Waldenstrom's macroglobulinemia (WM)

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Clinics and pathology

Disease
Waldenstrom's macroglobulinemia (corresponding to lymphoplasmacytoid lymphoma/Immunocytoma in the REAL and WHO classification).

Phenotype/cell stem origin
Stem cell origin: Post-germinal centre IgM-bearing memory B-cell. Phenotype: CD19+; CD20+; CD22+; FMC7+; CD38+; cytoplasmic IgM bright+; CD5-; CD23-; CD10-.

Epidemiology
It accounts for 1-2% of all nodal lymphomas. The annual incidence falls in the 0.2-0.6% range per 100 000.

Clinics
Indolent lymphoma characterized by the secretion of a monoclonal IgM protein and by the expansion of the neoplastic clone in the bone marrow, in the lymphoid tissue and in extra-nodal sites. The symptoms derive from tissue infiltration, bone marrow failure and from the presence of the IgM paraprotein (hyperviscosity syndrome, polyneuropathies, AL amyloidosis).

Pathology
Diffuse proliferation of small lymphocytes, plasmacytoid lymphocytes and plasma cells, infiltrating the bone marrow and the interfollicular areas of the lymph nodes. By definition, this lymphoma subtype lacks the diagnostic features chronic lymphocytic leukemia (CLL), mantle cell lymphoma and follicle centre cell lymphoma. Transformation into high grade lymphoma may occur in a minority (5%) of cases.

Treatment
Alkylating agents (chlorambucil), multiagent chemotherapy (vincristine cyclophosphamide, steroid, with or without anthracyclines); fludarabine, with or without mitoxantrone.

Prognosis
Median survival exceeds 5 years and there is no plateau in the survival curve, just as in other low-grade lymphomas.

Cytogenetics

Cytogenetics morphological
There is no chromosome anomaly that is specific for WM. The overall cytogenetic picture is similar to that of marginal zone B-cell lymphoma. Recurrent structural changes include:
- the t(9;14)(p13;q32)
- the t(11;18)(q21;q21)
- a 6q-chromosome
Numerical aberrations involving several chromosomes were also detected. Recurrent aberrations include trisomy 1q, and trisomy 3q.

Genes involved and proteins

Note
The t(9;14)(p13;q32), fuses PAX5 with the IgH gene. PAX5 was shown to code for the BSAP (B-cell specific activator protein). The t(11;18)(q21;q21) is cytogenetically indistinguishable from the classical t(11;18) found in MALT lymphoma, where involvement of the API2- MLT genes was demonstrated.
References


This article should be referenced as such: