RICHTER'S TRANSFORMATION

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The Richter transformation (Ricther syndrome) - RT is defined by the development of an aggressive large-cell lymphoma in a patient with chronic lymphocytic leukemia/small lymphocytic lymphoma (CLL/SLL). Diffuse large B cell lymphoma is the most common histology of RT, although Hodgkin lymphoma (LH) and T cell lymphomas have also been reported.

The incidence of RT from CLL/SLL to diffuse large B cell lymphoma varies between 2 and 9%, depending on the aggressiveness criteria for biopsy, a 0.4% being quoted for LH.

The RT occurs between 2 and 4 years from CLL/SLL diagnosis, the onset of is heralded by sudden clinical deterioration, the rapid increase in lymphadenopathy, or splenomegaly, worsening "B" symptoms.

Risk factors involved in RT are poorly defined: an increasing number of prior therapies, younger age, advanced Rai stage, elevated LDH and beta-2-microglobulin levels, lymph nodes >3cm, absence of del 13q14, CD38 expression and usage of IGHV4-39 and the G allele presence in the regulatory region of intron 1 of CD38 has been associated with the development of Richter's syndrome.

There are three possible origins of aggressive clone: arise from the underlying CLL/SLL, represent evolution from a biclonal phenotype in the CLL/SLL or may represent a new clone

It has been suggested that: P53 disruption aberrant c-myc expression, 11q, 17p, 8p and 20 deletions, 12 trisomy, unmutated IgVH genes, mutations of individual tumor-suppressor (P53, INK4a/ARP) and cell cycle regulatory genes, and EBV infections is associated with RT.