

Leukaemia Section

Short Communication

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

Chrystèle Bilhou-Nabera

Laboratoire d'Hématologie, Hôpital du Haut-Lévêque, CHU de Bordeaux, Avenue de Magellan, 33604 Pessac, France (CBN)

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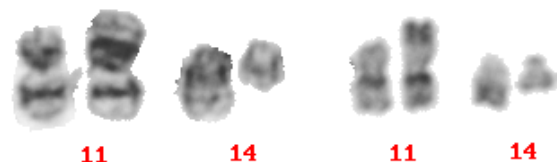
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Identity

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

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.

TRA/D or TRB

Location

14q11 and 7q35 respectively.

Result of the chromosomal anomaly

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).

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.

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