Atlas of Genetics and Cytogenetics in Oncology and Haematology

OPEN ACCESS JOURNAL AT INIST-CNRS

Leukaemia Section
Short Communication

B-cell prolymphocytic leukemia (B-PLL)

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Published in Atlas Database: October 1998

Online updated version : http://AtlasGeneticsOncology.org/Anomalies/BPLL.html
DOI: 10.4267/2042/37483

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 centrometric 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.

References

Pittman S, Catovsky D. Chromosome abnormalities in B-cell prolymphocytic leukemia: a study of nine cases. Cancer Genet Cytogenet. 1983 Aug;9(4):355-65

Sadamori N, Han T, Minowada J, Bloom ML, Henderson ES, Sandberg AA. Possible specific chromosome change in prolymphocytic leukemia. Blood. 1983 Oct;62(4):729-36

Bennett JM, Catovsky D, Daniel MT, Flandrin G, Galton DA, Gralnick HR, Sultan C. Proposals for the classification of chronic (mature) B and T lymphoid leukaemias. French-American-British (FAB) Cooperative Group. J Clin Pathol. 1989 Jun;42(6):567-84

Brito-Babapulle V, Catovsky D. Inversions and tandem translocations involving chromosome 14q11 and 14q32 in T-prolymphocytic leukemia and T-cell leukemias in patients with ataxia telangiectasia. Cancer Genet Cytogenet. 1991 Aug;55(1):1-9

Brito-Babapulle V, Ellis J, Matutes E, Oscier D, Khokhar T, MacLennan K, Catovsky D. Translocation t(11;14)(q13;q32) in chronic lymphoid disorders. Genes Chromosomes Cancer. 1992 Sep;5(2):158-65

Howell RT, Kitchen C, Standen GR. Telomeric associations in a patient with B-cell prolymphocytic leukaemia. Genes Chromosomes Cancer. 1993 Jun;7(2):116-8

Brennscheidt U, Eick D, Kunzmann R, Martens U, Kiehntopf M, Mertelsmann R, Herrmann F. Burkitt-like mutations in the c-myc gene locus in prolymphocytic leukemia. Leukemia. 1994 May;8(5):897-902

Döhner H, Pilz T, Fischer K, Cabot G, Diehl D, Fink T, Stilgenbauer S, Bentz M, Lichter P. Molecular cytogenetic analysis of RB-1 deletions in chronic B-cell leukemias. Leuk Lymphoma. 1994 Dec;16(1-2):97-103

Galiègue-Zouitina S, Collyn-d'Hooghe M, Denis C, Mainardi A, Hildebrand MP, Tilly H, Bastard C, Kerckaert JP. Molecular cloning of a t(11;14)(q13;q32) translocation breakpoint centromeric to the BCL1-MTC. Genes Chromosomes Cancer. 1994 Dec;11(4):246-55

Schröder M, Mathieu U, Dreyling MH, Bohlander SK, Hagemeijer A, Beverloo BH, Olopade OI, Stilgenbauer S, Fischer K, Bentz M. CDKN2 gene deletion is not found in chronic lymphoid leukaemias of B- and T-cell origin but is frequent in acute lymphoblastic leukaemia. Br J Haematol. 1995 Dec;91(4):865-70

Lens D, De Schouwer PJ, Hamoudi RA, Abdul-Rauf M, Farahat N, Matutes E, Crook T, Dyer MJ, Catovsky D. p53 abnormalities in B-cell prolymphocytic leukemia. Blood. 1997 Mar 15;89(6):2015-23

This article should be referenced as such:

Michaux L. B-cell prolymphocytic leukemia (B-PLL). Atlas Genet Cytogenet Oncol Haematol. 1999; 3(1):24-25.