Leukaemia Section
Short Communication

**t(2;3)(p16;q26) BCL11A/MECOM**

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Published in Atlas Database: September 2013

Online updated version: http://AtlasGeneticsOncology.org/Anomalies/t0203p16q26ID1664.html

DOI: 10.4267/2042/53539

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**Abstract**

Review on t(2;3)(p16;q26) BCL11A/MECOM, with data on clinics, and the genes implicated.

**Identity**

Note

This translocation is found in a subset of cases described in the card t(2;3)(p15-23;q26-27).

Other subsets involve other genes, such as THADA in the t(2;3)(p21;q26) THADA/MECOM.

**Clinics and pathology**

**Disease**

Acute myeloid leukemia (AML)

**Phenotype/cell stem origin**

There were one M1-AML, two M2-AMLs, and one M5-AML; patients presented with dysplasia of at least two myeloid cell lineages.

**Epidemiology**

Four cases to date; there were three male and one female patients; patients were aged 36, 36, 36, and 55 (in years) (Trubia et al., 2006).

**Prognosis**

Clinical outcome in cases with the t(2;3)(p16;q26) BCL11A/MECOM and the case with the t(2;3)(p21;q26) THADA/MECOM (plotted together) was severe: "One patient is alive with active disease at 12 months, five patients died after 4-14 months" (Trubia et al., 2006).

**Genetics**

Note

MECOM was overexpressed.

**Cytogenetics**

**Cytogenetics morphological**

The t(2;3)(p16;q26) was the sole anomaly in two of four cases (at least in a subclone), accompanied with -7 in one case, and +14 in another case.

**Genes involved and proteins**

**BCL11A**

**Location**

2p16

**Protein**

BCL11A is a Krüppel zinc-finger transcription factor, which has been shown to be essential for pre-B-cell development, thymocyte maturation, and globin switching, expressed in haematopoietic and neural tissues. BCL11A controls FLT3 and IL7R expression in early hematopoietic progenitors (Wu et al., 2013).

**MECOM**

**Location**

3q26

Note

MECOM is also known as EVI1 or PRDM3; MECOM symbol means: "MDS1 and EVI1 complex locus".
Protein
"EVI1" contains two domains of seven and three zinc finger motifs, respectively, a repression domain between the two sets of zinc fingers, and an acidic domain at its C-term. Sequence specific DNA binding protein. Interacts with transcriptional coactivators, corepressors, and other sequence specific transcription factors. MECOM ("MDS1-EVI1") also contains a PR domain from "MDS1" in N-term (Wieser, 2008).

Result of the chromosomal anomaly

Hybrid gene
Description
Regulatory elements were transferred at the 5' of MECOM.

Fusion protein
Description
The t(2;3) brings about the juxtaposition at 3q26 of the MECOM locus with regulatory elements normally located in proximity of the 2p breakpoints, with consequent EVI1 overexpression, without the formation of a fusion protein.

References


This article should be referenced as such: