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Leukaemia Section
Short Communication

t(14;14)(q11;q32) CEBPE/IGH, inv(14)(q11q32) CEBPE/IGH

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Identity

Note
This chromosome anomaly should not be confused with the t(14;14)(q11;q32)/inv(14)(q11q32) found in T-cell diseases, which implicates TCR alpha or TCR delta (14q11) and TCL1A (14q32).

Clinics and pathology

Disease
CD10+ acute lymphoblastic leukaemia (B-ALL).

Epidemiology
Only 4 cases to date of t(14;14)(q11;q32)/inv(14)(q11q32) with CEBPE and IGH involvements (Akasaka et al., 2007). Five other cases of t(14;14)(q11;q32)/inv(14)(q11q32) in B-cell leukemias are known (Denny et al., 1986; Speleman et al., 1991; Chervinsky et al., 1995; Wong et al., 1995; Thomas et al., 2001), but without proof that CEBPE was involved. As a matter of fact, a t(4;11)(q21;q23) was found in 2 of these cases, and a t(8;14)(q24;q32) in another case; this latter group is certainly heterogeneous.

Clinics
The four patients were male patients, aged 15, 25, 45, and 45 years, with a WBC under 50 x 10^9/l. Survival is available only for two cases: 19 mths+ and 48 months+, resembling the relatively fair survival of patients with a t(8;14)(q11;q32) CEBPD/IGH translocation.
One case was a Down syndrome patient; this may not be anecdotical, since more than 1/4 of t(8;14)(q11;q32) case are also Down syndrome patients.

Genes involved and proteins

CEBPE

Location
14q11

Protein
DNA-binding protein. CCAAT enhancer-binding protein (CEBP) transcription factors are a family of 6 multifunctional basic leucine zipper (bZIP) transcription factors. The 5 other CEBPs are: CEBPA (19q13), CEBPB (20q13), CEBPD (8q11), CEBPG (19q13), all four equally implicated in leukemias, and DDIT3/CHOP/CEBP zeta (12q13), so far known to be involved in solid tumours (liposarcoma). These transcription factors play a key role in cellular differentiation, in particular in the control of myeloid differentiation. CEBPE is composed of an N-term transactivation domain, a negative regulatory domain, a DNA-binding basic motif, and a leucine-zipper domain in C-term (Ramji et al., 2002; Nerlov et al., 2007).

IgH

Location
14q32

Result of the chromosomal anomaly

Fusion protein

Oncogenesis

Overexpression of the CEBP gene.

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