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# Clinical and pathological characterization of the t(14;19)(q32;q13)-positive splenic marginal zone lymphoma

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## Abstract

Splenic marginal zone lymphoma (SMZL) is a B-cell neoplasm which is characterized by splenomegaly, lymphocytosis, and involvement of bone marrow and—sometimes—an M–component. The most frequent findings in SMZL involve chromosomes 1, 3, 7 (usually deleted in 7q), and 18. The t(14;19)(q32;q13) is a rare cytogenetic abnormality with a bcl-3 rearrangement that has been reported in B-cell lymphomas and leukemia.

**Aim:** The aim of the study was to characterize the clinical and morphological changes in patients with t(14;19)(q32;q13)-positive SMZL.

**Patients, methods and results:** In our center between January 2005 and February 2009, 3 cases of SMZL with t(14;19)(q32;q13) were identified. All the patients (males, median age 58 years, range 51–67 years) presented with lymphoma with B-symptoms, a high level of lactate dehydrogenase (LDH), hepatosplenomegaly, and with splenic hilar lymphadenopathy only. The median hemoglobin was 110 g/l (range 92–122 g/l). All patients had a normal leukocyte count with an absolute lymphocytosis (median lymphocyte count $72.3x10^9$g/l, range $58–83x10^9$/l) and thrombocytopenia. A morphological examination of the peripheral blood and bone marrow lymphocytes showed that all lymphocytes were atypical with a wide cytoplasm and nuclear indentation. In all cases bone marrow involvement was nodular and composed of heterogeneous mixture cells, with the majority being medium sized. The results of the immunophenotypic analysis were the expression of mature B-cells antigens and an absence of CD10-, CD23-, CD5-, CD43-, CyclinD1-.

In debut two patients were to receive chemotherapy (CHOP–regimen). However there was progression: enlargement of the spleen and a decrease in the thrombocyte count. All patients underwent a splenectomy. The median weight of the spleen was 2083 g (range 1800–2850 g). The splenic section generally showed massive nodular patterns (involving the white and red pulp) associated with diffuse invasion of the sinuses. In each case, high Ki-67 was discovered. Progression occurred 3–6 months after the splenectomies, and was characterized by an increase in the leukocyte count (range $45.4–101.8x10^9$/l), a high level of LDH, and an appearance of peripheral and visceral lymph nodes. In consideration of increasing the leukocyte count and the presence of lymphadenopathy in all patients, alkylating agents were used (in one patient chlorambucil, and in two cyclophosphamide). There was a normal peripheral blood index, LDH level , and an absence of lymphadenopathy in all cases after 4–6 months of treatment with alkylating agents. Median observation was 19+ months (range 17–22 months). All patients were alive.

**Conclusion:** SMZL with t(14;19)(q32;q13) is a distinct variant that is characterized by transient progression after splenectomy and high efficiency of alkylating agents.

**Keywords:** splenic marginal zone lymphoma, t(14;19)(q32;q13), morphology, spleen, splenectomy, alkylating agents