Mucosa-associated lymphoid tissue (MALT) lymphoma
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Identity

(t(11;18)(q21;q21) FISH - Courtesy Charles Bangs, Ilana Galperin.
Clinics and pathology

Disease
MALT lymphoma is the extra-nodal presentation of marginal zone B-cell lymphomas (MZBCL).

Phenotype / cell stem origin
The morphologic and phenotypic characteristics of malignant cells correspond to those of lymphocytes belonging to the marginal zone, harbouring hypermutated IgV genes with the following immunophenotype: pan-B+; CD5-/+; CD10-; CD23-; CD11c+/-; cyIg+ (40% of the cells); sIgM+ bright; sIgD-.

Epidemiology
The incidence of extra-nodal MZBCL of MALT type in western countries is approximately 7% of all NHL diagnosed by histologic examination.

Clinics
Extra-nodal MZBCL of MALT type is an indolent disease involving most often the stomach, where it usually follows chronic gastritis due to Helicobacter pylori (HP) infection. The disease may also localize in the lung, the thyroid the salivary gland and in the orbit, where an association was documented with Chlamydia Psittaci infection.

Pathology
The tumour consists of a cytologically heterogeneous infiltrate including centrocyte-like cells, monocytoïd B-cells small lymphocytes and plasma cells. Large cells and/or blast-like cells may be present. Typically, lymphoepithelial lesions are seen in the stomach.

Treatment
Low grade MALT with limited disease involving the stomach is usually HP+ and respond to eradication of the HP infection. Cases presenting at a more advanced stage or with transformation into high grade lymphoma require single-agent or multi-agent chemotherapy. Rituximab (anti-CD20 monoclonal antibody) is an effective treatment. Gastrectomy is indicated in non-responding patients.

Prognosis
The patients usually have prolonged survival, as in other indolent lymphomas, but some cases may feature an aggressive disease.

Cytogenetics

Cytogenetics molecular
The most common anomalies in extra-nodal MZBCL of MALT type include:

Results of the chromosomal anomaly

Fusion protein
Oncogenesis
MALI overexpression and API2-MALT fusion confer constitutional NFkB activity. This, in turn, leads to enhanced proliferation and resistance to apoptosis by B lymphocytes. BCL10 functions in conjunction with intracellular proteins (Carmal and MALT1), producing the ubiquitination of NFkB inhibitor, leading to NFkB activation. These findings, along with the documented role of BCL10 in promoting survival of antigen-stimulated lymphocytes, suggest the IgH/BCL10 translocation may contribute to lymphomagenesis by enhancing BCL10 function.

References


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