**t(X;10)(p11;p12) DDX3X/MLLT10**

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**Abstract**

Review on t(X;10)(p11;p12), with data on clinics and the genes involved.

**Keywords**

chromosome X; chromosome 10; acute lymphoblastic leukemia; T-cell; DDX3X; MLLT10

**Clinics and pathology**

**Disease**

T-Acute lymphoblastic leukemia/lymphoma

**Phenotype/cell stem origin**

In two cases T-ALL was arrested at cortical stage. In the other 3 patients, immunophenotype was incomplete or not available [Brandimarte et al, 2013, 2014].

**Epidemiology**

t(X;10)(p11;p12) DDX3X/MLLT10 occurs in approximately 3% of adult T-ALL and characterizes a subgroup of NOTCH1 positive leukemias.

**All five T-ALL patients were males aged 11 - 38 years (median 24.4 years). Treatment**

All patients achieved hematologic remission but 3 relapsed and died. Two patients are alive, one of them was treated by HLA identical Hematopoietic stem cell transplantation (HSCT).

**Cytogenetics**

**Additional anomalies**

see above; notably: del(9p) in 4 cases, del(5q) and del(6q) in 1 case.

**Genes involved and proteins**

**DDX3X (DEAD-box helicase 3, X-linked)**

Location Xp11.4

**Protein**

DDX3X is a member of the large family of RNA helicases with a DEAD box domain that is involved in RNA transcription, splicing, mRNA transport, translation initiation, and cell-cycle regulation [Rosner et al, 2007].

**Somatic mutations**

DDX3X somatic mutations have recently been discovered in medulloblastoma, chronic lymphocytic leukemia, and Burkitt lymphoma. Recurrent DDX3X homozygous deletions were identified in gingivobuccal oral squamous cell carcinoma [Brandimarte L., et al, 2013].

**MLLT10 (myeloid/lymphoid or mixed-lineage leukemia (trithorax homolog, Drosophila); translocated to, 10)**

Location 10p12.31
**Protein**

MLLT10 contains 3 Zn fingers and a leucine zipper; nuclear localisation; transcription factor [Morerio and Panarello, 2005] 

Result of the chromosomal anomaly

**Fusion protein**

**Oncogenesis**

DDX3X is one of the genes that escapes X-inactivation in females [Lahn B.T. et al., 1997]. As all patients with DDX3X-MLLT10 positive T-ALL were males, no wild-type DDX3X allele was retained in the leukemic blasts, suggesting that the complete absence of a normally functional DDX3X protein might contribute to leukemogenesis. DDX3X appeared to have oncogenic as well as tumor suppressor functions [Change P.C. et al., 2006; Botlagunta M. et al., 2008; Brandimarte L., et al, 2014].

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### References


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This article should be referenced as such: