

Leukaemia Section

Short Communication

Splenic lymphoma with villous lymphocytes

Jean-Loup Huret, Hossain Mossafa

Genetics, Dept Medical Information, University of Poitiers, CHU Poitiers Hospital, F-86021 Poitiers, France (JLH), Laboratoire Pasteur-Cerba, 95066, Cergy-Pontoise, France (HM)

Published in Atlas Database: October 1998

Online updated version : <http://AtlasGeneticsOncology.org/Anomalies/splenvillousID2063.html>
DOI: 10.4267/2042/37484

This work is licensed under a Creative Commons Attribution-Noncommercial-No Derivative Works 2.0 France Licence.
© 1999 Atlas of Genetics and Cytogenetics in Oncology and Haematology

Clinics and pathology

Epidemiology

Occurs in the elderly (med 70 years); sex ratio 2M/1F.

Clinics

Splenomegaly without hepatomegaly nor enlarged lymph nodes; peripheral blood lymphocytes with villous projections; monoclonal Ig in half cases.

Cytology

B-cells expressing CD19+, CD20+, CD22+, CD24+ and DBA44+.

Treatment

Splenectomy.

Prognosis

5-year survival: 80%; adverse prognostic factors: WBC above $30 \times 10^9/l$, low lymphocyte count; cases treated with chemotherapy have shorter survival.

Cytogenetics

Cytogenetics morphological

The karyotype is often abnormal:

- del(7q) and translocations involving 7q (20% of cases),
- t(11;14)(q13;q32) (15%),
- other anomalies, in particular i(17q), 2p11 translocations.

Genes involved and proteins

Note

BCL1 in 11q13 and IgH in 14q32 are involved in 20% of cases, with or without a visible t(11;14); BCL1 encodes the cyclin D1; role in the cell cycle control (G1 progression and G1/S transition); 5' BCL1 translocated on chromosome 14 near JH, resulting in promoter exchange; the immunoglobulin gene enhancer stimulates the expression of BCL1, and overexpression of BCL1 which accelerates passage through the G1 phase; microdeletion in the RB1 region in half cases.

References

Oscier DG, Matutes E, Gardiner A, Glide S, Mould S, Brito-Mulligan SP, Matutes E, Dearden C, Catovsky D. Splenic lymphoma with villous lymphocytes: natural history and response to therapy in 50 cases. *Br J Haematol* 1991 Jun;78(2):206-9

Wong KF, Chu YC, Hui PK. Splenic lymphoma with villous lymphocytes showing del(7) and inv(10). *Cancer Genet Cytogenet* 1998 Apr 15;102(2):145-7. (Review)

Oscier DG, Matutes E, Gardiner A, Glide S, Mould S, Brito-Babapulle V, Ellis J, Catovsky D. Cytogenetic studies in splenic lymphoma with villous lymphocytes. *Br J Haematol* 1993 Nov;85(3):487-491

Matutes E, Morilla R, Owusu-Ankomah K, Houlihan A, Catovsky D. The immunophenotype of splenic lymphoma with villous lymphocytes and its relevance to the differential diagnosis with other B-cell disorders. *Blood* 1994 Mar 15;83(6):1558-62

Troussard X, Valensi F, Duchayne E, Garand R, Felman P, Tulliez M, Henry-Amar M, Bryon PA, Flandrin G. Splenic lymphoma with villous lymphocytes: clinical presentation, biology and prognostic factors in a series of 100 patients. Groupe Français d'Hématologie Cellulaire (GFHC). *Br J Haematol* 1996 Jun;93(3):731-736

García-Marco JA, Nouel A, Navarro B, Matutes E, Oscier D, Price CM, Catovsky D. Molecular cytogenetic analysis in splenic lymphoma with villous lymphocytes: frequent

allelic imbalance of the RB1 gene but not the D13S25 locus on chromosome 13q14. *Cancer Res* 1998 Apr 15;58(8):1736-1740

This article should be referenced as such:

Huret JL, Mossafa H. Splenic lymphoma with villous lymphocytes. *Atlas Genet Cytogenet Oncol Haematol*. 1999; 3(1):26-27.
