Nodal Marginal Zone Lymphoma

Nodal Marginal Zone Lymphoma

- Definition: NMZL is a primary nodal B-cell neoplasm that morphologically resembles lymph nodes involved by marginal zone lymphomas of extranodal or splenic disease. Monocytoid B-cells may be prominent.
- Epidemiology: nodal marginal zone lymphoma is a rare disease, comprising only 1.8% of lymphoid neoplasms.
- Sites of involvement: peripheral lymph nodes, occasionally bone marrow and peripheral blood.
- Clinical features: patients may present with localized or generalized peripheral lymphadenopathy, with good performance status.

Nodal Marginal Zone Lymphoma

- Morphology: the marginal zone and interfollicular areas of the lymph node are infiltrated by marginal zone (centrocyte-like) B-cells, monocytoid B-cells, or small B lymphocytes, with scattered centroblast-like cells and immunoblast-like cells.
- Two types described:

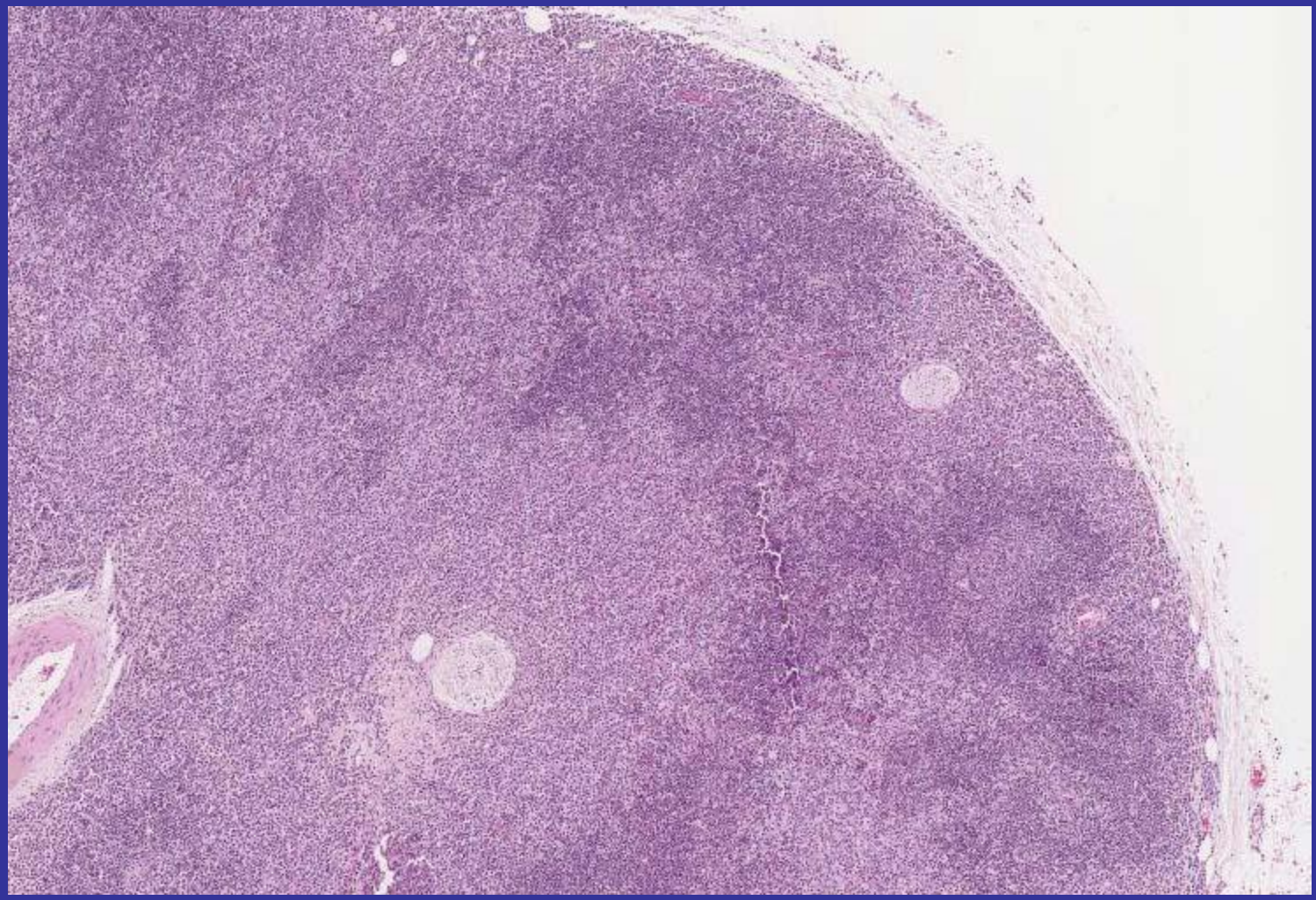
- One that closely resembles nodal involvement by MALT lymphoma

- One that resembles splenic marginal zone lymphoma.

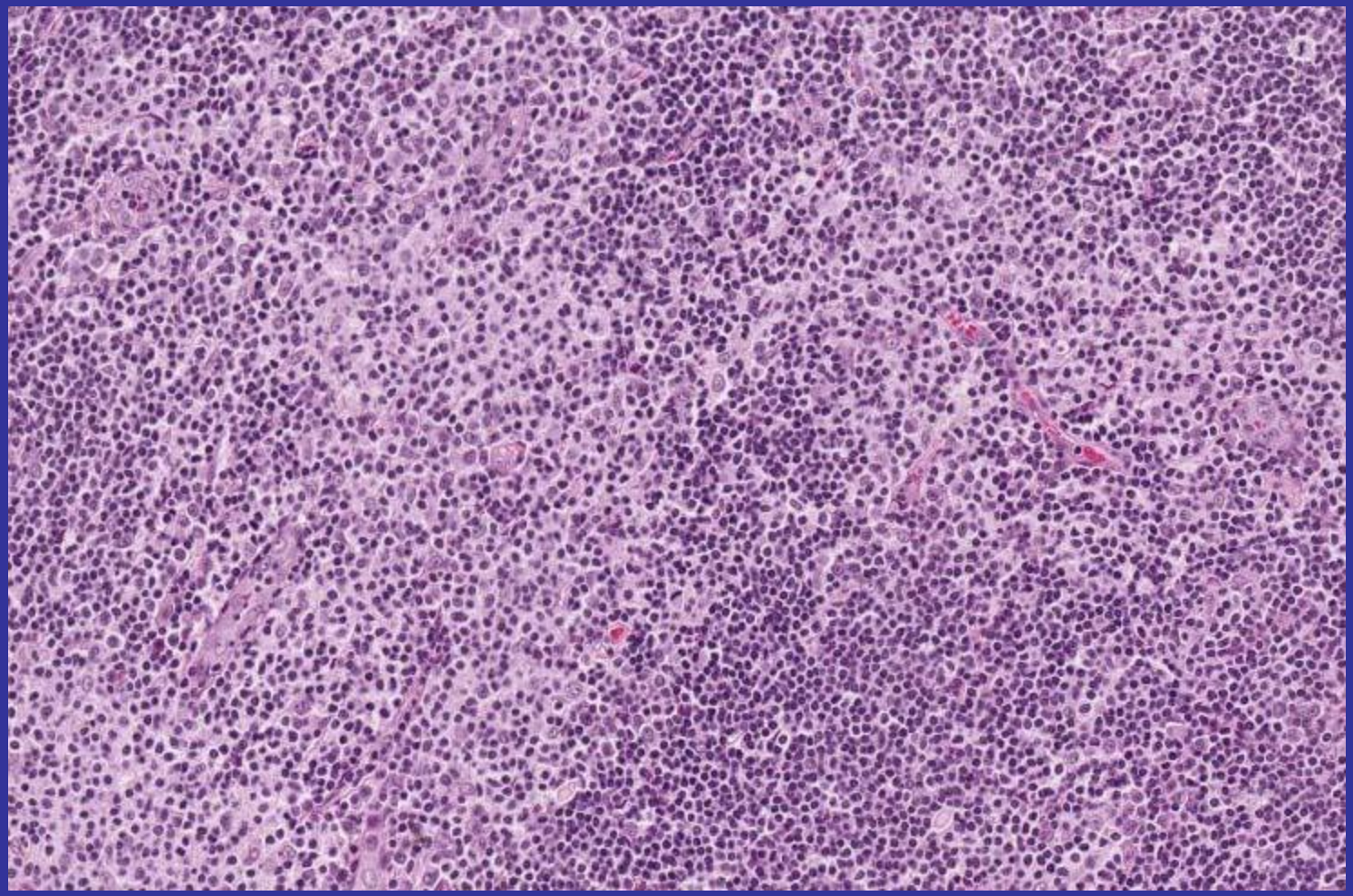
Nodal Marginal Zone Lymphoma

- Plasma cell differentiation is a feature of some cases.
- Follicular colonization may be present.
- Transformation to large B-cell lymphoma may occur.
- In patients with extranodal (MALT) lymphoma, Hashimoto thyroiditis or Sjögren syndrome, nodal involvement by marginal zone lymphoma should be considered secondary involvement by MALT lymphoma.

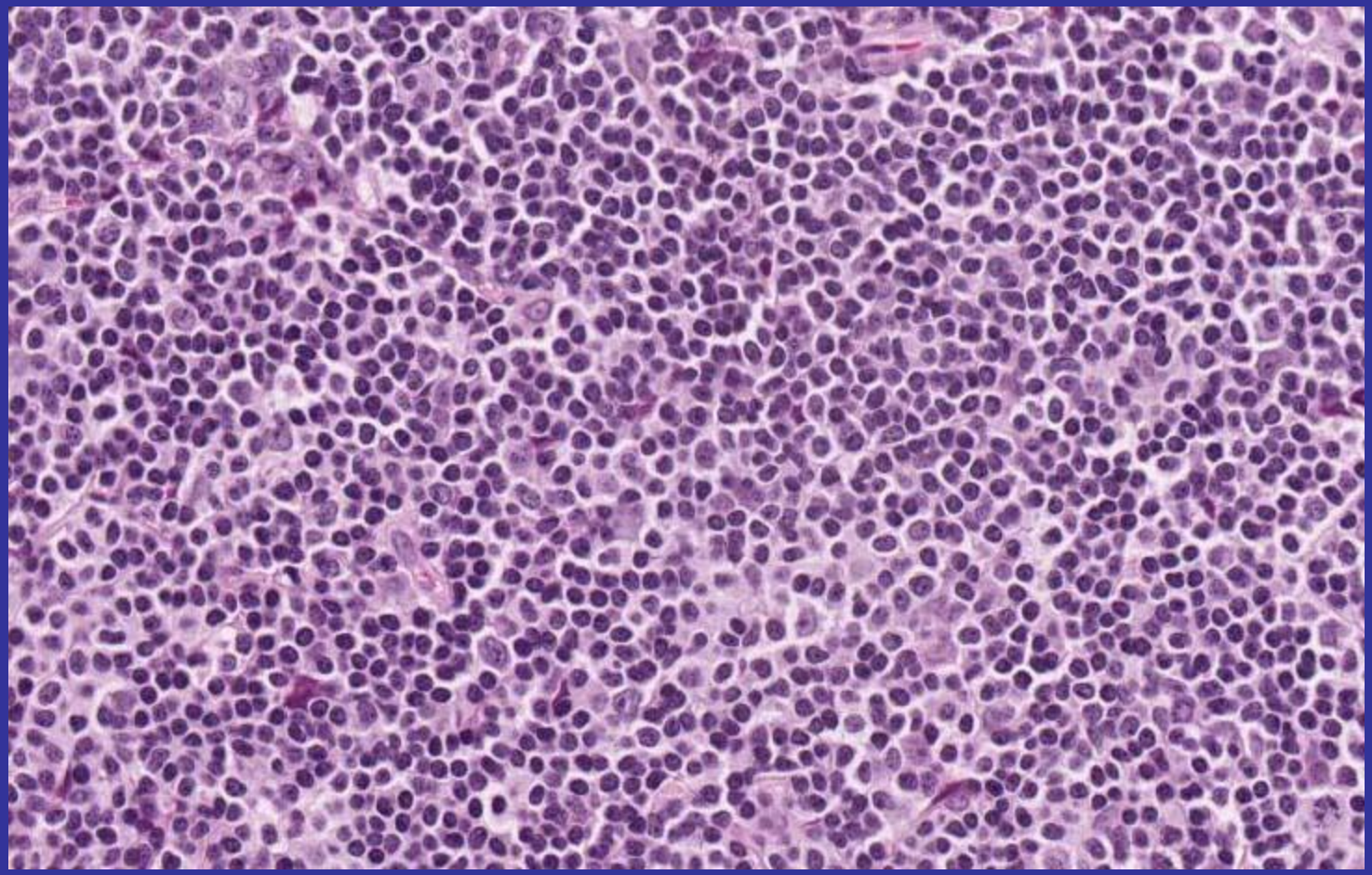
Nodal Marginal Zone Lymphoma



Nodal Marginal Zone Lymphoma



Nodal Marginal Zone Lymphoma



Nodal Marginal Zone Lymphoma

- Immunophenotype: similar to MALT lymphoma. Some are reported to be IgD+, CD43-, similar to splenic MZL.
- Genetics: not well studied. However, t(11;18)(q21;q21) and trisomy 3 associated with extranodal marginal zone lymphoma are not frequent.

