LYMPHOMA
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Getting the Facts

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Angioimmunoblastic T-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).

Angioimmunoblastic T-cell lymphoma (AITL) is a rare, aggressive (fast-growing) form of peripheral T-cell lymphoma (PTCL). While AITL only accounts for one to two percent of all NHL cases in the United States, it is one of the more common subtypes of mature T-cell lymphomas. Elderly patients are more likely to have AITL. Symptoms of AITL include high fever, night sweats, skin rash, and autoimmune disorders such as autoimmune hemolytic anemia (AIHA) and immune thrombocytopenia (ITP). As a result of these autoimmune disorders, the body's immune system does not recognize, and consequently destroys, its own cells and tissues, such as red blood cells (in the case of AIHA) or platelets (in the case of ITP).

Diagnosing AITL requires taking a small sample of the tumor tissue, called a biopsy, and looking at the cells under a microscope. A series of other tests may be done to determine the extent, or stage, of the disease. These 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.

The majority of patients with AITL are diagnosed with advanced-stage disease, which is either stage III or stage IV disease. In stage III, affected lymph nodes are found both above and below the diaphragm. In stage IV, one or more organs beyond the lymph nodes are affected, such as the bone, bone marrow, skin, or liver. Less extensive disease, stage I or II, is rare. Patients with stage I have localized disease that has not spread beyond the tumor; with stage II, if the cancer has spread, it has affected only a nearby lymph node.

Treatment Options

Patients with AITL may be treated with a steroid to relieve the symptoms caused by the immune system's reaction to the cancer cells, such as joint inflammation/pain and skin rash. Recommended first-line therapy for treatment of AITL is either a clinical trial or a multiagent chemotherapy regimen, such as CHOP (cyclophosphamide, doxorubicin, vincristine, and prednisone) and radiation therapy. Sometimes higher doses of chemotherapy followed by stem cell transplantation may be prescribed. Disease relapse (disease returns after treatment) is common with this cancer. Belinostat (Beleodaq) was approved by the U.S. Food and Drug Administration (FDA) for treatment of relapsed and/or refractory (disease does not respond to treatment) PTCL, including AITL. Belinostat is a histone deacetylase (HDAC) inhibitor that blocks tumor cells from growing and dividing, causing cell death. Similarly, another HDAC inhibitor, romidepsin (Istodax), has been approved to treat PTCL in patients who have received at least one prior therapy. Other recommended treatments following relapse may include high-dose chemotherapy followed by an autologous stem cell transplant (in which patients receive their own stem cells) or an allogeneic stem cell transplant (in which patients receive stem cells from a donor).

Treatments Under Investigation

Several drugs currently being tested in clinical trials (alone or in combination with current chemotherapy regimens) show promise for the treatment of AITL, including the following:

- Bortezomib (Velcade)
- Brentuximab vedotin (Adcetris)
- Fludarabine (Fludara)
- Lenalidomide (Revlimid)
- Panobinostat (Faridak)
- Rituximab (Rituxan)

These clinical trials are in various phases of development. 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.

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Supported through grants from:

Celgene
Genentech
biogen idec

Getting the Facts is published by the Lymphoma Research Foundation (LRF) for the purpose of informing and educating readers. Facts and statistics were obtained using published information, including data from the Surveillance, Epidemiology, and End Results (SEER) Program. Because each person's body and response to treatment is different, no individual should self-diagnose or embark upon any course of medical treatment without first consulting with his or her physician. The medical reviewer, the medical reviewer's institution, and LRF are not responsible for the medical care or treatment of any individual.

Last Updated December 2014

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Clinical Trials

Clinical trials are crucial in identifying effective drugs and determining optimal doses for patients with lymphoma. Because AITL is a rare disease, clinical trial enrollment is critical for establishing more effective, less toxic treatments. The rarity of the disease also means that the most novel treatments are often available only through clinical trials. 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

Once treatment is completed and AITL is in remission, physicians will continue to monitor the health and status of each patient during follow-up care. Patients in remission should have regular visits with a physician who is familiar with their medical history and the treatments they have received.

Disease relapse and infections are common with this cancer. It is important to seek medical attention for fever or other symptoms related to improper functioning of the immune system.

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. Visits may become less frequent the longer the disease remains in remission.

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.

Support

A lymphoma diagnosis often triggers a range of feelings and raises concerns. In addition, cancer treatment can cause physical discomfort. Support groups and online message boards can help patients connect with other people who have lymphoma. One-to-one peer support programs, such as the LRF Lymphoma Support Network, match lymphoma patients (or caregivers) with volunteers who have gone through similar experiences.

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. 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.FocusOnPTCL.org or www.lymphoma.org, or contact the LRF Helpline at (800) 500-9976 or helpline@lymphoma.org.