Chronic T-cell Leukemia/Lymphoma

This issue of Cancer Control focuses on the clinicopathologic features of chronic T-cell leukemia/lymphoma. Chronic leukemia of T-cell origin was first identified by Brouet and colleagues\(^1\) in 1975. Several classification systems for T-cell lymphoid malignancies have been proposed since then. The French-American-British (FAB) Cooperative Group’s proposal for classification designated four subgroups: (1) T-cell CLL or large granular lymphocyte (LGL) leukemia, (2) T-prolymphocytic leukemia, (3) adult T-cell leukemia/lymphoma, and (4) Sezary’s syndrome.\(^2\) In 1990, the Morphologic, Immunologic, Cytogenetic (MIC) Cooperative Study Group reviewed this FAB classification and determined that LGL leukemia instead of T-CLL was the preferable terminology.\(^3\) These four subgroups have also been recognized as distinct entities in the REAL classification.\(^4\)

Important biologic principles have been established by scientific study of these T-cell malignancies. The first human retrovirus, HTLV-I, is the causative agent of adult T-cell leukemia (ATL).\(^5\) ATL is characterized by skin rash, generalized lymphadenopathy, hypercalcemia, clonal proliferation of abnormal pleomorphic CD4+ lymphocytes, and usually a rapidly progressive clinical course.\(^6\) Studies of the other T-cell malignancies reviewed in this issue also have revealed insights into the normal T-cell counterparts of the neoplastic clones.

L. Frank Glass, MD, and colleagues review the diagnosis and staging of cutaneous T-cell lymphomas (CTCL). The leukemic phase of CTCL is known as Sezary’s syndrome. As Dr. Glass emphasizes, the diagnosis and treatment of CTCL remain challenging. At our center, a multidisciplinary team staffed by dermatology, dermatopathology, medical hematology/oncology, and radiation oncology evaluates each patient with CTCL. It is hoped that such an effort will lead to improved diagnosis and therapy for these patients who are often difficult to treat.

We are fortunate to have Estella Matutes, MD, PhD, review the characteristic presentation of T-prolymphocytic leukemia (T-PLL). Her group is the acknowledged world leader in studying this disease. The differences and similarities among T-PLL and the other post-hypertrophic T-cell malignancies are summarized nicely in her article. Dr. Matutes emphasizes that a key diagnostic test is a well-stained peripheral blood film. This point deserves repetition, as a careful review of the peripheral blood smear is an essential undertaking for the diagnosis of each of the four subgroups of T-cell leukemias. It is reassuring to the hematologist that such a simple test remains of utmost importance, even in the complex world of modern medicine.

Thierry Lamy, MD, and I discuss the presentation and differential diagnosis of LGL leukemia. It is likely that most cases referred to as T-CLL are LGL leukemia. LGL leukemia is a chronic lymphoproliferative disorder with autoimmune features. A model of pathogenesis is outlined in which leukemic LGL represents cytotoxic T lymphocytes that are stimulated to proliferate in an antigen-driven fashion. To further understand the natural history of this disease, a national registry for LGL leukemia based at our center is being established. Look for more details, including website access, in future issues of Cancer Control.

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References