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Leukaemia Section
Mini Review

del (13q) in chronic lymphoproliferative diseases

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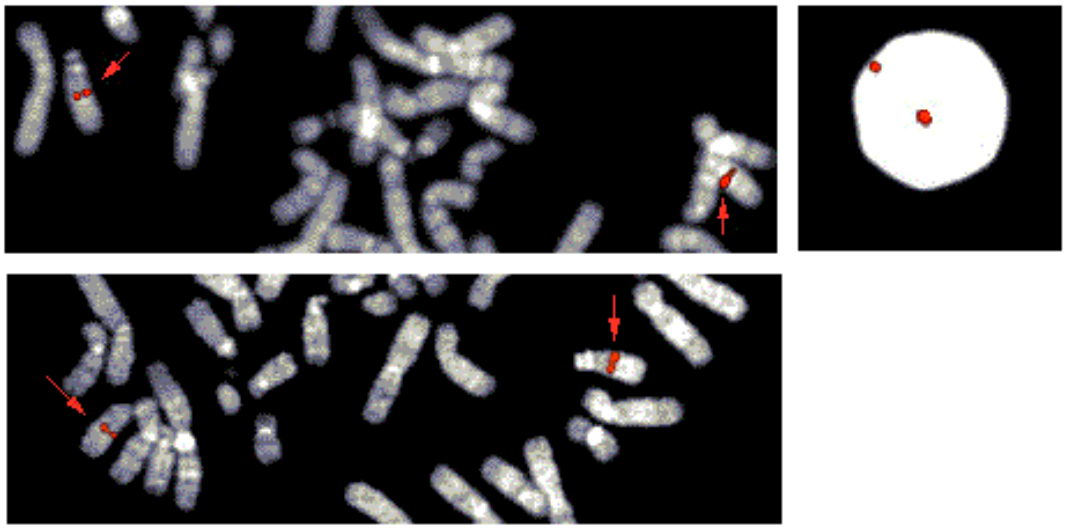
Identity

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 withour 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 condition, 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 appoximately 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.

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