t(3;6)(q27;p21)

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Identity

Note: In only two cases were the hybrid genes uncovered: a PIM1/BCL6 was found in one case (Yoshida et al., 1999), and a SFRS3/BCL6 in another case (Chen et al., 2001). These cases will also be described separately.

Clinics and pathology

Disease
Non Hodgkin lymphoma (NHL).

Epidemiology
Seven cases available: 3 cases of follicular lymphoma (FL), 3 cases of diffuse large B-cell lymphoma (DLBCL), and one NHL not otherwise specified. Very few data available: 4 male / 2 female patients, aged 49-79 years (Ohno et al., 1994; Miura et al., 1996; Yoshida et al., 1999; Chen et al., 2001; Varga et al., 2001; Keller et al., 2006).

Cytogenetics

Cytogenetics morphological
The t(3;6) was the sole anomaly in one case (a FL case), accompanied with +X in a subclone and +21 in another in the other FL case, found within a complex karyotype in the 4 remaining cases with cytogenetic data. Other additional anomalies of note were: del(7q), t(14;18)(q32;q21) (2 cases each) and t(1;14)(q21;q32), del(1q), del(5q) (1 case each).

Genes involved and proteins

BCL6
Location
3q27

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).

References


Chen W, Itoyama T, Chaganti RS. Splicing factor Srp20 is a novel partner of Bcl6 in a t(3;6)(q27;p21) translocation in transformed follicular lymphoma. Genes Chromosomes Cancer. 2001 Nov;32(3):281-4


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