

PRIMARY CUTANEOUS ANAPLASTIC LARGE CELL LYMPHOMA

What is Primary Cutaneous Anaplastic Large Cell Lymphoma?

Primary cutaneous anaplastic large cell lymphoma (PCALCL) is a subtype of cutaneous T-cell lymphoma (CTCL), and is a non-Hodgkin lymphoma (cancer of the white blood cells) that arises in the skin. PCALCL is named from the description of the size and shape of the cells under the microscope (“anaplastic large cell”), and is characterized by the presence of a molecule on the lymphoma cells called CD30.

It is important to know that there are two types of ALCL:

- **Systemic (or nodal) ALCL**, which can affect the skin and/or lymph nodes and other internal organs; and,
- **Primary cutaneous ALCL**, which affects the skin primarily.

PCALCL is considered a skin lymphoma, but is sometimes considered a variant of systemic ALCL. PCALCL is part of a family of primary cutaneous CD30+ lymphoproliferative disorders (CD30+ LPD). The CD30+ LPDs also includes lymphomatoid papulosis (LyP) and “borderline cases” that are somewhere between LyP and PCALCL. The CD30+ LPDs as a group account for approximately 10 percent of cutaneous lymphomas. PCALCL generally has an excellent prognosis when compared to systemic ALCL.

Who Gets Primary Cutaneous Anaplastic Large Cell Lymphoma?

There are no known risk factors for PCALCL. The disease can affect people of all ages, including children, but is most commonly found in adults 45 to 60 years old, and occurs more often in men than women.

What does Primary Cutaneous Anaplastic Large Cell Lymphoma Look Like?

The characteristic features of PCALCL are single or multiple raised red skin lesions, nodules or tumors. Any area of the skin can be affected, and lesions may be present for a long time before being diagnosed. Symptoms may include pain or itching, though many patients don’t notice any symptoms from their PCALCL.

Forty percent of people with PCALCL will have more than one episode or lesion in their lifetime. Some PCALCL (up to 40 percent) can spontaneously regress (go away) without any treatment. Most of the time, PCALCL remains only in the skin, but 10 percent of

people with PCALCL develop involvement of local lymph nodes. Extensive lymph node involvement or internal organ (visceral) disease is rare.

How is Primary Cutaneous Anaplastic Large Cell Lymphoma Diagnosed?

Typical procedures done to diagnose primary cutaneous anaplastic large cell lymphoma requires:

- A complete physical exam (including a thorough skin exam);
- 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; and,
- Imaging tests such as CT (computerized axial tomography) and/or PET (positron emission tomography) scans. If imaging tests show lymph node or other organ involvement, a bone marrow biopsy, in which soft tissue is removed from inside the bone, might also be performed.

It is important to know that there is no reliable way to tell systemic ALCL apart from primary cutaneous ALCL based only on a skin biopsy. The accurate diagnosis of PCALCL relies on examining the skin and imaging studies to verify that it is only in the skin and not lymph nodes or other organs at the time of diagnosis. It is also very important that any diagnosis of skin lymphoma is confirmed by a pathologist who has expertise in diagnosing cutaneous lymphomas.

How is Primary Cutaneous Anaplastic Large Cell Lymphoma Staged?

There are different stages of PCALCL. It’s very important to know that the staging system for PCALCL is different than the staging system for systemic ALCL. The staging of PCALCL is based on a TNM system:

T:

T1

Only a single spot or area of the skin is involved.

T2

Multiple skin lesions in one area or adjacent areas of the body. Either of the following may be true: the skin contains dry, red, scaly patches, but no tumors. Enlarged lymph nodes may be present, but they do not contain cancer cells. Or, there are tumors on the skin and lymph nodes are either normal or larger than normal, but they do not contain cancer cells.



T3

Skin lesions of widespread areas of skin, such as areas far apart on the body or three adjacent areas.

N:

N0: No lymph node involvement

N1 – 3: Lymph node involvement

M:

M0: No other organ involvement

M1: Organ (non-lymph node) involvement

For example, someone with a single lesion on the body and no lymph node or organ involvement would be stage T1N0M0.

Reference:

<http://www.bloodjournal.org/content/110/2/479.full>

What is the Prognosis for Primary Cutaneous Anaplastic Large Cell Lymphoma?

PCALCL has a generally excellent prognosis, with a 10-year survival of greater than 90 percent, even if there is local lymph node involvement. Factors that might worsen prognosis are the presence of multiple lesions in a limb (like the leg), or generalize (T3) skin involvement. While primary cutaneous anaplastic large cell lymphoma, like other non-Hodgkin lymphomas, is considered a chronic disease, the cancer can be cured in people with localized single lesions.

How Primary Cutaneous Anaplastic Large Cell Lymphoma is Treated

There are many options for treating PCALCL available, but localized treatment with either radiation therapy or surgical excision are the preferred therapies for single lesions of PCALCL. Radiation therapy is most commonly used, and has a response rate of 100 percent. Radiation therapy has been shown to be more effective and longer lasting than using multi-agent (“traditional”) chemotherapy.

For multiple or widespread lesions in which radiation therapy or surgery aren’t good options, there are several possible alternatives for treating PCALCL. As research is continuously changing, there may be other options that are not listed here:

- Bexarotene (Targretin®) capsules
- Brentuximab vedotin
- CHOP (cyclophosphamide, doxorubicin, vincristine, prednisone) chemotherapy
- CVP (cyclophosphamide, vincristine, prednisone) chemotherapy
- Imiquimod
- Low doses of methotrexate (Trexall®)
- Nitrogen mustard (Mustargen® topical)
- PUVA (Psoralen and ultraviolet-A light)
- Thalidomide

All of these therapies are effective in treating the disease, although relapse is common and, therefore, long-term follow-up care is required.

Participating in Clinical Trials

Clinical trials are crucial in identifying effective drugs, prognostic strategies and determining optimal doses for lymphoma patients. If you are interested in participating in a clinical trial, talk to your doctor about an appropriate trial for you. To learn more about clinical trials, visit the Cutaneous Lymphoma Foundation’s website: www.clfoundation.org.

Are Complementary and Alternative Therapies Safe and Effective?

Complementary and alternative medicines are nonstandard therapies that may help patients cope with their cancer and its treatment, but that should not be used in place of standard treatment. No alternative therapy has ever been proven effective against lymphoma. However, complementary therapies such as meditation, yoga, acupuncture, exercise, diet and relaxation techniques have been shown to be effective in combating some treatment side effects and for improving your overall health during therapy. Before embarking on any complementary therapies, patients should discuss the matter with their healthcare team. Certain unproven treatments, including some herbal supplements, can interfere with standard lymphoma treatments or may cause serious side effects.

How to Prepare for Follow-up Treatments

It is important to be proactive in your healthcare, including keeping a master file of medical records, writing down and asking questions, reporting new symptoms, exercising and eating a balanced diet. In addition, patients who smoke should strongly consider stopping. Follow-up visits for people with primary cutaneous anaplastic large cell lymphoma often depends on the stage of the disease and treatment and can range from as frequently as every few weeks when starting new therapies that require monitoring to as little as every six months.

Typically, follow-up visits include physical examinations, blood tests and occasionally imaging tests such as CT or PET scans. Besides determining disease recurrence, follow-up care can help identify and resolve unusual side effects of treatment.

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