

PRIMARY CUTANEOUS B-CELL LYMPHOMA

What is Primary Cutaneous B-Cell Lymphoma?

Primary cutaneous B-cell lymphoma (CBCL) is a form of “extranodal” (outside the lymph node) lymphoma. The skin is the second most common site of extranodal involvement after the gastrointestinal tract. 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. CBCLs originate in the skin, and the diagnosis of CBCL requires that there is no evidence of systemic (internal) disease. CBCLs are also called primary cutaneous B-cell lymphomas.

Most primary cutaneous B-cell lymphomas are 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 Primary Cutaneous B-Cell Lymphoma?

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

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.

The Different Types of Primary Cutaneous B-Cell Lymphoma

Recently, 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 develop slowly over months or years, and may look like a single tumor or several tumors of nodules grouped together. They typically have a pink or reddish 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 that is similar to the MALT (mucosa-associated lymphoid tissue) type lymphomas. 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, 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.

Primary cutaneous diffuse large B-cell lymphoma - other (LBCL-other)

Primary cutaneous diffuse large B-cell lymphoma - other (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 may need to be treated differently than other CBCLs.



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 and/or lymph node 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 and, depending on the number of lesions present, may be treated locally with either radiation or surgery.
- Topical steroid cream, topical chemotherapy, or injections may also be used.
- Rituximab is an infusion drug that is sometimes used for extensive indolent CBCL.
- 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.

There is no known best treatment strategy for primary cutaneous diffuse large-B cell lymphoma-leg type, but often treatments require radiation, chemotherapy, or a combination of both.

Reviewed and updated:

December 2019 - Cutaneous Lymphoma Foundation's Medical Advisory Council



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