Classification of T-Cell disorders

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Identity

Note
T-cell lymphoid disorders include a variety of disease entities which result from the clonal neoplastic expansion of an uncommitted (thymic) or a committed (post thymic) T-cell. Some of these diseases have distinct cytogenetic/molecular genetic features which allow to better define the various entities and understand their pathogenesis.

Clinics and pathology

Disease
T-prolymphocytic leukemia (T-PLL)
Variants: small cell and cerebriform cell.

Phenotype/cell stem origin
TdT-, CD1a-, CD4+ CD8-, CD4- CD8+, CD4+ CD8+.

Clinics
Aggressive course
Splenomegaly, high WBC with prolymphocytes.

Cytogenetics
inv(14)(q11q32), t(14;14)(q11;q32)
Xq28 abnormalities
idic(8)(p11), t(8;8)(p11;q1-2)
11q22-23 abnormalities
12p abnormalities
13q14.3 deletions

Genes
ATM gene (11q22-23) mutated. TCL1 (14q32.1) or MTCP1 (Xq28) activated.

Disease
Large granular lymphocyte leukemia (LGL) - T-cell Type.

Phenotype/cell stem origin
TdT-, CD1a, CD3+, CD2+, CD8+ CD4-, CD16+/Cytotoxic or suppressor activity.

Clinics
Indolent
Cytopenias, splenomegaly, lymphocytosis with granular lymphocytes.

Cytogenetics
Clonal abnormalities. In some cases, but no consistent specific abnormalities.

Genes
Clonality established by TCR rearrangements.

Disease
Large granular lymphocyte leukemia (LGL) - NK type.

Phenotype/cell stem origin
TdT-, CD1a, CD2+, CD56+, CD16+, CD7+/CD3-, CD5-, TCR-Natural killer Activity.

Clinics
Aggressive or indolent
Lymphocytosis, splenomegaly, hepatomegaly.

Cytogenetics
del(6)(q21-25).

Genes
TCR chain genes in germ line.

Disease
Sezary syndrome (SS).

Phenotype/cell stem origin
TdT-, CD1a-, CD3+, CD4+, CD8-, Helper or no functional activity.
Clinics
Variable clinical course with skin involvement and cells with cerebriform nuclei.

Cytogenetics
Complex, clonal, oligoclonal or nonclonal with variable ploidy. Abnormal.2p, Abnormal.6q, i(17q), del(13)(q14)

Genes
P53 gene deletion and protein expression in the absence of gene mutation. Few cases express MDM2.

Disease
Adult T-cell leukemia lymphoma (ATLL).

Phenotype/cell stem origin
TdT-, CD1a-, CD7- CD4+ CD8- CD25+, Suppressor activity.

Clinics
Aggressive, Hypercalcaemia, lymphadenopathy, Eflower cells', HTLV-1 Positive.

Cytogenetics
Complex and often oligoclonal. Numerical abnormalities: 3, 7, X. Structural abnormalities: 1q, 3q, 6q, 14q.

Genes
Oligoclonal/mono clonal integration of HTLV-1 in host DNA. Abnormalities of p53, p16 and p15 genes.

Disease
T-NHL hepatosplenic lymphoma.

Phenotype/cell stem origin
TdT-, CD1a-, CD3+/- CD56+, CD7+, granzyme A+, TCR g/d+

Clinics
Aggressive, Hepato splenomegaly

Cytogenetics
Abnormal.7q, i(7q)

Genes
TCR genes gamma/delta rearranged but alpha/beta not rearranged.

Disease
Peripheral/post-thymic T-cell lymphoma (pleomorphic and immunoblastic subtypes).

Phenotype/cell stem origin
TdT-, CD1a-, Variable expression of CD4 or CD8.

Clinics
Aggressive; advanced stages.

Cytogenetics
Variable.

Disease
Angio immunoblastic T-cell lymphoma.

Phenotype/cell stem origin
TdT-, CD1a-, CD2+, CD5+, CD3+ CD4+ CD8-.

Clinics
Disproteinemia, lymphadenopathy, immune abnormalities.

Cytogenetics
Complex with multiple related or unrelated clones. +3 or i(3q), +5, del(6q). Progression from normal karyotype to abnormal clone observed during transition from hyperplasia to neoplasia.

Genes
Integrated EBV sequences present in both B-and T-cells and is unlikely to be the etiological agent.

Disease
Angiocentric (nasal) T-cell lymphoma.

Phenotype/cell stem origin
TdT-, CD1a-, T-cell or NK phenotype.

Clinics
Prevalent in Asia and south America; extra nodal involvement.

Cytogenetics
i(1q), del(6q), i(6p)

Genes
Majority have no TCR rearrangement; EBV clonally integrated and plays a role in the etiology of the disease.

Disease
Anaplastic (Ki 1+) large cell lymphoma.

Phenotype/cell stem origin
TdT-, CD1a-, CD3+/ CD30+ (Ki 1+), CD15-, CD25+, HLA-Dr+, CD71+.

Clinics
Aggressive with skin nodes and extranodal involvement.

Cytogenetics
t(2;5)(p23;q35)

Genes
Fusion gene NPM-ALK; 2p23 -Nucleolar phosphoprotein NPM; 5q35 -Anaplastic lymphoma kinase- ALK.
**Disease**

Intestinal T-cell lymphoma.

**Phenotype/cell stem origin**

TdT, CD1a−, CD3+, CD8+, CD103+, CD4−, CD8−.

**Clinics**

Bone pain, coeliac disease, mesenteric nodes.

**Genes**

EBV genome present in mexican population but not in the europeans.

**Disease**

T-lymphoblastic Lymphoma/leukaemia (T-Lbly/T-ALL).

**Phenotype/cell stem origin**

TDT+, CD1a+, CD7+, cytCD3+ or +/-, other T-cell antigens. Thymic uncommitted T-cell.

**Clinics**

Aggressive; course similar to ALL. Mediastinal mass, high WBC.

**Cytogenetics**

del(6)(q21-q22)
t(11;14)(p13;q11)
t(1;14)(p34;q11); 1p34: tal-1gene; 14q11: TCR alpha.

**Genes**

TCR chain genes rearranged.

**References**


This article should be referenced as such: