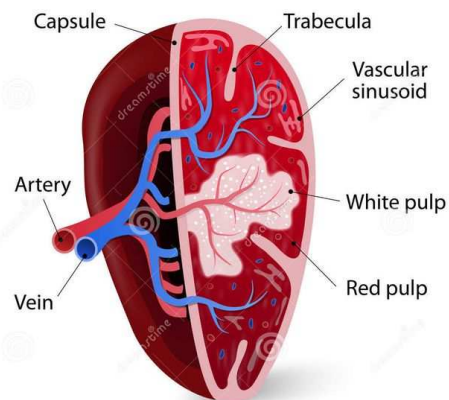
Filtration

Immunologic

Hematopoiesis

Reservoir

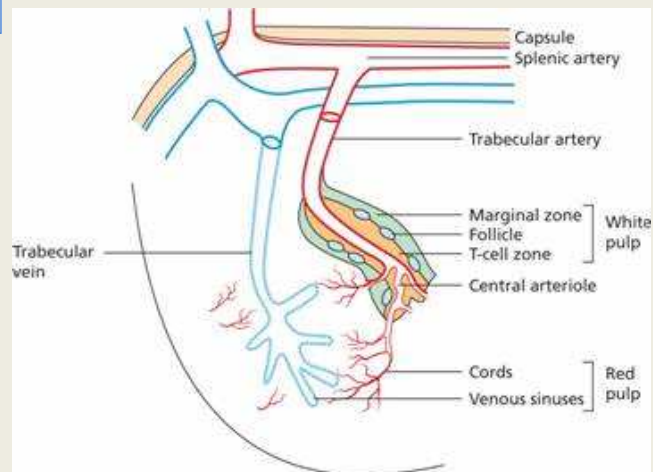
SPLEEN ANATOMY



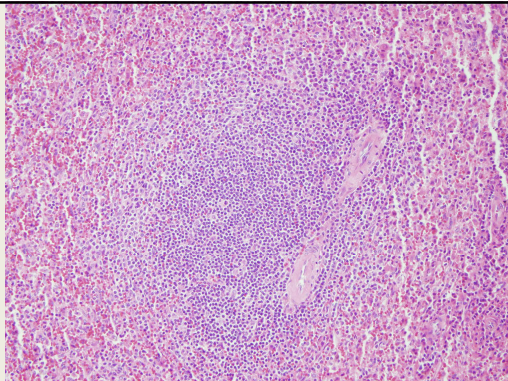
- White pulp
 - Central arteries
 - Lymphoid sheaths
 - Lymphoid follicles

- Stromal framework
 - Capsule
 - Trabecular arteries
 - Reticulin fiber network

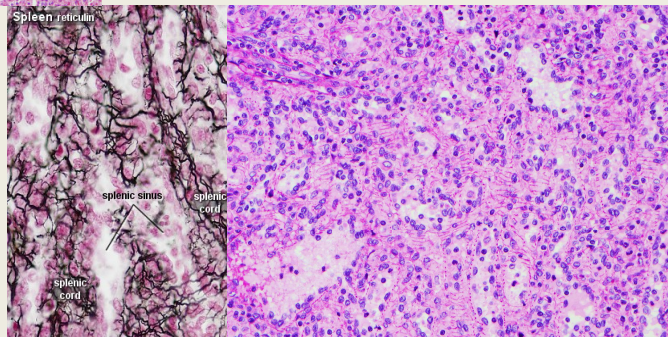
- Red pulp (75%)
 - Sinuses
 - Cords

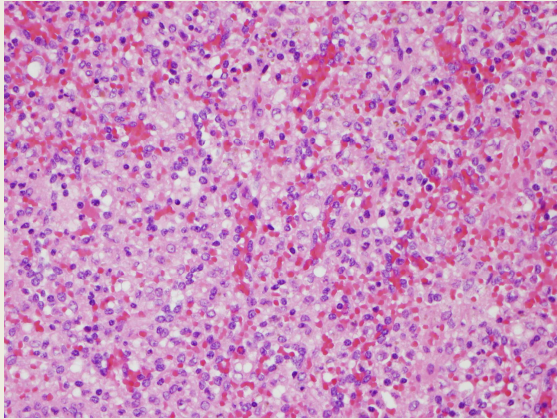


- Red & white pulp

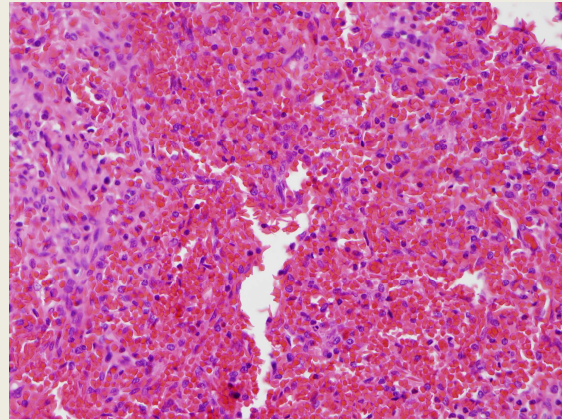


- Reticular (Ringed) fibers



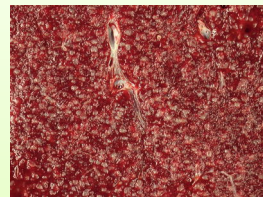


- Hereditary spherocytosis
- Sickle cell



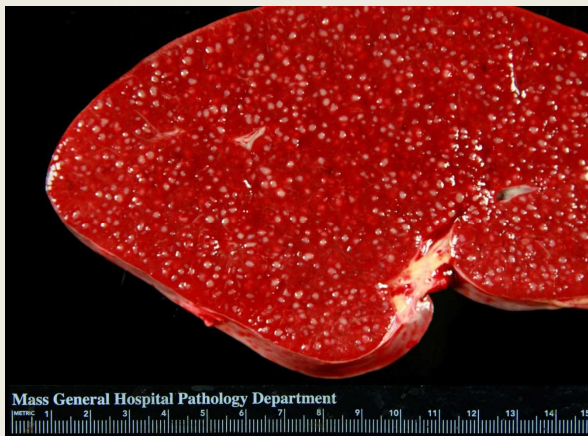
Patterns of Involvement

- Diffuse
 - White pulpmiliary appearance
 - Small B-cell lymphomas
 - Reactive lymphoid hyperplasia
 - Red pulp
 - Select B- & T-cell lymphomas
 - Leukemic infiltrates
 - Histiocytosis
- Focal / Nodular
 - Hematopoietic and non-hematopoietic



- Diffuse-white pulp involvement
 - Small B-cell lymphomas
 - Primary splenic marginal zone lymphoma
 - Involvement by LPL, FL, MCL, CLL
 - * Reactive hyperplasia (EBV)

