

# Mantle Cell Lymphoma (MCL)

## Clinical features and Diagnosis

# Lymphoma Epidemiology\*

- Surveillance Epidemiology and End Results (SEER)\* information from the NCI (2006-2010) show the following incidence results:
  - B-cell lymphomas: 16.6/100,000/yr
  - T-cell lymphomas: 1.8/100,000/y
  - Hodgkin lymphoma: 2.8/100,000/yr
  - Mantle cell lymphoma (estimate): ~0.4-1.2/100,000/yr [2- 6% of non-Hodgkin (B-cell+T-cell) lymphomas]

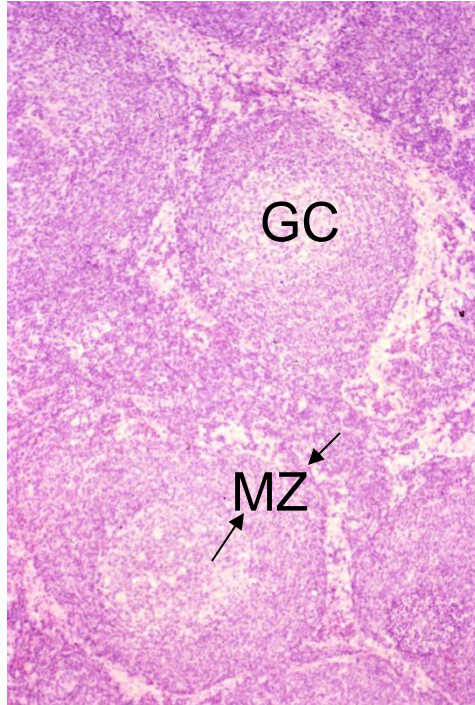
# Mantle Cell Lymphoma

## Clinical Features

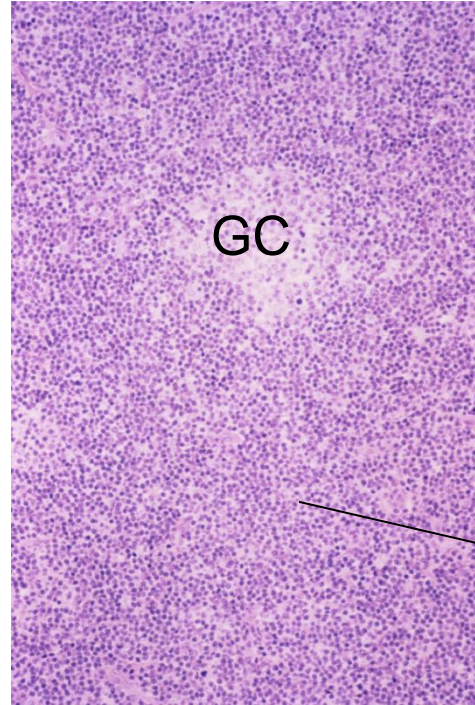
- Median age 60 with a male predominance
- Most patients present with generalized lymphadenopathy
- Blood/bone marrow involved in 20-40%
- Splenomegaly is common (60%)
- Gastrointestinal involvement in about 30% (“lymphomatous polyposis”)

# Mantle Cell Lymphoma

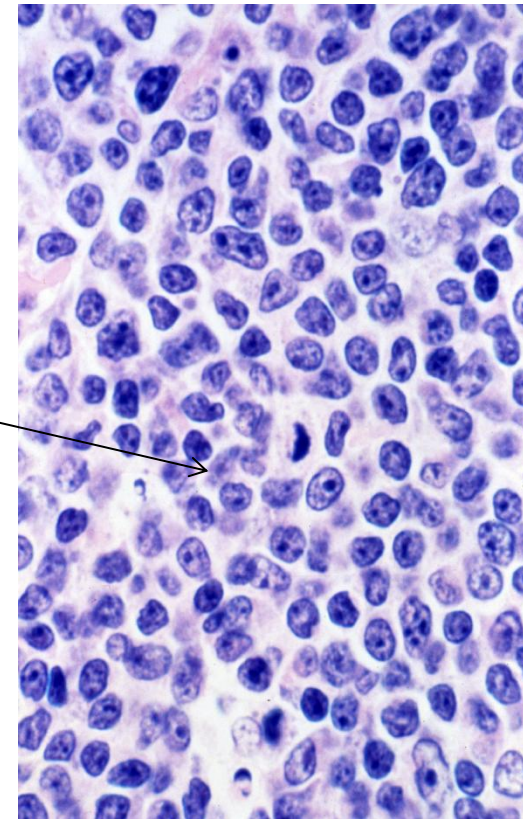
## Pathology



Normal Lymphoid follicle



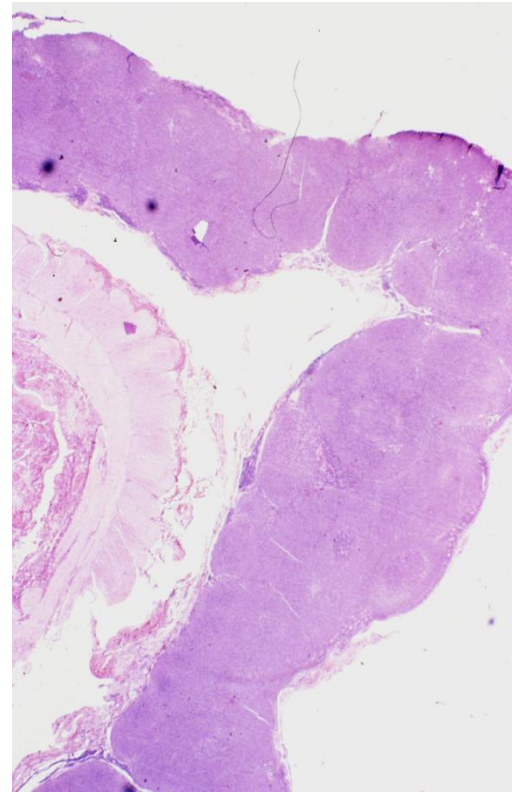
Confluent mantle zones  
with "naked" germinal centers



# Mantle Cell Lymphoma

## Pathology

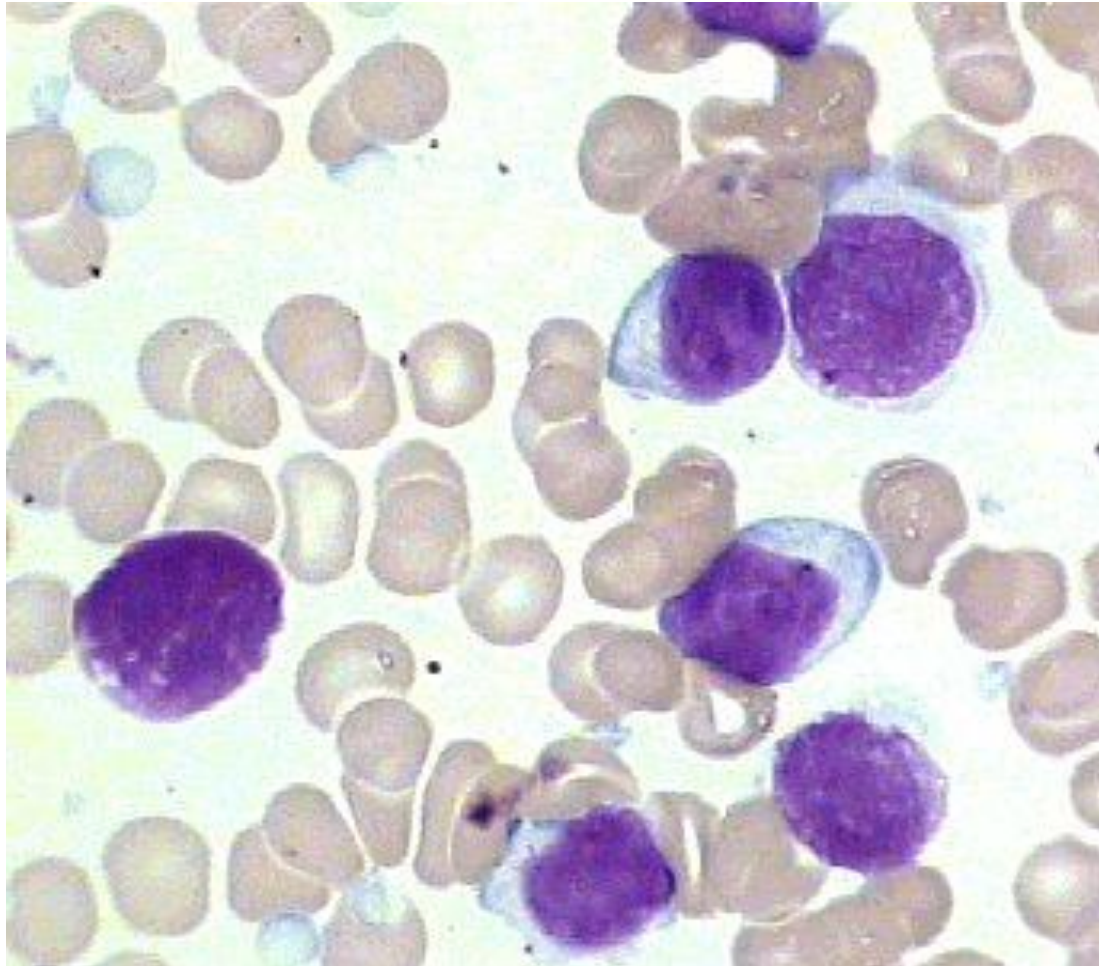
GI involvement may result in a condition called lymphomatous polyposis





