

Leukaemia Section

Short Communication

t(1;22)(p36;q11) IGL/PRDM16

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Abstract

Review on t(1;22)(p36;q11) IGL/PRDM16 translocations, with data on clinics, and the genes involved.

Keywords

chromosome 1; chromosome22; t(1;22)(p36;q11); IGL; PRDM16

Clinics and pathology

Disease

Splenic marginal zone B-cell lymphoma.

Clinics

Only one case to date: a 68-years old male patient, who died 38 months after diagnosis (Duhoux et al., 2012).

Cytogenetics

Cytogenetics morphological

Accompanying abnormalities were: +12, +18 and t(1;14)(p12;q32).

Genes involved and proteins

PRDM16 (PR domain containing 16)

Location

1p36.32

DNA/RNA

11 splice variants

Protein

1276 amino acids and smaller proteins. Contains a N-term PR domain; 7 Zinc fingers, a proline-rich domain, and 3 Zinc fingers in the C-term. Binds DNA. Transcription activator; PRDM16 has an intrinsic histone methyltransferase activity. PRDM16 forms a transcriptional complex with CEBPB. PRDM16 plays a downstream regulatory role in mediating TGF β signaling (Bjork et al., 2010). PRDM16 induces brown fat determination and differentiation. PRDM16 is expressed selectively in the earliest stem and progenitor hematopoietic cells, and is required for the maintenance of the hematopoietic stem cell pool during development. PRDM16 is also required for survival, cell-cycle regulation and self-renewal in neural stem cells (Chuikov et al., 2010; Kajimura et al., 2010; Aguilo et al., 2011; Chi and Cohen, 2016).

IGL (Immunoglobulin Lambda)

Location

22q11.22

Result of the chromosomal anomaly

Fusion protein

Oncogenesis

IGL may act as an enhancer of PRDM16.

References

- Aguilo F, Avagyan S, Labar A, Sevilla A, Lee DF, Kumar P, Lemischka IR, Zhou BY, Snoeck HW. Prdm16 is a physiologic regulator of hematopoietic stem cells. *Blood*. 2011 May 12;117(19):5057-66

Bjork BC, Turbe-Doan A, Prysak M, Herron BJ, Beier DR. Prdm16 is required for normal palatogenesis in mice. *Hum Mol Genet.* 2010 Mar 1;19(5):774-89

Chi J, Cohen P. The Multifaceted Roles of PRDM16: Adipose Biology and Beyond. *Trends Endocrinol Metab.* 2016 Jan;27(1):11-23

Chuikov S, Levi BP, Smith ML, Morrison SJ. Prdm16 promotes stem cell maintenance in multiple tissues, partly by regulating oxidative stress. *Nat Cell Biol.* 2010 Oct;12(10):999-1006

Duhoux FP, Ameye G, Montano-Almendras CP, Bahloula K, Mozziconacci MJ, Laibe S, Wlodarska I, Michaux L,

Talmant P, Richebourg S, Lippert E, Speleman F, Herens C, Struski S, Raynaud S, Auger N, Nadal N, Rack K,

Mugneret F, Tigaud I, Lafage M, Taviaux S, Roche-Lestienne C, Latinne D, Libouton JM, Demoulin JB, Poirrel HA. PRDM16 (1p36) translocations define a distinct entity of myeloid malignancies with poor prognosis but may also occur in lymphoid malignancies. *Br J Haematol.* 2012 Jan;156(1):76-88

Kajimura S, Seale P, Kubota K, Lunsford E, Frangioni JV, Gygi SP, Spiegelman BM. Initiation of myoblast to brown fat switch by a PRDM16-C/EBP-beta transcriptional complex. *Nature.* 2009 Aug 27;460(7259):1154-8

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