Nodal Marginal Zone Lymphoma

- Postulated cell of origin: marginal zone B-cell of nodal type.
- Prognosis: the clinical course has not been well studied. In two recent series, the majority of the patients responded to chemotherapy, but with a high early relapse rate; nonetheless, the median survival was approximately 5 years, consistent with an indolent lymphoma.

Follicular Lymphoma

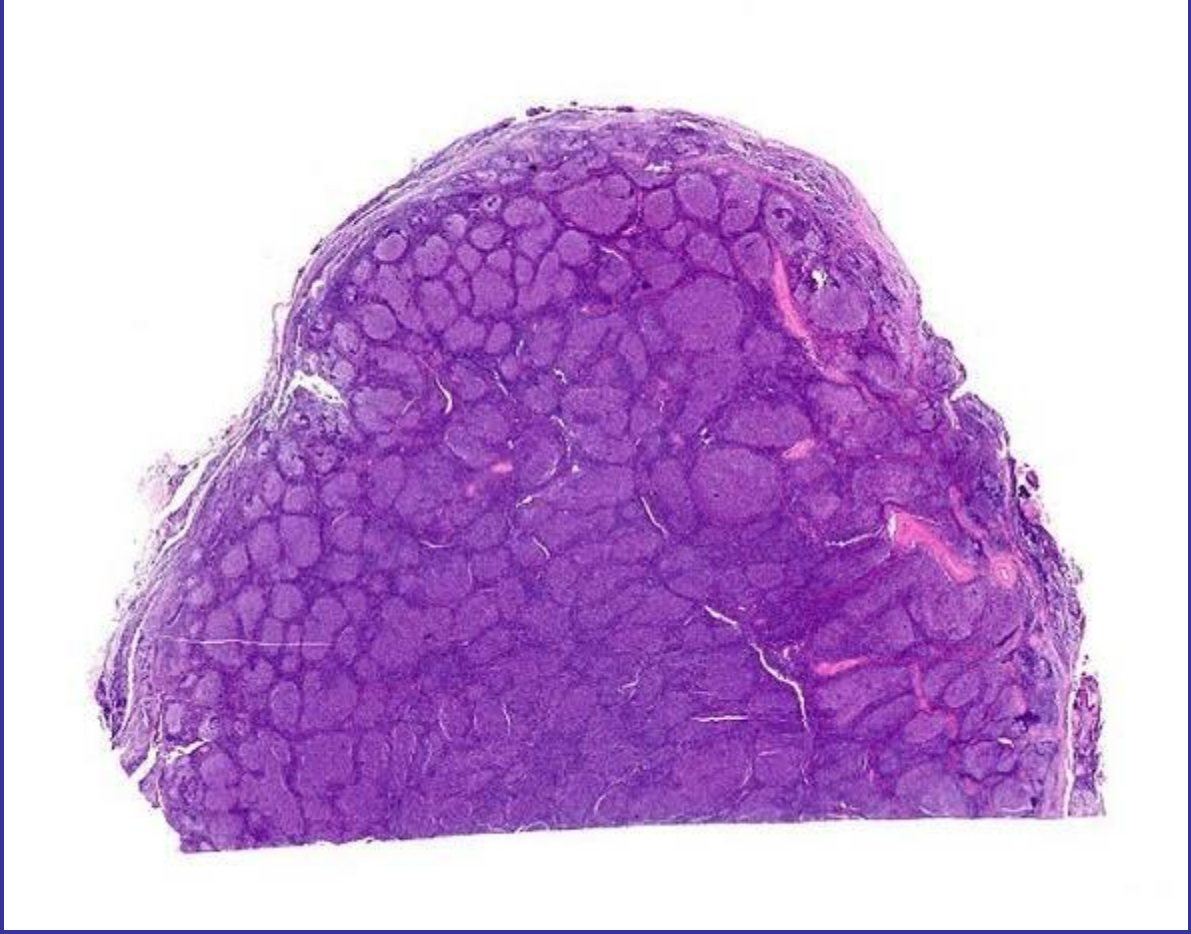
Follicular Lymphoma, Definition

- Neoplasm of follicular centre B cells, with at least a partially follicular pattern. The lymphoma cells consist of two types: centrocytes (cleaved follicle centre B cells), and centroblasts (non-cleaved follicle centre B cells)
- Predominantly adults
- 70% of low grade lymphomas
- Most patients have widespread disease at diagnosis (bone marrow involvement in 40-50%)
- Patients are usually asymptomatic at diagnosis, except for lymph node enlargement

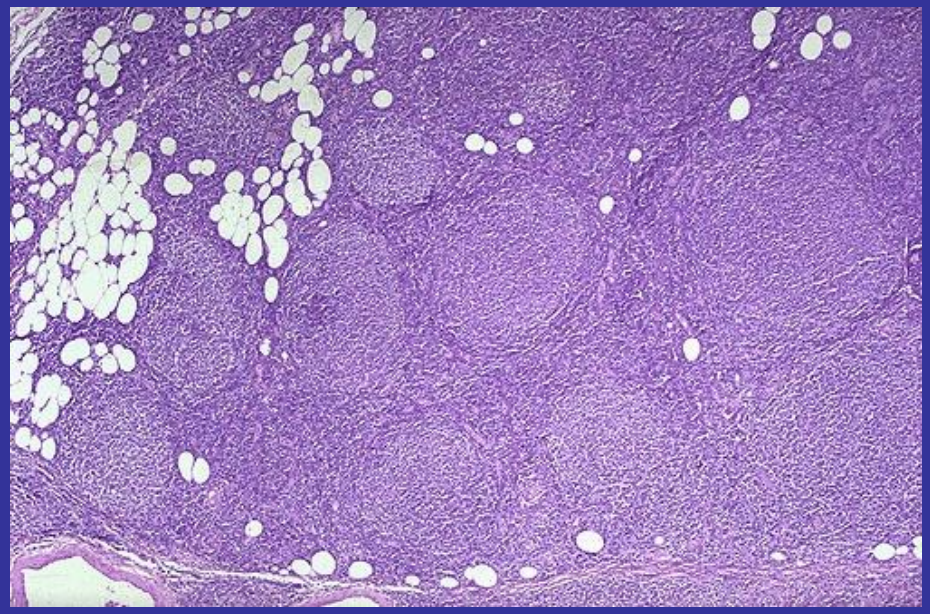
Follicular Lymphoma, Morphology

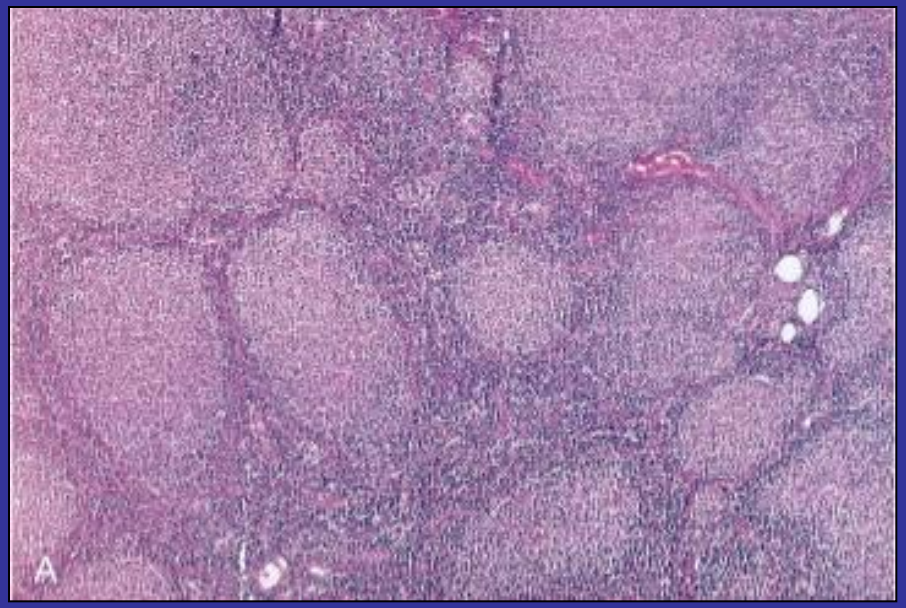
- Follicular architecture
- Neoplastic follicles are: poorly defined and closely packed, no mantle zone, no polarization, no tingible-body macrophages

Follicular Lymphoma



Follicular Lymphoma





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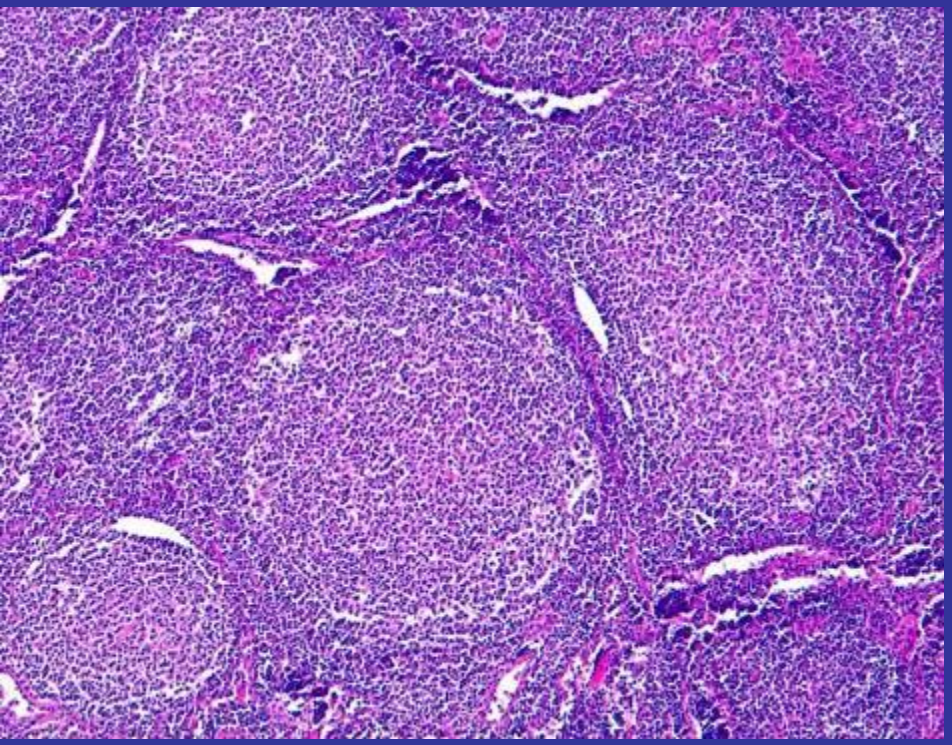
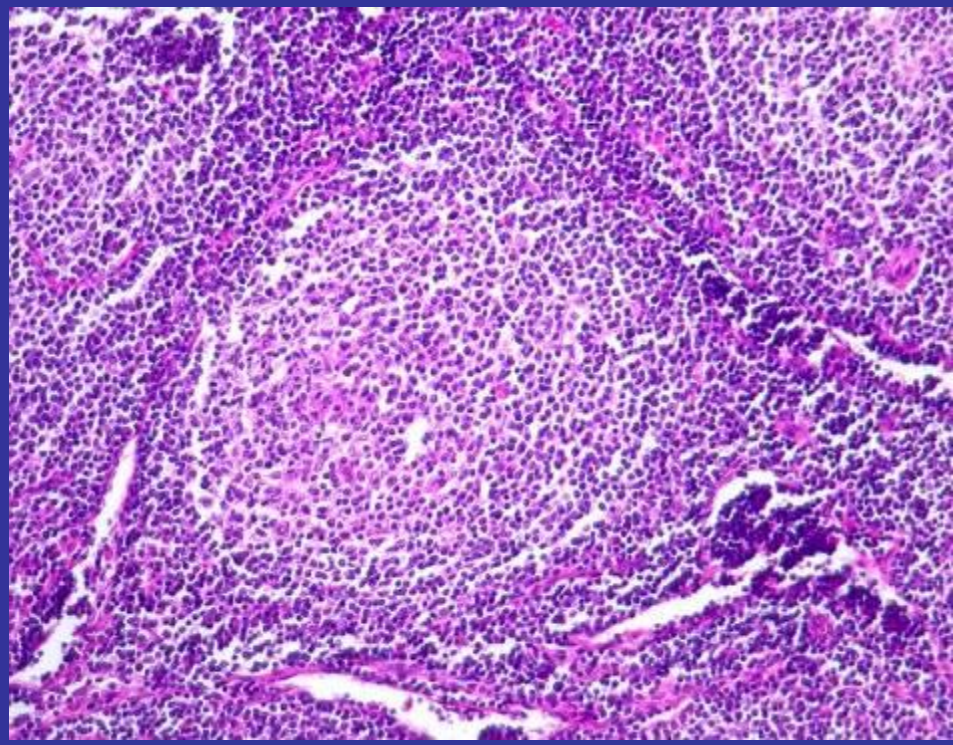
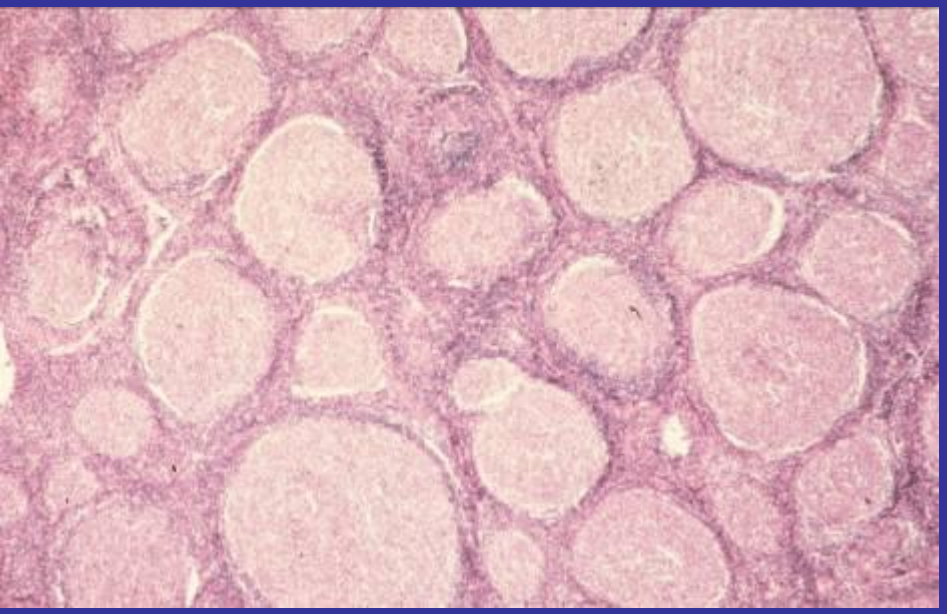
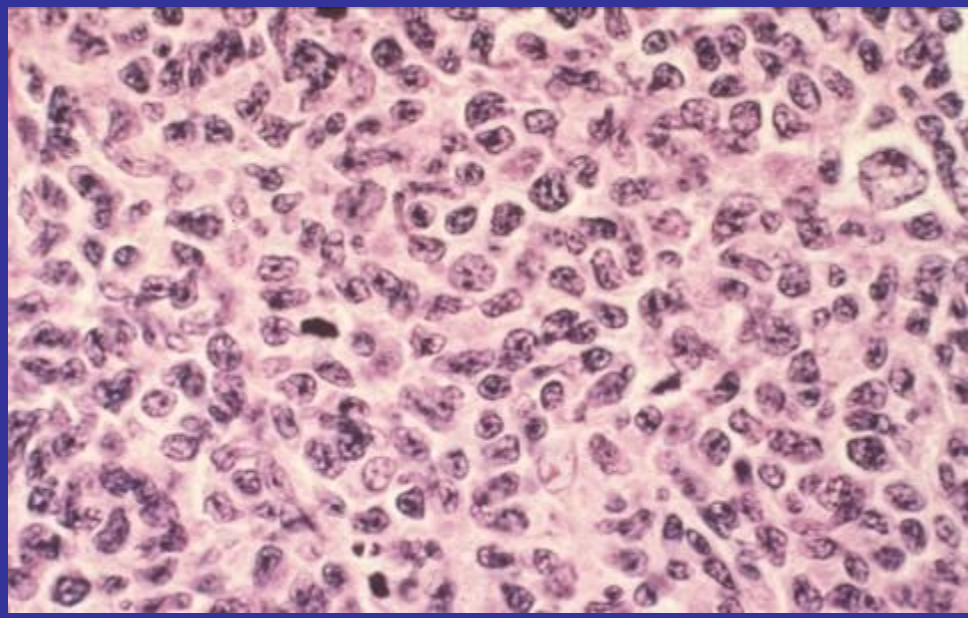