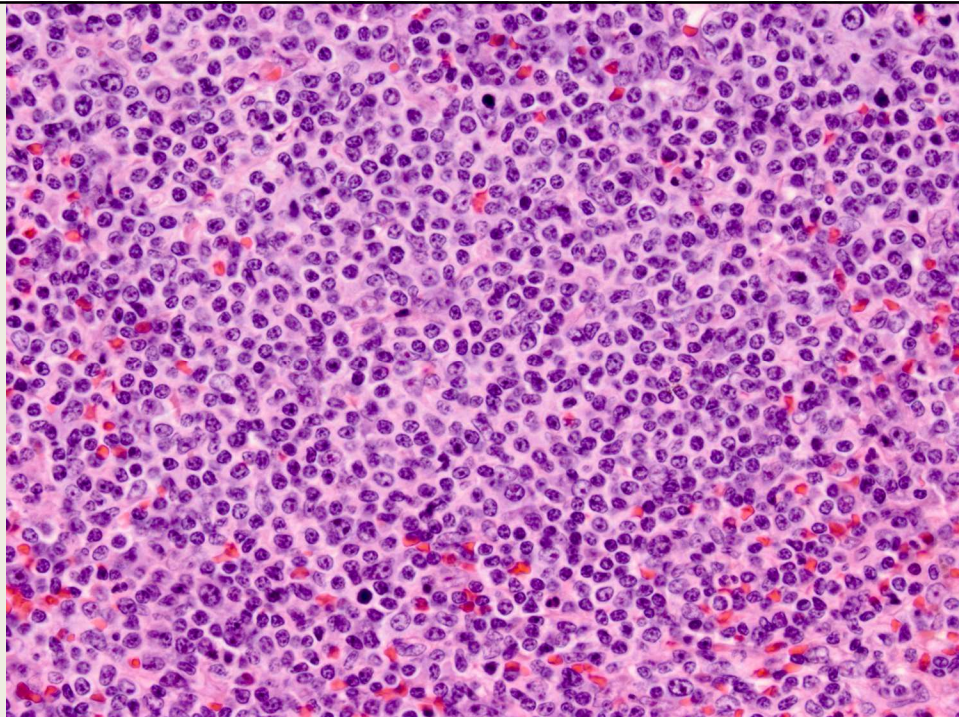
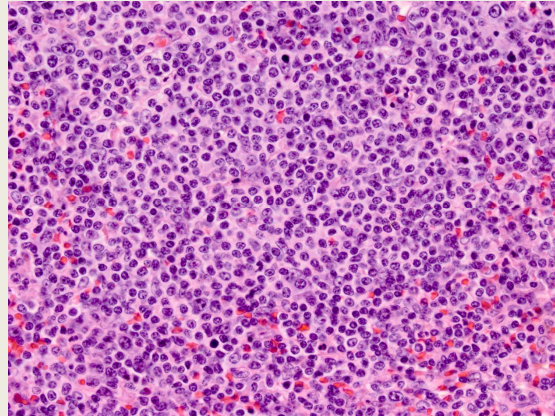
Splenic Marginal Zone Lymphoma

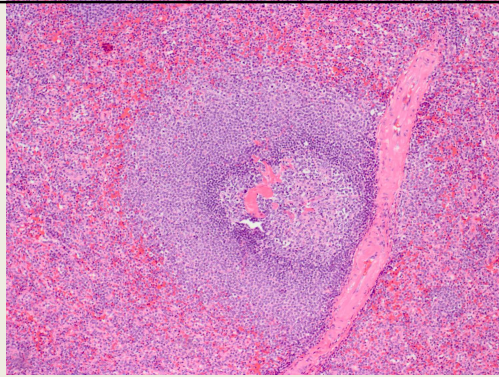


- Associated with an enlarged spleen and enlarged splenic hilar lymph nodes
- Diffuse, prominent, discrete white nodules in a miliary pattern
 - Expanded white pulp

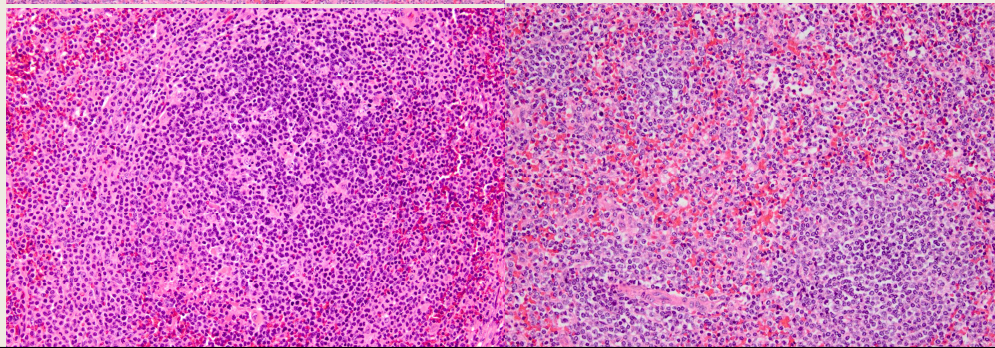
Splenic marginal zone lymphoma

- Prominent splenomegaly
- Spleen white pulp, bone marrow, liver and blood
- No peripheral adenopathy
- Indolent lymphoma
- B-cell markers , IgM, IgD
- Negative CD5, CD10, BCL6, cyclin D1, CD43, DBA44
- Loss 7q21, trisomy 3
- Notch 2 mutations
- DDX: lymphoid hyperplasia and other small B-cell lymphomas

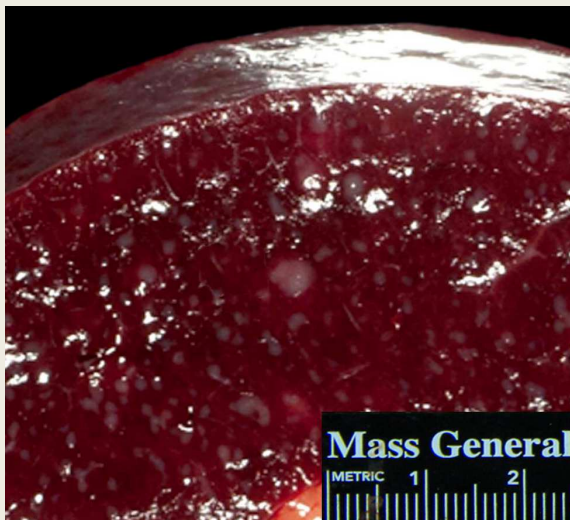




Biphasic
Disrupted and colonized germinal
centers
Monophasic



Follicular / marginal zone hyperplasia

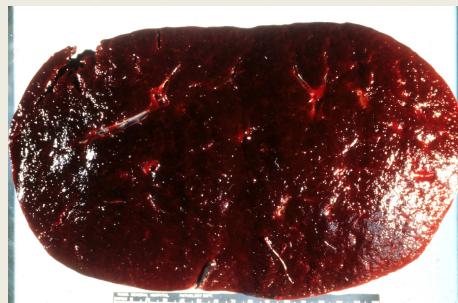


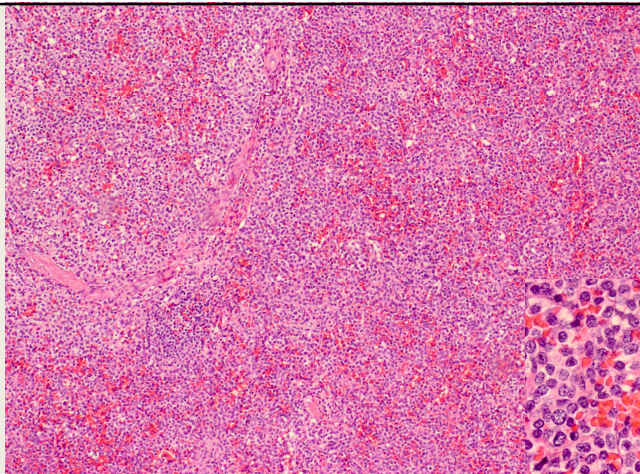
- Often found in enlarged spleens (>500g)
- Expansion of splenic white pulp (proliferation of B cells)
- Diffuse, white, gelatinous nodules in miliary pattern
- Focal aggregation can mimic lymphoma

- **Diffuse-red pulp involvement**
 - **Hairy cell leukemia**
 - **Diffuse red pulp small B-cell lymphoma**
 - **Hepatosplenic T-cell lymphoma**
 - **Histiocytosis**
 - **Extramedullary hematopoiesis**
 - **Leukemia**

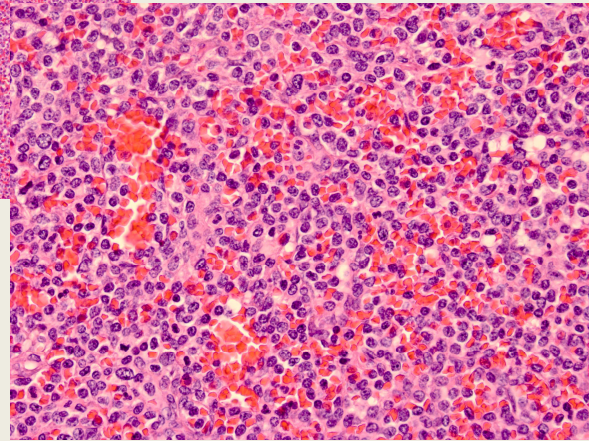
Hairy cell leukemia

- Prominent splenomegaly & pancytopenia
- Spleen red pulp, bone marrow, blood
- No follicles or white pulp involvement
- No peripheral adenopathy
- Responsive to interferon alpha and nucleosides
- B-cell markers , CD25, CD103, CD103, Annexin A1, cyclin D1, CD11c, CD123, DBA44
- Negative CD5, CD10
- BRAF V600E mutation (>95%)
- Splenic B-cell lymphoma with hairy cell features (new name for hairy cell leukemia variant).





