LYMPHOMA RESEARCH • FOUNDATION

Understanding Cutaneous B-Cell Lymphoma

B-cell lymphomas account for approximately 90 percent of all non-Hodgkin lymphoma (NHL) cases in the United States. This 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 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 often *indolent* (slow growing) and may appear on the skin as a reddish bump, lump, or nodule, usually with a raised and smooth appearance. They can appear as a single lesion or multiple lesions in either one body region or several. Sometimes they can look like a rash. The disease can *recur* (return after treatment) or occur in new places on the skin, but it rarely develops into a disease that affects other non-skin areas of the body. About 50 percent of patients with single lesions are cured after radiation therapy. However, patients with multiple lesions are more likely to continue to have new lesions appear.

TYPES OF CBCL

Primary Cutaneous Follicle Center Lymphoma is the most common type of CBCL. These skin lymphomas are indolent, developing slowly over months or years. They usually appear on the head, neck, or torso of the body as a red-brown bumpy skin eruption, nodule(s), or tumor(s).

Primary Cutaneous Marginal Zone B-Cell Lymphoma is the second most common form of CBCL. These indolent lymphomas can have a similar appearance as cutaneous follicle center lymphoma, often as pink or red bumpy lesion(s) or nodule(s), 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, but can involve non-leg areas, such as the arms and/or torso. The lesions may ulcerate and spread outside the skin more frequently than slow-growing CBCLs.

Primary Cutaneous Diffuse Large B-Cell Lymphoma, Other describes a group of very rare, aggressive 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

Upon diagnosis, appropriate staging work-up should be done to make sure that the disease is limited to the skin. In general, this includes routine laboratory tests and whole-body imaging studies. Bone marrow biopsies are not recommended for indolent CBCLs.

Treatment selection for CBCL depends on the type of CBCL, and whether the skin lesion is solitary/regional or multifocal (widespread). For the aggressive CBCLs, the patient's overall health is taken into account for optimal management. The two indolent CBCLs are managed very similarly and are thus discussed together in most guidelines. If the lesion is solitary or localized, the most common treatment is local radiation therapy. Surgical treatment can be an option, but may result in wide, unnecessary scars. Indolent CBCLs that present as multiple lesions may be observed through an approach known as "active surveillance" or "watchful waiting," in which patients' overall health and disease are monitored through regular checkup visits and various evaluation procedures, such as laboratory and imaging tests. Symptomatic lesions can be treated locally with radiation therapy. Sometimes, intralesional steroids, topical therapies, or surgical treatment can be considered. If lesions are very widespread and symptomatic, systemic therapies such as rituximab (Rituxan), which targets B cells, may be appropriate. Regular skin examinations are very important, especially for indolent CBCLs, as skin is the most common site of new lesions. General laboratory tests may also be done, but imaging is not needed unless there is a concern of systemic (widespread) disease.

For aggressive CBCLs, such as DLBCL, Leg-Type, systemic chemotherapy (with or without rituximab) is often more appropriate for initial treatment, with or without radiation therapy. Regular imaging studies are usually done to evaluate treatment response or disease status.

Treatment for *relapse* (disease returns after treatment) of indolent CBCL can include observation, surgery, topical treatments, injected steroids, or radiation (low-dose). Indolent CBCLs usually remain indolent and relapse in the skin. Very rarely, indolent CBCLs relapse as systemic disease, most commonly in regional lymph nodes. In extremely rare cases, indolent CBCLs can transform into more aggressive types of lymphoma.

Relapsed aggressive CBCLs may be treated with chemotherapy (with or without rituximab), targeted therapies such as brentuximab vedotin (Adcetris), ibrutinib (Imbruvica), lenalidomide (Revlimid), radiation therapy, and/or radioimmunotherapy.

TREATMENTS UNDER INVESTIGATION

Many treatments are currently being tested in clinical trials for patients with both newly diagnosed and relapsed/refractory B-cell lymphomas. 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 view the *Understanding Clinical Trials* fact sheet on LRF's website at lymphoma.org/publications, and the *Clinical Trials Search Request Form* at 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 with lymphoma 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, computed tomography [CT] scans, and positron emission tomography [PET] scans) may be required at various times during remission (disappearance of signs and symptoms) 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 side effects resulting from treatment or potential disease recurrences. LRF's award-winning mobile app *Focus On Lymphoma* (lymphoma.org/mobileapp) and the *Lymphoma Care Plan* (lymphoma.org/publications) can help manage this documentation.

SUPPORT SERVICES

A lymphoma diagnosis often triggers a range of feelings and concerns. In addition, cancer treatment can cause physical discomfort. One-to-one peer support programs, such as *LRF's Lymphoma Support Network*, connect patients and caregivers with volunteers who have experience with lymphoma, similar treatments, or challenges, for mutual emotional support and encouragement. Patients and loved ones may find this information useful whether the patient 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. LRF also provides many educational activities, from in-person meetings to webinars for people with CBCL, as well as patient guides and e-Updates that provide the latest disease-specific news and treatment options. To learn more about any of these resources, visit our websites lymphoma.org/CBCL or lymphoma.org, or contact the LRF Helpline at (800) 500-9976 or helpline@lymphoma.org.

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Supported through grants from: Bristol Myers Squibb | Genentech, A Member of the Roche Group | Biogen

Last updated 2021

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