Splenic lymphoma with villous lymphocytes

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Clinical and pathologic

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 x 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 t(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


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