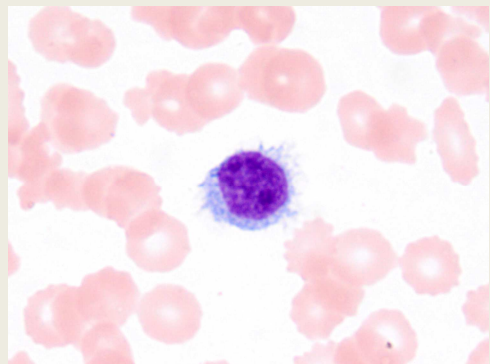
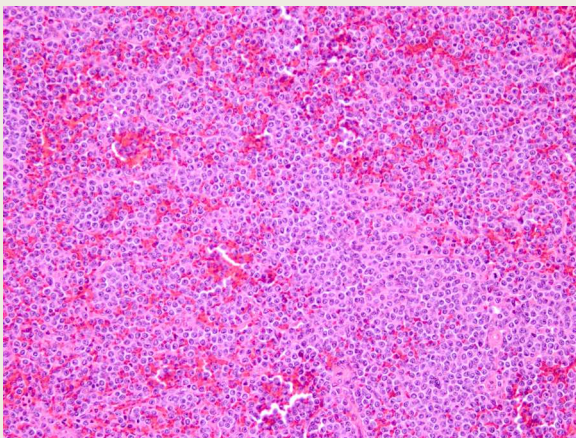
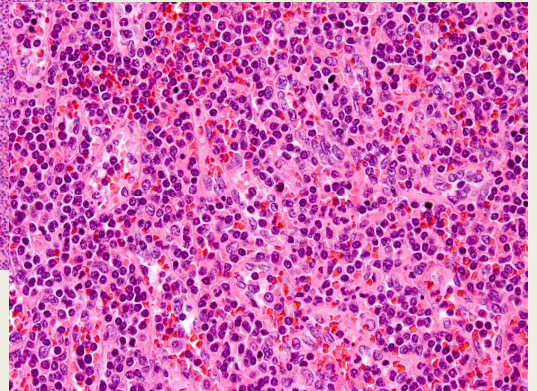
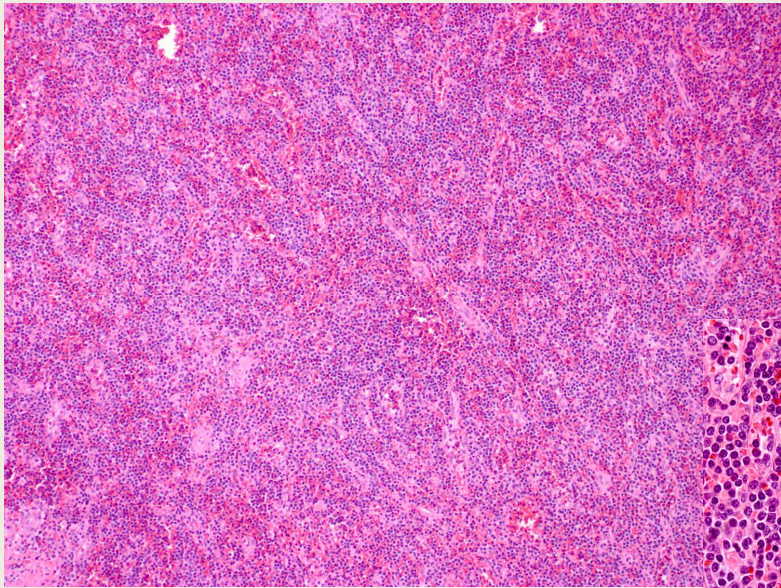
Atrophic white pulp
Cords filled
Red blood cell lakes
Widely spaced cells
oval or indented nuclei



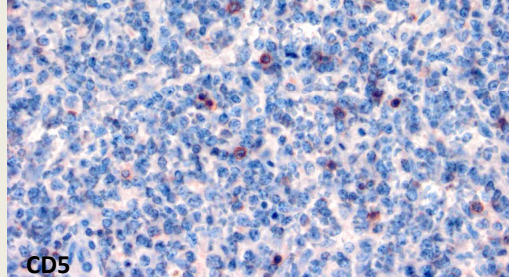
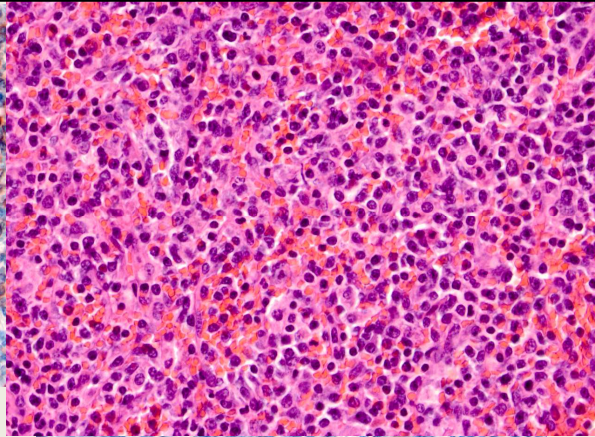
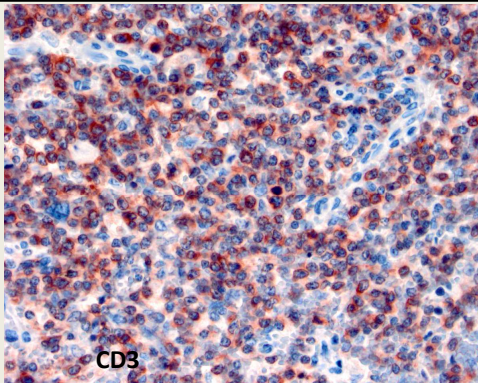
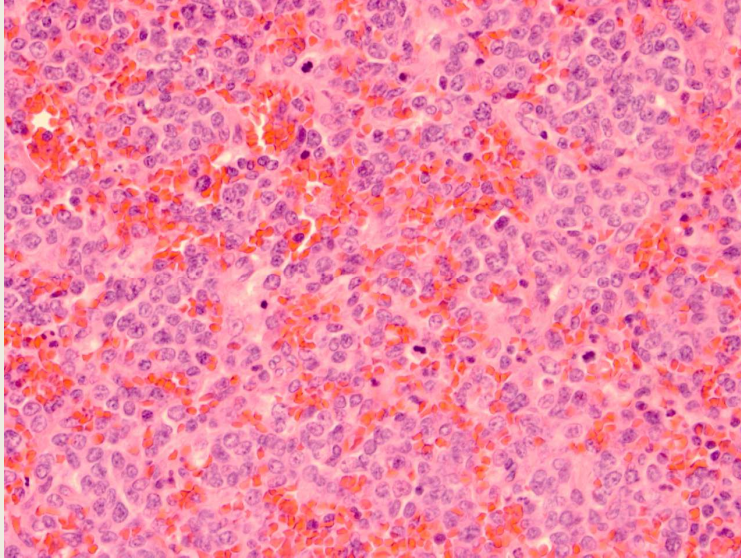
Diffuse red pulp small B-cell lymphoma

- Prominent splenomegaly
- Spleen red pulp, bone marrow sinusoids, blood
- No follicles or white pulp involvement
- No peripheral adenopathy
- Indolent but incurable lymphoma
- B-cell markers , DBA44, IgG, IgD-/+
- Negative CD5, CD10, BCL6, cyclin D1, CD43, CD25, CD103
- Complex cytogenetics involving PAX5 & IGH
- Cyclin D3 abnormalities
- DDX: Other small B-cell lymphomas

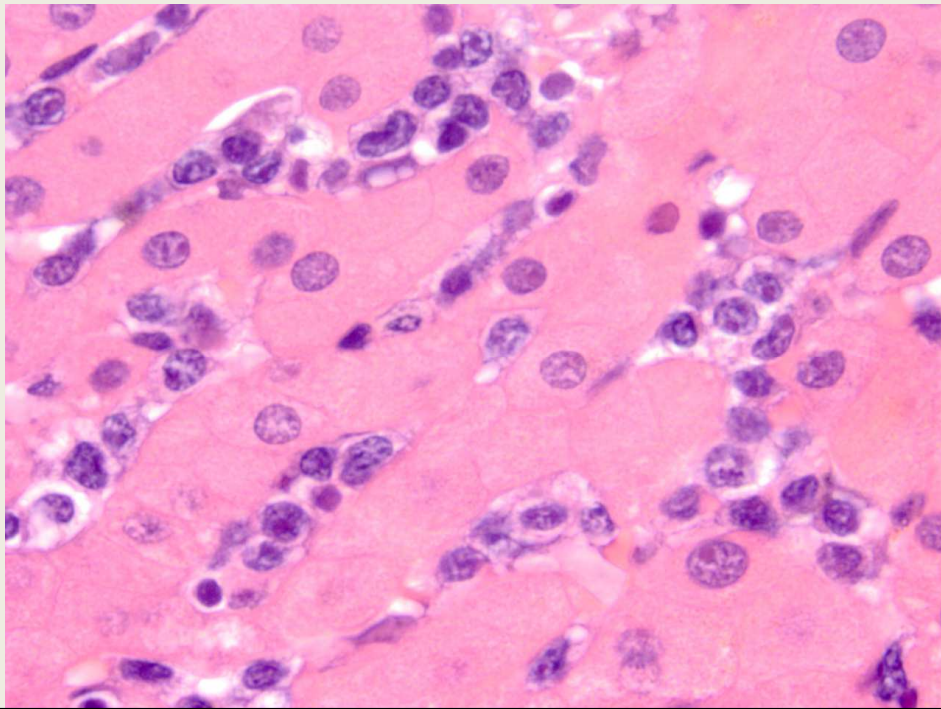




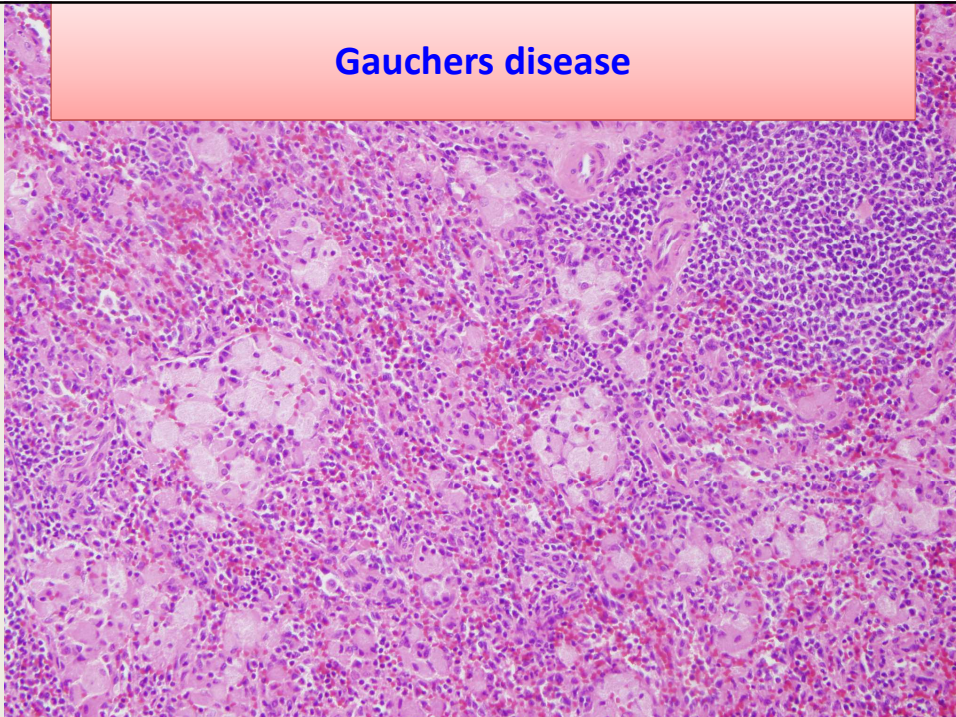
Hepatosplenic T-cell lymphoma



Aggressive extranodal lymphoma usually of cytotoxic T-cells of Gamma-Delta type
Marked splenomegaly with diffuse red pulp disease
Monotonous cells with medium sized nuclei with atypia at close examination

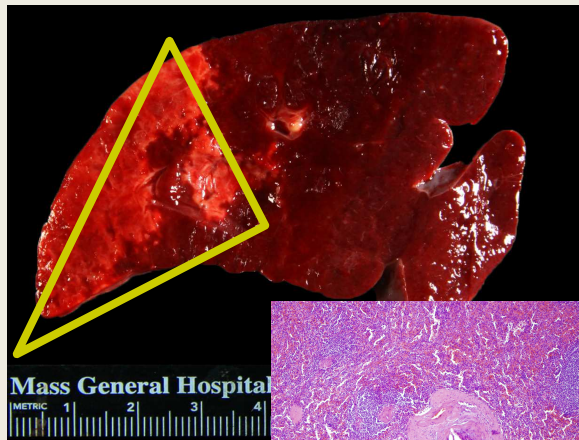


Gauchers disease

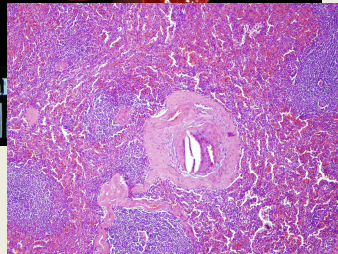


- Focal splenic nodules / involvement
 - Diffuse large B-cell lymphoma
 - Hodgkin lymphoma
 - Splenic infarct
 - Splenic hamartoma
 - Hemangioma
 - Angiosarcoma
 - SANT
 - Dendritic cell tumors
 - Metastases

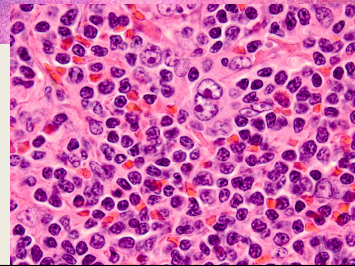
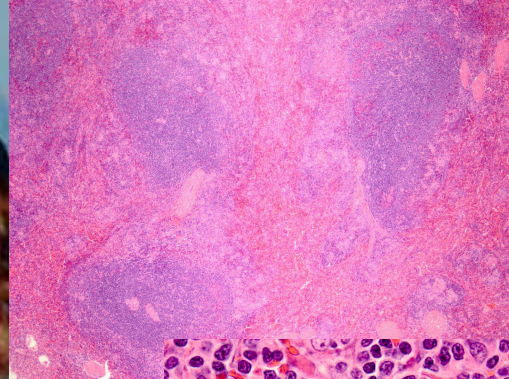
Splenic Infarct



- Wedge-shaped lesion corresponding to vascular distribution
 - Capsule = base
 - Apex = artery
- Well-circumscribed
- Initially pale (necrosis, inflammation)
- Hyperemic rim (necrosis, macrophages)
- Tan-firm (fibroblast repair and scar)



Hodgkin lymphoma



Early lesions in PALS and marginal zones
Later lesions in red pulp and follicles
Granulomas may be clue

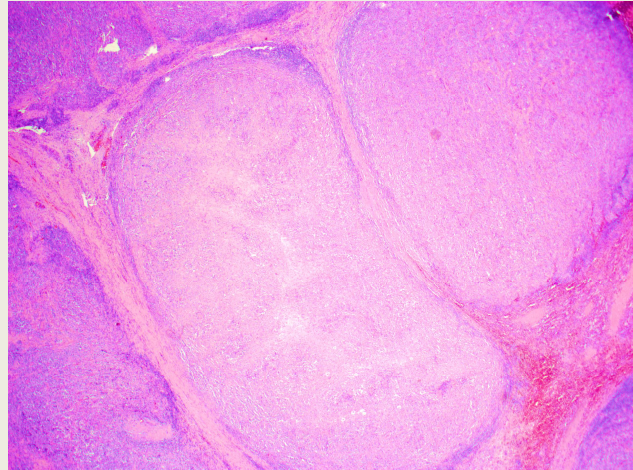
Diffuse Large B-Cell Lymphoma



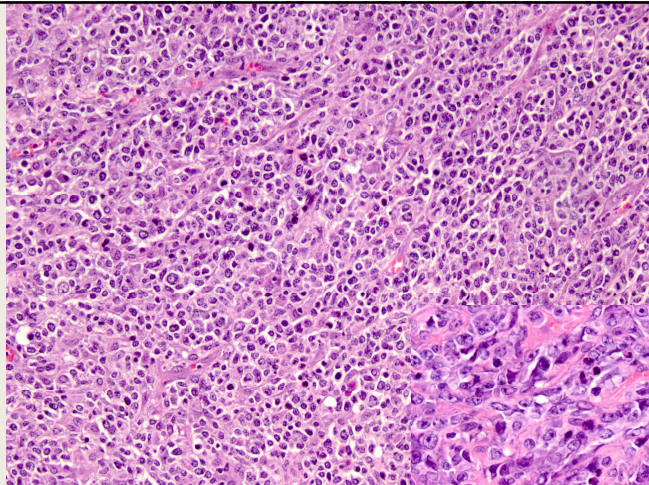
- Generally associated with splenomegaly and lymphadenopathy
- Distinct, solitary mass
- Can have multiple distinct masses
- Background of non-neoplastic spleen
- Well-circumscribed, nodular with tan-white to fleshy cut surfaces
- Central necrosis and hemorrhage

DLBCL

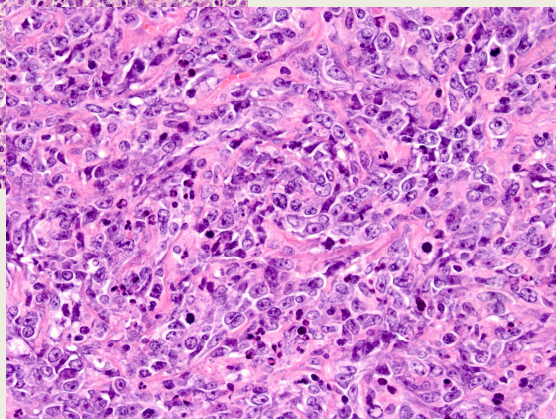
- Macronodules (favorable outcome)
- Micronodules [TCHRLBCL] (poor outcome)
- Diffuse red pulp involvement (poor outcome)

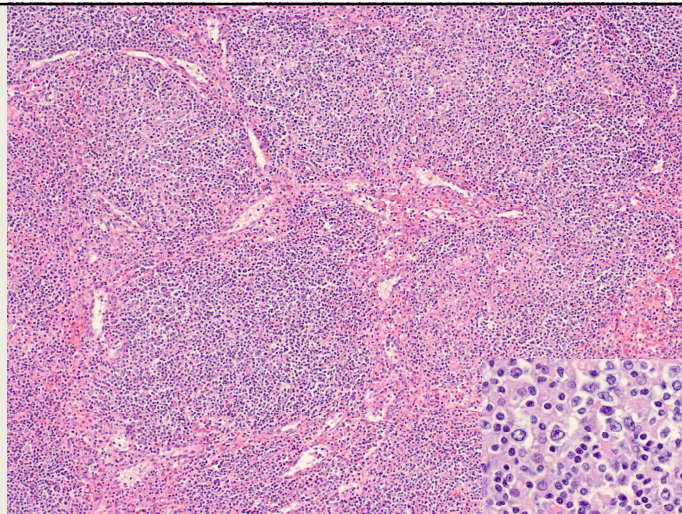


DLBCL

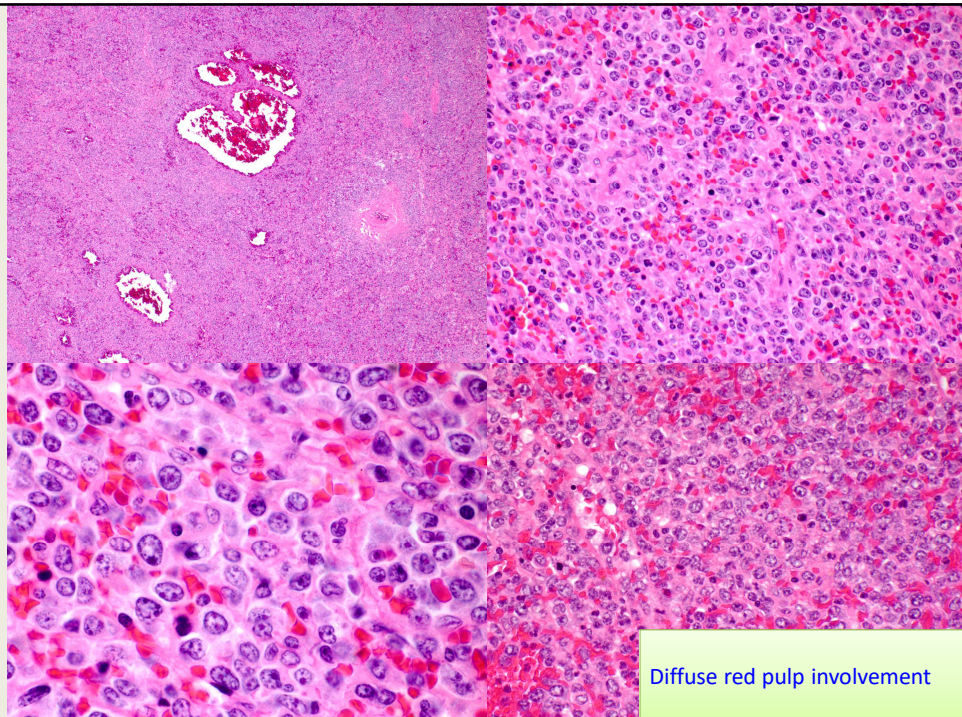
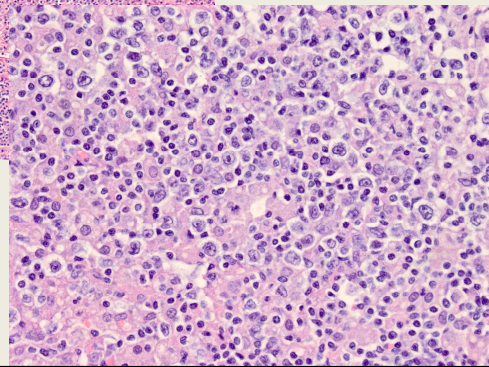


macronodules





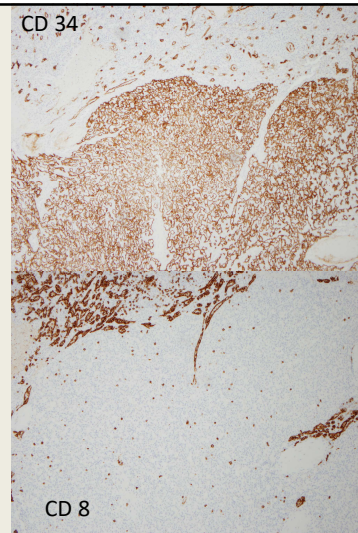
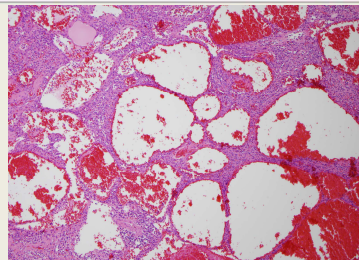
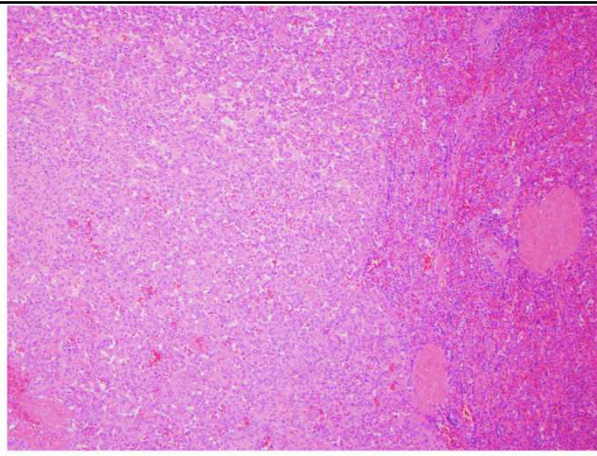
micronodular



