



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 Anaplastic Large Cell Lymphoma

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What is Lymphoma?

Lymphoma is a cancer of the white blood cells, namely lymphocytes, which constitute the lymphatic system. The two main types of lymphoma are Hodgkin lymphoma and non-

Hodgkin lymphoma (NHL). 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 main types of lymphocytes: B lymphocytes, or B-cells, and T lymphocytes, or T-cells. B-cells play an important role in making antibodies to fight bacterial infections and T-cells play a role in fighting viruses and organ rejection in transplant patients. Although both cell types can develop into lymphomas, B-cell lymphomas are more common, comprising nearly 85 percent of all non-Hodgkin lymphomas. Like normal lymphocytes, those that become malignant can grow in any part of the body, including the lymph nodes, spleen, bone marrow, blood or other organs.

What is Non-Hodgkin Lymphoma?

Together, Hodgkin lymphoma and non-Hodgkin lymphoma comprise more than 67 subtypes of lymphoma. There are six primary types of

Hodgkin lymphoma and as many as 61 types of non-Hodgkin lymphoma.

Nearly all non-Hodgkin lymphoma cases occur in adults, with the average age of diagnosis occurring in the 60s, but lymphomas can develop at any age. While scientists do not know the exact causes of non-Hodgkin lymphoma, they do know that it is not caused by injury or by coming in contact with someone with the disease. Most people diagnosed with NHL have no known risk factors, although increasingly many scientists believe infections may contribute to the development of certain types of non-Hodgkin lymphoma. According to the American Cancer Society, there were approximately 66,120 new cases of NHL diagnosed in 2008. The disease is more common in men than in women.

What is Primary Cutaneous Anaplastic Large Cell Lymphoma?

Primary cutaneous T-cell lymphomas (CTCL) are part of a group of rare non-Hodgkin lymphomas that arise from the T-cell type lymphocytes. Included in this group is primary cutaneous anaplastic large cell lymphoma. Anaplastic large cell lymphoma (ALCL) comprises only about three percent of all lymphomas in adults and between ten percent and 30 percent of all lymphomas in children. The disease is characterized by the size

and shape of the cells under the microscope and by the uniform expression of a special marker on the lymphoma cells called CD30. There are two types of anaplastic large cell lymphoma: systemic, which can affect the skin and/or lymph nodes and other internal organs; and primary cutaneous, which affects the skin only. However, in rare instances, over time, primary cutaneous ALCL may extend beyond the skin to internal organs.

Primary cutaneous anaplastic large cell lymphoma is an indolent, or slow growing, lymphoma and is associated with a rare condition called lymphomatoid papulosis (LyP), which, while classified as a lymphoma, always goes away by itself. Lymphomatoid papulosis is occasionally a precursor to the development of cutaneous anaplastic large cell lymphoma or other lymphomas. There are no known risk factors for primary cutaneous anaplastic large cell lymphoma. Although the disease can affect people of all ages, including children, it is most commonly found in adults 45 to 60 years old, and typically occurs more often in men than women.

The characteristic features of primary cutaneous anaplastic large cell lymphoma include the appearance of solitary or multiple raised red skin lesions, nodules or tumors, which do not go away and have a tendency to ulcerate and may itch. The lesions can appear on any part of the body and often grow very slowly and may be present for a long time before being diagnosed.

How Primary Cutaneous Anaplastic Large Cell Lymphoma Is Diagnosed

Distinguishing primary cutaneous anaplastic large cell lymphoma from systemic anaplastic

large cell lymphoma requires a biopsy (removal of a small piece of tissue) of the skin lesion, followed by a series of other diagnostic tests, including blood, CT (computerized axial tomography), MRI (magnetic resonance imaging) and/or PET (positron emission tomography) scans. After a sample of the skin lesion is removed, it is examined by a pathologist (a doctor who studies tissues and cells to identify diseases) under a microscope to determine a diagnosis. Because a pathological diagnosis of cutaneous anaplastic large cell lymphoma can sometimes be difficult to determine, it may be necessary to have the pathology report reviewed by a dermatopathologist or a hematopathologist, a pathologist who is an expert in diagnosing lymphomas.

In addition to these diagnostic tests, in some instances, for example, 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 to determine the extent, or stage, of the disease.

