

# PRIMARY CUTANEOUS B-CELL LYMPHOMA

## What is Primary Cutaneous B-Cell Lymphoma?

Primary cutaneous B-cell lymphoma (CBCL) is a form of cancer that originates from B cells that present in the skin and nowhere else in the body, often referred to as an “extranodal” (outside the lymph node) non-Hodgkin’s lymphoma. The skin is the second most common site of extranodal presentation, after lymphomas in the gastrointestinal tract. CBCLs represent about one quarter, 20-25%, of primary cutaneous lymphomas and are different in presentation and behavior from cutaneous T cell lymphomas of the skin (75-80% of primary cutaneous lymphomas). To be diagnosed with a primary cutaneous B cell lymphoma, you must have no evidence of systemic (internal) disease, usually done by medical imaging of the body at the time of presentation. However, in cases with a classic clinical presentation and pathology findings on biopsy, imaging may not be needed.

Most primary cutaneous B-cell lymphomas are indolent or slow growing. They typically appear on the skin as a single to several firm pink-to-purplish thin to very raised small bumps, or lumps or nodules of varying sizes. They tend to develop in the dermis, or second layer of the skin, giving it a raised, smooth appearance (no scale like dry skin). After treatment they may recur on the skin but rarely will a primary CBCL develop into a systemic lymphoma.

## Who Gets Primary Cutaneous B-Cell Lymphoma?

The disease affects men and women equally and can affect any age group. CBCL is an acquired disease. The disease is not inherited or contagious. There are no proven risk factors that make it more likely one will develop CBCL. However, underlying autoimmune disease, type II diabetes mellitus, and history of other cancers has been proposed as possible risk factors.

## What is the Prognosis for Primary Cutaneous B-Cell Lymphoma?

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 for the most common types of CBCLs.

## The Different Types of Primary Cutaneous B-Cell Lymphoma

The World Health Organization and European Organization for Research and Treatment of Cancer Classification reached a consensus on a classification system for CBCL and determined that there are three main types of primary cutaneous B-cell lymphomas:

- Primary cutaneous follicle center lymphoma
- Primary cutaneous marginal zone B-cell lymphoma
- Primary cutaneous diffuse large cell lymphoma, leg type

### Primary cutaneous follicle center lymphoma (CFCL)

Primary cutaneous follicle center lymphoma (CFCL) is the most common cutaneous B-cell lymphoma. CFCL develops slowly over months or years, and may look like a single bump or several grouped together. They typically have a pink or purplish appearance that is slightly raised and smooth, and usually do not ulcerate. CFCL is most common on the head, neck or trunk of the body.

### Primary cutaneous marginal zone B-cell lymphoma (CMZL)

Primary cutaneous marginal zone B-cell lymphoma (CMZL) is the second most common form of CBCL. CMZL is a low-grade B-cell lymphoma. This slow-growing B-cell lymphoma appears as pink or purplish papules, nodules and/or tumors. Although CMZL can occur anywhere on the body, they are commonly found on the upper extremities, especially the arms, trunk, or head.

### Primary cutaneous diffuse large B-cell lymphoma, leg type (LBCL-L)

Primary cutaneous diffuse large B-cell lymphoma, leg type (LBCL-L) is less common than other types of CBCL and can be more aggressive. LBCL-L usually appears 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 quickly into large tumors that can ulcerate.

## How Primary Cutaneous B-Cell Lymphoma Is Diagnosed

Typical procedures done to diagnose CBCL include:

- A complete physical exam (including a thorough



- skin exam);
- A skin biopsy (removal of a small piece of tissue) for examination under the microscope by a pathologist (a doctor who studies tissues and cells to identify diseases);
- Blood tests;
- Imaging tests such as CT (computerized axial tomography) and/or PET (positron emission tomography) scans.

When a skin biopsy shows B-cell lymphoma it is very important to make sure that the lymphoma is truly coming from the skin, and not from a systemic lymphoma that has spread to the skin. It is also important that any diagnosis of CBCL is confirmed by a pathologist who has expertise in diagnosing cutaneous lymphomas.

### **Treating Primary Cutaneous B-Cell Lymphoma**

Primary cutaneous follicle center lymphoma and cutaneous marginal zone lymphoma are among the most slow growing or indolent lymphomas:

- Depending on the number of lesions present, solitary or few lesions may be treated locally with either radiation or surgery.
- Topical steroid cream, topical chemotherapy, or injections 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.
- Numerous and highly bothersome lesions may be eligible for treatment with rituximab.

Primary cutaneous diffuse large-B cell lymphoma-leg type, often require treatment chemotherapy, radiation, or a combination of both.

#### **Reviewed and updated:**

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