Leukaemia Section
Mini Review

T-cell prolymphocytic leukemia (T-PLL)
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Clinics and pathology

Disease
Chronic T-cell lymphoproliferative syndrome

Phenotype / cell stem origin
Disease affecting mature T-cells.
T-cell prolymphocytes usually express CD3, CD5 and CD7; they have either a T-helper (CD4+/CD8-) or a T-suppressor (CD4-/CD8+) phenotype; a small number of cases may co-express CD4 and CD8; this finding is more prevalent in the small cell variant of T-PLL than in classic T-PLL.

Epidemiology
Very rare disease; represents 20% of prolymphocytic leukemias; the disease occurs at advanced age, typically in the 7th or 8th decade; slight male predominance.

Clinics
Splenomegaly is common; lymphadenopathy at presentation is unusual but more frequent than in B-PLL; blood data: high leucocyte counts usually exceeding 100 x 10^9/l; T-cell prolymphocytes have the same morphologic features than B-cell prolymphocytes; a small cell variant of T-PLL has been described.

Prognosis
Evolution: progresses rapidly and is generally more aggressive than B-PLL; prognosis: poor response to chemotherapy is observed; median survival is approximatively 7 months from diagnosis.

Cytogenetics

Cytogenetics, morphological
Few cases have been reported in the literature so far; karyotypes are usually complex.
-14q11 abnormalities: very frequent, either as an inv(14)(q11q32) or as a translocation t(14;14)(q11;q32); another reported change involving 14q11 is a translocation t(X;14)(q28;q11), similar to the translocation observed in ataxia-telangectasia, involving the Mature T-cell Prolymphocyte 1 (MTCP1) gene located at Xq28.
-Other recurrent changes involve chromosome 8 either as i(8)(q10) or as der(8) t(8;8).
-Finally, some aberrations involving 12p have been reported.

Genes involved and Proteins

Note: as with other T-cell neoplasms, T-PLL exhibits clonal rearrangement of T-cell receptor genes; translocation t(X;14)(q28;q11) may result into fusion of MTCP1 with TCR-δ genes; finally, the TCL1 locus on chromosome 14q32 might also been involved; biallelic mutation in ATM can occur.

References


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