t(10;11)(q22;q23)
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Disease
Acute myeloid leukemia (AML)

Phenotype/cell stem origin
Described in subtypes AML-M2, -M4 and -M5. Cell lineage dysplasia may be associated.

Epidemiology
Less than 10 cases described mainly adults and one child.

Prognosis
Undetermined, possibly intermediate.

Cytogenetics

Cytogenetics morphological
Easy to detect, evident 10q- and 11q+ derivatives.

Cytogenetics molecular
Commercial dual color MLL FISH probes are splitted by the translocation. 10q22 breakpoint may be detected with RP11-119F7 BAC probe.

Additional anomalies
Sole anomaly in half published cases, complex karyotype in others.

Genes involved and proteins

MLL
Location
11q23

Note
MLL (mixed-lineage leukemia or myeloid-lymphoid leukaemia) is also called ALL-1 or HRX.

DNA/RNA
36 exons, multiple transcripts 13-15 kb.

Protein
430 kDa, contains two DNA binding motifs (a AT hook and a CXXC domain), a DNA methyl transferase motif, a bromodomain; transcriptional regulatory factor involved in maintenance of Hox gene expression during embryogenesis and during the process of haematopoietic progenitors expansion and differentiation.

CXXC6
Location
10q22

Note
CXXC6 (CXXC finger 6) is also called LCX (leukemia-associated protein with a CXXC domain) or TET1.

DNA/RNA
8497 bp representing the whole coding sequence. At least 12 exons. Contains 3 bipartite nuclear localization sites, 1 alpha helice coiled-coil region and 1 cysteine rich domain with high level homology with a CXXC DNA binding site.

Protein
Predicted size of 2136 amino acids, expression restricted to some fetal tissues, mainly lung, heart and brain; not expressed in hematopoietic tissues, except in spleen; unknown function.
Result of the chromosomal anomaly

Hybrid gene

Description
Breakpoint within MLL intron 6 and LCX intron 8; MLL exon 8 is fused in frame with LCX exon 9 and transcripts from the 5' MLL-LCX 3' fusion gene on der(11) are expressed; transcripts from the 5' LCX-MLL3' counterpart are not detected.

Fusion protein

Description
Predicted molecular weight of 204.4 kDa.

Oncogenesis
Unknown; the alpha helice coiled-coil region retained at the COOH extremity might be involved in the leukemogenesis.

References

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