Diffuse red pulp involvement

Splenic hemangioma

- Single mass
- Well-demarcated from surrounding spleen
- Cystic cut surfaces
- CD31+ CD34+ CD8-

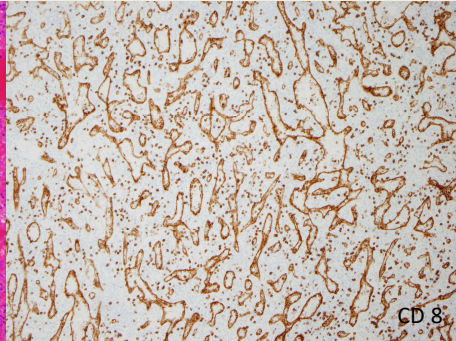
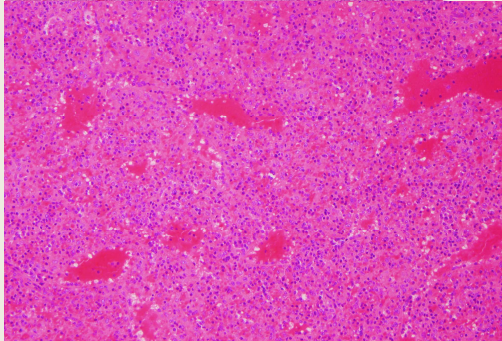
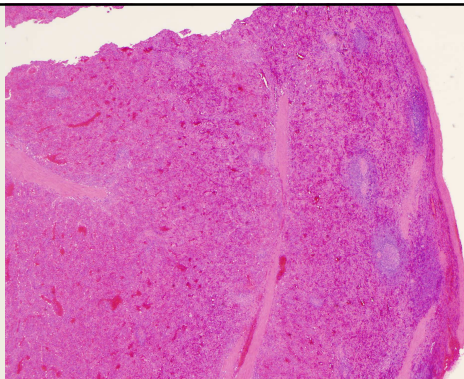


Splenic hamartoma

- Single mass
- Well-demarcated from surrounding spleen grossly
- Red cut surfaces
- CD8 staining

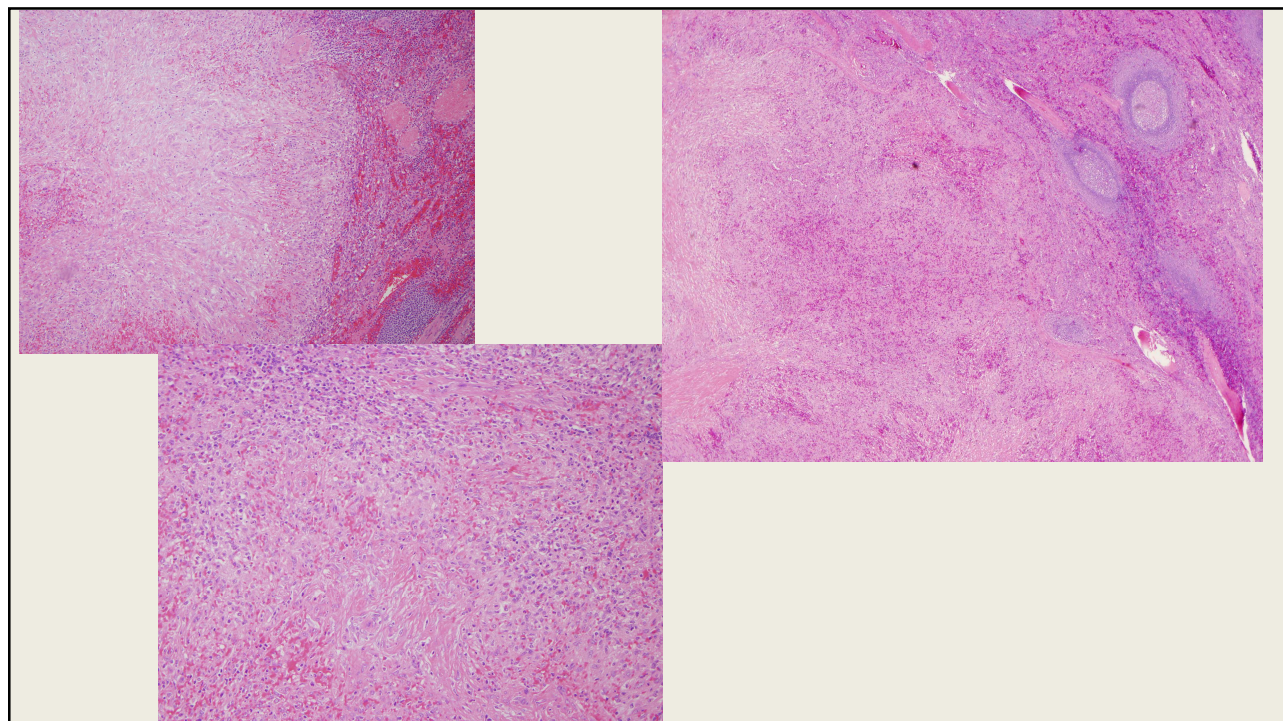


Splenic hamartoma

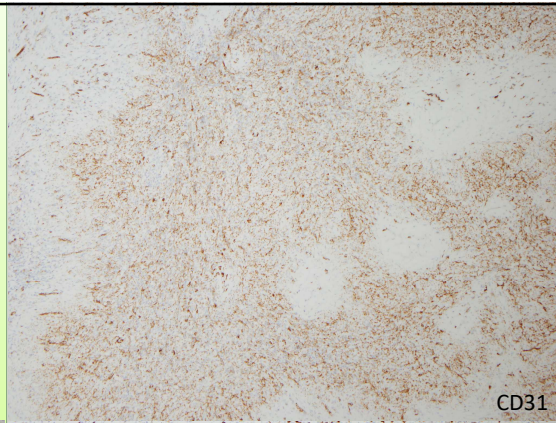


Sclerosing angiomatoid nodular transformation (SANT)

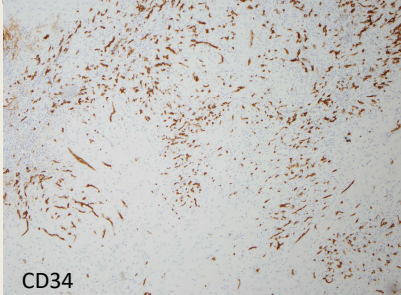
- Single central mass
 - Stellate sclerosis
- Well-demarcated from surrounding spleen
- Tan-white cut surfaces with areas of entrapped red pulp
- Peripheral nodularity



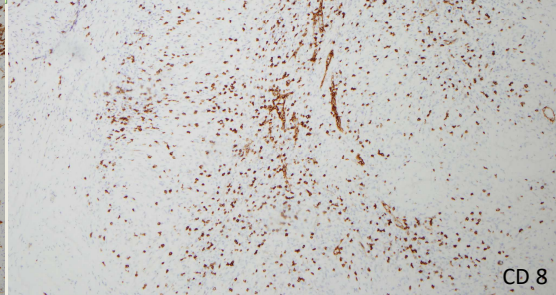
Female predominance
Multinodular lesion surrounded
by inflamed collagen sclerosis
Nodules contain sinusoids, and
vessels
Sinusoids (CD31+ CD34- CD8+)
Small arteries (CD31+ CD34+
CD8-)
Small veins (CD31+ CD34- CD8-)
CD68 in macrophages
Benign
Unclear what lesion is



