Chronic lymphocytic leukaemia (CLL)
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Clinics and pathology

Disease
Chronic lymphoproliferation

Phenotype / cell stem origin
B-cell disease; the existence of rare cases of T-CLL has been debated.

Epidemiology
Annual incidence 30/10^6; represents 70% of lymphoid leukaemias, 1/4 of all leukaemias; median age: 60-80 yrs, 2M/1F.

Clinics
Diagnosis is often delayed, due to the lack of symptoms (therefore, median survival from the beginning of the disease may be much more than median survival from diagnosis); enlarged lymph nodes; splenomegaly; blood data: lymphocytosis > 4 X 10^9/l; hypogammaglobulinemia in 60%.

Cytology
Typically, proliferation of mature small lymphocytes of normal morphology; lymphocytes with more abundant cytoplasm can be present; prolymphocytes must represent less than 10% of the lymphocytes (otherwise, the diagnosis of ‘chronic lymphocytic leukaemia-prolymphocytic leukaemia’ should be made); expression of sIg with monotypy (monoclonality); CD19+, CD20+, and CD5+ most often.

Treatment
None in early stage; chemotherapy afterwards.

Evolution
Unrelated causes and disease-related infections are the 2 major causes of death.

Other: autoimmune hemolytic anaemia and thrombocytopenia; transformation into Richter's disease or into prolymphocytic leukaemia (in 10%).

Prognosis
According to the staging: A (less than 3 lymph nodes, Hb < 10g/dl, platelets < 100 X 10^9/l): survival not reduced compared to age matched population; B (3 or more lymph nodes; Hb and platelets maintained): median survival of 5 yrs; C (Hb < 10g/dl and/or platelets < 100 X 10^9/l): median survival of 2 yrs; according to the karyotype: survival is better in cases with a normal karyotype (median: 15 yrs vs 8 yrs with an abnormal karyotype), worse in the 10% of cases where a complex karyotype is found (median: 6 yrs); specific chromosome anomalies have specific prognoses (see below).

Cytogenetics

Cytogenetics, morphological
Clonal anomaly is found in about 50% of cases; complex karyotypes are found in 10%; unrelated clones demonstrating the existence of cells subpopulations are frequent findings in this disease.

+12: is found in 15-20% of cases, depending on the use of interphase cytogenetics methods (FISH) and the cell morphology of the cases under study (trisomy 12 is typically found in atypical lymphocyte morphology and CD5- cases, often with an increased number of prolymphocytes, in advanced stages, and is associated with disease progression); trisomy 12 is an adverse prognostic factor (median survival: 5 yrs); found either as the sole anomaly, as an anomaly accompanied by others, or even as an accompanying (secondary) anomaly; present only in a subset of the malignant cell population; region q13-q22 might be of particular pathogenetic importance.
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**del(13q) and t(13;Var):** found in 10-20% of cases; q14 and Rb gene and also DNA sequences telomeric and centromeric to Rb are often involved; deletion may be hetero- or homozygous; good prognostic feature (median survival > 15 yrs);

**14q32 involvement:** is frequent in CLL, as in other B-cell chronic leukaemias or lymphomas; t(11;14)(q13;q32), typical of mantle cell lymphoma, with BCL1/IgH rearrangement, may occasionally be found in CLL;

t(14;19)(q32;q13), with BCL3/IgH rearrangement, may be associated with short survival;

Other t(14; var) have been found;

de(6q), del(11q), +3, +18: are the most frequent other anomalies.

**Genes involved and Proteins**

**Note:** Genes involved as a primary event are still unknown. P53 has been found mutated in 10-15% of cases; adverse prognostic indicator.

**References**


**This article should be referenced as such:**