LYMPHOMA
RESEARCH • FOUNDATION

Getting the Facts

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Cutaneous B-Cell Lymphoma

Overview

Lymphoma is the most common blood cancer. The two main forms of lymphoma are Hodgkin lymphoma (HL) and non-Hodgkin lymphoma (NHL). Lymphoma occurs when cells of the immune system called lymphocytes, a type of white blood cell, grow and multiply uncontrollably. Cancerous lymphocytes can travel to many parts of the body, including the lymph nodes, spleen, bone marrow, blood, or other organs, and form a mass called a tumor. The body has two main types of lymphocytes that can develop into lymphomas: B lymphocytes (B cells) and T lymphocytes (T cells).

B-cell lymphomas account for 85 percent of all NHL cases in the United States. The disease affects men and women equally, and any age group can be affected. There are many different forms of B-cell lymphoma.

Lymphomas that arise in tissues or organs outside of the lymphatic system are called extranodal lymphomas. When extranodal lymphomas originate in the skin (comprising 18 percent of all extranodal lymphomas and five percent of all NHLs) and there is no evidence of disease outside of the skin, they are called primary cutaneous lymphomas. Primary cutaneous B-cell lymphomas (CBCLs) occur when the lymphoma cells originate in B cells.

CBCLs are most often indolent (slow-growing). They may appear on the skin as a reddish rash, lump, or nodule and may have a slightly raised and smooth appearance. The disease tends to go away and then return on the skin, but it rarely develops into a disease that affects other areas of the body. Nearly 50 percent of patients diagnosed with CBCL experience a recurrence after an initial complete response to treatment. Prognosis is usually very good.

Types of CBCL

Primary Cutaneous Follicle Center Lymphoma is the most common type of CBCL. These skin lymphomas develop slowly over months or years. They usually appear on the head, neck, or torso of the body as a red-brown pimply rash or nodules.

Primary Cutaneous Marginal Zone B-Cell Lymphoma is the second most common form of CBCL. This slow-growing lymphoma appears as pink or red lesions, nodules, and/or tumors, most commonly found on the torso or arms.

Primary Cutaneous Diffuse Large B-Cell Lymphoma, Leg-Type is less common than other CBCLs but is usually more aggressive (fast-growing), developing over weeks or months. This lymphoma usually appears as solitary or multiple tumor nodules on the legs and/or torso. The lesions frequently grow into large tumors that extend deep into the body. The lesions may become open sores and spread outside the skin more frequently than the slow-growing CBCLs.

Primary Cutaneous Diffuse Large B-Cell Lymphoma, Other describes a group of very rare lymphomas, including intravascular large B-cell lymphoma, T-cell–rich large B-cell lymphoma, plasmablastic lymphoma, and anaplastic B-cell lymphoma. These lymphomas usually appear on the head, torso, and extremities.

Treatment Options

Treatment selection for CBCL depends on the type of CBCL, the patient's symptoms and overall health, and the stage of the disease. Depending on the number of lesions present, primary cutaneous follicle center lymphoma and cutaneous marginal zone B-cell lymphoma may be observed (a method known as "watch and wait") or treated locally with either radiation therapy or surgery. Secondary therapy for relapsed (disease returns after treatment) CBCL can include observation, surgery, topical treatments, injected steroids, or radiation. Slow-growing CBCLs rarely transform into more aggressive types of lymphoma. For diseases that become widespread (systemic), a variety of regimens may be used. In these cases, therapies such as rituximab (Rituxan), either alone or in combination with chemotherapy, are often used.

There is no known best treatment strategy for the more aggressive types of CBCL, such as cutaneous diffuse large B-cell lymphoma, leg type. For single tumors, radiation is often effective, as are combinations of chemotherapy or radiation and chemotherapy given sequentially. Similar to other B-cell NHLs, R-CHOP (rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisone) plus radiation therapy is commonly used to treat CBCL that has spread and become a more generalized lymphoma. Clinical trials are also often recommended. Although all of these treatments usually result in disease remission, relapse is common.

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Treatments Under Investigation

Many treatments at various stages of drug development are currently being tested in clinical trials for relapsed or refractory (disease does not respond to treatment) CBCL, including panobinostat (Farydak), lenalidomide (Revlimid), bendamustine (Treanda), and others. It is critical to remember that today's scientific research is continuously evolving. Treatment options may change as new treatments are discovered and current treatments are improved. Therefore, it is important that patients check with their physician or with the Lymphoma Research Foundation (LRF) for any treatment updates that may have recently emerged.

Clinical Trials

Clinical trials are crucial in identifying effective drugs and determining optimal doses for patients with lymphoma. Patients interested in participating in a clinical trial should talk to their physician or contact the LRF Helpline for an individualized clinical trial search by calling (800) 500-9976 or emailing helpline@lymphoma.org.

Follow-up

Patients in remission should have regular visits with a physician who is familiar with their medical history and the treatments they have received.

Some treatments can cause long-term effects or late effects, which can vary based on duration and frequency of treatments, age, gender, and the overall health of each patient at the time of treatment. A physician will check for these effects during follow-up care.

Patients and their caregivers are encouraged to keep copies of all medical records and test results as well as information on the types, amounts, and duration of all treatments received. This documentation will be important for keeping track of any effects resulting from treatment or potential disease recurrences.

Support

A lymphoma diagnosis often triggers a range of feelings and raises concerns. In addition, cancer treatment can cause physical discomfort. Support groups and online message boards can help patients connect with other people who have lymphoma. One-to-one peer support programs, such as the LRF Lymphoma Support Network, match lymphoma patients (or caregivers) with volunteers who have gone through similar experiences.

Resources

LRF offers a wide range of resources that address treatment options, the latest research advances, and ways to cope with all aspects of lymphoma, including our award-winning mobile app. LRF also provides many educational activities, from in-person meetings to teleconferences and webcasts, as well as E-Updates that provide the latest disease-specific news and treatment options. For more information about any of these resources, visit our website at www.lymphoma.org, or contact the LRF Helpline at (800) 500-9976 or helpline@lymphoma.org.