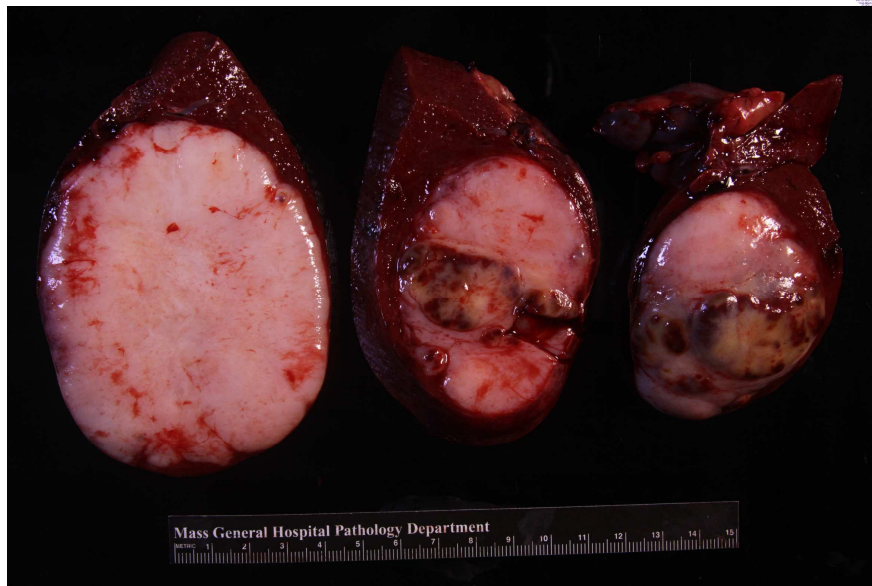
CD31

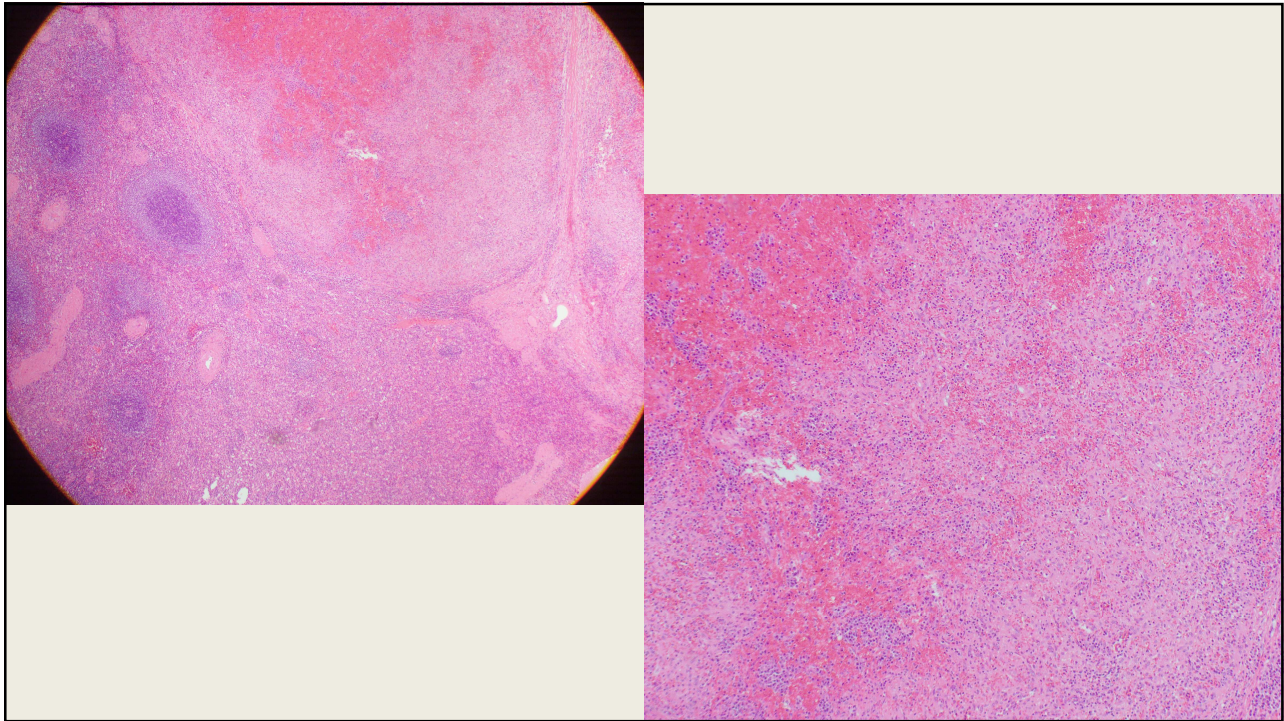


CD34



CD 8

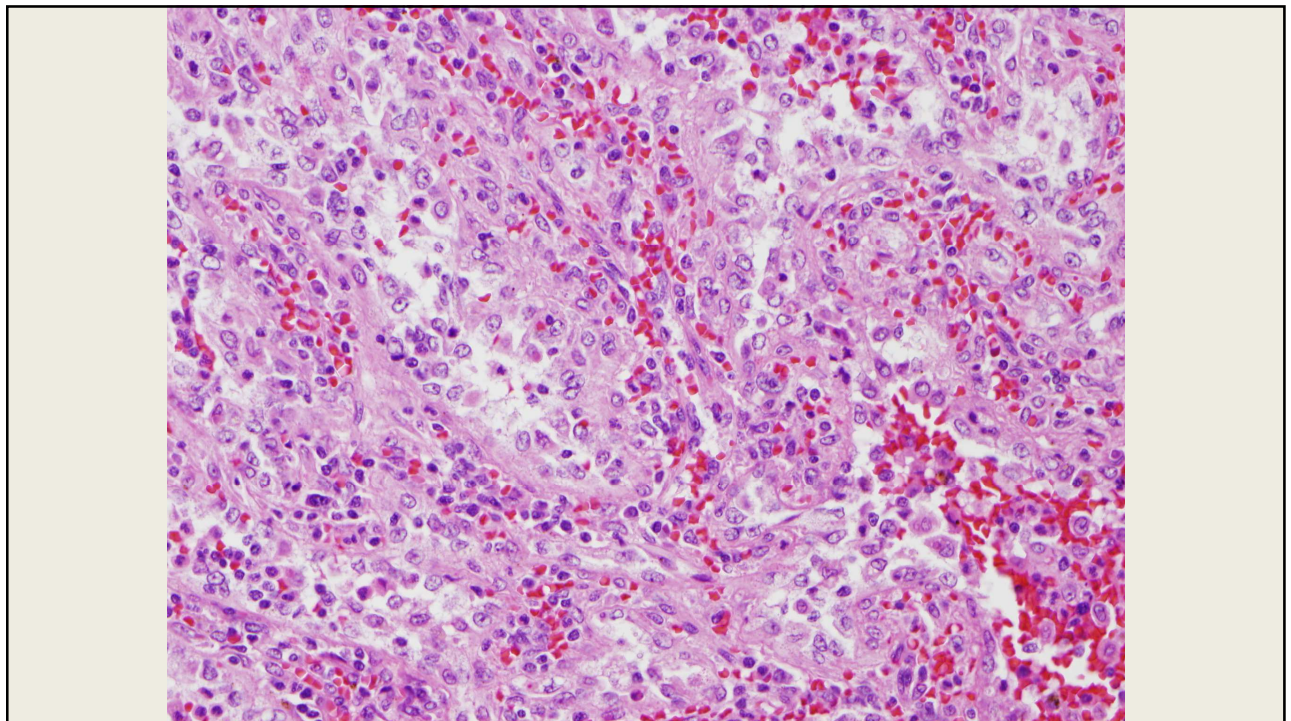
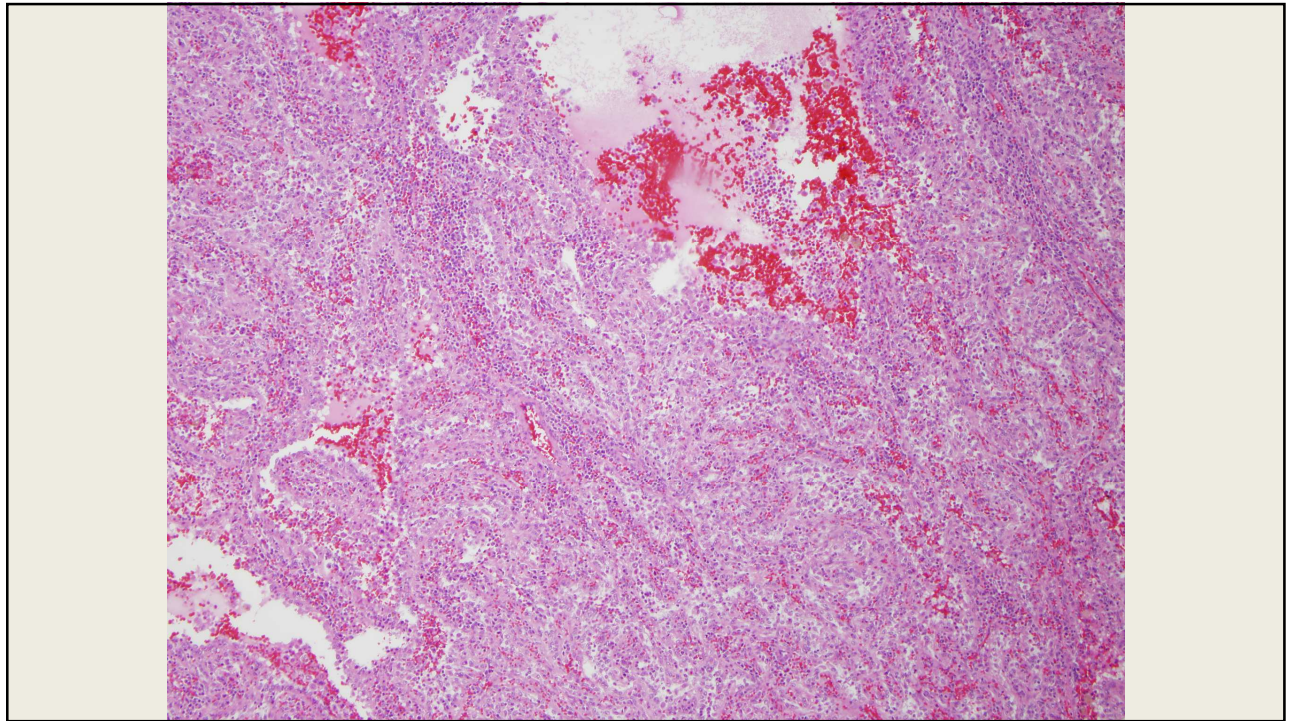


