

Leukaemia Section

Mini Review

t(11;14)(q23;q32)

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Clinics and pathology

Disease

Treatment related leukemia (treatment related acute myeloid leukemia, t-AML)

Epidemiology

The involvement of MLL in 11q23 and KIAA0284 in 14q32 was shown in only 2 cases (Burmeister et al., 2008; De Braekeleer et al., 2009). These 2 cases were treatment related leukemia cases (t-AML for: treatment related acute myeloid leukemia). These t-AML cases occurred in a 45-year-old male patient (a M1 case) and in a 65-year-old female patient, 2 years after an urothelial carcinoma and 5 years after a ductal mammary carcinoma respectively. In 1 other case of t(11;14)(q23;q32), a myelodysplastic syndrome case, the involvement of MLL was excluded, and IGH in 14q32 was rearranged (Yujiri et al., 2009). Finally, in 2 other cases of t(11;14)(q23;q32), no molecular studies were available (Kaneko et al., 1982; Hanson et al., 1993). The two latter cases were a biphenotypic leukemia (BAL) case and an acute lymphoblastic leukemia (ALL) in a 33-year-old male patient.

Prognosis

One of the t-AML cases died 2 months after diagnosis, while the other one was lost to follow-up 5 years after diagnosis of the t-AML. The ALL case died 7 months after diagnosis.

Cytogenetics S

Cytogenetics morphological

The t(11;14)(q23;q32) was the sole anomaly in the 2 cases were MLL and KIAA0284 involvements

were ascertained. The BAL case showed a complex karyotype with -7 and i(17q), the ALL case also had a complex karyotype, with +12.

Genes involved and proteins

MLL

Location

11q23

DNA/RNA

36 exons, multiple transcripts 13-15 kb.

Protein

3969 amino acids; 431 kDa; contains two DNA binding motifs (a AT hook and a CXXC domain), a DNA methyl transferase motif, a bromodomain. MLL is cleaved by *taspase 1* into 2 proteins before entering the nucleus, called MLL-N and MLL-C. The FYRN and a FRYC domains of native MLL associate MLL-N and MLL-C in a stable complex; they form a multiprotein complex with transcription factor TFIID. MLL is a transcriptional regulatory factor. MLL can be associated with more than 30 proteins, including the core components of the SWI/SNF chromatin remodeling complex and the transcription complex TFIID. MLL binds promoters of HOX genes through acetylation and methylation of histones. MLL is a major regulator of hematopoiesis and embryonic development.

KIAA0284

Location

14q32.33

Protein

KIAA0284 presents an amino acids similarity of 30% with CEP170. CEP170 is a forkhead-associated

domain protein which associates with centrosomes during interphase and with spindle microtubules during mitosis (Guarguaglini et al., 2005).

Result of the chromosomal anomaly

Hybrid gene

Description

5' MLL - 3' KIAA0284

Transcript

The breakpoint was located in intron 9 of MLL. The breakpoint in KIAA0284 was located in intron 2 in one case, and intron 4 in the other case.

Fusion protein

Description

N-term MLL - C-term KIAA0284

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