

t(4;11)(q21;p15)

Identity

Note not to be confused with a variant of the classical [t\(4;11\)\(q21;q23\)](#) translocation

Clinics and Pathology

Disease T-cell acute lymphoblastic leukemia

Phenotype / cell stem origin lymphoblasts, either L1 or L2 in the FAB classification; mature T and myeloid markers variably co-expressed

Epidemiology rare, approximately 10 cases described ; evaluated to 2-5% of adult T-ALL ; not evaluated in childhood ALL; both sexes equally involved; found in children or young adults

Clinics no notable particular aspect

Prognosis probably unfavorable, median survival below 18 months; improved by allogeneic bone marrow transplantation

Cytogenetics

Cytogenetics Morphological in approximately 2/3 of cases; 2 cases with [12p-](#)

Cytogenetics Molecular two BAC clones 290A12 and 118H17 (California Institute of Technology BAC library) encompasses all NUP98 gene and are split by translocation

Variants not described

Genes involved and Proteins

Gene Name [RAP1GDS1](#)

Location 4q22.3

Protein smgGDS, 558 amino acids; stimulates GDP --> GTP transition in a series of small GTP-binding proteins (g proteins) including [rap1a](#), [rap1b](#), [K-ras](#), rac1, rac2, rhoA and ralB.

Somatic mutations not involved in other known clonal rearrangement associated with tumoral proliferation.

Gene Name [NUP98](#)

Location 11p15.4

Protein nucleoporin 98, a 98 kDa component of the nuclear pore complex implicated in nucleo-cytoplasmic transport

Somatic mutations involved in different types of acute myeloid leukemia, as fusion gene with [HOX A9](#), [DDX10](#), HOX D13, [TOP1](#), PMX1 and LEDGF, resulting respectively from [t\(7;11\)\(p15;p15\)](#), inv(11)(p15q22), t(2;11)(q31;p15), [t\(11;20\)\(p15;q11\)](#), t(1;11)(q23;p15) and t(9;11)(p22;p15)

Result of the chromosomal anomaly

Hybrid gene NUP98 breakpoint in the intron between exons B and C; 5'-part of NUP98 is fused in frame with the whole coding sequence of

Description RAP1GDS1; fusion gene called NRG: 5'-NUP-RAP1GDS1-3'. Variant described with breakpoint in NUP98 before exon A.

Fusion Protein t(4 ;11) generates only one chimeric protein 5' NUP98 - RAP1GDS1 - 3' which contains a variable part of NUP98 and the totality of smgGDS

Description except for the initial methionine

External links

Other database [t\(4;11\)\(q21;p15\)](#) [Mitelman database \(CGAP - NCBI\)](#)

Other database [t\(4;11\)\(q21;p15\)](#) [CancerChromosomes \(NCBI\)](#)

To be noted

Additional cases are needed to delineate the epidemiology of this rare entity:

you are welcome to submit a paper to our new [Case Report section](#).

Bibliography

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Molecular evaluation of the NUP98/RAP1GDS1 gene frequency in adults with T-acute lymphoblastic leukemia.

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Citation

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<http://www.infobiogen.fr/services/chromcancer/Anomalies/t0411q21p15ID1191.html>

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