The four stages of cutaneous anaplastic large cell lymphoma include:

- Stage 1—The cancer affects only a single spot or single area of the skin. Lymph nodes are not enlarged.
- Stage 2—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.
- Stage 3—Nearly all the skin is covered in dry, red, scaly lesions. The lymph nodes

are either normal or larger than normal, but they do not contain cancer cells.

- State IV—In addition to the skin being involved, cancer cells are found in the lymph nodes or the cancer has spread to other organs such as the liver or lung.

How Primary Cutaneous Anaplastic Large Cell Lymphoma Is Treated

While primary cutaneous anaplastic large cell lymphoma, like other non-Hodgkin lymphomas, is considered a chronic disease, the cancer is occasionally cured in some people with localized single lesions. Even when multiple skin sites are involved, treatment for primary cutaneous ALCL is generally nonaggressive and the prognosis is usually excellent. While most people will have disease that is always confined to the skin, over a lifetime, between five percent and 15 percent of people with primary cutaneous anaplastic large cell lymphoma may develop involvement of the lymph nodes or other sites outside the skin, in which case the cancer is treated more aggressively with combination regimens of chemotherapy and sometimes radiation therapy. Very aggressive treatments, such as bone marrow transplantation, are rarely considered.

There are many therapies available to treat primary cutaneous ALCL both for single and multiple lesions. For localized disease, meaning the existence of a single or a few clustered lesions, nodules or tumors limited to one area of the body, radiotherapy is preferred, although surgical excision or topical treatments such as bexarotene (Targretin) or nitrogen

mustard (Mustargen) may be used. For people with multiple sites, systemic treatment is usually needed and may include:

- Bexarotene (Targretin) capsules
- CVP (cyclophosphamide, vincristine, prednisone) chemotherapy
- Low doses of methotrexate (Trexall) in either oral or injection form
- The monoclonal antibody denileukin difitox (Ontak)
- PUVA (Psoralen and ultraviolet-A light), a type of photodynamic therapy
- Vorinostat (Zolinza), a histone deacetylase inhibitor

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

Treatments Under Investigation

Because primary cutaneous ALCL is such a rare disease and few people will need systemic therapy, finding enough patients to enroll in clinical trials is difficult. Oftentimes, people with primary cutaneous ALCL may be included in trials for cutaneous T-cell lymphoma or for other forms of T-cell lymphoma. Some agents showing promising results in clinical trials include:

- Anti-CD30 monoclonal antibodies
- PDX (pralatrexate)

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 at 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. 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 Appointments

It is important for patients both during and after treatment to be proactive in their healthcare, including keeping a master file of medical records, 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.

Finding Support

Getting a lymphoma diagnosis can be frightening and treatment can cause physical and emotional discomfort. Connecting with other people who have primary cutaneous anaplastic large cell lymphoma can be extremely helpful. 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 website at www.clfoundation.org. 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 at www.lymphoma.org, matches lymphoma survivors (or caregivers) with volunteers who have gone through similar experiences.

Staying 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

Website: www.clfoundation.org

The Lymphoma Research Foundation offers a wide range of resources on the latest treatment and research advances, and services for coping with lymphoma. For more information about any of these resources:

Call: 800-500-9976

E-mail: helpline@lymphoma.org

Website: www.lymphoma.org



Contact Us

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

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Who We Are

The Cutaneous Lymphoma Foundation is an independent, nonprofit patient advocacy organization dedicated to supporting patients with cutaneous lymphomas by promoting awareness and education, advancing patient care and facilitating research.

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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.

Chemotherapy regimen

Combinations of anti-cancer drugs given at a certain dose in a specific sequence according to a strict schedule.

CT or CAT (computerized axial tomography) scan

This imaging test provides a series of detailed pictures of the inside of the body using an X-ray machine linked to a computer.

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.

PET (positron emission tomography) scan

PET scans may be used instead of gallium scans to identify areas in the body that are affected by lymphoma. This test evaluates metabolic activity in different parts of the body using a radioisotope.

Relapse

The return of cancer after treatment. Lymphoma may recur in the area where it first started or it may relapse in another area of the skin.

Stage

The extent of cancer on the skin or in the body, including whether the disease has spread from the original site to other body parts.