Chronic small lymphocytic neoplasm: proposal for nomenclature change in order to clinically integrate chronic lymphocytic leukemia and small lymphocytic lymphoma.

We propose that chronic lymphocytic leukemia (CLL) and small lymphocytic lymphoma (SLL) be called chronic small lymphocytic neoplasm (CSLN) in order to fully integrate them clinically into one disease entity. This would streamline the clinical approach to patients with CLL/SLL.

CLL and SLL have been considered one disease entity by the World Health Organization (WHO) since 2001, essentially because it became clear that CLL and SLL are identical phenotypically and morphologically[1,2]. This was facilitated by a general approach to classification which ignored clinical groupings on the rather sound basis that treatment of patients depends on the biology of disease[3]. Thus CLL and SLL were not immediately fully integrated into one disease entity clinically. This created some clinical inconsistencies in the approach to CLL/SLL. Even WHO conceded that CLL is likely to be managed by hematologists and SLL by medical oncologists in many centers[3]. It also took some time before it became clear that CLL and SLL patients can be offered the same treatment options[4]. SLL and CLL continue to have different staging systems (Ann Arbor vs Rai/Binet staging systems). The Ann Arbor staging system has been attempted in CLL, but was found to be unsuitable[5]. In the presence of lymphadenopathy/splenomegaly, a B cell lymphocytosis exceeding 5 x10^9/L implies CLL rather than SLL[6]. This means that an SLL patient who later develops a B cell lymphocytosis exceeding 5 x10^9/L would need to move from Ann Arbor staging to Rai/Binet staging; CLL and SLL also have different response criteria[6]. It is not uncommon for a discussion of CLL to exclude SLL[7-8] and in some instances it is implied that SLL is a differential of CLL[9]. It is more common for CLL to be discussed alone without reference to SLL than vice versa. Where CLL and SLL are discussed together, they are often referred to as "CLL/SLL" which semantically means "CLL and or SLL" and which implies similarity but not necessarily entity identicalness. The approach to a patient with CLL is not necessarily the same when CLL is considered a leukemia[6] and when it is considered a lymphoma[5]. Symptomatic B cell lymphocytosis of <5 x10^9/L could be either CLL or SLL[6] and additional time and costs may be required in order to make the distinction between the two.

All the foregoing considerations mean that differing amounts of time and cost may be involved in the standard approach to CLL/SLL patients with identical presentation. This appears unnecessary for patients with the same disease entity. CLL and SLL are considered different phases or manifestations of the same disease[3,5]. But which disease? We suggest that the disease in question be called CSLN. This would cement the identicalness of CLL and SLL, eliminate the inconsistencies inherent in the CLL/SLL designation, and encourage a simplified approach to CLL/SLL patients.

CSLN would be part of small B-cell neoplasms (SBCNs) which currently comprise follicular lymphoma (FL), CLL/SLL, mantle cell lymphoma (MCL), marginal zone B-cell lymphoma of nodal and extranodal sites, and lymphoplasmacytic lymphoma. As they currently stand, distinguishing among these can be challenging[10]. Increasing reliance on immunophenotyping for classification of these neoplasms[11] suggests that the main difference among them is that they arise from different "stages of maturation" of small B-cells. The main difference, therefore, between the term "small B-cell neoplasms" and "chronic small lymphocytic neoplasm" in terms of terminology, would be the words "lymphocytic" as opposed to "B-cell". The former would imply a more specific characterisation of the latter. This would clarify the difference between CSLN and SBCN.

CSLN would be defined as a neoplasm composed of monomorphic, small, round to slightly irregular B lymphocytes in the peripheral blood, bone marrow,

spleen and/or lymph nodes admixed with prolymphocytes and paraimmunoblasts forming proliferation centers in tissue infiltrates. Peripheral B-cell lymphocytosis of 5 x10^9/L (for at least 3 months) would be required for diagnosis of CSLN if there was no extramedullary tissue involvement.

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