

Anaplastic Large Cell Lymphoma

Overview

Lymphoma is the most common blood cancer. The two main forms of lymphoma are Hodgkin lymphoma (HL) and non-Hodgkin lymphoma (NHL). Lymphoma occurs when lymphocytes, a type of white blood cell, grow abnormally. The body has two main types of lymphocytes that can develop into lymphomas: B-lymphocytes (B-cells) and T-lymphocytes (T-cells). Cancerous lymphocytes can travel to many parts of the body, including the lymph nodes, spleen, bone marrow, blood or other organs, and can accumulate to form tumors.

Anaplastic large cell lymphoma (ALCL) is a rare type of NHL, but the second or third most common subtype of T-cell lymphoma. There are three types of ALCL and altogether they comprise about 3 percent of all NHLs in adults and between 10 percent and 30 percent of all NHLs in children. It can present either systemically (in lymph nodes or organs throughout the body) or in the skin.

When ALCL presents in the skin it is called primary cutaneous ALCL and follows a less aggressive course. In almost all cases of primary cutaneous ALCL, the disease is confined to the skin. Despite a tendency to relapse, the relapses are usually in the skin only. As long as it is confined to the skin, it is usually managed as an indolent (slow-growing) lymphoma. Approximately 10 percent of the time, primary cutaneous ALCL extends beyond the skin to lymph nodes or organs. If this occurs, it is usually managed like the systemic forms of ALCL.

Occasionally, primary cutaneous ALCL is associated with another rare condition called lymphomatoid papulosis (LyP). LyP is a skin condition with similar features to primary cutaneous ALCL. While LyP is classified as a lymphoma, the skin lesions always go away by themselves, usually over a four to eight week period, and, therefore, do not behave like a malignancy.

The characteristic features of primary cutaneous ALCL include the appearance of solitary or multiple raised red skin lesions, nodules or tumors, which do not go away, have a tendency to

ulcerate and may itch. The lesions can appear on any part of the body, often grow very slowly and may be present for a long time before being diagnosed.

Patients with systemic ALCL are divided into two groups, depending on the expression of a protein called anaplastic lymphoma kinase (ALK). While both lymphomas are treated as aggressive lymphomas, the prognosis for ALCL depends on whether a patient is ALK positive (expresses the protein) or ALK negative (does not express the protein). ALK positive disease responds well to chemotherapy, putting most patients in long-term remission or cure. Most people with ALK negative ALCL respond to chemotherapy, but many will relapse within five years. Because of this, they are sometimes treated more aggressively, often with stem cell transplant. The ALK positive subtype usually affects children and young adults. The ALK negative subtype is more commonly found in older patients over age 60.

Symptoms of ALCL include weight loss, night sweats, enlarged lymph nodes throughout the body (especially in the neck or armpits) and skin lesions in the cutaneous form.

Treatment Options

Although standard chemotherapy, such as CHOP (cyclophosphamide, doxorubicin, vincristine and prednisone), is given to newly diagnosed systemic ALCL patients, long-term disease outcome varies depending on the subtype. The ALK positive type responds extremely well to CHOP, rendering over a 70 percent long-term disease-free survival, whereas the ALK negative type has a poorer prognosis with less than 45 percent to 50 percent achieving long-term disease-free survival. Higher doses of chemotherapy followed by a stem cell transplant may be prescribed for relapsed patients or patients with a low chance of being cured by CHOP chemotherapy. In 2011, the U.S. Food and Drug Administration (FDA) granted accelerated approval of brentuximab vedotin (Adcetris), an antibody-drug conjugate which targets CD30, for the treatment of patients with relapsed or refractory systemic anaplastic large cell lymphoma (ALCL).

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The Lymphoma Research Foundation offers the following patient education and support programs:

- *Lymphoma Helpline*
- Clinical Trials Information Service
- Lymphoma Support Network
- Publications
- Teleconferences
- Webcasts & podcasts
- In-person conferences

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Primary cutaneous ALCL is very different than systemic ALCL in terms of prognosis and management. If the disease is localized to a single lesion or single area, radiation therapy will leave about half of people in long-term remission. If there are multiple lesions or relapsed disease in the skin, radiation can eradicate the skin lesions, but will not reduce the likelihood of new lesions developing in the future. Despite this tendency to relapse, as long as the relapses are confined to the skin, the long-term prognosis remains excellent. At relapse, radiation, topical treatments, mild chemotherapies, biologic therapies, or excisions for small lesions can all be used successfully.

For those with primary cutaneous ALCL appearing in multiple sites on the body, systemic treatment is usually needed and may include:

- Mild chemotherapy (single agents or mild combinations)
- Bexarotene (Targretin) capsules
- CVP (cyclophosphamide, vincristine, prednisone) chemotherapy
- Methotrexate (Trexall), oral or injection form

All of the therapies listed above are often effective in treating the disease, however, relapse is common. Therefore, long-term follow-up care is required.

Treatments Under Investigation

Because ALCL is such a rare disease, finding enough patients to enroll in clinical trials is difficult. However, there are several new drugs now in clinical trials that are showing promising results, including Pralatrexate (Folotyn).

Novel combination chemotherapy regimens are also being studied in ongoing trials for patients with systemic T-cell lymphomas:

- CHOP chemotherapy, plus bevacizumab (Avastin)
- PEGS (cisplatin, etoposide, gemcitabine and solumedrol) chemotherapy

Participating in Clinical Trials

Clinical trials are crucial in identifying effective drugs and determining optimal doses for lymphoma patients. Patients interested in participating in a clinical trial should talk to their physician. Contact the Lymphoma Research Foundation's *Helpline* for an individualized clinical trial search by calling (800) 500-9976 or emailing helpline@lymphoma.org.

Resources

The Lymphoma Research Foundation offers a wide range of resources that address treatment options, the latest research advances and coping with all aspects of lymphoma. For a more comprehensive source of NHL information, please visit the Foundation's website to view or order the publication entitled *Understanding Non-Hodgkin Lymphoma: A Guide for Patients, Survivors and Loved Ones*. The Foundation also provides many educational activities, from in-person meetings to teleconferences and webcasts.

For more information about any of these resources, visit the website at lymphoma.org, e-mail the *Helpline* at helpline@lymphoma.org or call at (800) 500-9976.