Leukaemia Section

Short Communication

\(t(11;14)(p13;q11), t(7;11)(q35;p13)\)

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Identity

Note
This \(t(11;14)\) must be not confused with the \(t(11;14)(p15;q11)\) associated with an immature immunophenotype (CD3-, CD4-, CD8-) and involving respectively RTBN1 gene and TRD locus.

\(t(11;14)(p13;q11)\) G- banding (left) and R- banding (right) - Courtesy Jean-Luc Lai and Alain Vanderhaegen.

Clinics and pathology

Disease
T-ALL

Epidemiology
5-10\% of childhood T-ALL.

Cytogenetics

Additional anomalies
+17; to be noted that a Ph chromosome (m-BCR) has been found in one case of T-ALL.

Variants
\(t(11;14)(p13;q11)\) and \(t(7;11)(q35;p13)\) are variant translocations of each other.

Genes involved and proteins

\textbf{RBTN2}

Location
11p13

Protein
Cystein-rich protein with two tandemly arranged zinc binding LIM-domain motifs: named Lom2; Lmo2 directly interacts with the basic-loop-helix protein Tal1/Scl and the GATA DNA protein Gata-1; central role in adult hematopoietic pathway regulation.

\textbf{TRA/D or TRB}

Location
14q11 and 7q35 respectively.

Result of the chromosomal anomaly

\textbf{Hybrid gene}

Description
Chromosomal breakpoints occur 25 kb upstream RBTN2 gene, in a presumed transcriptional start site, inducing truncation of the promoter/control region and leading to inappropriate Lmo2 level especially in T-cells (abnormal T-cell differentiation).

\textbf{Fusion protein}

Oncogenesis
Lmo2 is activated after chromosomal translocation by association with either the TRA/D or the TRB.
To be noted

Note
CELL LINE with t(11;14)(p13;q11): KOPT-K1; the breakpoints occur: - on chromosome 11 in an Alu-rich region, between two Alu sequences, 160 kb-closed from RTBN2; - on chromosome 14 within Jd1; RTBN2 is highly expressed in KOPT-K1.

References
Wadman IA, Osada H, Grütz GG, Agulnick AD, Westphal H, Forster A, Rabbitts TH. The LIM-only protein Lmo2 is a bridging molecule assembling an erythroid DNA-binding complex which includes the TAL1, E47, GATA-1 and Ldb1/LNI proteins. EMBO J. 1997 Jun 2;16(11):3145-57

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