



The Cutaneous Lymphoma Foundation is an independent, non-profit patient advocacy organization dedicated to supporting every person with cutaneous lymphoma by promoting awareness and education, advancing patient care, and facilitating research.

## CUTANEOUS LYMPHOMA FOUNDATION

# Fast Facts: Primary Cutaneous B-Cell Lymphoma

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### What is Lymphoma?

Lymphoma is a cancer of the white blood cells, namely lymphocytes, that constitute the lymphatic system. The two main types of lymphoma are Hodgkin lymphoma and non-Hodgkin lymphoma. Together they comprise more than 35 subtypes of lymphoma. There are five types of Hodgkin lymphoma and at least 30 types of non-Hodgkin lymphoma. Lymphoma is the most common blood cancer and the third most common cancer of childhood. Lymphoma occurs when lymphocytes grow abnormally. The body has two types of lymphocytes: B lymphocytes, or B-cells, and T lymphocytes, or T-cells. Although both cell types can develop into lymphomas, B-cell lymphomas are more common. Like normal lymphocytes, those that turn malignant can grow in many parts of the body, including the lymph nodes, spleen, bone marrow, blood or other organs.

### What is Primary Cutaneous B-Cell Lymphoma?

Lymphomas that grow outside the nodal system, also called extranodal lymphomas, are tumors that occur in organs or tissues outside the lymphatic system. When extranodal lymphomas originate in the skin and there is no evidence of systemic or extracutaneous disease, they are called primary cutaneous lymphomas and prima-

ry cutaneous B-cell lymphomas (CBCLs) when B-cells are involved. Skin is the second most common site of extranodal involvement after the gastrointestinal tract, making up 18 percent of all extranodal lymphomas, and 5 percent of all non-Hodgkin lymphomas. The majority of primary cutaneous lymphomas are of the T-cell type, with CBCL comprising 20 percent to 25 percent of all primary cutaneous lymphomas.

Primary cutaneous B-cell lymphomas are nearly always indolent or slow growing. They may appear on the skin as a reddish rash, lump or nodule and, because they tend to develop in the dermis, or second layer of the skin, may have a slightly raised and smooth appearance. They may recur on the skin but rarely will a primary CBCL develop into a systemic lymphoma.

### Who Gets CBCL?

The disease affects men and women equally and can affect any age group. Like all lymphomas, CBCL is an acquired disease and there are no known risk factors. The disease is not inherited or contagious.

### The Different Types of CBCL

Recently, the World Health Organization and European Organization for Research and Treatment of Cancer Classification reached a con-

sensus on a classification system for CBCL and determined that there are three main types of primary cutaneous B-cell lymphomas:

- 1) primary cutaneous follicle center lymphoma,
- 2) primary cutaneous marginal zone B-cell lymphoma and
- 3) primary cutaneous diffuse large cell lymphoma, leg type.

Prognosis is usually very good, although relapse is common, with nearly 50 percent of CBCL patients experiencing recurrence after an initial complete response to treatment.

Brief descriptions of the main types of CBCL appear below.

**Primary cutaneous follicle center lymphoma (CFCL)** is the most common B-cell lymphoma to develop as a primary tumor of the skin. These skin lymphomas develop slowly over months or years and may manifest as a single tumor or nodule or be grouped together and have a pink or reddish appearance that is slightly raised and smooth and usually do not ulcerate. Cutaneous follicle center lymphoma usually appears on the head, neck or trunk of the body.

**Primary cutaneous marginal zone B-cell lymphoma (CMZL)** is a low-grade malignant B-cell lymphoma that is similar to the MALT (mucosa-associated lymphoid tissue) type and is the second most common form of CBCL. This slow-growing B-cell lymphoma appears as pink or red papules, nodules and/or tumors. Although CMZL can occur anywhere on the body, they are commonly found on the extremities, especially the arms and trunk.

## **Primary cutaneous diffuse large B-cell lymphoma**

**Primary cutaneous diffuse large B-cell lymphoma, leg type (LBCL-L)** is less common than other types of CBCL and more aggressive, usually appearing on the lower legs of elderly women, although lesions can occur on any part of the body. The lesions are red or bluish-red and frequently grow into large tumors that extend deep into the fat. Unlike cutaneous follicle center lymphoma, LBCL-L tumors develop quickly over weeks and months, usually becoming open sores and spreading outside the skin.

