t(3;19)(q27;q13) NAPA/BCL6

Jean-Loup Huret
Genetics, Dept Medical Information, University of Poitiers, CHU Poitiers Hospital, F-86021 Poitiers, France (JLH)

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Clinics and pathology

Disease
Non Hodgkin lymphoma

Clinics
Two cases available: a case of follicular lymphoma transformed to diffuse aggressive lymphoma, from a study with no individual data (Akasaka et al., 2003), and a male patient aged 60 years with a diffuse large B-cell lymphoma (Yunis et al., 1984).

Cytogenetics

Additional anomalies
The case described by Yunis et al., 1984 showed a complex karyotype.

Genes involved and proteins

Note
Only the more recent case (Akasaka et al., 2003) was studied for gene rearrangements.

BCL6
Location
3q27.3

Protein
706 amino acids; composed of a NH2-term BTB/POZ domain (amino acids 1-130 (32-99 according to Swiss-Prot) which mediates homodimerization and protein-protein interactions with other corepressors (including HDAC1 and NCOR2/SMRT to constitute a large repressing complex, another transcription repression domain (191-386), PEST sequences (300-417) with a KKYK motif (375-379), and six zinc finger at the C-term (518-541, 546-568, 574-596, 602-624, 630-652, 658-681), responsible for sequence specific DNA binding.

Transcription repressor; recognizes the consensus sequence: TTCCT(A/C)GAA (Albagli-Curiel, 2003); Role in germinal centers of lymphoid follicles. BCL6 prevents ATM and TP53 to induce apoptosis in response to DNA rearrangements such as somatic hypermutation and class switch recombination. Therefore essential for normal B cell development.

NAPA
Location
19q13.33

Protein
295 amino acids; SNARE protein found in the endoplasmic reticulum and implicated in protein trafficking, which possesses anti-apoptotic properties by promoting resistance to cisplatin in cancer cells by inducing the degradation of TP53 (Wu et al., 2011).

References


This article should be referenced as such: