

Marginal Zone Lymphoma

What is Marginal Zone Lymphoma?

Marginal zone lymphoma (MZL) is a group of **indolent non-Hodgkin's B-cell lymphomas** that begin in the marginal zones of lymph tissue.¹

Common symptoms include:



fever without infection



night sweats



unexplained weight loss



skin rash



chest or abdominal pain



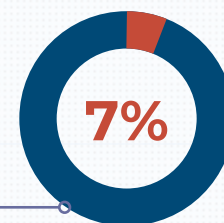
tiredness²

Who does MZL affect?



There are approximately **7,500** **newly diagnosed** patients in the United States every year.³

MZL is the **3rd most common** type of indolent non-Hodgkin's lymphoma, accounting for approximately **7% of all B-cell lymphomas**.^{3,4}



Subtypes of MZL

Marginal zone B-cells wait at the interface between lymphoid tissue and the circulation, so that they can quickly respond to antigens.⁵ There are **three subtypes of MZL**, and they each differ in **epidemiology, etiology, diagnosis, and treatment strategies**.

Extranodal MZL (EMZL) or Mucosa-Associated Lymphoid Tissue (MALT) is the most common form of MZL, accounting for approximately 60% of cases. MALT lymphoma occurs outside the lymph nodes and is further divided into two categories: gastric, which develops in the stomach and is the most common site, and non-gastric, which develops outside the stomach.⁶

Cause

EMZL forms when B-cells accumulate in the MALT organs in response to infections or autoimmune mechanisms.^{6,7}

Diagnosis

Lymphoepithelial lesions may signify EMZL, and if the presence of active H. pylori infection is not shown in immunohistochemistry, a serology test, urea breath test, and/or stool antigen test must be performed to rule it out.⁸

Treatment

Antibiotics are most commonly used in gastric MZL, whereas anti-infective therapy is commonly used in non-gastric extranodal MZL, and radiation therapy is commonly used in localized EMZL.⁸

Nodal MZL (NMZL) occurs in the lymph nodes and accounts for approximately 30% of cases.⁶

Cause

NMZL tends to be associated with hepatitis C virus infection and chronic inflammation.⁹

Diagnosis

NMZL shares many similarities with other types of MZLs which makes it difficult to diagnose unless monocytoid B-cell morphology is prominent. A small monoclonal component, usually IgM, may also signify this type of MZL.⁸

Treatment

In asymptomatic patients, it is typically recommended to “watch and wait” with frequent follow-up evaluation, although radiation therapy is suggested in patients presenting with localized disease.⁸

Splenic MZL (SMZL) occurs most often in the spleen and blood and accounts for approximately 10% of cases.⁶

Cause

SMZL is also associated with hepatitis C virus infection.¹⁰

Diagnosis

A combination of peripheral blood or bone marrow aspirate morphology and flow cytometry, as well as bone marrow biopsy histology and immunohistochemistry can be used to diagnose SMZL.⁸

Treatment

In asymptomatic patients, it is also common to “watch and wait” with frequent follow-up evaluation. However, in patients with chronic hepatitis C virus infection, antiviral therapy is considered.⁸

Note: Genetic mutations are seen in as many as 80% of splenic MZL cases; the most common are complete or partial trisomy of the q arm of chromosome 3, deletion or translocation of chromosome 7q32, and gains of the q arm of chromosome 12.¹¹

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