**Primary cutaneous diffuse LBCL-other** describes a group of very rare diffuse LBCLs that do not fit into the other LBCL categories. They include intravascular LBCL, T-cell-rich LBCL, plasmablastic lymphoma and anaplastic B-cell lymphoma. Although these lymphomas usually appear on the head, trunk or extremities, they are often cutaneous manifestations of systemic lymphomas and have to be treated the same way.

## **How CBCL Is Diagnosed and Staged**

A biopsy of the tumor tissue is needed to render a diagnosis and determine CBCL classification. Once a diagnosis is made, CBCL patients are either referred to an oncologist or a dermatologist specializing in CBCL for clinical staging to eliminate the possibility that the disease originated outside the skin. A comprehensive CBCL staging evaluation includes:

- A physical examination of the entire skin

and lymphatic system

- Laboratory tests, including complete blood and serum counts and serum lactate dehydrogenase (LDH) measurements
- Imaging tests, including a CT (computerized axial tomography) or PET (positron emission tomography) scan of the chest, abdomen and pelvis
- A bone marrow biopsy may be recommended for more aggressive subtypes of CBCL. A bone marrow biopsy may not be necessary for patients with cutaneous follicle center lymphoma or marginal zone lymphoma if systemic staging is negative

### **Treating CBCL**

Primary cutaneous follicle center lymphoma and cutaneous marginal zone lymphoma are among the most slow growing or indolent lymphomas and, depending on the number of lesions present, may be treated locally with either radiation or surgery. Topical steroid cream or topical chemotherapy may also be used. Sometimes no treatment is needed. Most often these slow-growing lymphomas recur over time, usually only on the skin. As many as 50 percent of people with single lesions that get radiation or excision will never get another lesion. Indolent CBCLs rarely transform into more aggressive types of lymphoma. For diseases that do transform, become widespread or in rare cases when they become systemic, a variety of chemotherapy regimens may be used. Other therapies such as the monoclonal antibody Rituxan (rituximab), either alone or in combination with chemotherapy, and interferon may also be recommended.

There is no known best treatment strategy for the

more aggressive type of CBCL, primary 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. All of these treatments usually put the disease in remission, although relapse is common. Research is underway to test the most effective strategies for treating cutaneous B-cell lymphomas.

### **How to Find Support**

Getting a lymphoma diagnosis can be frightening and treatment can cause physical and emotional discomfort. Connecting with other people who have primary cutaneous B-cell lymphoma can be extremely helpful. Support groups and online message boards are often useful. One-to-one peer support programs, such as the Lymphoma Research Foundation's Lymphoma Support Network, matches lymphoma survivors (or caregivers) with volunteers who have gone through similar experiences. The Cutaneous Lymphoma Foundation offers information on patient assistance programs, free e-mail listservs, where to find clinical trials and the latest research on its Web site at [www.clfoundation.org](http://www.clfoundation.org).

### **How to Stay Informed**

The Cutaneous Lymphoma Foundation is a patient advocacy organization dedicated to educating patients and caregivers about cutaneous lymphomas and improving access to safe and effective treatments. For more information about these resources:

Call: 248-644-9014

Web site: [www.clfoundation.org](http://www.clfoundation.org)



## Contact Us

For more information about the Cutaneous Lymphoma Foundation, please contact:

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The Cutaneous Lymphoma Foundation offers a comprehensive array of patient education and support programs, including:

- Comprehensive patient guides
- Treatment Information
- Patient Referral Information
- Select publications in Spanish
- Forum (newsletter)
- On-line support group
- Patient Assistance Resources
- Information teleconferences and webcasts
- Regional Educational Forums

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Because each person's body and response to treatment is different, no individual should indulge in self-diagnosis or embark upon any course of medical treatment without first consulting with his or her physician.

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## Glossary of Terms

### Biopsy

Removal of a small piece of tissue (for example, a lymph node) for evaluation under a microscope.

### Chemotherapy

Treatment with "chemo" drugs to stop the growth of rapidly dividing cancer cells, including lymphoma cells.

### Complete remission

The term used when all signs of disease have disappeared after treatment.

### Electron beam radiation

Radiation of the skin that does not penetrate to internal organs.

### Lymph nodes

Small bean-shaped glands located in the small vessels of the lymphatic system. Thousands are located throughout the body and are most easily felt in the neck, armpits and groin.

### Lymphatic system

The vessels, tissues and organs that store and carry lymphocytes that fight infection and other diseases.

### Lymphocyte

A type of white blood cell.

### Ultraviolet light therapy

UVB ultraviolet light is used to slow the rapid growth of skin cells. Light boxes with full-body exposure are used to deliver ultraviolet rays that can treat cutaneous T-cell lymphoma.