Leukaemia Section
Mini Review

t(3;14)(q27;q32)
t(2;3)(p12;q27)
t(3;22)(q27;q11)

Christian Bastard
Department of Hematology, Centre Henri Becquerel, Rouen, France

Published in Atlas Database: April 1998
DOI: 10.4267/2042/37454

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Identity

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Disease
NHL; mainly diffuse large cell lymphoma (DLCL); high grade lymphoma; rare cases with low grade/follicular lymphoma; in those cases, 3q rearrangements can be isolated or associated with a t(14;18) translocation.

Phenotype / cell stem origin
B-cell.

Epidemiology
3q27 rearrangements present in 25 to 30% of DLCL; about 10% of follicular lymphoma.

Clinics
Mainly adult aggressive lymphoma.

Treatment
Intensive chemotherapy.

Prognosis
Controversial.

Cytogenetics

Cytogenetics, morphological
(t(2;3) and t(3;22) are easily identified; t(3;14) is telomeric and difficult to see, especially when using G-banding.

Cytogenetics, molecular
Some cosmid and YAC probes have been developed.

Additional anomalies
Very frequent, as usual in NHL, involving chromosome 1, del(6q), 10q, 11q, 12q, 13q, 17p arms.

Variants
Large number of recurring defects involving the 3q27 region:
t(3;4)(q27;p13), t(3;6)(q27;p22), t(3;7)(q27;p13),
t(3;8)(q27;q24), t(3;11)(q27;q23), t(3;13)(q27;q14),
t(3;15)(q27;q22), t(3;17)(q27;q11).

Genes involved and Proteins

Note: BCL6 (see below) with various partners: immunoglobulin genes IgH (mainly Sµ region) in 14q32, IgK in 2p12, and IgL in 22q11; (for variant translocations, partners are: RHOH in the t(3;4), histone H1F1 in the t(3;6), OBFI in the t(3;11), LCP1 in the t(3;13); in these cases, the first non-coding exon of the partner gene fuses with the second exon of BCL6, resulting in the deregulated expression of a chimerical transcript and the production of a normal BCL6 protein).

BCL6
Location: 3q27

DNA / RNA
10 exons; alternative splicing of exons 1 (1a and 1b), without modification of the open reading frame.

Protein
Transcription factor; belongs to the Krüppel family, with a N-term BTB/POZ domain and 6 zinc fingers; transcription repressor.

Results of the chromosomal anomaly

Fusion protein
Description
No fusion protein, but promoter exchange; the immunoglobulin gene promoter controls the expression of the BCL6 gene.

Oncogenesis
Deregulated protein expression; alternative deregulation mechanisms: deletion of regulating sequences, mutations.

References

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This article should be referenced as such:
Bastard C. t(3;14)(q27;q32); t(2;3)(p12;q27); t(3;22)(q27;q11). Atlas Genet Cytogenet Oncol Haematol.1998;2(4):122-124.