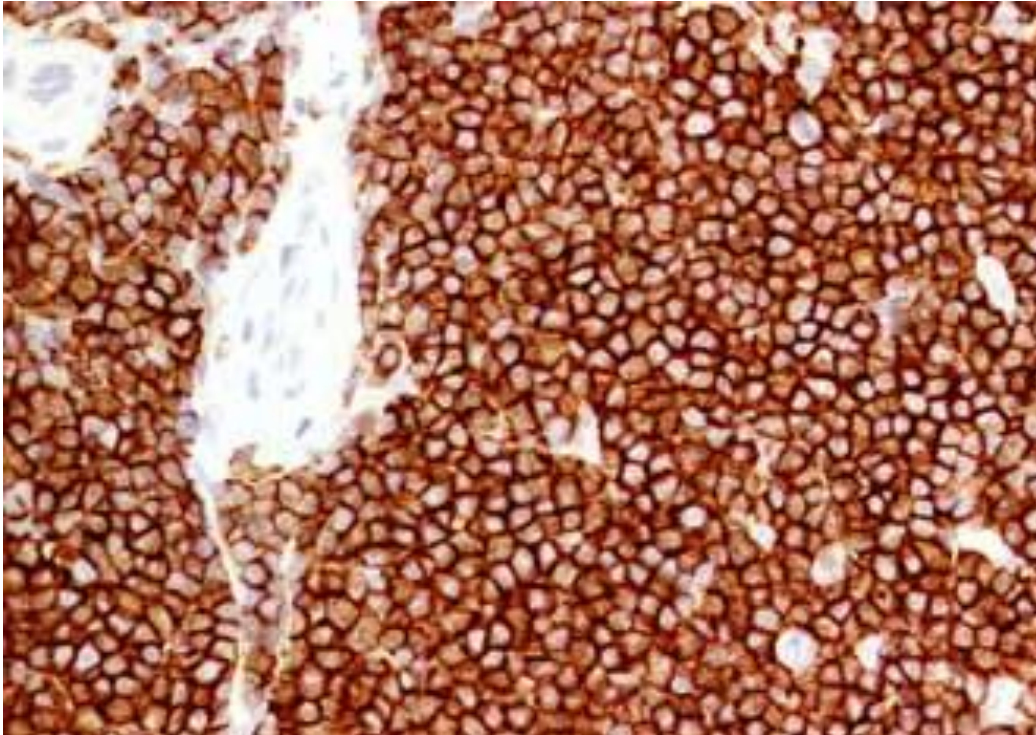
# Mantle Cell Lymphoma

## Blood Involvement



# Mantle Cell Lymphoma

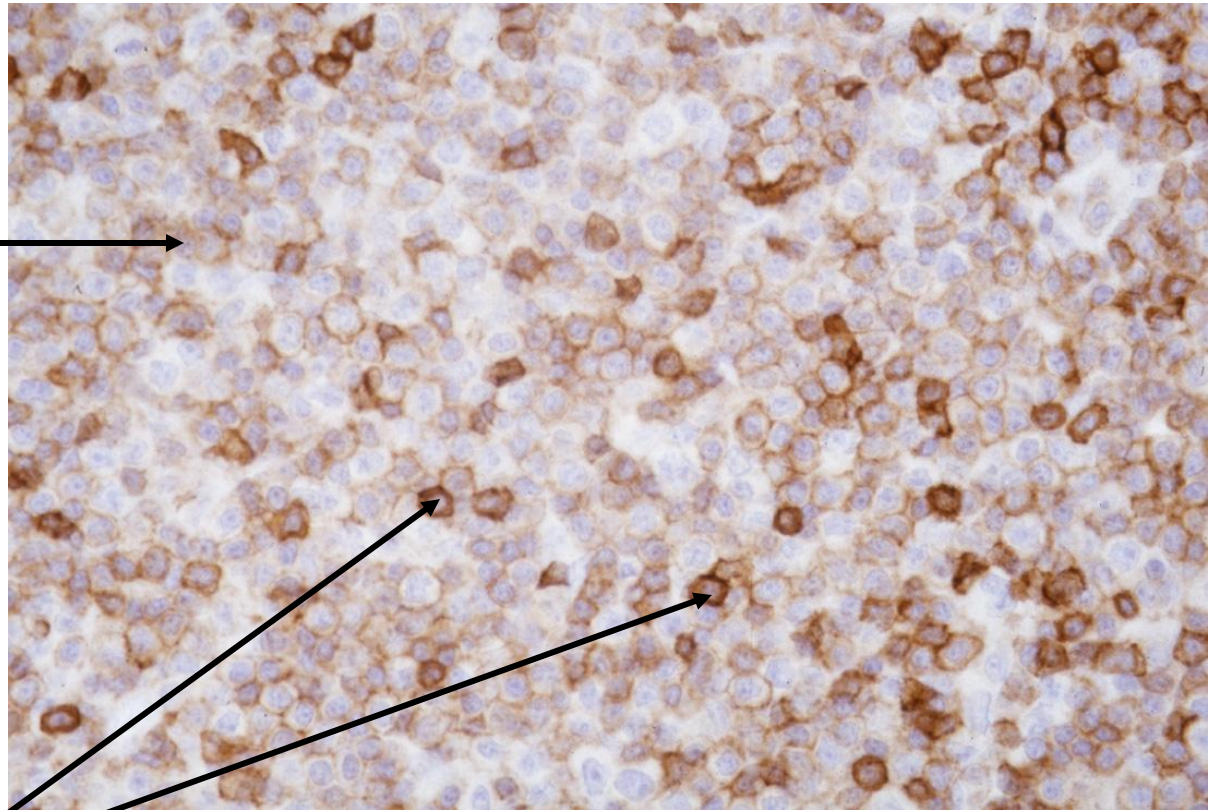
## Immunophenotype



**CD20+ (B cell)**

# Mantle Cell Lymphoma Immunophenotype

**MCL: CD5+  
(weak)**



**T cells: CD5+(strong)**



# MCL: Differential Diagnosis

## CLL/SLL

slg intensity (flow): **dim**

CD5: positive

CD23: **positive**

Cyclin D1: **negative**

t(11;14): **negative**

## Mantle Cell

slg intensity(flow): **bright**

CD5: positive

CD23: **negative\***

Cyclin D1: **positive**

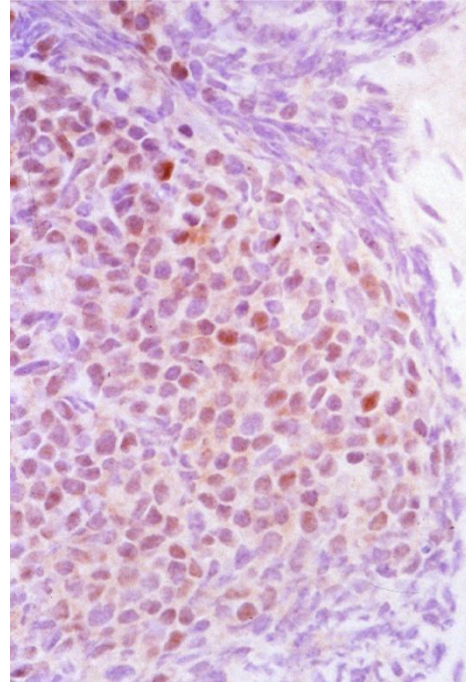
t(11;14): **positive**

\*Some moderately positive by flow cytometry

# Mantle Cell Lymphoma

## Cytogenetics

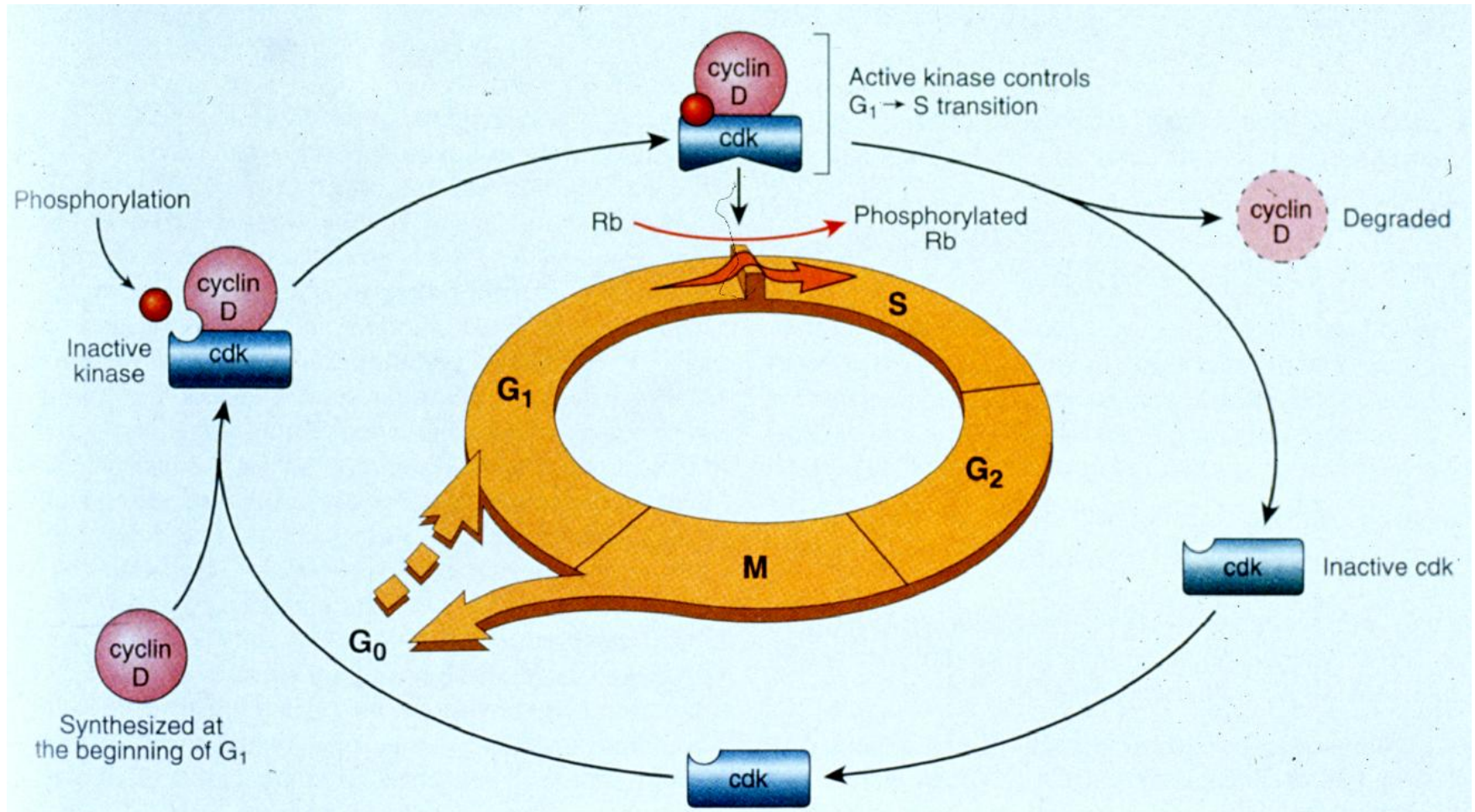
t(11;14)(q12;q32)→



Overexpression of cyclin D1

# MCL: Disease Mechanism

## Overexpression of Cyclin D1(+?)



# MCL: Disease Mechanism

## Overexpression of Cyclin D1(+?)

- In addition to dysregulation of cyclin D1 and the cell cycle, recent studies suggest that disruption of DNA damage response pathways and activation of cell survival mechanisms **contribute to oncogenesis** (Molecular pathogenesis of mantle cell lymphoma. Jares P, Colomer D, Campo E. 2012. J Clin Invest 122:3416-3423)



# Mantle Cell Lymphoma

## Prognosis

- Median survival about 3 years
- Rarely curable with current therapies