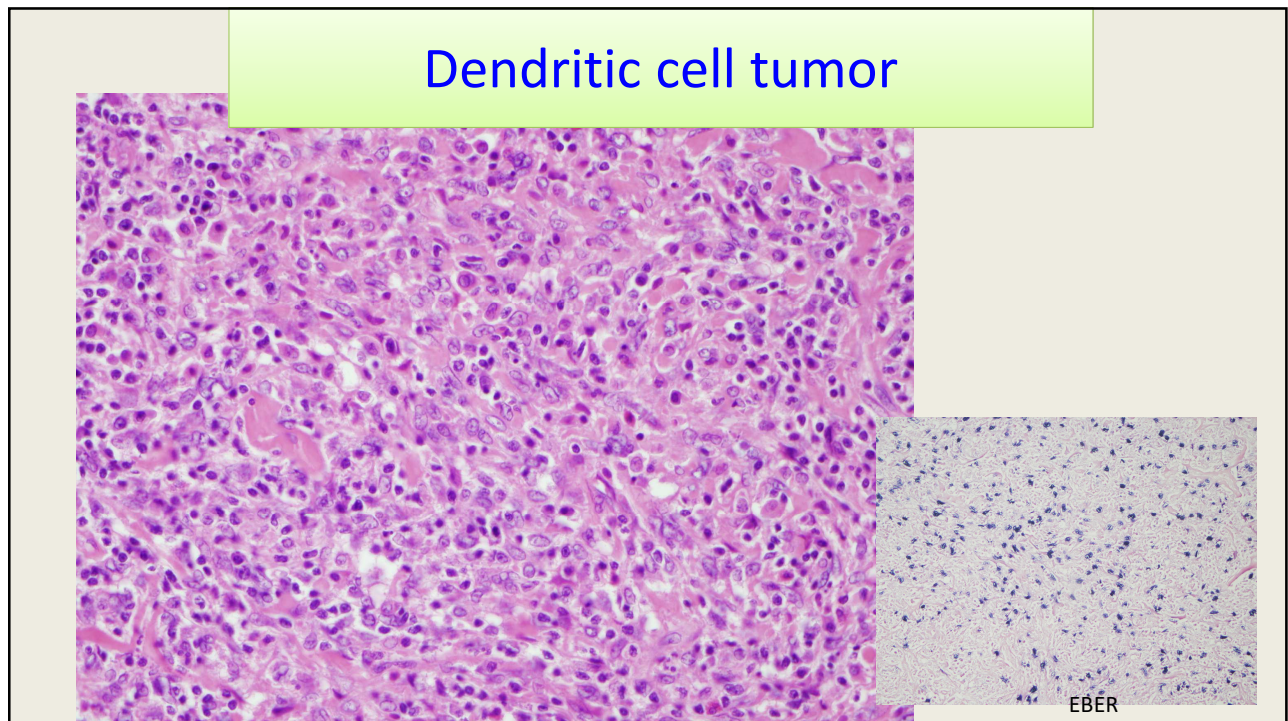
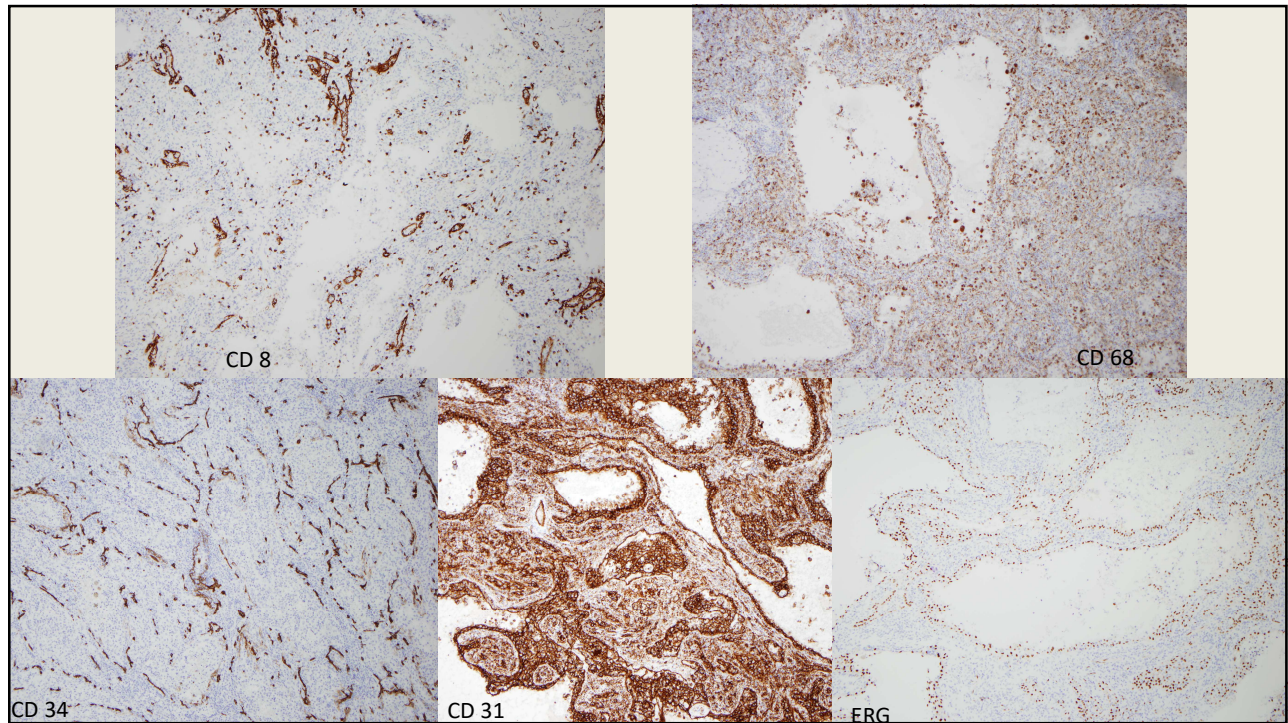


Littoral cell angioma

- Vascular tumor derived littoral cells of splenic sinuses
- Anastomosing channels with cystic spaces and papillary projections
- Tall endothelial cells
- Often hemophagocytosis
- Endothelial differentiation (CD31+ Factor VIII + CD34-/+)
 - Histiocytic differentiation (CD68+ Lysozyme+)
- CD8-
- Usually benign







Spleen is fascinating and involved by a large number of primary and secondary neoplasms, especially lymphomas and vascular tumors, many of which are unique to the spleen.

Immunostains, flow cytometry, and molecular studies helpful in separating B-cell lymphomas.

Most vascular lesions are benign.

Questions email: lzukerberg@partners.org