Leukaemia Section
Short Communication

\textbf{t(8;9)(q24;p13) /MYC}

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\textbf{Abstract}

Review on \textit{t(8;9)(q24;p13) /MYC}, with data on clinics, and the genes implicated.

\textbf{Clinics and pathology}

\textit{Disease}

B-cell acute lymphoblastic leukemia (ALL) and non-Hodgkin lymphoma (NHL)

\textit{Phenotype/cell stem origin}

There were 5 cases of ALL (L3-ALLs and CD10+ ALLs in 2 cases each), 11 cases of diffuse large B-cell lymphoma (DLBCL), 7 cases of unclassifiable B-cell lymphoma, one case of follicular lymphoma, and one case of diffuse mixed small and large cell lymphoma (Levine et al., 1989; Nacheva et al., 1993; Dunphy et al., 2003; D'Achille et al., 2006; Bertrand et al., 2007; Le Gouill et al., 2007; Christie et al., 2008; Johnson et al., 2009; Subramaniyam et al., 2011).

\textit{Epidemiology}

Twenty five cases are available.

There were 12 male and 13 female patients, median age was 63 years (range 42-90 years).

In a review of NHLs with a t(14;18)(q32;q21), the incidence of MYC translocations was 5%, and the incidence of t(8;9)(q24;p13) was 1% (Johansson et al., 1995).

\textit{Prognosis}

Median survival was 4 months (see figure). However, patients with an ALL died at 1, 1, 1.5, 1.5 months.

Median survival of patients with NHL was 6 months, and 3 out of 19 NHL patients were still alive 5 years after diagnosis (16%).

\textbf{Cytogenetics}

\textit{Cytogenetics morphological}

A t(14;18)(q32;q21) was present in 24 of 25 cases, including 5 of the 5 cases of ALL, a complex karyotype was found in 24 of 25 cases.

Trisomy 7 and 12 were found in seven cases each, trisomy 11 and 21, and del(6q) in four cases each, trisomy 20 in three cases, trisomy 8, 13, and 18, t(1:22)(q21;q11), del(3p), and del(3q) in two cases each.

\textit{Genes involved and proteins}

Breakpoints occurred close to MYC on chromosome 8 and close to: ZBTB5, ZCCHC7, and PAX5 on chromosome 9 (Bertrand et al., 2007; Le Gouill et al., 2007; Johnson et al., 2009).

- ZBTB5 is a POZ domain Krüppel-like zinc finger transcription repressor. ZBTB5 interacted with co-repressor-histone deacetylase complexes such as BCOR, NCO1, and NCO2 (SMRT), resulting in deacetylation of histones Ac-H3 and Ac-H4, and transcriptional repression of CDKN1A (p21) (Koh et al., 2009).

- ZCCHC7 is the equivalent of the yeast AIR1. In yeast, a number of unwanted RNA transcripts are first recognized by the nuclear exosome cofactor Trf4/5p-Air1/2p-Mtr4p polyadenylation (TRAMP) complex before subsequent nuclear-exosome-mediated degradation.
TRAMP facilitates pre-mRNA splicing (Kong et al., 2013).

- PAX5 is a lineage-specific transcription factor; recognizes the consensus recognition sequence GNCCANTGAAGCGTGAC, where N is any nucleotide. Involved in B-cell differentiation.

Entry of common lymphoid progenitors into the B cell lineage depends on E2A, EBF1, and PAX5; activates B-cell specific genes and represses genes involved in other lineage commitments.

Activates the surface cell receptor CD19 and represses FLT3.

PAX5 physically interacts with the RAG1/RAG2 complex, and removes the inhibitory signal of the lysine-9-methylated histone H3, and induces V-to-DJ rearrangements.

Genes repressed by PAX5 expression in early B cells are restored in their function in mature B cells and plasma cells, and PAX5 repressed (Fuxa et al., 2004; Johnson et al., 2004; Zhang et al., 2006; Cobaleda et al., 2007).

**MYC**

**Location**

8q24.2

**Protein**

DNA binding protein.

Binds DNA as a heterodimer with MAX.

Involved in various cellular processes including cell growth, proliferation, cell adhesion, apoptosis, angiogenesis, and stem cell behaviour modulation.

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**Result of the chromosomal anomaly**

**Fusion protein**

**Expression / Localisation**

There was abundant MYC expression in all the cases studied.

**References**


This article should be referenced as such: