I See Mantle Cell Lymphoma

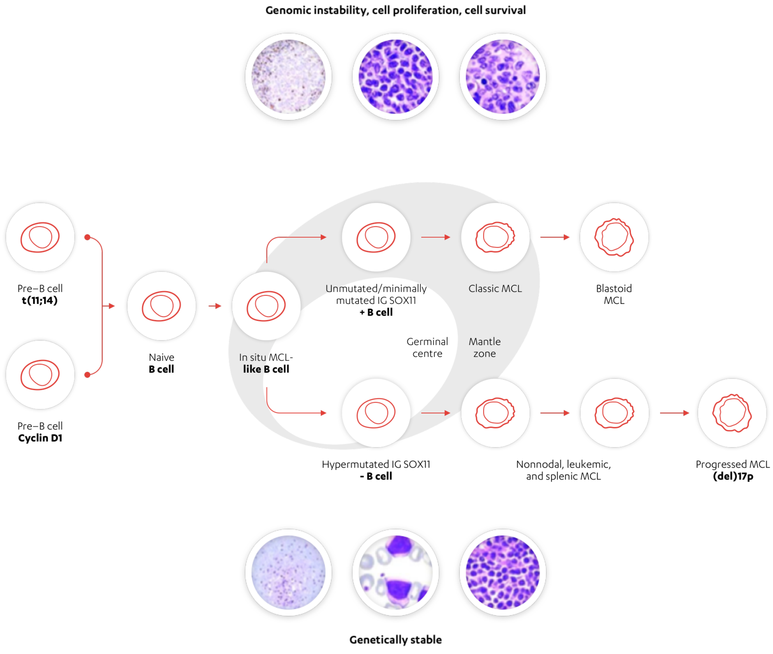
Welcome to I See Mantle Cell Lymphoma, a dedicated resource to help healthcare practitioners find the latest information about the signs, symptoms and diagnosis of MCL.

What is mantle cell lymphoma?

Mantle cell lymphoma (MCL) is a relatively rare, unique subcategory of B-cell non-Hodgkin lymphoma (NHL) with a generally aggressive clinical course.[1]

Originally, MCL was classified with other types of lymphomas using different histology-based classification systems.[2] MCL was only classified as a distinct subtype of B-cell NHL in the 1994 Revised European-American Lymphoma Classification (REAL). It was then confirmed in the World Health Organization (WHO) classification system.[3]

MCL develops from changes to the outer edge, or mantle, of lymph nodes, causing the development of abnormal B-cells from that region.[4] It can also affect the spleen, blood and bone marrow.[4] MCL encompasses a wide range of biological and clinical variants.[5] The majority of patients present with advanced stage disease and require systemic therapy.[5]



A proposed model of molecular pathogenesis and progression of MCL.[6]
Adapted from Dreyling *et al.* 2017.

Statistics surrounding MCL

3–10%

of non-Hodgkin lymphomas[1]

60–70 years

Median age at diagnosis[7]

3x more common in men[7]

MCL incidence rates

MCL has an annual incidence of one case per 200,000 people[7]
MCL represents approximately 5–7% of all malignant lymphomas in Western Europe[6]
MCL is typically sporadic, but it may have a higher incidence in some families[7]

Risk factors for MCL

The causes of MCL are mostly unknown; however, there are certain risk factors that are associated with the disease. These include[8]:

- infection with certain viruses or bacteria
- a weakened immune system
- autoimmune disease
- a previous cancer
- having a close blood relative with MCL

MCL immunophenotype

MCL is a well-characterised **B-cell lymphoma** with markers readily detectable by immunohistochemistry[9]

t(11;14)(q13;q32) translocation associated with overexpression of cyclin D1. Cyclin D1 overexpression is a key event in MCL pathogenesis[1]
Overexpression of SOX-11 is observed in a majority of patients with MCL[1]

The majority of **MCL cases also show expression of**[10]:

CD20
CD5
BCL2

The more aggressive **MCL subtypes,** such as **blastoid variants,** display features such as[1,4]:

High Ki-67 proliferation index
p53 mutations and p16 deletions

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References

[1] Jain P, Wang M. Mantle cell lymphoma: 2019 update on the diagnosis, pathogenesis, prognostication, and management. *Am J Hematol.* 2019;94:710–725.

[2] Harris N, *et al.* A revised European-American classification of lymphoid neoplasms: a proposal from the International Lymphoma Study Group. *Blood.* 1994;84(5):1361–1392.

[3] Campo E, Pileri SA. The classification of lymphoma: updating the WHO classification. In: Hoffbrand A, *et al*, eds. *Postgraduate Haematology*. Wiley-Blackwell; 2011:614–638.

[4] Vose J. Mantle cell lymphoma: 2017 update on diagnosis, risk-stratification, and clinical management. *Am J Hematol.* 2017;92:806–813.

[5] Kim L, *et al.* EHA Endorsement of ESMO clinical practice guidelines for newly diagnosed and relapsed mantle cell lymphoma. *HemaSphere.* 2020;4(5):464.

[6] Dreyling M, *et al.* Newly diagnosed and relapsed mantle cell lymphoma: ESMO Clinical Practice Guidelines for diagnosis, treatment and follow-up. *Ann Oncol.* 2017;28(S4):iv62–iv71.

[7] Lynch DT, Acharya U. Mantle cell lymphoma. In: StatPearls [Internet]. StatPearls Publishing; 2021 Jan. Updated 2020 August 12. Accessed May 2021. https://www.ncbi.nlm.nih.gov/books/NBK536985/

[8] Macmillan Cancer Support. Cancer information and support. Causes and risk factors of non-Hodgkin Lymphoma (NHL). Accessed May 2021. https://www.macmillan.org.uk/cancer-information-and-support/lymphoma/causes-and-risk-factors-of-non-hodgkin-lymphoma

[9] Aqil B, *et al.* Immunophenotypic variations in mantle cell lymphoma and their impact on clinical behavior and outcome. *Arch Pathol Lab Med.* 2018;142(10):1268–1274.

[10] McKay P, *et al.* A British Society for haematology good practice paper on the diagnosis and investigation of patients with mantle cell lymphoma. *Br J Haematol.* 2018;182:63–70.

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