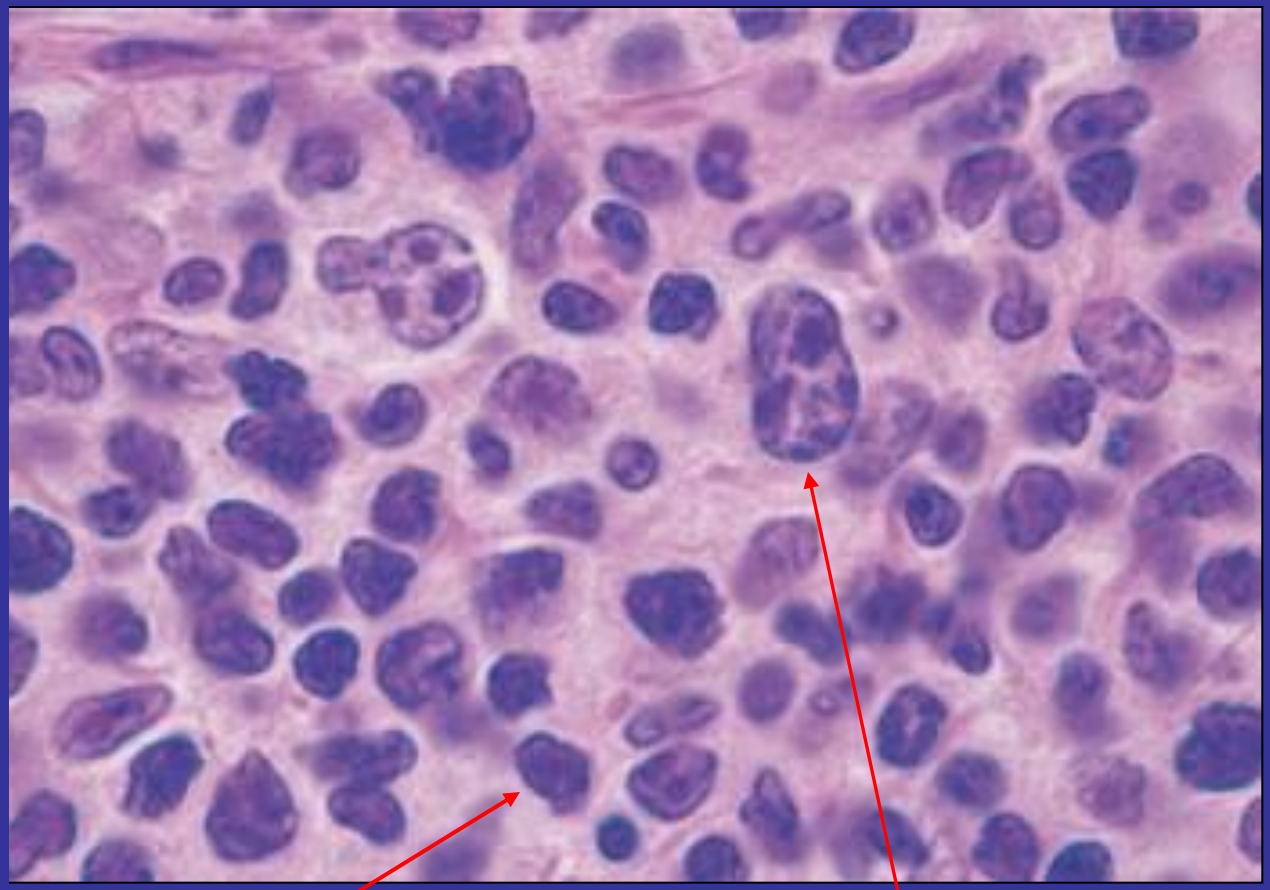
Follicular Lymphoma



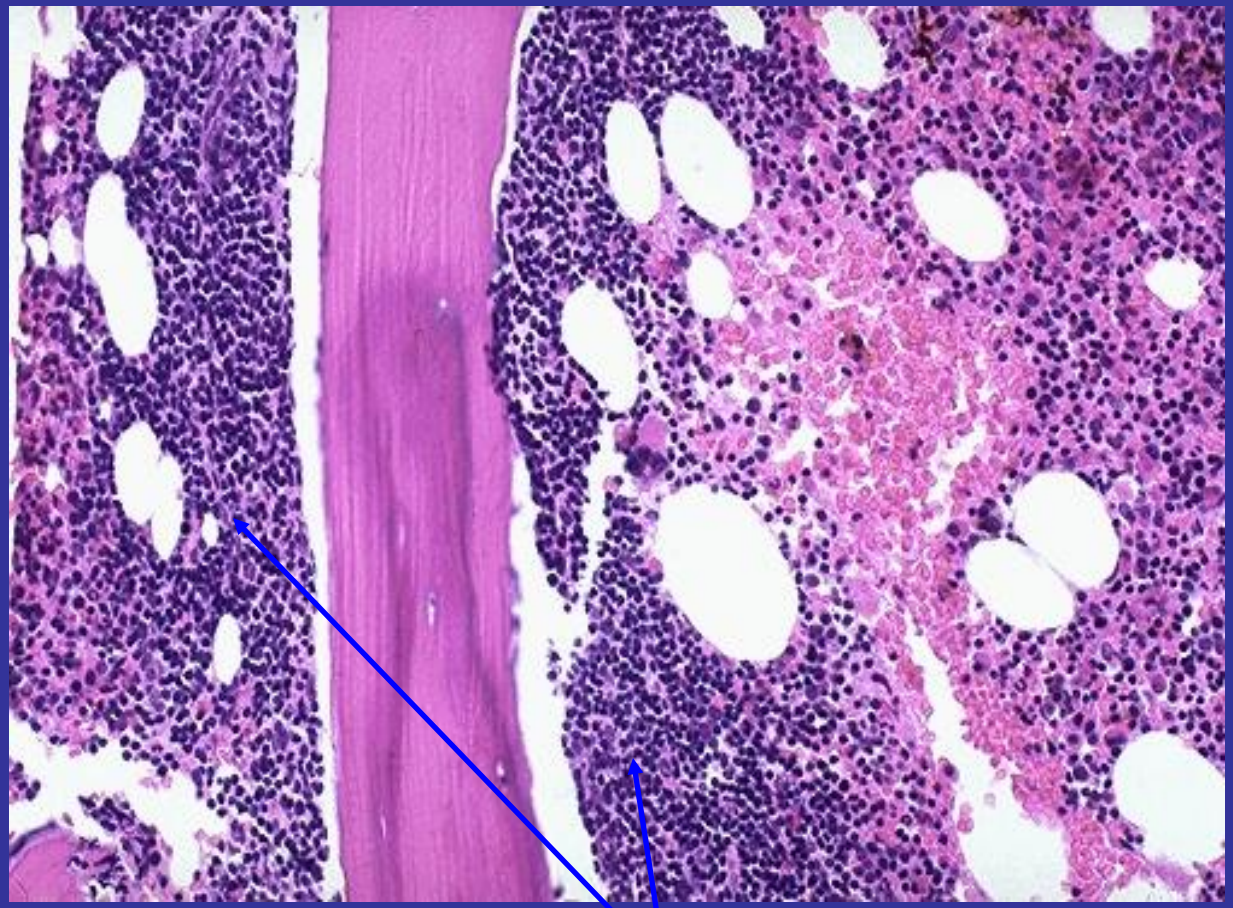


L.N.: Follicular lymphoma

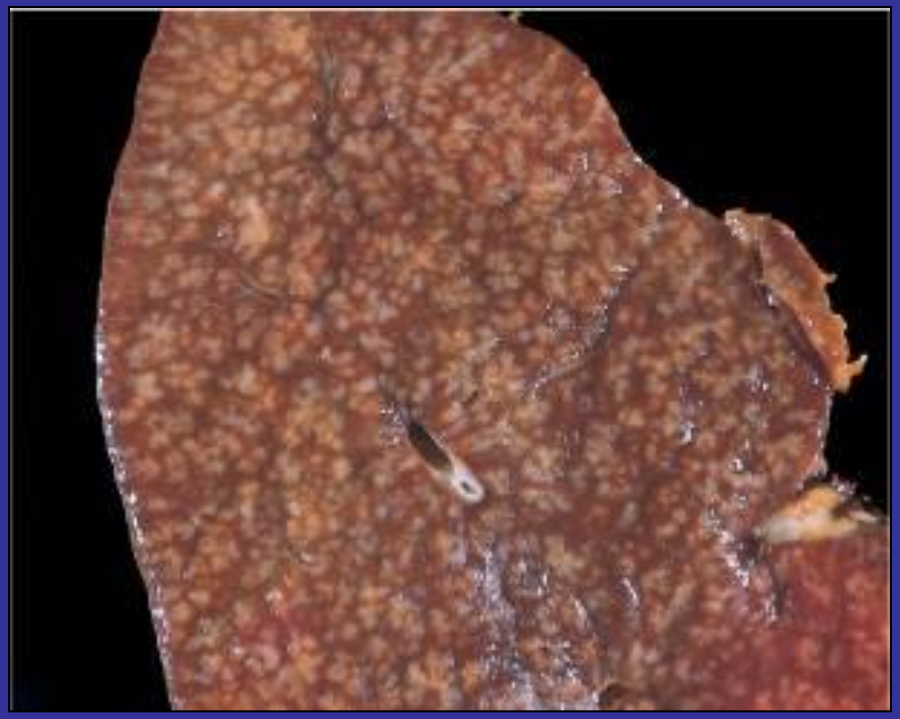
Follicular Lyphoma

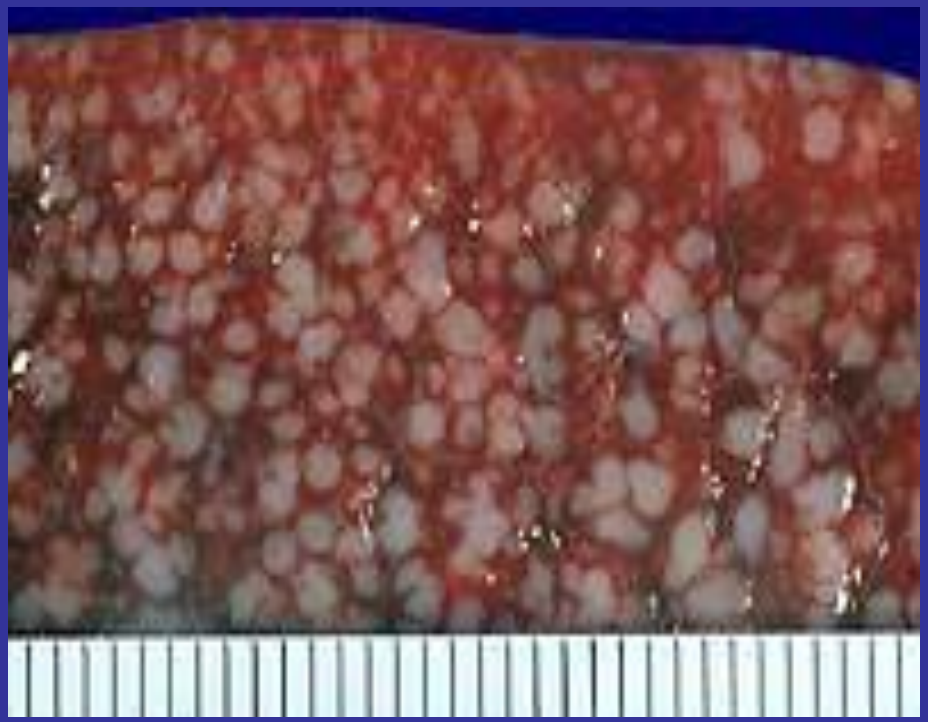


Follicular Lymphoma in Bone Marrow



Follicular Lymphoma in Spleen

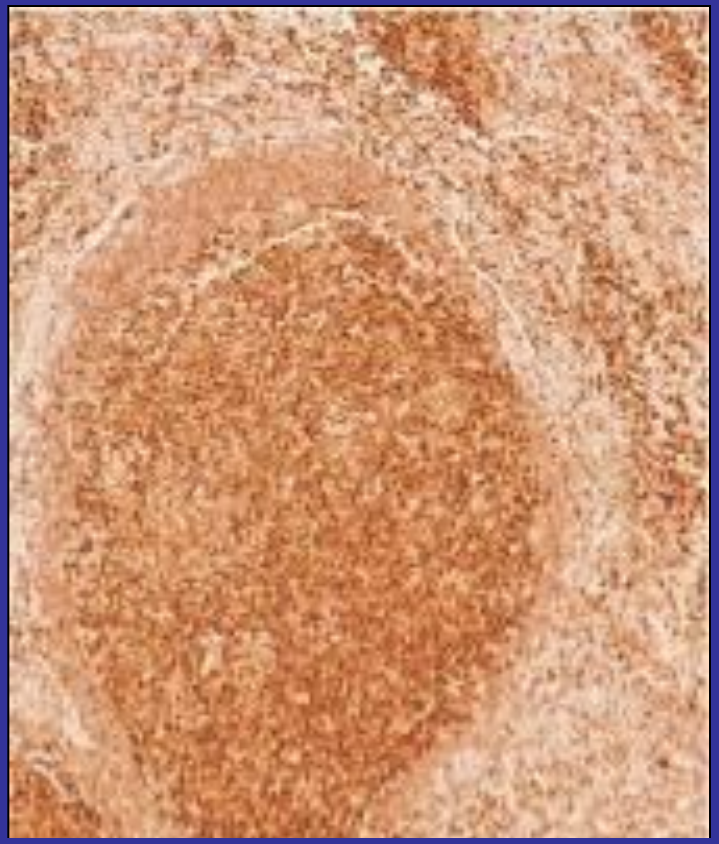




BCL2 Staining

Use in distinguishing reactive follicular hyperplasia from follicular lymphoma

Reactive germinal center



Follicular lymphoma

Follicular Lymphoma, Pattern

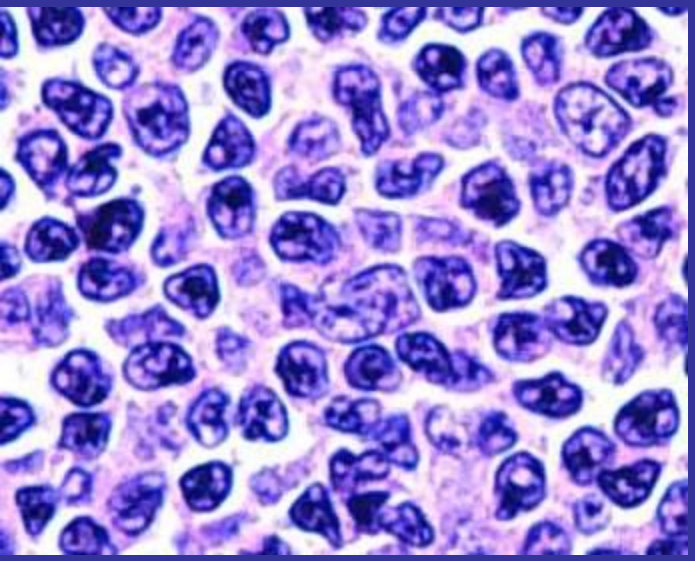
- Follicular > 75% follicular
- Follicular and diffuse 25-75% follicular
- Minimally follicular < 25% follicular

Follicular Lymphoma, Grading

- Grade 1: 0-5 centroblasts / hpf
- Grade 2: 6-15 centroblasts / hpf
- Grade 3: > 15 centroblasts / hpf
- 3a: Some centrocytes present
- 3b: Solid sheets of centroblasts

Follicular Lymphoma Grading

Grade 1



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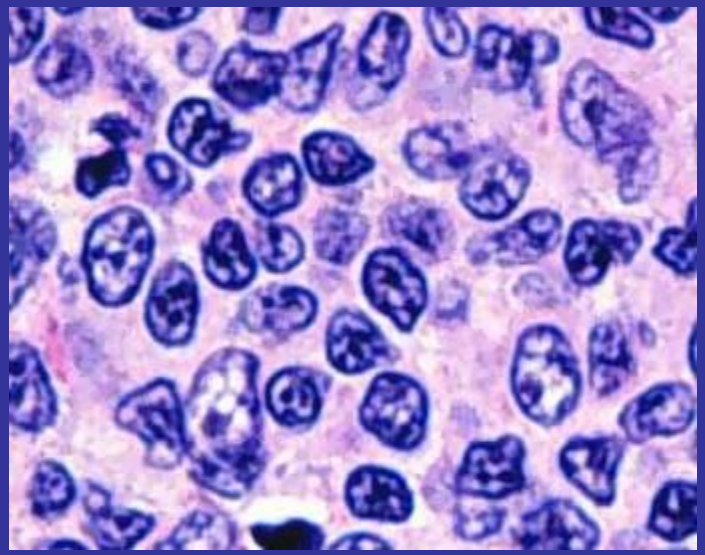
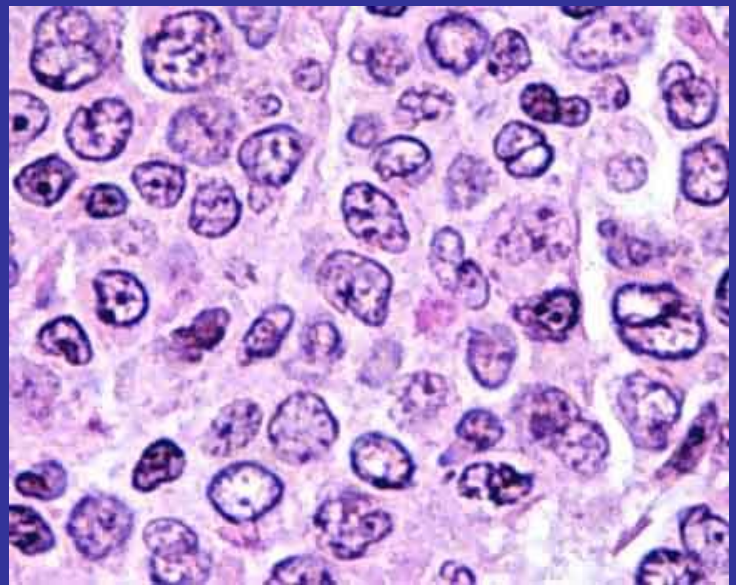
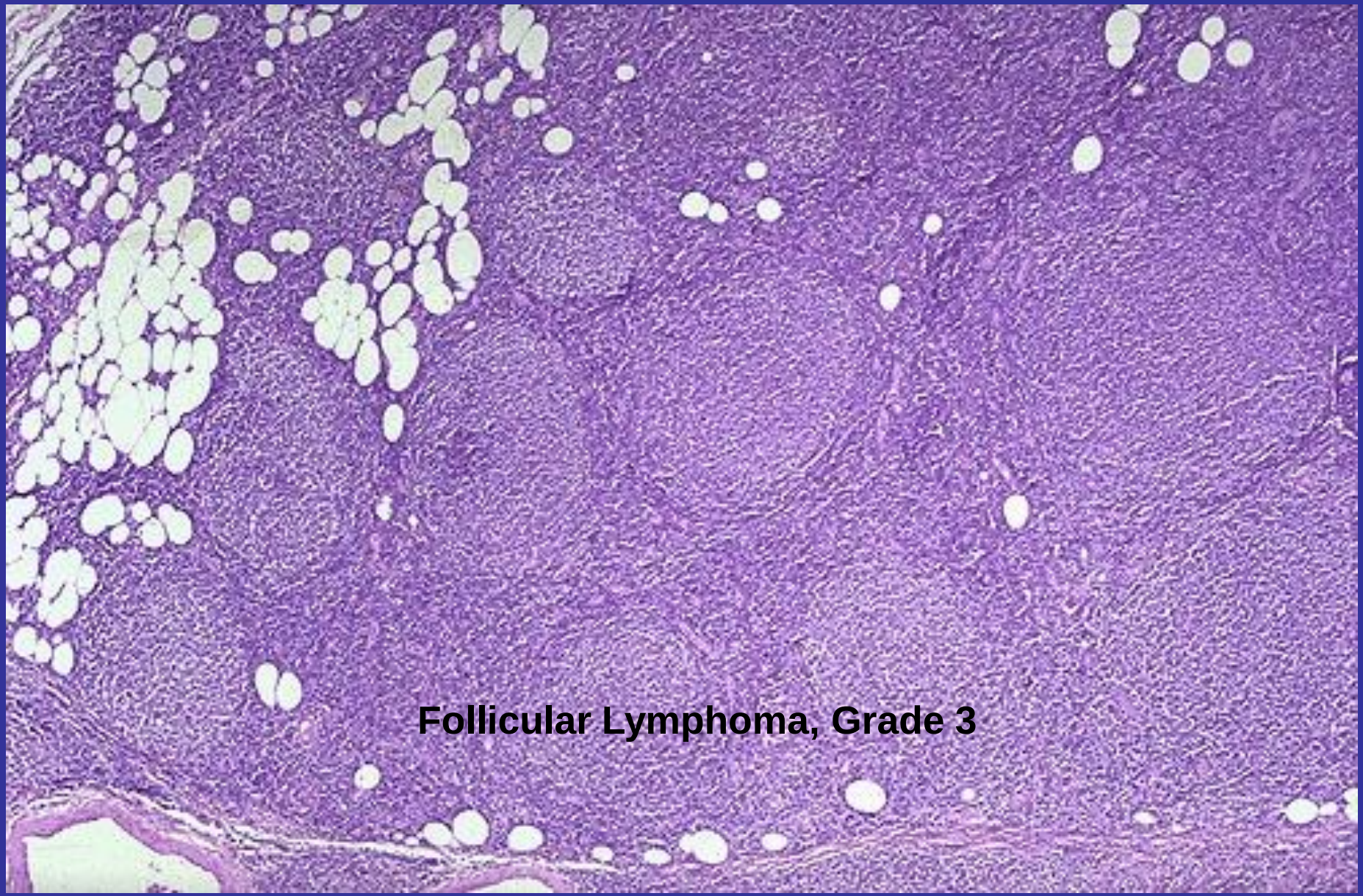
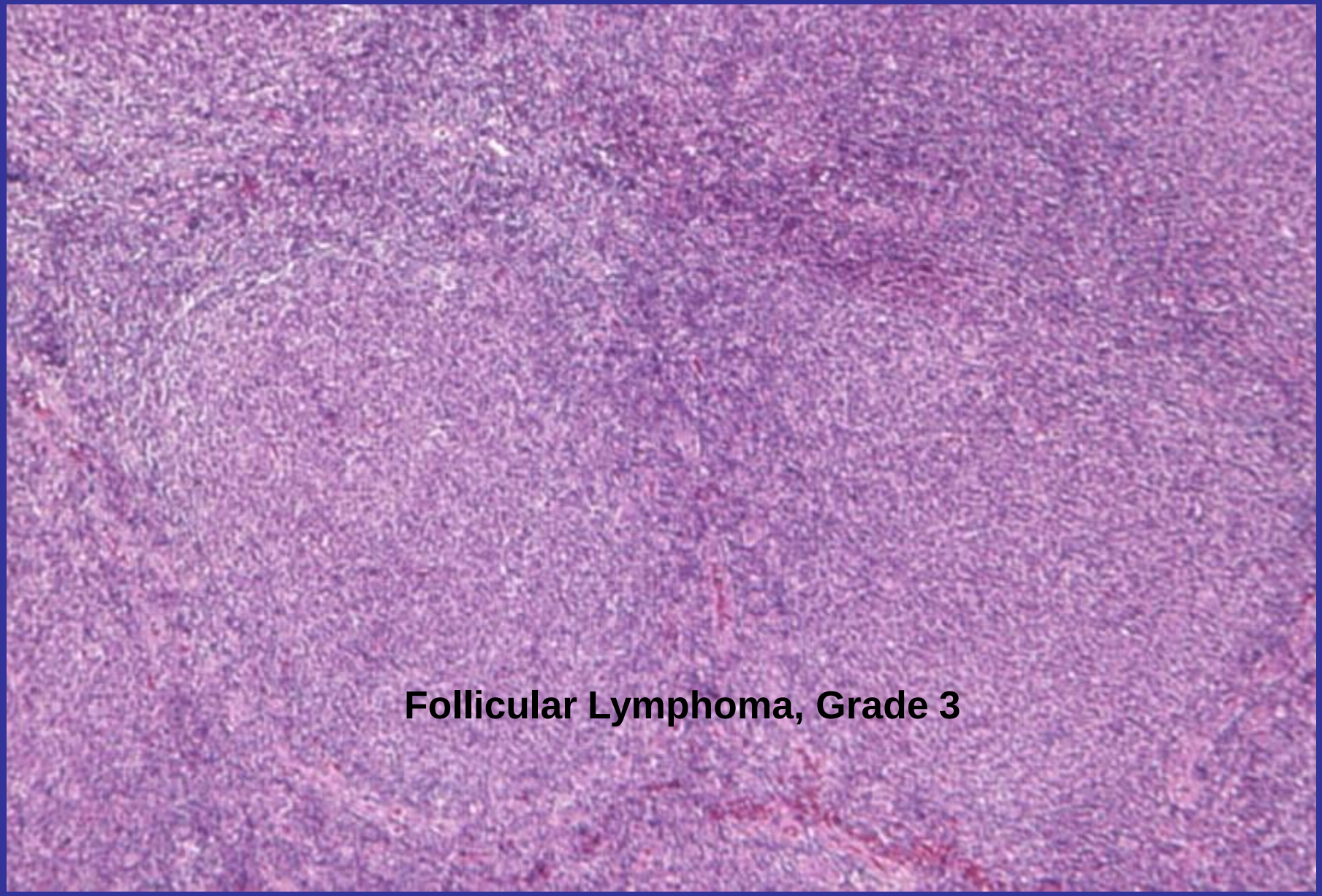
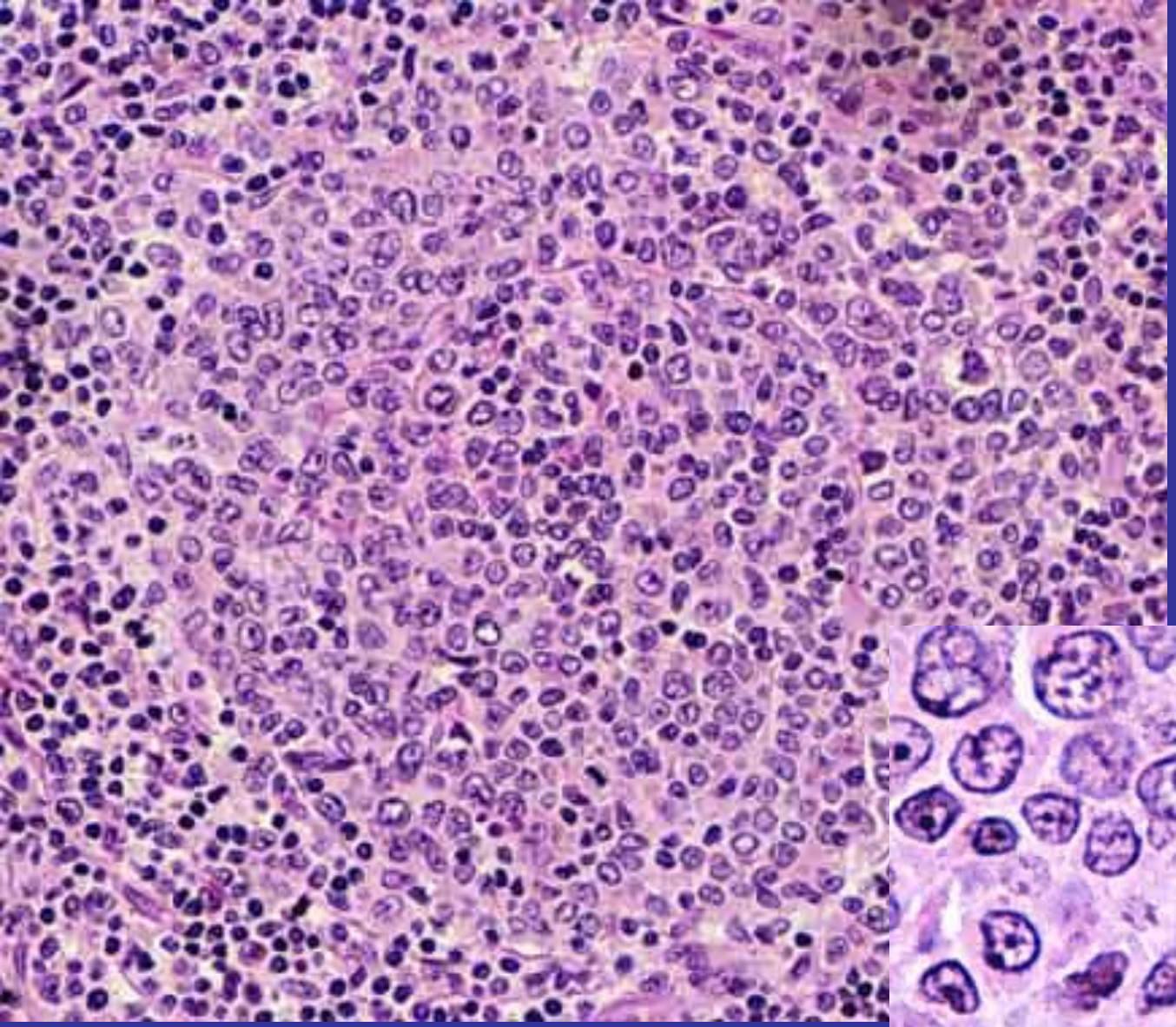


