Hairy cell leukemia

Hairy cell leukemia is an uncommon hematological malignancy characterized by an accumulation of abnormal B lymphocytes. it's always classified as a sub-type of leukemia (CLL). Hairy cell leukemia makes up approximately 2% of all leukemias, with fewer than 2,000 new cases diagnosed annually in North America and Western Europe combined.

Hairy cell leukemia was originally described as monocytic leukaemia , malignant reticulosis, or lymphoid myelofibrosis in publications dating back to the 1920s. The disease was formally named leukemic reticuloendotheliosis and its characterization significantly advanced by Bertha Bouroncle and colleagues at The Ohio State University College of medicine in 1958. Its common name, which was coined in 1966, springs from the "hairy" appearance of the malignant B cells under a microscope.

Signs and symptoms

In hairy cell leukemia, the "hairy cells" (malignant B lymphocytes) accumulate within the bone marrow, interfering with the assembly of normal white blood cells, red blood cells, and platelets. Consequently, patients may develop infections related to low white blood cell count, anemia and fatigue because of a scarcity of red blood cells, or easy bleeding because of a coffee platelet count. Leukemic cells may absorb the spleen and cause it to swell; this may have the side effect of making the person feel full even when he or she has not eaten much.

Hairy cell leukemia is typically diagnosed after a routine blood count shows unexpectedly low numbers of 1 or more kinds of normal blood cells, or after unexplained bruises or recurrent infections in an otherwise apparently healthy patient.

Platelet function could even be somewhat impaired in HCL patients, although this does not appear to possess any significant practical effect. it's getting to end in somewhat more mild bruises than would preferably be expected for a given platelet count or a mildly increased bleeding time for a minor cut. it's likely the results of manufacturing slightly abnormal platelets within the overstressed bone marrow tissue.

Patients with a high tumor burden also can have somewhat reduced levels of cholesterol, especially in patients with an enlarged spleen Cholesterol levels return to more normal values with successful treatment of HCL.

Cause

As with many cancers, the reason for hairy cell leukemia is unknown. Exposure to tobacco smoke, radiation , or industrial chemicals (with the possible exception of diesel) doesn't appear to increase the danger of developing HCL. Farming and gardening correlate with an increased risk of HCL development in some studies which does not necessarily imply causation.

A 2011 study identified somatic BRAF V600E mutations altogether 47 hairy cell leukemia (HCL) patients studied, and no such mutations within the 193 peripheral B-cell lymphomas/leukemias apart from HCL.

The U.S. Institute of medicine (IOM) sees a correlation which allows an association between exposure to herbicides and later development of chronic B-cell leukemias and lymphomas generally . The IOM report emphasizes that neither animal nor human studies indicate an association of herbicides with HCL specifically. However, the IOM extrapolated data from leukemia and non- Hodgkin lymphoma to conclude that HCL and other rare B-cell neoplasms may share this risk factor. As a results of the IOM report, the U.S. Department of Veterans Affairs considers HCL an illness presumed to be a service-related disability (see Agent Orange).

Human T-lymphotropic virus 2 (HTLV-2) has been isolated during alittle number of patients with the variant kind of HCL. within the 1980s, HTLV-2 was identified during a patient with a T-cell lymphoproliferative disease; this patient later developed hairy cell leukemia (a B cell disease), but HTLV-2 wasn't found within the hairy cell clones. there is no evidence that HTLV-II causes any quite hematological malignancy, including HCL.

Pathophysiology

Pancytopenia in HCL is caused primarily by marrow failure and splenomegaly. Bone marrow failure is caused by the buildup of hairy cells and reticulin fibrosis within the bone marrow, also as by the detrimental effects of dysregulated cytokine production.

Splenomegaly reduces blood counts through sequestration, marginalization, and destruction of healthy blood cells inside the spleen.

Hairy cells are nearly mature B cells, which are activated clonal cells with signs of VH gene differentiation. they'll be related to pre- plasma marginal zone B cells or memory cells.

Cytokine production is disturbed in HCL. Hairy cells produce and thrive on TNF-alpha. This cytokine also suppresses normal production of healthy blood cells within the bone marrow.

Unlike healthy B cells, hairy cells express and secrete an system protein called Interleukin-2 receptor (IL-2R). In HCL-V, only a neighborhood of this receptor is expressed. As a result, disease status are often monitored by measuring changes within the quantity of IL-2R within the serum . the extent increases as hairy cells proliferate, and reduces once they're killed. Although uncommonly utilized in North America and northern Europe , this test correlates better with disease status and predicts relapse more accurately than the opposite test.

Hairy cells answer normal production of some cytokines by T cells with increased growth. Treatment with Interferon-alpha suppresses the assembly of this pro-growth cytokine from T cells. a coffee level of T cells, which is typically seen after treatment with cladribine or pentostatin, and thus the resultant reduction of these cytokines, is additionally associated with reduced levels of hairy cells.