LYMPHOMA
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Getting the Facts

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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 cells of the immune system called lymphocytes, a type of white blood cell, grow and multiply uncontrollably. Cancerous lymphocytes can travel to many parts of the body, including the lymph nodes, spleen, bone marrow, blood, or other organs, and form a mass called a tumor. The body has two main types of lymphocytes that can develop into lymphomas: B lymphocytes (B cells) and T lymphocytes (T cells).

Anaplastic large cell lymphoma (ALCL) is a rare type of NHL, but one of the more common subtypes of T-cell lymphoma. ALCL comprises about two percent of all NHLs and approximately 20 percent of all T-cell lymphomas. Initial symptoms of ALCL can include fever, backache, painless swelling of lymph nodes, loss of appetite, and tiredness.

ALCL can initially appear either in the skin, in lymph nodes, or in organs throughout the body. ALCL that appears in the skin is called primary cutaneous ALCL, which has a less aggressive disease course than the systemic (throughout the body) types. The characteristic features of primary cutaneous ALCL include the appearance of solitary or multiple raised, red skin lesions that do not go away, have a tendency to ulcerate, and may itch. These ALCL lesions are tumors, and they can appear on any part of the body, often grow very slowly, and may be present for a long time before being diagnosed. Approximately 10 percent of the time, primary cutaneous ALCL extends beyond the skin to lymph nodes or organs. If this occurs, it is usually treated like the systemic forms of ALCL.

Patients with systemic ALCL are divided into two groups, depending on whether or not their cells have an abnormal form of a protein on their surface called “anaplastic lymphoma kinase” (ALK). Although both lymphomas are treated as aggressive (fast-growing) lymphomas, the disease course may be different in patients who have ALK-positive (have the abnormal protein) or ALK-negative (do not have the abnormal protein) ALCL. ALK-positive ALCL responds well to standard chemotherapy treatments, putting most patients in long-term remission. Most people with ALK-negative ALCL initially respond to treatment, but more people will relapse within five years than those with ALK-positive ALCL. ALK-negative patients (and some ALK-positive patients with certain risk factors) are sometimes treated more aggressively, often with a stem cell transplantation after remission. ALK-positive ALCL usually affects children and young adults. ALK-negative ALCL is more common in patients over the age of 55 years.

A diagnosis of ALCL requires taking a small sample of tumor tissue or abnormal skin tissue, called a biopsy, and looking at the cells under a microscope. Additional tests may be conducted that give physicians more information about the disease and whether or not it has spread (and if so, how far) in the body. The assessments can include blood tests, a computed axial tomography (CAT) scan, a positron emission tomography (PET) scan, a magnetic resonance imaging (MRI) scan, and a bone marrow biopsy.

Treatment Options

Treatment of primary cutaneous ALCL depends on the extent of skin lesions. If the disease is confined to a single lesion or area, radiation therapy or surgical excision will result in complete remission in approximately 95 percent of patients. If there are multiple lesions or relapsed (disease returns after treatment) disease in the skin, radiation can eradicate the skin lesions but will not reduce the likelihood of new lesions developing. Although primary cutaneous ALCL tends to relapse in about 40 percent of cases, the long-term prognosis remains excellent as long as relapses are confined to the skin. At relapse, radiation, topical treatments, mild chemotherapies, biologic therapies, or excisions for small lesions have all been shown to be successful in controlling the disease.

For those with primary cutaneous ALCL appearing in multiple sites on the body, systemic treatment, which travels through the blood and affects many parts of the body, is usually needed.

Although patients with newly diagnosed systemic ALCL respond well to common first-line chemotherapy regimens such as CHOP (cyclophosphamide, doxorubicin, vincristine, and prednisone), long-term disease outcome varies depending on the subtype. Patients with ALK-positive disease usually respond very well to CHOP or CHOEP (CHOP including etoposide), with a three-year overall survival rate of about 90 percent. In contrast, patients with ALK-negative disease have three-year survival rates of about 60 percent with similar treatments. In some circumstances, higher doses of chemotherapy followed by stem cell transplantation may be prescribed. Vorinostat (Zolinza) and romidepsin (Istodax), which are histone deacetylase inhibitors, and bexarotene (Targretin), a retinoid, are approved for cutaneous T-cell lymphoma and could be used to treat the cutaneous form of ALCL.

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Pralatrexate (Folotyn) is approved for treatment of relapsed/refractory (disease does not respond to treatment) peripheral T-cell lymphoma, which includes ALCL. This is based on the overall treatment response rate of patients in a clinical trial; however, clinical benefit such as progression-free survival or overall survival was not demonstrated. Pralatrexate is a folate analogue metabolic inhibitor that interferes with cellular metabolism.

Brentuximab vedotin (Adcetris) was approved by the U.S. Food and Drug Administration for the treatment of patients with relapsed/refractory systemic ALCL. Both ALK-positive and ALK-negative subtypes of ALCL have been shown to respond well to brentuximab vedotin.

Treatments Under Investigation

New treatments for ALCL are being researched all the time. There are several new drugs currently in clinical trials that are showing promising results, including:

- Alisertib (MLN8237)
- Bortezomib (Velcade)
- Combination of brentuximab vedotin and chemotherapy
- Crizotinib (Xalkori)

It is critical to remember that today's scientific research is continuously evolving. Treatment options may change as new treatments are discovered and current treatments are improved. Therefore, it is important that patients check with their physician or with the Lymphoma Research Foundation (LRF) for any treatment updates that may have recently emerged.

Clinical Trials

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

Follow-up

Patients in remission should have regular visits with a physician who is familiar with their medical history and the treatments they have received. Medical tests (such as blood tests and CAT scans) may be required at various times during remission to evaluate the need for additional treatment.

Some treatments can cause long-term effects or late effects, which can vary based on duration and frequency of treatments, age, gender, and the overall health of each patient at the time of treatment. A physician will check for these effects during follow-up care.

Patients and their caregivers are encouraged to keep copies of all medical records and test results as well as information on the types, amounts, and duration of all treatments received. This documentation will be important for keeping track of any effects resulting from treatment or potential disease recurrences.

Resources

LRF offers a wide range of resources that address treatment options, the latest research advances, and ways to cope with all aspects of lymphoma, including our award-winning mobile app. LRF also provides many educational activities, from in-person meetings to teleconferences and webcasts, as well as E-Updates that provide the latest disease-specific news and treatment options. For more information about any of these resources, visit our websites at www.FocusOnALCL.org or www.lymphoma.org, or contact the LRF Helpline at (800) 500-9976 or helpline@lymphoma.org.