FEATURE ARTICLE

The Disease With Hope: Hairy Cell Leukemia

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Hairy cell leukemia (HCL), comprising 2% of all leukemias, is a chronic disorder characterized by mononuclear cells with prominent cytoplasmic projections. For years, patients with HCL underwent splenectomies and then interferon alpha for treatment, which provided high response rates but low percentages of complete remission. More recent treatments with 2-chlorodeoxyadenosine result in 85%–90% complete remission, minimal toxicity, and lower rates of relapse using a single course of therapy. A second course of therapy can be administered if HCL continues to be resistant or recurs. New research using anti-CD22 recombinant immunotoxin BL22 is proving successful. With these latest chemotherapy options, patients’ prognoses are optimistic.

Background

HCL is a monoclonal proliferation of relatively mature B lymphocytes, typically expressing monoclonal immunoglobulin G on their cell surfaces and having unique immunoglobulin gene arrangement. Hairy cells also coexpress the pan B-cell antigens CD19, DC20, and CD22 (Goodman, Bethel, et al., 2003). B lymphocytes in adults are processed in bone marrow and manufacture antibodies. Each B lymphocyte has on the surface of its cell membrane 100,000 antibody molecules that react specifically to one type of antigen. With the disruption of the B lymphocyte in HCL, immunity is disturbed and infections are common (Guyton, 1991).

Serum levels of soluble interleukin-2 (IL-2) are high in HCL and correlate with disease activity. The abnormal cells do not produce IL-2; however, they do produce tumor necrosis factor and a B-cell growth factor (Goodman, Bethel, et al., 2003).

The cause of HCL is unknown. Genetic and viral origins have been studied without any associations noted. Patients with HCL have been found to have a higher previous occupational exposure to ionizing radiation and organic chemicals (Goodman, Bethel, et al., 2003).