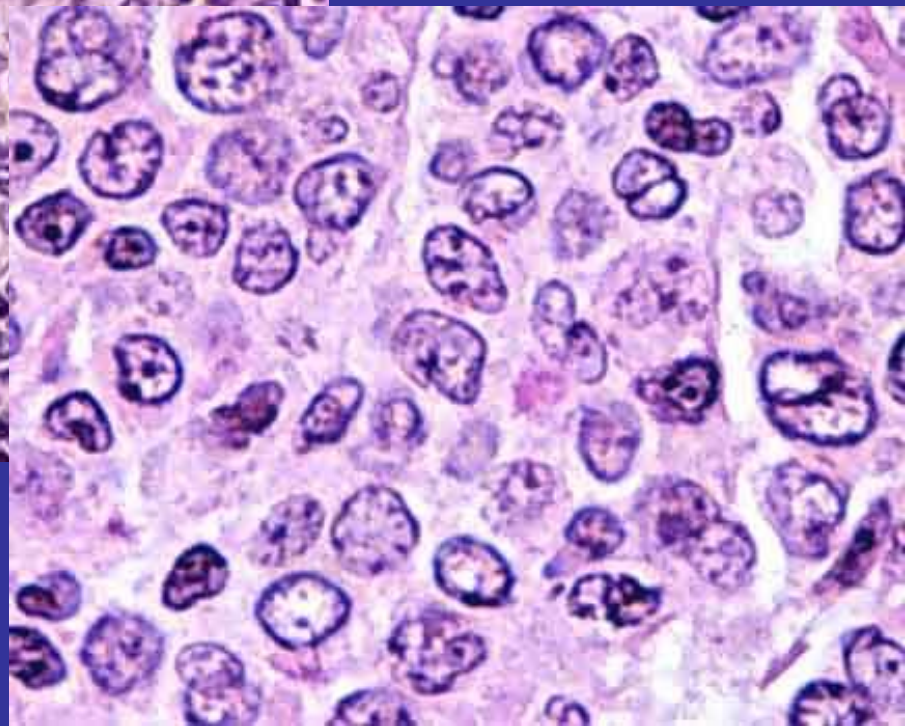
Grade 3



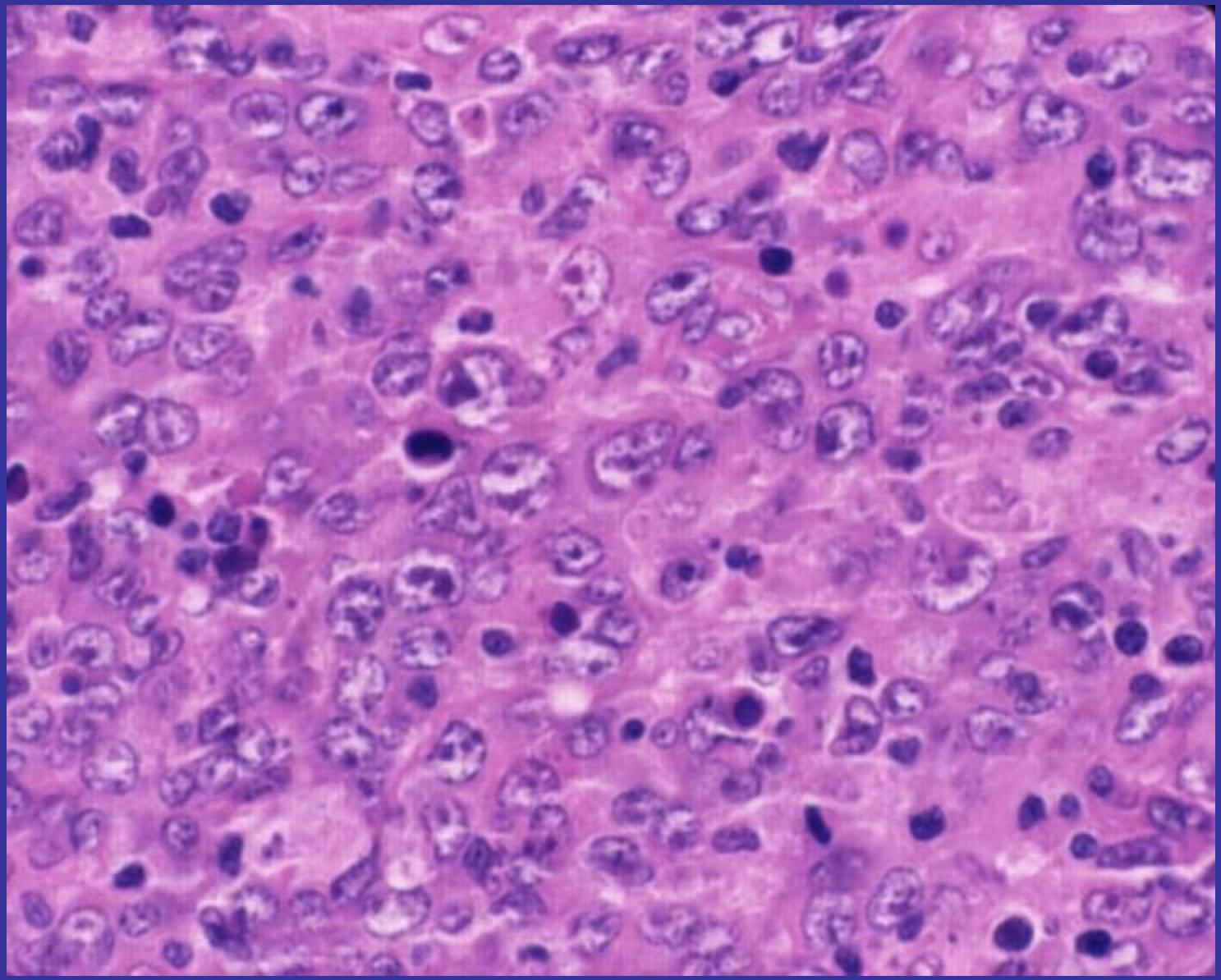

Follicular Lymphoma, Grade 3


Follicular Lymphoma, Grade 3

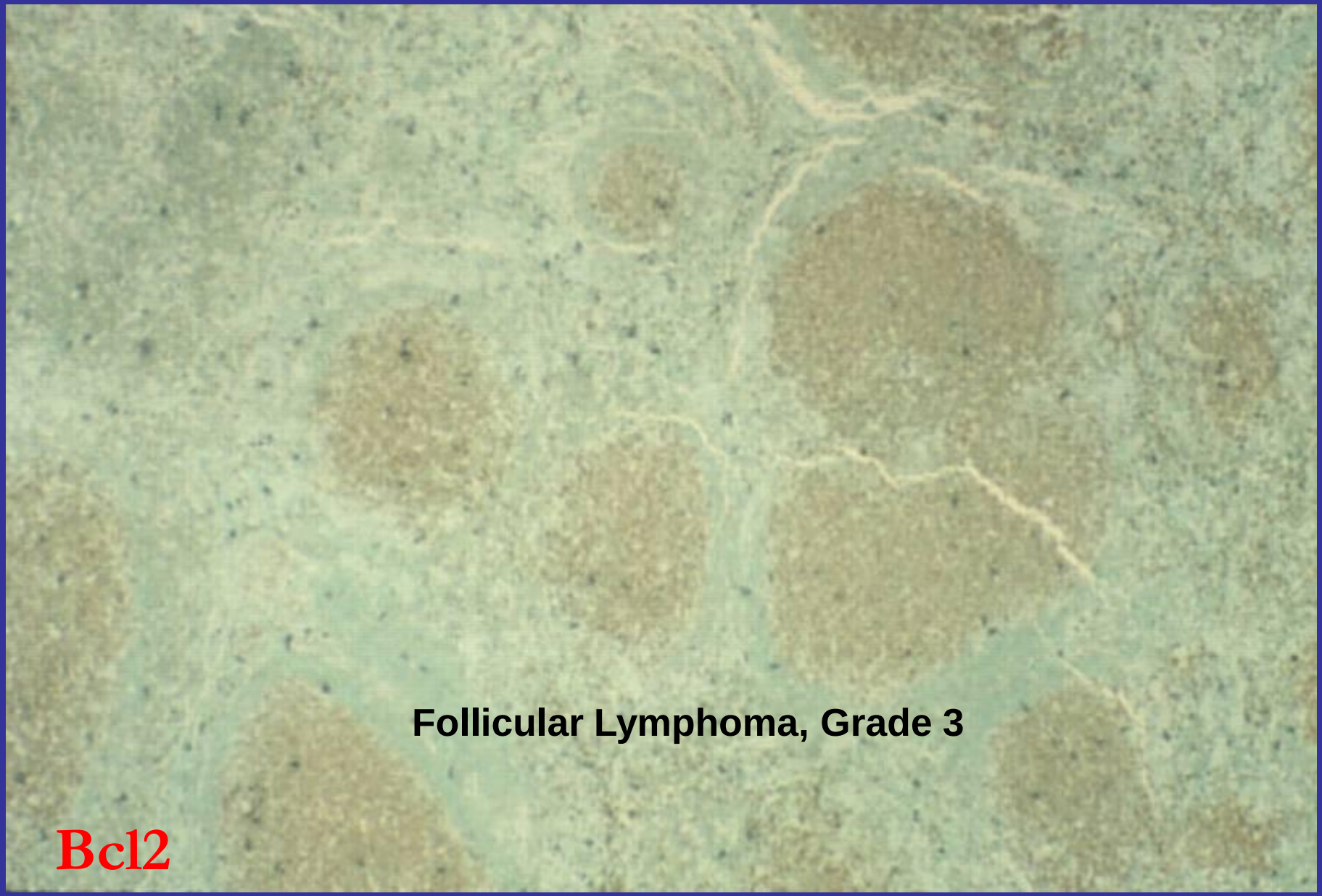




Follicular Lymphoma, Grade 3

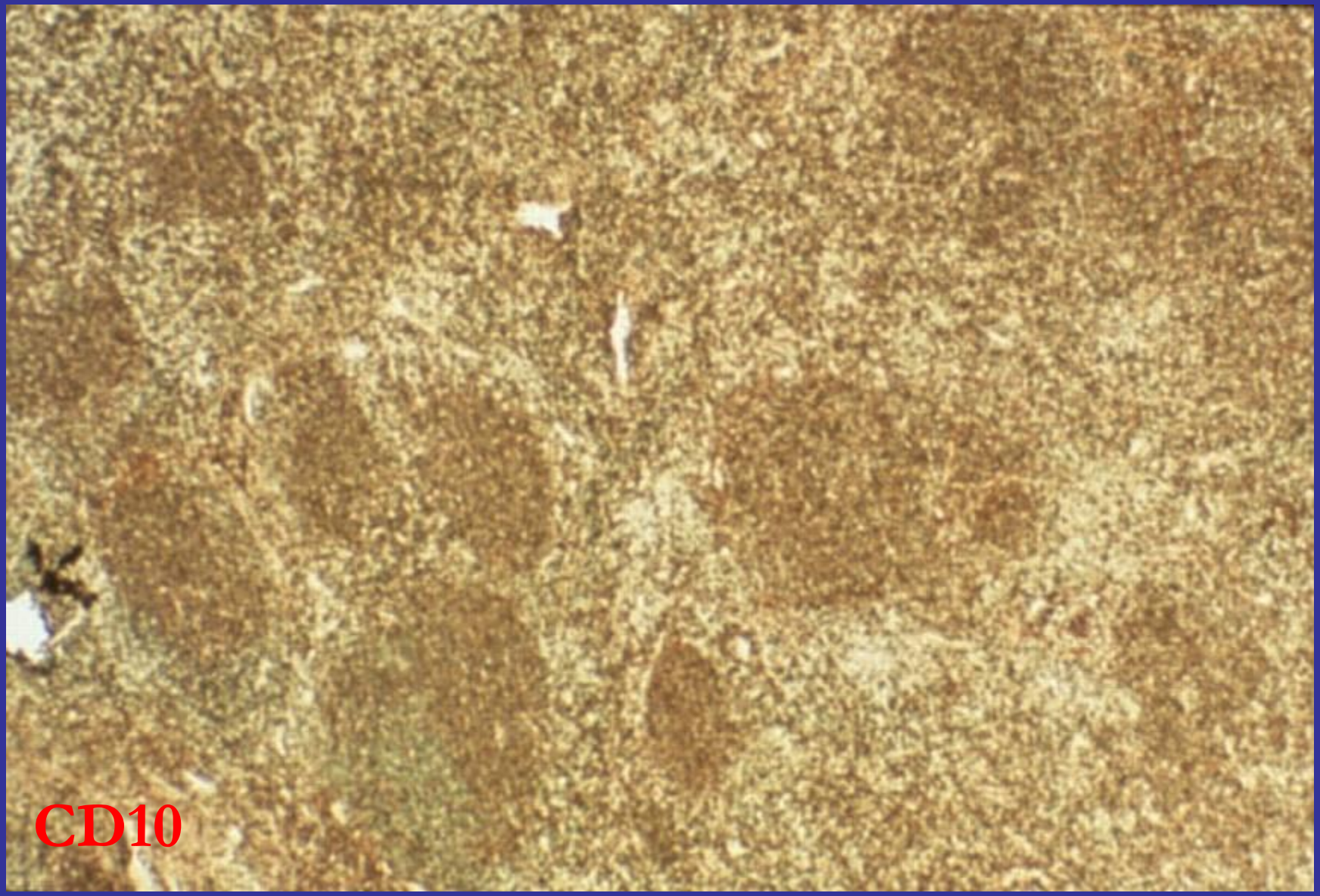


Follicular Lymphoma, Grade 3

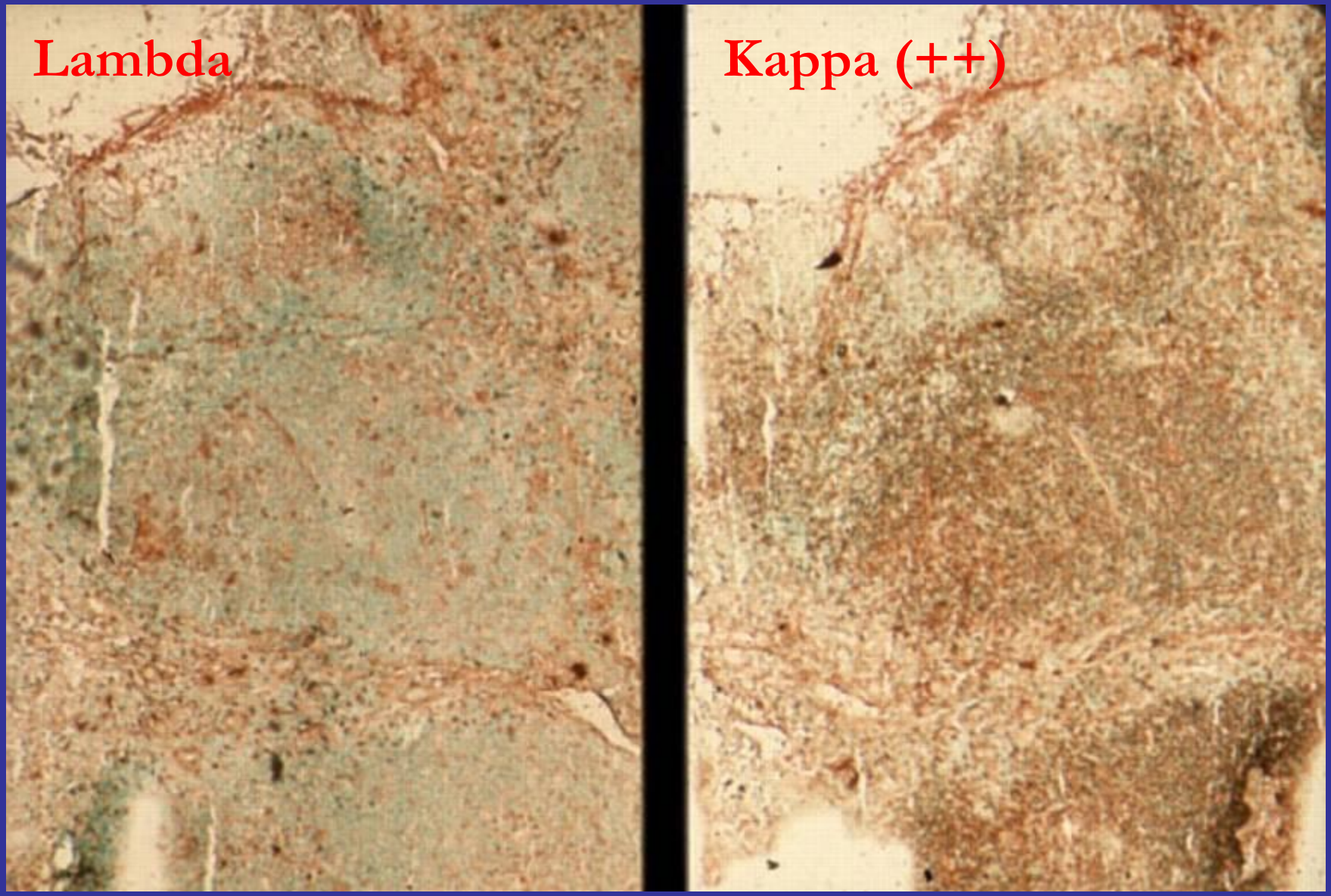

Follicular Lymphoma, Grade 3
Bcl2

Follicular Lymphoma, Immunophenotype

- Surface Ig +
- Express B-cell antigens: CD19, CD20, CD22, CD79a
- CD 10 +
- BCL-2 + (can help distinguishing from reactive follicles; however, grade 3 and cutaneous type may be negative)
- BCL 6 +

