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.

Hairy cell leukemia (HCL) first was described in 1958 by Bouroncle, Wiseman, and Doan, who called it leukemic reticuloendotheliosis. In 1966, Schrek and Donnelly changed the name to hairy cell leukemia, describing its unique appearance. The hairy cell is a B lymphocyte; little is known about its pathology (Goodman, Burian, Koziol, & Saven, 2003). HCL accounts for 2% of all leukemias, and 600 new cases are diagnosed each year. Men are four times more likely to be diagnosed with HCL than women, and the mean age of onset is 52 years. HCL occurs primarily in Caucasians; Jewish men are overrepresented (Goodman, Bethel, et al., 2003).

Background

HCL is a monoclonal proliferation of relatively mature B lymphocytes, typically expressing monoclonal immunoglobin 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).

At a Glance

- Hairy cell leukemia (HCL) is a rare form of leukemia.
- Early symptoms include fatigue, infection, and bleeding.
- Treatment with 2-chlorodeoxyadenosine is used for initial pharmacologic management of HCL.

Signs and Symptoms

Early signs and symptoms of HCL are related to pancytopenias, including fatigue, infection, and, less commonly, bleeding. All HCL cases present with anemia; 75% of patients have thrombocytopenia (Schroeder, Tierney, McPhee, Papadakis, & Krupp, 1992). Splenomegaly can be massive in 90% of patients and hepatomegaly occurs in 40%, both causing abdominal discomfort (Goodman, Bethel, et al., 2003). HCL is progressive, with a median survival rate of 53 months, if left untreated (Saven & Piro, 1994).

Identification and Diagnosis

The hairy cell is mononuclear with prominent cytoplasmic projections, irregular cytoplasmic outlines, and villi of various lengths. They are relatively large cells with abundant pale blue cytoplasm and a low nuclear and cytoplasmic ratio. Hairy cells may be identified by Wright's stained peripheral blood films in approximately 90% of patients. The cells appear as round, oval,

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