

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 a cell of the immune system called a lymphocyte, a type of white blood cell, grows and multiplies uncontrollably. Cancerous lymphocytes can travel to many parts of the body, including the lymph nodes, spleen, bone marrow, blood, or other organs. 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 lymphocytes.

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 *recur* (return after treatment) in new places 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 bumpy 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, arms, and/or torso. 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. With this strategy, patients' overall health and disease are monitored through regular checkup visits and various evaluating procedures, such as laboratory and imaging tests. Active treatment is started if the patient begins to develop lymphoma-related symptoms or there are signs that the disease is progressing based on testing during follow-up visits. 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 with or without chemotherapy is recommended. Similar to other B-cell NHLs, R-CHOP (rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisone) with or without 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.

Contact the
Lymphoma Research Foundation

Helpline: (800) 500-9976

National

Headquarters: (212) 349-2910

Email: LRF@lymphoma.org

Website: www.lymphoma.org

Medical reviewer:

Lauren Pinter-Brown, MD
UCI Chao Comprehensive Cancer
Center School of Medicine

Supported through grants from:



Genentech
A Member of the Roche Group



© 2016 Lymphoma Research Foundation

Getting the Facts is published by the Lymphoma Research Foundation (LRF) for the purpose of informing and educating readers. Facts and statistics were obtained using published information, including data from the Surveillance, Epidemiology, and End Results (SEER) Program. Because each person's body and response to treatment is different, no individual should self-diagnose or embark upon any course of medical treatment without first consulting with his or her physician. The medical reviewer, the medical reviewer's institution, and LRF are not responsible for the medical care or treatment of any individual.

Last Updated June 2016

Stay Connected through our social media



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 lenalidomide (Revlimid), bendamustine (Treanda), rituximab (Rituxan), 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. The Lymphoma Research Foundation (LRF) provides a Clinical Trials Information Service to increase awareness about investigational treatments for lymphoma being evaluated at cancer treatment centers nationwide. Our LRF Helpline staff will conduct a search for potential lymphoma treatment trials based upon medical information that is provided. Patients are strongly encouraged to discuss with their physician the summaries emailed or mailed by LRF. The patient's cancer specialist will be familiar with their medical history and can best evaluate all of the study criteria to determine if the clinical trial is appropriate.

Patients interested in participating in a clinical trial should view the *Understanding Clinical Trials* fact sheet on LRF's website at www.lymphoma.org, 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. Medical tests (such as blood tests and computed tomography [CT] scans) may be required at various times during remission to evaluate the need for additional treatment.

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 concerns. One-to-one peer support programs, such as LRF's Lymphoma Support Network, connects patients and caregivers with volunteers who have experience with CBCL, similar treatments, or challenges, for mutual emotional support and encouragement. You may find this useful whether you or a loved one is newly diagnosed, in treatment, or in remission.

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, *Focus on Lymphoma*. LRF also provides many educational activities, from in-person meetings to teleconferences and webcasts for people with CBCL, as well as disease-specific websites, videos, and e-Updates for current lymphoma information and treatment options. To learn more 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.