t(8;17)(q24;q22) ???BCL3/MYC

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Identity

Note
It is unlikely that the BCL3 gene (HGNC official name) is involved in this translocation with a breakpoint in 17q22, since BCL3 sits in 19q13.32 (coordinates: starts at 45251978 and ends at 45263301 bp from 19pter); the alternative would be a cryptic translocation, involving a cryptic inserted fragment of 19q13.32, including BCL3, within 17q22.

Clinics and pathology

Disease
Aggressive prolymphocytic leukemia

Epidemiology
Only one case to date, with no clinical data.

Cytogenetics

Cytogenetics morphological
The karyotype also showed the classical t(14;18)(q32;q21), usually found in follicular lymphoma, a 12q+ and a Xp+, not otherwise described.

Genes involved and proteins

Note
As said above, it is unprobable that the MYC partner is BCL3.

MYC
Protein
MYC regulates the transcription of genes required to coordinate a range of cellular processes, including those essential for proliferation, growth, differentiation, apoptosis and self-renewal, and protein synthesis through ribosome biogenesis (van Riggelen et al., 2010).

BCL3
Location
19q13.32

Protein
BCL3 is mainly found in the nucleus. Protein which contains seven ankyrin repeats. Ankyrin repeats are found in IkB family members, including IkBa, IkBb, and IkBe. BCL3 is a member of the IkappaB family, whose proteins regulate the NFKappaB family of transcription factors. Component of a complex with a NF-kB p52-p52 homodimer. Down-regulates inflammatory responses through limiting the transcription of NF-kB-dependent genes. Binds to NF-kB p50 and p52, Jab1, Pirin, Tip60 and Bard1. Bcl-3 is an adaptor protein (Dechend et al., 1999; Kreisel et al., 2011). Regulates genes involved in cell proliferation and apoptosis. NFKappaB plays a major role in B-cell development.

Result of the chromosomal anomaly

Hybrid gene
Description
Disruption of MYC close to the first intron, with the decapitation of the first intron, replaced by a sequence of 1.7 kb, that the authors have called "BCL3".

References


This article should be referenced as such: