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, often but not always *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 attacks its own cells and tissues, such as red blood cells (AIHA) or platelets (ITP).

Diagnosing AITL requires taking a *biopsy* (sample of the tumor tissue) 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 tomography (CT) 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, either Stage III or Stage IV. 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 in AITL. Patients with Stage I have localized disease that has not spread beyond the tumor; Stage II disease has spread only to a nearby lymph node.

Treatment Options

Patients with AITL may be treated with a steroid that can temporarily relieve the symptoms caused by the immune system's reaction to the cancer cells, such as joint inflammation or pain and skin rash, and is a mild treatment for the lymphoma. 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). Sometimes higher doses of chemotherapy followed by stem cell transplantation may be added to multiagent chemotherapy.

Disease *relapse* (disease returns after treatment) is common with this cancer. If the cancer returns or does not go away with initial therapy, there are several other treatment options available. Belinostat (Beleodaq) was approved by the U.S. Food and Drug Administration (FDA) for treatment of relapsed 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 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).

Other Treatment Possibilities

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

- Brentuximab vedotin (Adcetris)
- Lenalidomide (Revlimid)
- Panobinostat (Farydak)

Some other drugs that are used for other lymphomas may at times be considered for use in patients with AITL that has relapsed or is refractory to other treatments, including the following:

- Alemtuzumab (Campath)
- Bendamustine (Treanda)
- Bortezomib (Velcade)
- Cyclosporine
- Fludarabine (Fludara)
- Gemcitabine (Gemzar)
- Pralatrexate (Folotyn)
- Rituximab (Rituxan)

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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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 view the *Understanding Clinical Trials* fact sheet on LRF's website at www.lymphoma.org, 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

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.

Patients with lymphoma 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 CT scans or at times PET 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. 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, including our award-winning mobile app. LRF also provides many educational activities, from in-person meetings to teleconferences and webcasts, as well as disease-specific websites, videos, and eNewsletters for current lymphoma information and treatment options. LRF's T-Cell Lymphoma Transportation Assistance Fund grant program provides financial assistance to T-cell lymphoma patients who are uninsured or have adequate medical insurance but struggle to pay for transportation costs (e.g., mileage, parking, tolls, gas, train tickets, etc.) to get to and from treatment. 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.