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")

Patrick J. Buckley, MD. PhD
Mantle Cell Lymphoma

Pathology

Normal Lymphoid follicle

Confluent mantle zones with “naked” germinal centers

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Pathology

GI involvement may result in a condition called lymphomatous polyposis
Mantle Cell Lymphoma
Blood Involvement

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Immunophenotype

CD20+ (B cell)
Mantle Cell Lymphoma

Immunophenotype

MCL: CD5+ (weak)

T cells: CD5+ (strong)

Patrick J. Buckley, MD, PhD
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