**Mantle cell lymphoma**

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**Clinics and pathology**

**Disease**
NHL of the low to intermediate grade.

**Phenotype / cell stem origin**
B-cell lineage.

**Epidemiology**
5% of NHL; sex ratio: 3M/1F; median age: 65 yrs.

**Clinics**
Advanced disease (Ann Harbor stage III-IV) with generalized lymphadenopathies in 90%, bone marrow involvement (70%), frequent splenomegaly (50%), hepatomegaly (30%), and gastro-intestinal involvement (20%), and lymphocytosis (30%); elevated LDH in 50%.

**Pathology**
Small cleaved cell B-lymphocytes with inconspicuous nucleoli and pale cytoplasm; with a pan B-cell, CD5+, CD10−, CD23− (in contrast with CLL) phenotype; diffuse, nodular and blastoid types, the latter having large cells and higher mitotic count.
Treatment
According to the age and the disease stage.

Prognosis
Median survival is 3 to 4 yrs; less than 2 yrs in the case of a leukaemic form (blastoid); in contrast with the good prognosis of the MALT lymphoma, a closely related disease.

Cytogenetics

Cytogenetics, morphological
Knowledge is still scarce, and data complex. t(11;14)(q13;q32) is found in 50-70% of cases, but this translocation may also, at a much lesser frequency, be found in other diseases; the genes involved in this translocation are described below; t(11;14) is found in complex karyotypes. Other frequent findings: deletion of parts of: 1p, 6q, 9p, 11q (in particular 11q22-23), 13q; gains of parts of: 3q, 8q, 15q; frequent markers; tetraploidy may be found, especially in the blastoid subtype.

Cytogenetics, molecular
CGH (comparative genomic hybridization) may disclose important events in this disease, taken into account that the t(11;14) may be insufficient for oncogenesis; furthermore, this and other molecular cytogenetic techniques may come in addition to the usual techniques to ‘dissect’ complex karyotypes.

Genes involved and Proteins

BCL1
Location: 11q13
DNA / RNA
5 exons.
Protein
Encodes the cyclin D1; role in the cell cycle control: G1 progression and G1/S transition.

IgH
Location: 14q32

Results of the chromosomal anomaly

Hybrid gene
Description
5′ BCL1 translocated on chromosome 14 near JH (junctions genes of IgH) and C in 3′; the breakpoint in BCL1 is in MTC (major translocation cluster), centromeric to the gene (in 5′), in 80% of cases, or dispersed in mTC1, 2, or 3 in 5′ of the gene or in the 3′ untranslated region of exon 5.

Fusion protein
Description
No fusion protein, but promoter exchange; the immunoglobulin gene enhancer stimulates the expression of BCL1.

Oncogenesis
Overexpression of BCL1 accelerates passage through the G1 phase.

References

This article should be referenced as such: