

## Leukaemia Section

### Short Communication

## B-cell prolymphocytic leukemia (B-PLL)

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### Clinics and pathology

#### Disease

Chronic lymphoproliferative disorder (CLD)

#### Phenotype/cell stem origin

Disease affecting mature B-cells.

Immunophenotypically, B-PLL is characterized by reactivity with pan B-cell markers CD19, CD20 and CD24.

B-PLL cells are distinct from B-CLL cells in that they express bright surface immunoglobulin, unfrequently express CD5, fail to form rosettes with mouse erythrocytes and react strongly with FMC7. Expression of CD22 is often observed whereas CD23 is usually not expressed.

#### Epidemiology

Rare disease; slight male predominance with median age of 69 years.

#### Clinics

Patients often present with advanced stage disease.

B-PLL is characterized by high white blood cell counts and splenomegaly without adenopathy.

Bone marrow infiltration pattern is either diffuse or mixed.

Blood data: elevated white blood cell counts with prolymphocytes representing more than 55% of the circulating lymphoid cells.

Anemia and thrombocytopenia may be observed.

#### Prognosis

Evolution: this disease is always progressive.

Prognosis: poor response to therapy is often observed; median survival is 3 years.

### Cytogenetics

#### Cytogenetics morphological

Few studies focused on B-PLL; the use of B-cell mitogens might increase the detection rate of cytogenetic changes; the most frequent aberrations involve chromosomes 14, 6 and 1; 14q+ changes are the most commonly observed and are often the consequence of a translocation t(11;14)(q13;q32); structural abnormalities of chromosome 6 are primary or secondary; deletion 6q, as well as translocation t(6;12)(q15;p13) are described; structural aberrations of chromosome 1 involve both p and q arms; trisomy 12 represents a secondary change in this disease; finally, i(17)(q10), as well as telomeric associations have been reported; karyotypic evolution has been documented in some cases and seems to be associated with disease progression.

### Genes involved and proteins

#### Note

Little is known about underlying genetic mechanisms in B-PLL.

Immunoglobulin gene rearrangements are always observed.

BCL-1 gene is involved in some cases bearing t(11;14)(q13;q32), with breakpoints located centromeric to the major translocation cluster.

Overall, abnormalities of P53 occur in 75% cases, representing the highest reported frequency in B-cell malignancies.

No CDKNL-2 or RB1 gene involvement has been documented so far.

C-MYC rearrangement has been described in PLL.

## To be noted

### Note

T-cell prolymphocytic leukaemia also exists and account for 1/4 of cases of PLL.

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