

Waldenstrom's macroglobulinemia (WM)

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	There is no chromosome anomaly that is specific for WM. The overall Morphological 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
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Genes involved and

Proteins

Note The t(9;14)(p13;q32), fuses [PAX5](#) with the [IqH](#) 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.

Bibliography

t(9,14)(p13;q32) denotes a subset of low-grade non Hodgkin's lymphoma with plasmacytoid differentiation.

Offitt KA, Parsa NZ, Filippa D, Jhanwar SC, Chaganti RSK.
Blood 1992; 80: 2594-2599.
Medline [1384792](#)

A revised European-American classification of lymphoid neoplasms: a proposal from the International Lymphoma Study Group.

Harris NL, Jaffe ES, Stein H, Banks PM, Chan JK, Cleary ML, Delsol G, De Wolf-Peeters C, Falini B, Gatter KC.
Blood 1994; 84: 1361-92.

World Health Organization classification of neoplastic diseases of the hematopoietic and lymphoid tissues: report of the Clinical Advisory Committee meeting-Airlie House, Virginia, November 1997.

Harris NL, Jaffe ES, Diebold J, Flandrin G, Muller-Hermelink HK, Vardiman J, Lister TA, Bloomfield CD.
J Clin Oncol 1999; 17: 3835-3849.
Medline [10577857](#)

Waldenstrom's macroglobulinemia. Clinical manifestations and prognosis.

Merlini G.
Education Program Book, American Society of Hematology, New Orleans, 3-7 December 1999, pp. 358-369.

Indolent lymphomas. Different entities and diagnostic problems.

Berger F.
Educational book. pp 1-6. Fifth Congress of the European Hematology Association, Birmingham UK, 25-28 June 2000.

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Citation

This paper should be referenced as such :

Cuneo A, Castoldi G . Waldenstrom's macroglobulinemia (WM). Atlas Genet Cytogenet Oncol Haematol. January 2002 .

URL : <http://www.infobiogen.fr/services/chromcancer/Anomalies/WaldensID2043.html>

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