**del (13q) in chronic lymphoproliferative diseases**

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**Identity**

*Note:* A spectrum of B-cell chronic lymphoproliferative disorders (CLD) may carry a chromosome 13q deletion; among these, three forms other than chronic lymphocytic leukemia (CLL) were identified by the FAB group which may frequently carry a 13q- chromosome: atypical CLL, splenic lymphoma with villous lymphocytes, corresponding to splenic marginal zone B-cell lymphoma, and mantle cell lymphoma (MCL) in leukemic phase.

Clones dJ1154H7 (top) and dJ1013C9 (bottom) for 13q14 deletions, in normal cells - Courtesy Mariano Rocchi, Resources for Molecular Cytogenetics.

**Clinics and pathology**

**Disease**

Atypical CLL, including the CLL/PL (prolymphocytic leukemia) or CLL mixed-cell-type variant by FAB criteria.

**Phenotype/cell stem origin**

Virgin CD5+ recirculating B-cell.

**Epidemiology**

del(13q) is found in approximately 10-15% of all CLLs.

**Clinics**

The clinical course may be more aggressive than in typical CLL, depending on stage at presentation and % of prolymphocytes.
**Disease**
Splenic lymphoma with villous lymphocytes.

**Phenotype/cell stem origin**
Chronic proliferation originating from the marginal zone B-lymphocytes.

**Epidemiology**
The disorder appears to be relatively rare, but it is probably underdiagnosed.

**Clinics**
The clinical course is indolent.

**Disease**
Leukemic mantle cell lymphoma.

**Note**
The majority of mantle cell lymphomas show peripheral blood (PB) involvement at diagnosis or at disease evolution; there is a disease variant presenting as a de novo leukemic condition, presenting heterogeneous cytological features with PB and BM lymphocytosis, without adenopathy, with or without splenomegaly; some of these cases may fulfill the FAB criteria for the diagnosis of atypical CLL; because these cases usually carry the t(11;14)(q13;q32) and a mantle-cell phenotype, they have also been referred to as 'mantle cell leukemia'; it is reasonable to assume that the transformation of a mantle cell may give rise to a spectrum of diseases ranging from the classical lymphomatous form of MCL to an overt leukemic disease, as is the case with small lymphocytic lymphoma and chronic lymphocytic leukemia.

**Phenotype/cell stem origin**
Proliferation of cells of follicle mantle lineage (CD5/CD19/CD22 positive, CD23 negative, bright sIg expression).

**Cytogenetics**

**Cytogenetics morphological**
The frequency of 13q- as an isolated chromosome change in atypical CLL is much lower than in typical CLL; however FISH studies detected an approximately 40% incidence for this anomaly using a 13q14 probe; additional chromosome anomaly included +12, 6q- and complex karyotypes.

The incidence of 13q- in splenic marginal zone B-cell lymphoma is low by conventional cytogenetic analysis. FISH studies detected a 12-47% incidence for cryptic 13q deletion, the highest frequency having been reported using a 13q14 Rb probe; the 13q- is usually associated with other chromosome changes, including +12, 14q+.

As is the case with classical MCL, a 40-60% incidence for 13q14 deletion was reported in leukemic MCL/mantle cell leukemia by interphase FISH.

**References**


This article should be referenced as such: