LYMPHOMA
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Getting the Facts

Cutaneous T-Cell Lymphoma (CTCL)

Overview

Lymphoma is the most common blood cancer. The two main forms of lymphoma are Hodgkin lymphoma 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).

T-cell lymphomas account for approximately 15 percent of all NHLs in the United States. There are many different forms of T-cell lymphomas, some of which are extremely rare. Most T-cell lymphomas can be classified into two broad categories: aggressive (fast-growing) or indolent (slow-growing).

One of the most common forms of T-cell lymphoma is cutaneous T-cell lymphoma (CTCL), a general term for T-cell lymphomas that involve the skin. CTCL also can involve the blood, the lymph nodes, and other internal organs. Symptoms can include dry skin, itching (which can be severe), a red rash, and enlarged lymph nodes. The disease affects men more often than women and usually occurs in men in their 50s and 60s.

Most patients with CTCL experience only skin symptoms, without serious complications; however, approximately 10 percent of those who progress to later stages develop serious complications. Early stage CTCL is typically indolent; some patients with early-stage CTCL might not progress to later stages at all, while others might progress rapidly, with the cancer spreading to lymph nodes and/or internal organs.

Types of Cutaneous T-Cell Lymphoma

CTCL describes many different disorders with various symptoms, outcomes, and treatment considerations. The two most common types are mycosis fungoides and Sézary syndrome.

Mycosis fungoides is the most common type of CTCL, with approximately 16,000 to 20,000 cases across the United States, accounting for half of all CTCLs. The disease looks different in each patient, with skin symptoms that can appear as patches, plaques, or tumors. Patches are usually flat, possibly scaly, and look like a rash; plaques are thicker, raised, usually itchy lesions that are often mistaken for eczema, psoriasis, or dermatitis; and tumors are raised bumps, which may or may not ulcerate. It is possible to have more than one type of lesion.

A medical history, physical exam, and skin biopsy are essential for diagnosis. A physician will examine lymph nodes, order various blood tests, and may conduct other screening tests, such as a chest x-ray or a computed axial tomography (CAT) scan. Scans are usually not needed for those with the earliest stages of the disease.

Mycosis fungoides is difficult to diagnose in its early stages because the symptoms and skin biopsy findings are similar to those of other skin conditions.

Sézary syndrome is an advanced, variant form of mycosis fungoides, which is characterized by the presence of lymphoma cells in the blood. Extensive thin, red, itchy rashes usually cover over 80 percent of the body. In certain patients, patches and tumors appear. Patients may also experience changes in the nails, hair, or eyelids, or have enlarged lymph nodes.

Many of the same procedures used to diagnose and stage other types of cutaneous T-cell lymphomas are used in Sézary syndrome. In addition, a series of imaging tests may be needed to determine if the cancer has spread to the lymph nodes or other organs (although that uncommonly occurs). These tests may include a CAT scan, a positron emission tomography (PET) scan, and/or a magnetic resonance imaging (MRI) scan. A bone marrow biopsy may also be done, but is usually not necessary.

Treatment Options

Treatment selection for CTCL depends on the extent of skin involvement, the type of skin lesion, and whether the cancer has spread to the lymph nodes or other internal organs.

For mycosis fungoides, treatment is either directed at the skin or the entire body (systemic). Because Sézary syndrome is chronic and systemic (affecting the entire body), it is usually not treated with skin-directed therapies alone. Treatments may be prescribed alone or in combination to achieve the best long-term benefit. Many patients live normal lives while they treat their disease, and some are able to remain in remission for long periods of time.

Skin-directed therapies are useful for patch and limited plaque disease and include topical treatments such as corticosteroids, retinoids, or imiquimod (which activates immune cells), topical chemotherapy, local radiation, methotrexate, photopheresis, ultraviolet light (phototherapy).

Systemic therapies for more advanced disease are usually deferred until patients have not responded well to topical therapies. More advanced disease is commonly treated with radiation,

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chemotherapy, and/or therapies such as:

- Bortezomib (Velcade)
- Denileukin diftitox (Ontak)
- Pralatrexate (Folotyn)
- Romidepsin (Istodax)
- Vorinostat (Zolinza)

Combination chemotherapy regimens are generally reserved for when patients have not responded well to several single-agent therapies. Options for refractory disease (disease that no longer responds to the initial therapy) include alemtuzumab (Campath), liposomal doxorubicin (Doxil), and gemcitabine (Gemzar)

Treatments Under Investigation

Many treatments at various stages of drug development are currently being tested in clinical trials and for various stages of CTCL, including everolimus (Afinitor), lenalidomide (Revlimid), brentuximab vedotin (Adcetris), panobinostat, forodesine, APO866, KW0761, and others. 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 LRF or with their physician for any treatment updates that may have recently emerged.

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 or contact LRF's 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 as well as with the treatments they have received.

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

The **Cutaneous Lymphoma Foundation** is an independent nonprofit patient advocacy organization dedicated to supporting every person with cutaneous lymphoma by promoting awareness and education, advancing patient care, and facilitating research. To find out more information about the resources that the Cutaneous Lymphoma Foundation provides, please visit the website (www.clfoundation.org).