FAST FACTS:

ANAPLASTIC LARGE CELL LYMPHOMA (ALCL)

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 papulois (LyP), which, while classified as a lymphoma, always goes away by itself. Lymphomatoid papulois 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 and Staged

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.
- Stage 4—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 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

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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
- Brentuximab Vedotin
- CVP (cyclophosphamide, vincristine, prednisone) chemotherapy
- Low doses of methotrexate (Trexall)
- PUVA (Psoralen and ultraviolet-A light), a type of photodynamic therapy
- PDX (pralatrexate)
- Romidepsin
- Vorinostat (Zolinza), a histone deacety-lase inhibitor

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

Prognosis

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.

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 Treatments

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.

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