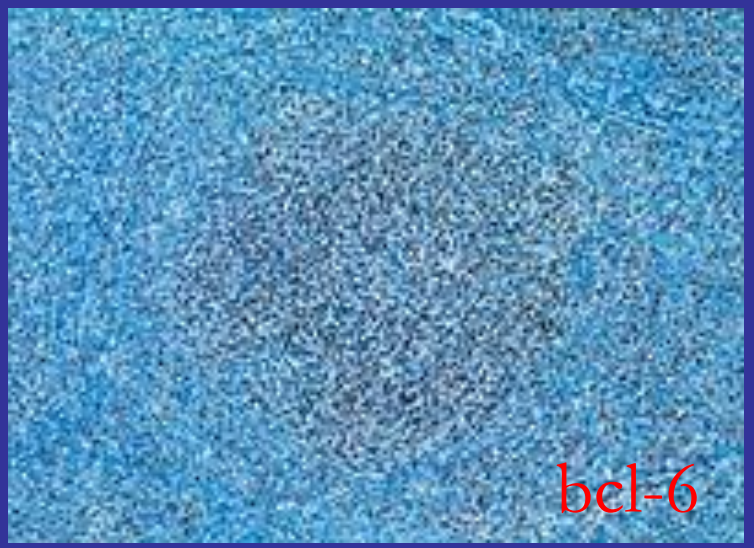


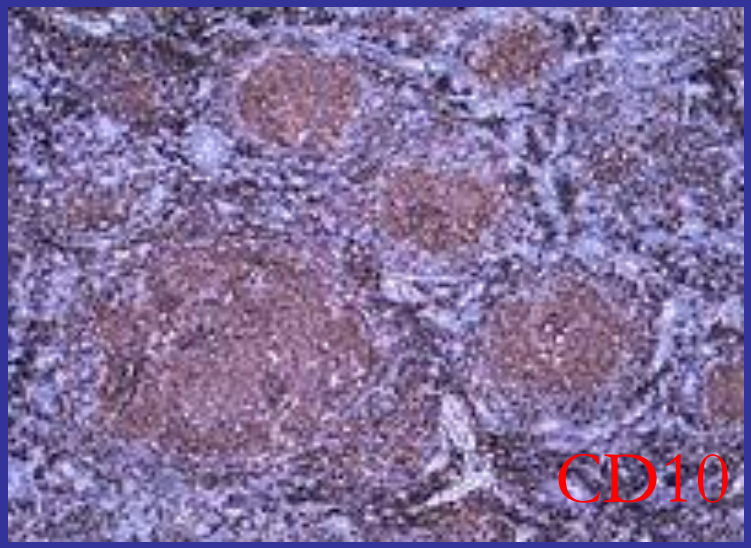
Follicular Lymphoma, Grade 3

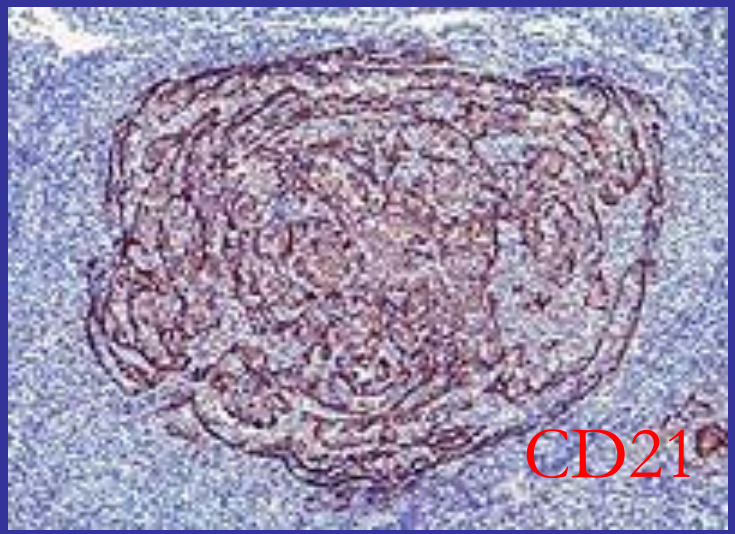


Follicular Lymphoma, Grade 3

Follicular Lymphoma







Follicular Lymphoma, Genetics

- t(14;18) (q32;q21)
- BCL 2 rearrangement, present is 70-95% cases
- Confers a survival advantage on B cells; failure to switch off BCL 2 during blast transformation may contribute to development of lymphoma by preventing apoptosis

Follicular Lymphoma, Prognosis

- Grades 1 and 2: indolent
- Grade 3: aggressive; treatment as for DLBCL, 25-33% of cases progress to DLBCL

Follicular Lymphoma, Variants

- (1) Diffuse Follicle Centre Lymphoma:
  - Centrocytes and centroblasts (minority) but no follicles
  - Both cell types must have follicle-centre cell phenotype (sIg+, CD 10+, BCL2+, BCL6+)
  - If centroblasts predominate, or if the small cells are T cells, DLBCL should be considered

- (2) Cutaneous Follicle Centre Lymphoma:
  - Partially follicular pattern
  - Composed of cells that resemble centrocytes and centroblasts
  - Often BCL2 –

Mantle Cell Lymphoma

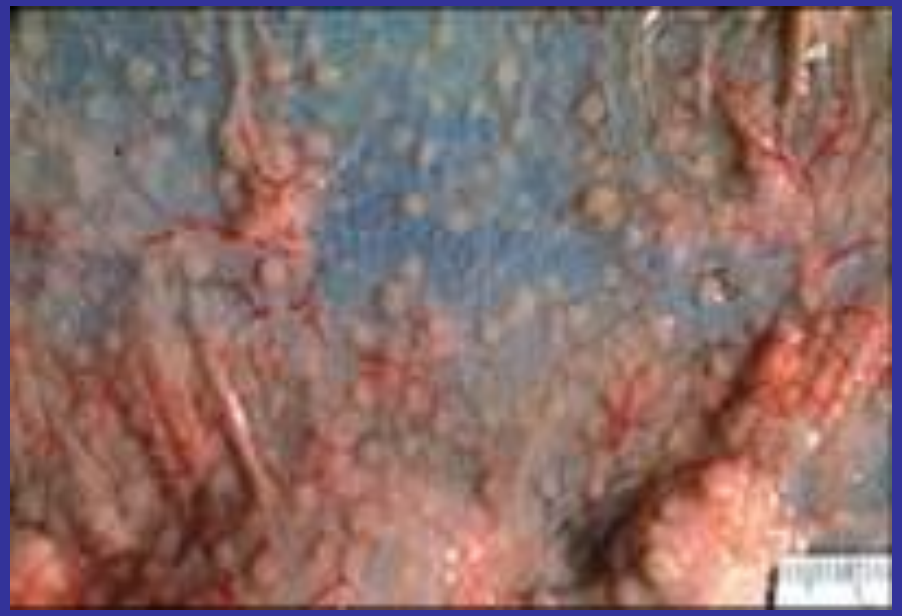
Mantle Cell Lymphoma, Definition

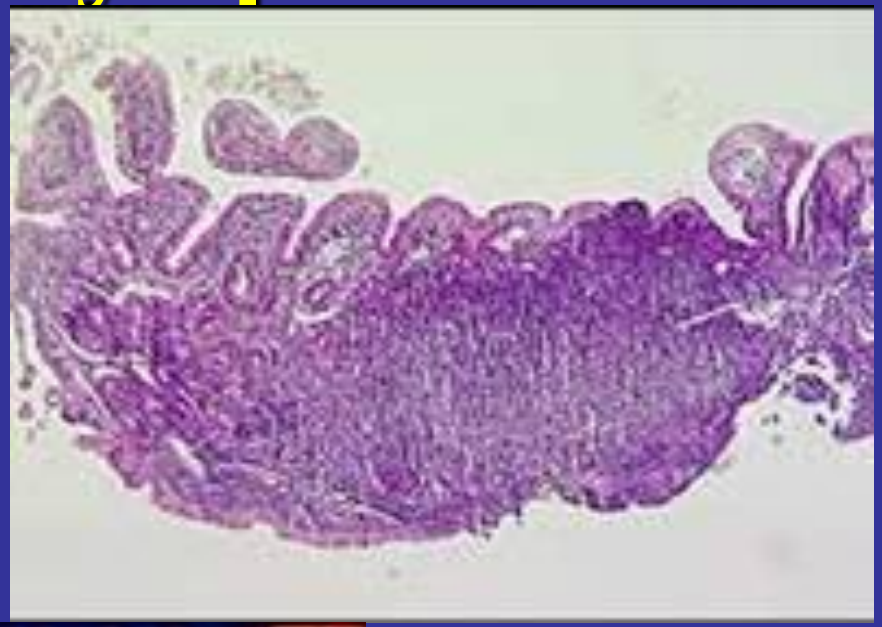
- B-cell neoplasm of monomorphous small to medium-sized cells that resemble centrocytes
- Median age: 60 yrs
- Male predominance
- Extranodal sites: bone marrow (50-60%), GI (30% with lymphomatous polyposis in large intestine), and Waldeyer's ring
- Most patients present with lymphadenopathy, hepatosplenomegaly

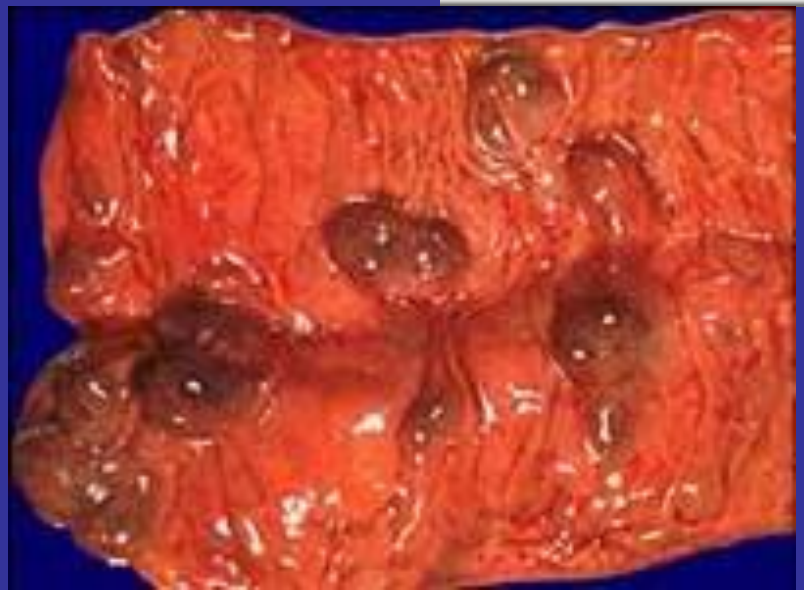
Mantle Cell Lymphoma, Morphology

- Monomorphic proliferation of small to medium-sized lymphoid cells that resemble centrocytes
- Vague nodular, or diffuse, or mantle zone growth pattern
- Hyalinized small blood vessels

Mantle Cell Lymphoma

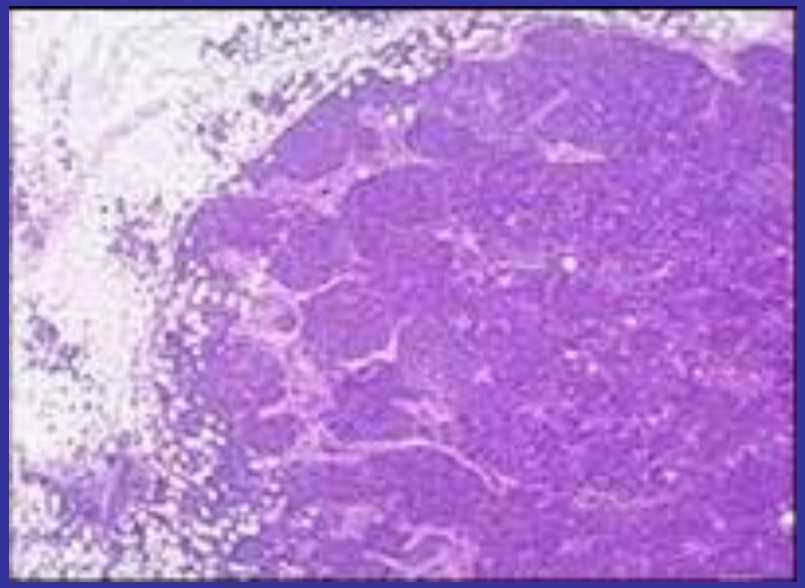


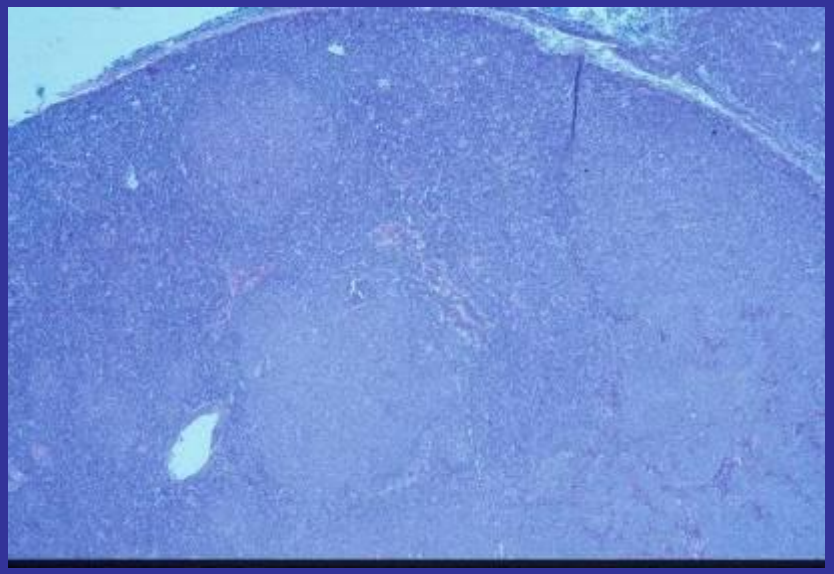


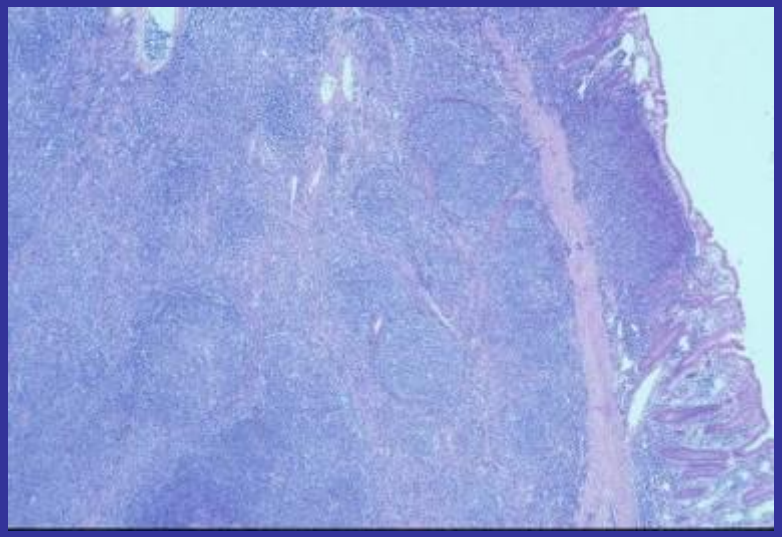


Lymphomatous polyposis

Mantle Cell Lymphoma

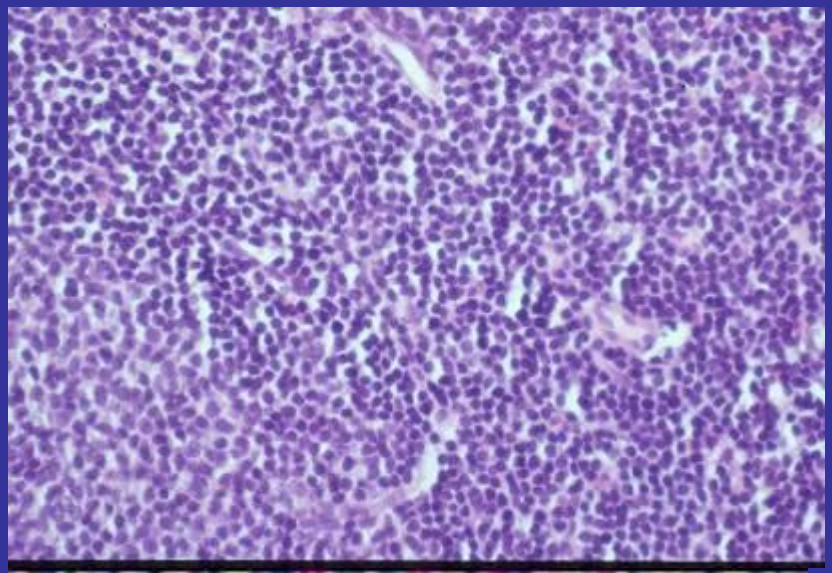


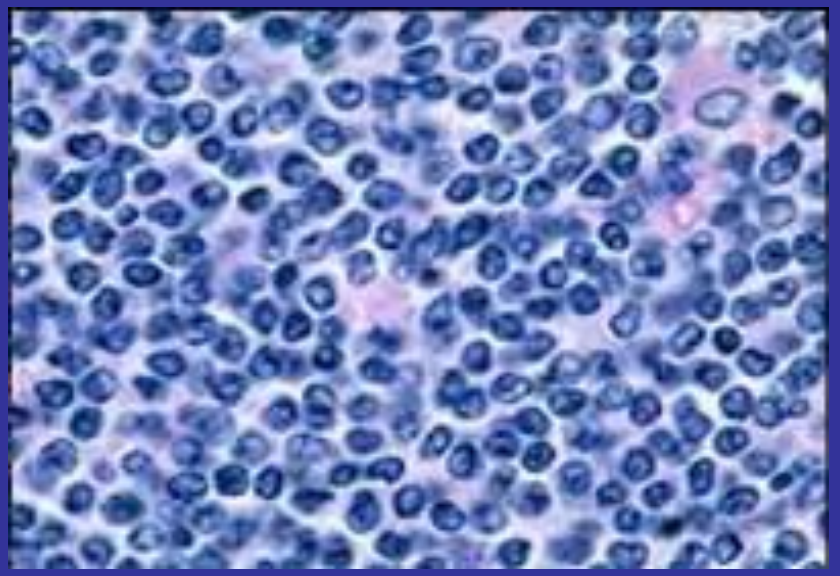


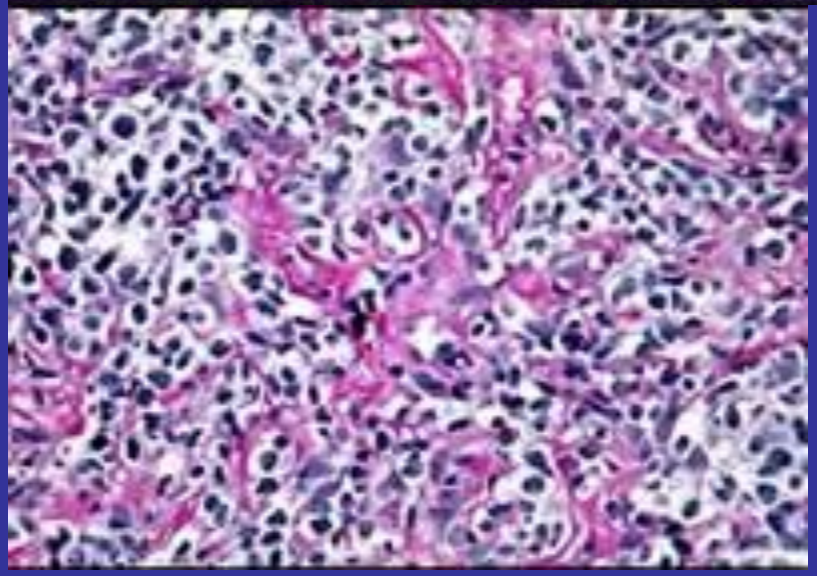


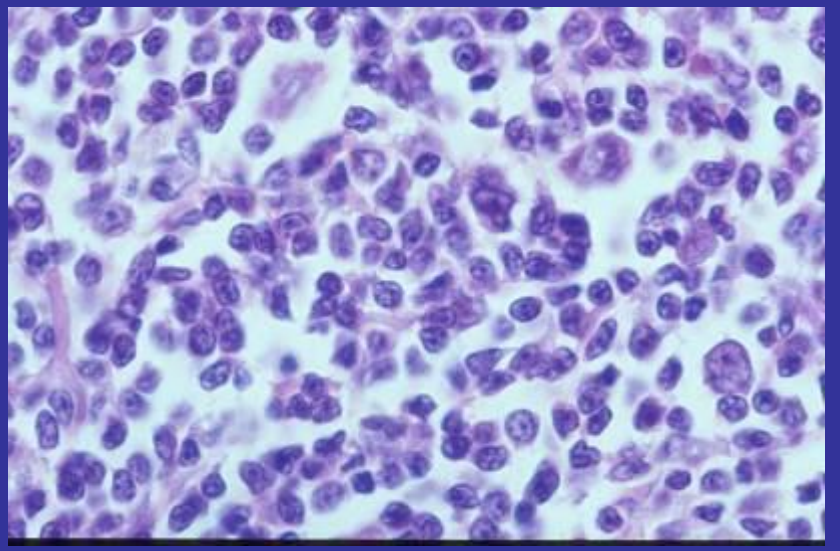
Nodular pattern

Mantle Cell Lymphoma





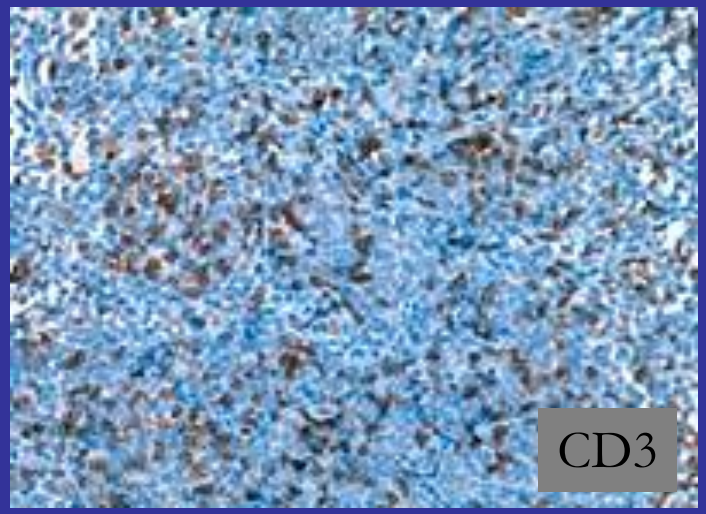


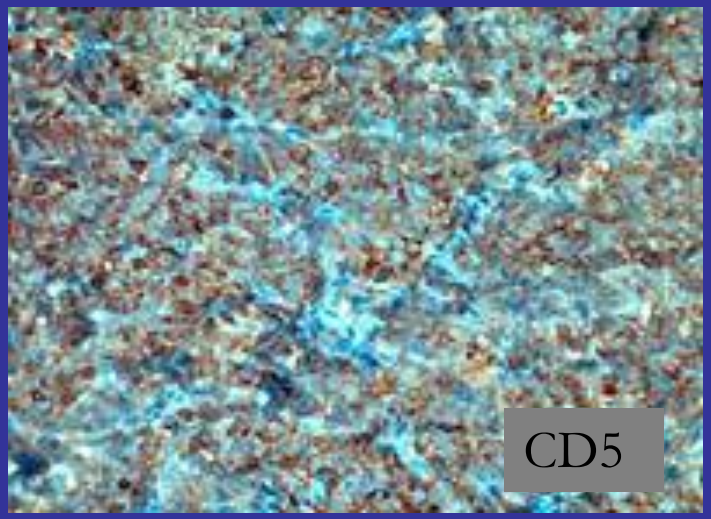


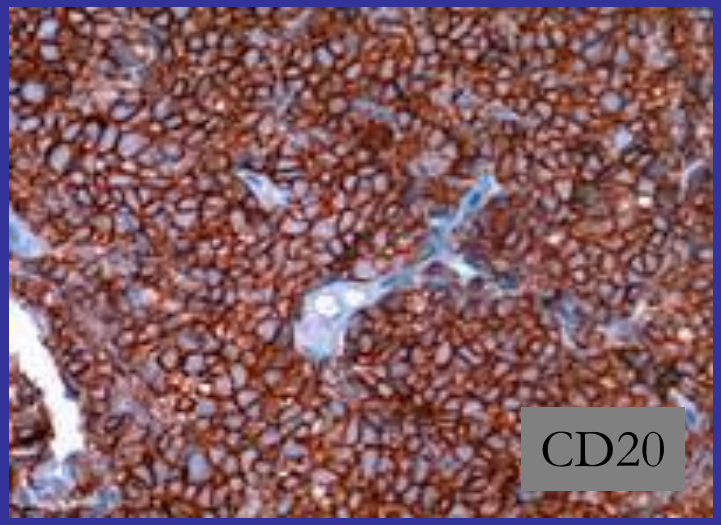
Mantle Cell Lymphoma, Immunophenotype

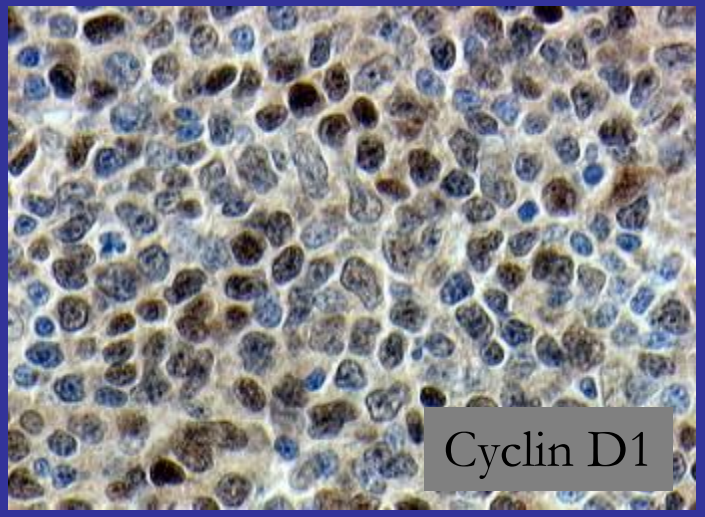
- Intense sIg (IgM +/- IgD)
- CD5 +, CD43 +, BCL-2 +
- Cyclin D1 seen in 70-80% of cases
- CD10 -, BCL-6 –

Mantle Cell Lymphoma









Mantle Cell Lymphoma, Genetics

- t(11;14) (q13;q32)
  chromosome 11: Cyclin D1
  chromosome 14: Ig heavy chain

Mantle Cell Lymphoma, Blastoid Variant

- Cells resemble lymphoblasts with dispersed chromatin
- High mitotic rate (>20-30/ 10 hpf)

Mantle Cell Lymphoma
Blastoid Variant